MASTERING PEDIATRIC ECHOCARDIOGRAPHY

A Practical, Stepwise Approach — from Normal Physiology to Complex Congenital Heart Disease

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MBBS, FCPS

FIRST EDITION

Mastering Pediatric Echocardiography: A Practical, Stepwise Approach — from Normal Physiology to Complex Congenital Heart Disease

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First Edition

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Dedication

This book is dedicated to

The children with courageous hearts

who teach us resilience beyond textbooks, strength beyond physiology, and hope beyond diagnosis.

To the tiny neonates fighting their first battles, to the families who stand beside them with unwavering faith, and to every child whose heartbeat inspires us to learn more, think deeper, and care better.

I also dedicate this work to

My teachers and mentors

who transformed complex anatomy into understanding, and responsibility into purpose.

And to

My family

for their patience, encouragement, and constant support through the long hours of study, practice, and writing.

May this work serve the noble mission of healing young hearts.

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Preface

Pediatric echocardiography is more than an imaging technique—it is the language through which we understand congenital and acquired heart disease in children. From the fragile neonate in the intensive care unit to the growing child with complex congenital anatomy, echocardiography remains the most powerful bedside diagnostic tool in pediatric cardiology.

Despite its importance, many trainees find pediatric echo overwhelming. The anatomy is complex. The physiology is dynamic. The decisions are often urgent. Traditional teaching frequently emphasizes structural details but under-emphasizes clinical application.

This ECHO MODULE was created to bridge that gap.

The purpose of this book is to simplify pediatric echocardiography without diluting its depth. Each chapter focuses on clinically relevant interpretation, practical scanning strategy, hemodynamic understanding, and decision-making logic. The aim is not just to recognize anatomy—but to understand physiology and guide management.

This module is designed for:

- Pediatric cardiology fellows
- Neonatologists and pediatric intensivists
- Pediatricians involved in cardiac care
- Sonographers seeking clinical integration
- Trainees preparing for examinations

If this book helps you diagnose faster, think clearer, and manage more confidently—its purpose is fulfilled.

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MODULE 1

Foundations of Pediatric Echocardiography



Module 1 Chapters	Content
1. Basics of Pediatric Echocardiography	What pediatric echo is, why children are different, modalities, five questions, overall workflow
2. Ultrasound Physics	Frequency, wavelength, resolution, Doppler, aliasing, artifacts
3. Image Optimization	Depth, sector, gain, focus, frame rate, color and spectral Doppler settings
4. Standard Echo Views	One comprehensive version
5. Sequential Segmental Analysis	Situs → atria → ventricles → great arteries
6. Measurements & Z-Scores	Measurement technique + pediatric indexing
End-of-Module summary	Master Pediatric Echo Protocol

CHAPTER 1

Basics of Pediatric Echocardiography



Learning Objectives

By the end of this chapter, the learner should be able to:

1. **Define pediatric echocardiography** and explain its role in assessing cardiac anatomy, connections, function, and hemodynamics in neonates, infants, children, and adolescents.
2. **Explain why pediatric echocardiography differs from adult echocardiography**.
3. **Describe the major echocardiographic modalities**.
4. **Apply the principle of "2D anatomy first"** before using color and spectral Doppler to interpret blood flow and hemodynamics.
5. **Recognize the importance of sequential anatomical assessment**.
6. **Perform a systematic pediatric echocardiographic examination** using a reproducible sequence through the subcostal, apical, parasternal, and suprasternal windows.
7. **Understand the importance of systematic sweeps**.
8. **Recognize the importance of pediatric measurements, body surface area, reference values, and Z-scores** when interpreting cardiac dimensions.
9. **Integrate anatomical and hemodynamic findings with the clinical presentation**.

Pediatric echocardiography is much more than performing an adult echocardiogram on a smaller patient. Children differ substantially in **body size, heart rate, loading conditions, physiology, acoustic windows, and spectrum of congenital heart disease**. In addition, congenital heart disease often alters the basic arrangement of cardiac chambers and great vessels, so the echocardiographer cannot begin by assuming normal anatomy.

A high-quality pediatric echocardiogram therefore has three major purposes:

1. **Define cardiac anatomy.**
2. **Understand cardiovascular physiology and hemodynamics.**
3. **Answer the clinical question for the individual child.**

The central philosophy is:

> **Do not merely obtain images. Reconstruct the child's heart, understand the physiology, and determine the clinical significance.**

1. What Is Pediatric Echocardiography?

Pediatric echocardiography is the use of ultrasound to evaluate the **structure, connections, function, and hemodynamics of the heart and great vessels in neonates, infants, children, and adolescents**.

The examination should answer not only:

"Is there a structural abnormality?"

but also:

"How are the cardiac segments connected, what is the physiological consequence, and how should this finding influence management?"

A comprehensive examination combines several echocardiographic modalities, each providing a different part of the diagnostic picture.

2. The Main Echocardiographic Modalities

2.1 Two-Dimensional Echocardiography

2D imaging is the foundation of the examination.

It defines:

- cardiac position
- chamber morphology
- septal anatomy
- valve morphology
- ventricular outflow tracts
- great arteries
- systemic and pulmonary veins
- aortic arch
- coronary origins where technically feasible

The beginner should develop the habit:

Understand the anatomy in 2D before turning on color Doppler.
Color should confirm anatomy, not replace it.

2.2 M-Mode

M-mode displays movement along a single ultrasound line with excellent temporal resolution.

It may be useful for:

- ventricular dimensions
- wall thickness
- selected systolic function measurements
- valve motion
- timing of cardiac events

Its usefulness depends on correct alignment. An oblique M-mode cursor can produce inaccurate measurements.

3. Color Doppler

Color Doppler superimposes blood-flow information on the 2D image.

It helps identify:

- ASD and VSD flow
- PDA
- valve regurgitation
- stenotic/turbulent jets
- anomalous venous connections
- coronary flow
- collateral vessels

Color Doppler is highly dependent on machine settings.

A lesion may appear very different when:

- gain is changed,
- velocity scale is changed,
- color-box size is changed.

Therefore:

Color Doppler is a diagnostic tool—not a substitute for anatomical thinking.

4. Pulsed-Wave Doppler

PW Doppler measures velocity at a **specific sampling location**.

Typical applications include:

- mitral inflow
- tricuspid inflow
- pulmonary veins
- systemic veins
- LVOT
- RVOT
- pulmonary arteries
- descending aorta

Its major advantage is:

Location specificity

Its major limitation is:

Aliasing at high velocities

A useful memory rule is:

PW = Where is the velocity coming from?

5. Continuous-Wave Doppler

CW Doppler measures high velocities along the entire ultrasound beam.

It is particularly useful for:

- pulmonary stenosis
- aortic stenosis
- restrictive VSD
- coarctation
- high-velocity regurgitant jets

Its main limitation is reduced depth localization.

Remember:

PW tells you where.
CW tells you how fast.

6. Tissue Doppler Imaging

Tissue Doppler measures myocardial rather than blood velocities.

Common measurements include:

- **s′** — systolic myocardial velocity
- **e′** — early diastolic velocity
- **a′** — late diastolic velocity

Tissue Doppler can support assessment of systolic and diastolic ventricular function.

7. Three-Dimensional Echocardiography

Three-dimensional echo can provide additional anatomical information in selected patients.

Potential applications include:

- AV valve anatomy
- mitral valve morphology
- AVSD
- ASD morphology
- procedural planning
- postoperative assessment

It complements rather than replaces high-quality 2D imaging.

8. Speckle-Tracking and Strain Imaging

Strain imaging evaluates myocardial deformation.

It may help identify subtle ventricular dysfunction before conventional measures such as ejection fraction become abnormal.

It is particularly useful in selected patients with:

- cardiomyopathy
- chemotherapy exposure
- complex CHD
- postoperative ventricular dysfunction

For the beginner, however, conventional anatomy and hemodynamics should be mastered first.

9. Why Is Pediatric Echocardiography Different?

There are several important differences from routine adult echocardiography.

9.1 Patient Size Varies Enormously

A pediatric echocardiography service may evaluate:

- an extremely premature infant,
- a 3-kg neonate,
- a school-aged child,
- an adolescent,

- an infant,
- a toddler,
- or an adult with congenital heart disease.

Therefore, no single machine preset is suitable for all children.

The operator must adjust:

Frequency → Depth → Focus → Gain → Sector Width → Frame Rate → Doppler Settings

10. Heart Rates Are Faster

A neonate may have a heart rate of approximately:

140–180 beats/min

Therefor temporal resolution is particularly important.

Poor frame rate may make it difficult to assess:

- valve motion
- ventricular contraction
- rapid septal movement
- short Doppler events

Frame rate can often be improved by:

- reducing depth
- narrowing sector width
- reducing unnecessary focal zones
- reducing color-box size

Pediatric Echo Rule
Small heart + fast heart rate = prioritize frame rate.

11. Children Often Have Better Acoustic Windows

Children generally have:

- thinner chest walls
- less lung interference
- less body fat
- excellent subcostal access

This frequently permits high-resolution imaging that may not be possible in adults.

The **subcostal window** is especially powerful in infants and young children for:

- situs
- systemic venous anatomy
- atrial septum
- AV connections
- ventricular septum

12. Congenital Anatomy May Not Follow Normal Expectations

This is perhaps the most important difference.

A child may have:

- situs abnormality
- ventricular inversion
- TGA
- DORV
- single-ventricle physiology
- abnormal venous return
- abnormal great artery relationship
- abnormal coronary origins

Therefore:

Never assume anatomy. Demonstrate it.

A chamber should not be called the left ventricle simply because it is on the left side of the image.

A great vessel should not be called the aorta merely because it appears posterior.

Identify structures according to **morphology and connections**.

13. Pediatric Measurements Depend on Body Size

In adult echo, fixed measurement thresholds are commonly used.

In children, interpretation depends strongly on body size.

A dimension that is normal in a teenager may be severely enlarged in a newborn.

Therefore pediatric measurements frequently require:

- accurate height
- accurate weight
- body surface area
- validated pediatric reference values
- Z-scores where appropriate

The practical approach is:

Measure → Index → Interpret.

The module appropriately emphasizes Z-scores as a key difference between pediatric and adult echocardiography.

14. The Five Questions of Every Pediatric Echocardiogram

Before searching for a specific congenital lesion, ask five fundamental questions.

Question 1 — Where Is the Heart?

Determine:

- abdominal situs
- atrial arrangement
- cardiac position
- direction of the apex

Ask:

- Is the heart normally positioned?
- Is the apex leftward or rightward?
- What is the relationship between the aorta and IVC?

This establishes orientation before detailed anatomy is interpreted.

15. Question 2 — What Connects to What?

This is fundamental in congenital cardiology.

Follow blood flow in sequence:

Veins → Atria → Ventricles → Great Arteries

Establish:

Systemic venous return

Does the:

- IVC,
- SVC,

enter the appropriate atrium?

Pulmonary venous return

Can pulmonary veins be demonstrated entering the LA?

If one or more pulmonary veins are not confidently demonstrated, actively exclude **anomalous pulmonary venous connection**.

AV connections

Which atrium connects to which ventricle?

VA connections

Which great artery arises from each ventricle?

This single systematic habit prevents many diagnostic errors.

16. Question 3 — Is There an Abnormal Communication?

Search systematically for:

- PFO
- ASD
- VSD
- PDA
- AV septal communication
- abnormal vascular connections

Do not simply identify a communication.

Determine:

- location
- size
- direction of flow
- restriction
- hemodynamic effect

For example, the diagnosis is not merely:

"VSD."

A clinically useful evaluation asks:

What type of VSD? How large? What is the gradient? Is there pulmonary hypertension? Is the LV volume loaded? Is there aortic cusp prolapse?

17. Question 4 — Is There Obstruction or Regurgitation?

Systematically examine:

AV valves

- mitral valve
- tricuspid valve

Ventricular outflow tracts

- LVOT
- RVOT

Semilunar valves

- aortic valve
- pulmonary valve

Great vessels

- pulmonary arteries
- ascending aorta
- aortic arch

Use:

2D → Color Doppler → PW/CW Doppler

to determine:

- site of obstruction
- maximum velocity
- pressure gradient
- valve regurgitation
- physiological consequence

18. Question 5 — How Is the Heart Coping?

Once the anatomy is understood, assess its impact.

Evaluate:

- RA size
- LA size
- RV size
- LV size
- RV hypertrophy
- LV hypertrophy
- ventricular systolic function
- ventricular filling
- pressure loading
- volume loading
- pulmonary pressure
- pericardial effusion

This is the step that converts an anatomical diagnosis into a clinically useful echocardiographic assessment.

19. The Five-Question Framework

The entire pediatric echo can therefore be simplified into:

1. Where is the heart? — Orientation.

2. What connects to what? — Connections.

3. Is there an abnormal communication? — Shunts.

4. Is there obstruction or regurgitation? — Hemodynamics.

5. How is the heart coping? — Function and physiological impact.

Or more simply:

Anatomy → Connections → Communications → Hemodynamics → Function

20. The Overall Pediatric Echo Workflow

A high-quality study should proceed in a deliberate sequence.

STEP 1 — REVIEW THE CLINICAL QUESTION

Before touching the probe, know:

- age
- weight
- height
- oxygen saturation
- clinical presentation
- previous diagnosis
- surgery/intervention history
- reason for examination

21. Step 2 — Optimize the Machine

Select:

- appropriate transducer
- appropriate frequency, appropriate depth
- sector width, focus
- gain, frame rate

Do not accept the default preset without adjustment.

22. Step 3 — Establish Orientation and Situs

Begin by determining:

- abdominal situs
- position of the aorta and IVC
- cardiac position
- apex direction

If orientation is misunderstood at the beginning, the entire examination may subsequently be mislabeled.

23. Step 4 — Perform Sequential Segmental Analysis

Assess:

Atria → AV connections → Ventricular morphology → VA connections → Great artery relationship

This should precede lesion-specific labeling in complex CHD.

Avoid:

"This looks like TGA."

Instead establish:

- situs
- AV connection
- ventricular morphology
- VA connection
- great artery relationship

Then make the diagnosis.

24. Step 5 — Perform a Systematic Window-Based Examination

A practical sequence is:

Subcostal → Apical → Parasternal → Suprasternal

The exact order may vary, but the examination should never become random. Each window provides complementary information.

SUBCOSTAL

Situs, septa, venous return.

APICAL

AV connections, chamber size, function, valve regurgitation.

PARASTERNAL

Valves, outflows, ventricular septum, coronary origins.

SUPRASTERNAL

Arch, great vessels, selected venous anatomy.

The module repeatedly emphasizes the importance of a fixed scanning sequence.

25. Step 6 — Start With 2D Anatomy

Before Doppler:

- identify the structure
- determine morphology
- establish relationships
- sweep through it

Then:

Add color Doppler
to establish flow direction and turbulence.

Then:

Add spectral Doppler
to quantify velocity and hemodynamics.

A simple rule:

2D → Color → PW/CW → Measurement → Interpretation

26. Step 7 — Sweep, Do Not Photograph

Congenital heart anatomy is three-dimensional. A single still image is only one plane through the heart.

Therefore:

Do not photograph the heart. Sweep through it.
For every important structure:

Find → Sweep → Optimize → Color → Doppler → Measure

This is particularly important for:

- atrial septum, ventricular septum
- pulmonary veins, coronary arteries
- RVOT, LVOT
- branch pulmonary arteries, aortic arch

27. Step 8 — Quantify

Measurements should answer a clinical question.

Assess, where relevant:

- valve annuli
- ventricular dimensions
- aortic root
- ascending aorta
- pulmonary arteries
- coronary dimensions
- Doppler velocities
- pressure gradients
- ventricular function

Use pediatric reference data and Z-scores where appropriate.

Avoid measuring simply because the machine provides a button for it.

Good measurement begins with a correct imaging plane.

28. Step 9 — Integrate Hemodynamics

Do not stop after describing anatomy.

Ask:

- Is there pressure overload?
- Is there volume overload?
- What is the shunt direction?
- What is the estimated RV pressure?
- Is pulmonary blood flow increased or reduced?
- Is systemic output compromised?
- Is the lesion restrictive or nonrestrictive?
- Is ventricular function preserved?

29. Step 10 — Complete the Entire Examination

One obvious abnormality should not stop the examination.

For example:

A child referred with VSD may also have:

- additional muscular VSD
- aortic cusp prolapse
- coarctation
- pulmonary stenosis
- arch abnormality
- abnormal venous anatomy

Once the dominant lesion is found:

Return to the systematic sequence and complete the study.

30. The MASTER Approach

To create a consistent framework throughout this textbook, every lesion can be approached using:

M — MORPHOLOGY

What does the abnormal structure look like?

A — ANATOMY & CONNECTIONS

Where is the lesion and what connects to what?

S — SHUNTS

Is there abnormal communication or abnormal flow?

T — TURBULENCE & PRESSURE

Is there obstruction, regurgitation or abnormal pressure?

E — EFFECT ON THE HEART

What is happening to chamber size and ventricular function?

R — RELEVANCE

31. Image Quality Comes Before Diagnosis

A useful way to understand pediatric echo is as a diagnostic pyramid:

LEVEL 5 — CLINICAL DECISION
What does this mean for the child?

↑

LEVEL 4 — HEMODYNAMICS
Pressure, flow, shunt and obstruction.

↑

LEVEL 3 — DOPPLER
Direction and velocity of blood flow.

↑

LEVEL 2 — ANATOMY
What structure is abnormal?

↑

LEVEL 1 — IMAGE QUALITY
Can the structure actually be seen properly?

A technical error at the bottom can propagate upward.

Poor image → wrong measurement → wrong physiological interpretation → wrong clinical decision.

Therefore:

Optimize before you interpret.

32. What Should a "Normal Pediatric Echo" Establish?

A normal pediatric echocardiogram should be more than a normal four-chamber image.

As technically feasible, establish:

Situs → Systemic venous return, Pulmonary venous return → Atrial anatomy, Atrial septum → AV connections

Ventricular morphology, Ventricular septum → VA connections → AV valves → RVOT and LVOT

Semilunar valves → Main and branch pulmonary arteries → Aortic root and arch → Coronary origins where appropriate/adequately visualized

33. Common Beginner Mistakes

MISTAKE 1 — STARTING WITH COLOR DOPPLER

Color draws attention immediately, but it can distract from anatomy.

Correction: 2D first.

MISTAKE 2 — ASSUMING NORMAL CONNECTIONS

Correction: Demonstrate veins → atria → ventricles → arteries.

MISTAKE 3 — IDENTIFYING VENTRICLES BY POSITION

Correction: Use ventricular morphology:

RV	LV
moderator band	smoother cavity
coarse trabeculation	no moderator band
more apical AV valve insertion	more basal AV valve insertion

MISTAKE 4 — STOPPING AFTER FINDING THE MAIN LESION

Correction: Complete the entire sequential study.

MISTAKE 5 — USING ONE PRESET FOR EVERY CHILD

Correction: Adapt the machine according to:

- body size
- heart rate
- acoustic window
- diagnostic question

MISTAKE 6 — TRUSTING A SINGLE VIEW

Correction: Confirm important findings in multiple planes.

34. Clinical Example — VSD

A 2-month-old infant presents with tachypnea, feeding difficulty and a murmur. Echo shows a VSD.

The beginner may report:

"VSD present."

The pediatric echocardiographer asks:

- What type of VSD?
- What is its anatomical location?
- How large is it?
- Is the shunt restrictive?
- What is the Doppler velocity?
- Is the shunt left-to-right or bidirectional?
- Is the LA enlarged?
- Is the LV volume loaded?
- What is the RV pressure?
- Is there aortic valve prolapse?
- Is there RVOT obstruction?
- Is the aortic arch normal?
- Are there additional defects?

The difference is fundamental:

Finding the lesion is the beginning of the diagnosis. Understanding its physiology makes the echocardiogram clinically useful.

35. Ten Rules for Beginning Pediatric Echocardiography

1. Never assume anatomy.
2. Use a fixed scanning sequence.
3. Establish 2D anatomy before relying on color Doppler.
4. Identify ventricles by morphology, not position.
5. Sweep every important structure.
6. Optimize the machine for each child and each diagnostic question.
7. Use Doppler only when alignment is appropriate.
8. Interpret measurements using pediatric reference values where appropriate.
9. Never stop after finding the first abnormality.
10. Always ask whether the echo findings explain the clinical presentation.

36. Clinical Pearls

CLINICAL PEARL 1

A beautiful image is not necessarily a complete echocardiogram.

CLINICAL PEARL 2

If anatomy does not make sense, return to situs and reconstruct the heart from the beginning.

CLINICAL PEARL 3

Color Doppler should answer a question generated by the 2D anatomy.

CLINICAL PEARL 4

In congenital heart disease, connections are as important as structures.

CLINICAL PEARL 5

A lesion should always be described anatomically, hemodynamically and functionally.

37. The Complete Pediatric Echo Workflow

For practical daily use:

Before the Study: Clinical history and indication

Optimize → Orient → Connect → Inspect

Interrogate → Quantify → Evaluate Effect

Integrate → Report

- **Optimize** — Probe + frequency + depth + focus + gain + sector
- **Orient** — Situs + position + apex
- **Connect** — Venous → atrial → ventricular → arterial connections
- **Inspect** — Septa + valves + outflow tracts + vessels
- **Interrogate** — Color + PW + CW
- **Quantify** — Measurements + Z-scores + pressure estimates
- **Evaluate Effect** — Chambers + ventricular function + pressure/volume loading
- **Integrate** — Does the echo explain the clinical picture?
- **Report** — Clear structured impression

Chapter Summary

Pediatric echocardiography is a **systematic anatomical and physiological examination**, not simply a method of producing ultrasound images.

Children differ from adults because of:

- wide variation in body size,
- faster heart rates,
- excellent but age-dependent acoustic windows,
- the need for body-size-adjusted measurements,
- and, most importantly, the possibility of fundamentally abnormal cardiac connections.

A complete examination uses:

- 2D imaging, M-mode
- Color Doppler, PW Doppler, CW Doppler
- selected tissue Doppler, strain and 3D imaging

but the foundation remains:

2D anatomy first.

The five questions that should guide every pediatric echo are:

1. Where is the heart?
2. What connects to what?
3. Is there an abnormal communication?
4. Is there obstruction or regurgitation?
5. How is the heart coping?

The overall workflow is:

Optimize → Orient → Connect → Inspect → Interrogate → Quantify → Interpret → Report

GOLDEN RULE

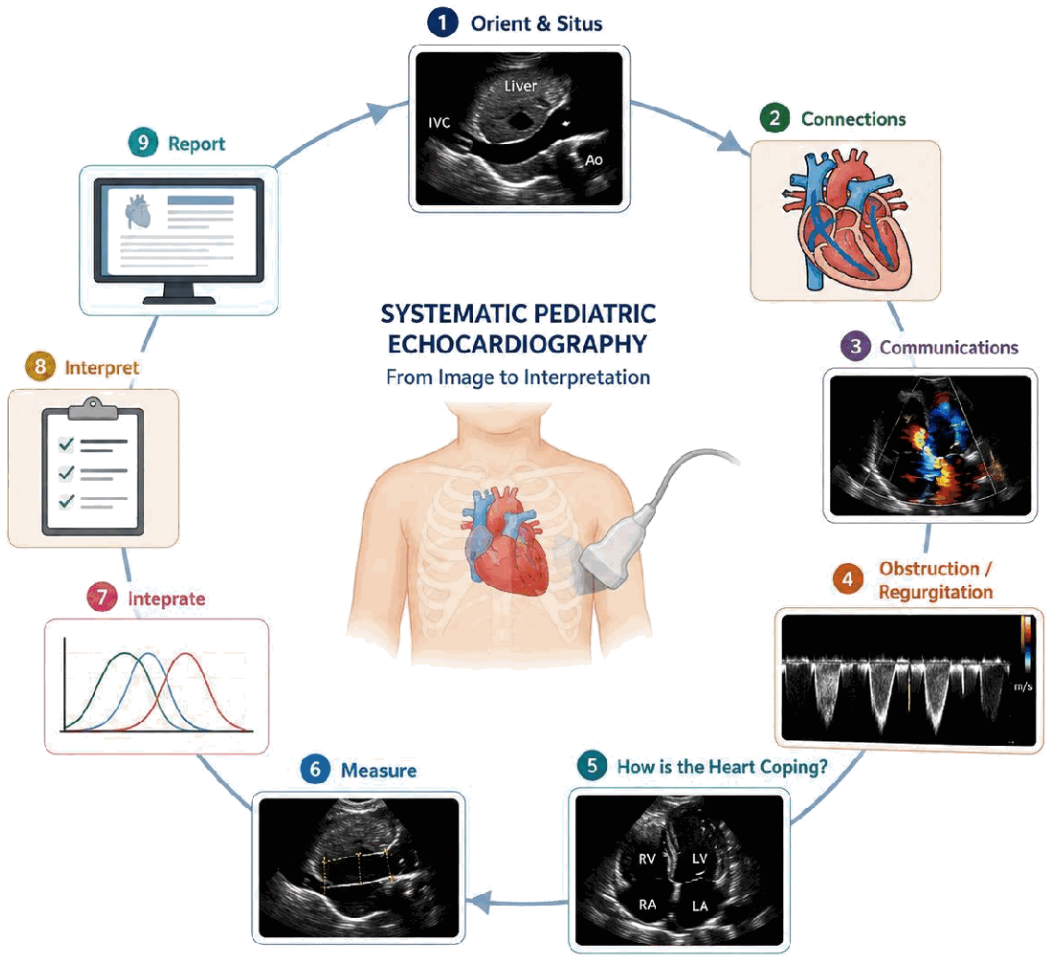
Never simply look for a diagnosis. Reconstruct the anatomy systematically, define the physiology, and determine what the findings mean for the child.

Table 1.2 — Key Differences Between Pediatric and Adult Echocardiography

Aspect	Pediatric Echocardiography	Adult Echocardiography
Patient size	Wide range (premature neonate to adolescent)	Relatively uniform
Heart rate	Faster (e.g., 120–180 bpm in neonates)	Slower (typically 60–100 bpm)
Transducer frequency	Higher frequency often used (≈ 10–12 MHz in neonates, 1–3 MHz in older children)	Lower frequency (≈ 2–5 MHz)
Acoustic windows	Generally excellent (thin chest wall, less lung interference, less body fat)	Often more limited (lung, rib and body habitus interference)
Spectrum of disease	Predominantly congenital heart disease (wide anatomical variability)	Predominantly acquired heart disease (coronary, valvular, cardiomyopathy, etc.)
Anatomical assumptions	Cannot assume normal anatomy (connections may be abnormal)	Usually normal segmental anatomy
Measurement interpretation	Depends on body size (Z-scores, indexed values)	Often uses fixed reference ranges
Hemodynamics	Physiology influenced by transitional circulation, shunts and congenital lesions	Mostly acquired lesions and comorbidities
Typical clinical questions	Define anatomy, connections, shunts, obstruction, and physiological impact	Assess ventricular function, ischemia, valvular disease, cardiomyopathy, etc.
Examination duration	May be longer (systematic segmental analysis)	Often shorter, focused studies
Longitudinal follow-up	Lifelong follow-up often required (growth, repair, residual lesions)	Follow-up depends on specific disease

KEY MESSAGE

Pediatric echocardiography is not simply an adult echocardiogram on a smaller patient. Children have different hearts, different physiology and different questions.

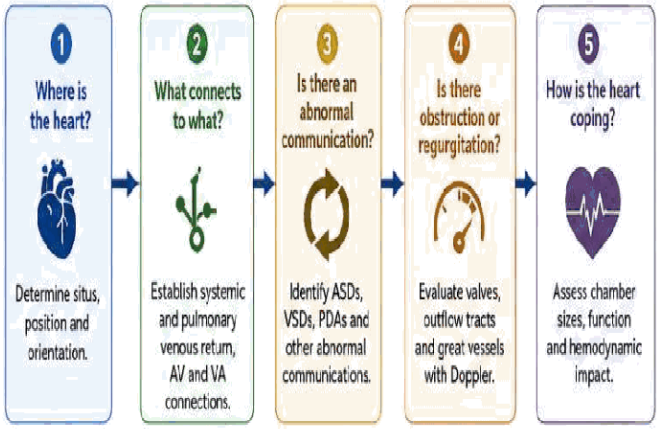

1 Orient & Situs
Liver
IVC
Ao
2 Connections
3 Communications
4 Obstruction / Regurgitation
m/s
5 How is the Heart Coping?
RV
LV
RA
LA
6 Measure
7 Inteprate
8 Interpret
9 Report
SYSTEMATIC PEDIATRIC ECHOCARDIOGRAPHY
From Image to Interpretation

The Overall Pediatric Echocardiography Workflow

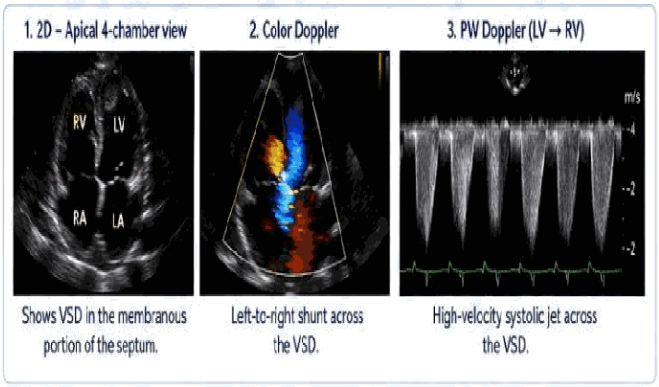
A high-quality pediatric echocardiogram is a systematic process that moves from patient information to image acquisition, interpretation and clinical integration. The workflow below ensures that important anatomy is not missed and that the final report answers the clinical question for the child.

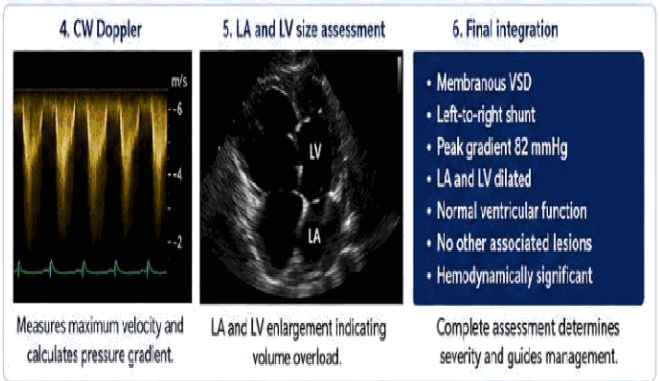
1 Before the Study	• Review clinical history, indication, previous echo/surgery • Note age, weight, height, saturation, and current support
2 Optimize the Machine	• Select appropriate transducer and frequency • Adjust depth, sector width, focus, gain, and frame rate (children → small heart + fast heart rate = prioritize frame rate)
3 Orient and Assess Situs	• Determine abdominal situs (stomach, liver, IVC) • Identify cardiac position and apex direction • Establish relationship of IVC and aorta
4 Define Connections (Systematic Anatomy)	• Systemic venous return (SVC, IVC → right or left atrium?) • Pulmonary venous return (all veins to left atrium?) • AV connections (which atrium → which ventricle?) • VA connections (which ventricle → which great artery?)
5 Inspect Structures	• Atrial and ventricular septa • AV valves • Ventricular outflow tracts • Semilunar valves • Great arteries and aortic arch • Coronary origins (when feasible) and pericardium
6 Interrogate with Doppler	• Color Doppler to assess flow direction and turbulence • PW Doppler for specific locations (inflow/outflow, veins) • CW Doppler for high-velocity jets and pressure gradients • Tissue Doppler as needed for function assessment
7 Quantify	• Measure relevant dimensions • Calculate Doppler gradients and estimate pressures • Index measurements to body size (Z-scores when possible)
8 Evaluate the Effect (How is the Heart Coping?)	• Chamber sizes and hypertrophy • Ventricular systolic and diastolic function • Pressure and volume loading • Pulmonary pressure and flow • Pericardial effusion
9 Integrate and Report	• Does the echo explain the clinical presentation? • Provide clear, structured, and clinically relevant report • Suggest further evaluation if needed

The Five Questions of Every Pediatric Echocardiogram



Example: Sequential Evaluation in a Child with Ventricular Septal Defect (VSD)





Remember: 2D anatomy first → then Doppler → then hemodynamic interpretation.
Never stop after finding the first abnormality. Complete the entire systematic study.

CHAPTER 2

Ultrasound Physics



Learning Objectives

By the end of this chapter, the learner should be able to:

1. Explain the basic principles of ultrasound generation and image formation.
2. Describe frequency, wavelength, propagation speed, amplitude, power, and intensity.
3. Explain the relationship between **frequency, resolution, and penetration**.
4. Understand reflection, scattering, refraction, absorption, and attenuation.
5. Differentiate **axial, lateral, and temporal resolution**.
6. Explain how depth, sector width, focus, and frequency influence image quality and frame rate.
7. Understand the basic principles of **2D, M-mode, color, PW, and CW Doppler**.
8. Explain the Doppler equation, Doppler angle, PRF, Nyquist limit, and aliasing.
9. Apply the simplified Bernoulli equation appropriately in pediatric echocardiography.
10. Recognize common ultrasound artifacts and avoid mistaking them for congenital pathology.

1. Why Does a Pediatric Echocardiographer Need Physics?

The purpose of learning ultrasound physics is not to memorize formulas. It is to understand **why the image looks the way it does and what to change when it is inadequate**.

Physics helps answer practical questions:

- Why is a coronary artery poorly defined?
- Why can I see the near field but not the far field?
- Why did reducing depth improve the neonatal image?
- Why does a large color box make the image sluggish?
- Why is a stenotic gradient unexpectedly low?
- Why does a septum appear to have a defect in one view but not another?
- Why can pulmonary venous or coronary color flow disappear with inappropriate settings?

The central principle is:

Learn the physics that changes your image and your diagnosis.

2. What Is Ultrasound?

Sound is a **mechanical wave** that requires a medium through which to travel.

Human hearing extends approximately from:

20 Hz to 20,000 Hz
Sound above 20 kHz is called **ultrasound**.

Diagnostic echocardiography uses frequencies in the **megahertz (MHz)** range:

1 MHz = 1,000,000 Hz
Pediatric echocardiography frequently uses relatively high-frequency transducers because the heart is generally closer to the transducer and the anatomical structures are small.

3. Basic Properties of an Ultrasound Wave

The important physical characteristics are:

Property	Meaning	Clinical Relevance
Frequency	Cycles per second	Resolution vs penetration
Wavelength	Length of one cycle	Influences axial resolution
Propagation speed	Speed through tissue	Used to calculate depth
Amplitude	Strength of the wave	Related to echo strength
Power	Energy delivered per unit time	Acoustic output

Property	Meaning	Clinical Relevance
Intensity	Power per unit area	Related to acoustic exposure

For daily pediatric echocardiography, **frequency and wavelength** are particularly important.

4. Frequency

Frequency is the number of ultrasound cycles occurring each second.

It is measured in Hertz:

- 1 MHz = 1 million cycles/s
- 5 MHz = 5 million cycles/s
- 10 MHz = 10 million cycles/s

The most important clinical relationship is:

Higher frequency → Better resolution → Less penetration
Lower frequency → Greater penetration → Lower resolution
This is the fundamental trade-off in transducer selection.

5. Frequency Selection in Children

A premature neonate may allow very high-frequency imaging because the heart is superficial.

A larger adolescent may require a lower-frequency probe to reach deeper structures.

Therefore, do not choose frequency simply according to age.

Consider:

- body size
- chest-wall thickness
- imaging depth
- acoustic window
- structure of interest

GOLDEN RULE
Use the highest frequency that still provides adequate penetration.

PRACTICAL EXAMPLE

A neonate has excellent penetration, but the coronary origins look poorly defined. **Think:** Can I increase the frequency?

A large child has a bright near field, but the deeper LV and LA are poorly visualized. **Think:** Do I need a lower frequency rather than simply more gain?

6. Wavelength

Wavelength (λ) is the physical distance occupied by one complete ultrasound cycle.

The relationship is:

λ = c / f

where **λ** = wavelength, **c** = speed of sound in the medium, and **f** = ultrasound frequency.

So the key rule is:

Higher frequency → shorter wavelength → better spatial resolution, but less tissue penetration.

Frequency ↑ → Wavelength ↓

Shorter wavelengths improve the ability to distinguish small structures. This helps explain why high-frequency probes are valuable for visualizing:

- coronary origins
- valve leaflets
- small VSDs
- atrial septal margins
- PDA anatomy
- small branch pulmonary arteries

7. Propagation Speed

The ultrasound machine assumes that sound travels through soft tissue at approximately:

1540 m/s

The machine determines the depth of a structure by measuring the time required for the pulse to travel to that structure and return.

Because the ultrasound travels **to the structure and back**, depth is approximately:

(c × time) / 2 = Depth

This is the basis of pulse-echo imaging.

8. How Is an Ultrasound Image Produced?

The process can be simplified as:

Electrical energy → Transducer generates ultrasound → Ultrasound travels through tissue → Tissue interfaces reflect/scatter part of the sound → Returning echoes reach the transducer

Transducer converts them into electrical signals → Machine calculates location and echo strength → Image appears on the screen

The time taken for the echo to return determines **depth**. The strength of the returning echo contributes to **brightness**.

9. The Piezoelectric Effect

The transducer contains piezoelectric elements. When an electrical voltage is applied, these elements deform and generate ultrasound.

The reverse also occurs. Returning ultrasound waves deform the elements and generate electrical signals.

Therefore, the transducer works as both:

Transmitter + Receiver

This conversion between electrical and mechanical energy is called the **piezoelectric effect**.

10. What Happens When Ultrasound Meets Tissue?

Ultrasound may undergo:

- reflection
- scattering
- refraction
- absorption
- transmission

The final image is the result of these interactions.

11. Reflection

Reflection occurs when ultrasound reaches an interface between tissues with different acoustic properties.

Some of the sound returns toward the transducer. Strong reflectors produce stronger echoes and appear brighter.

Examples include:

- pericardium
- valve tissue
- calcification
- prosthetic material
- surgical patches
- occlusion devices

The degree of reflection is influenced by differences in **acoustic impedance**.

12. Acoustic Impedance

Acoustic impedance is defined as:

Z = ρc = rho c

where:

- Z = acoustic impedance
- ρ (rho) = tissue density
- c = propagation speed

A large difference in acoustic impedance between two tissues produces greater reflection at their interface.

WHY IS GEL NECESSARY?

Air creates a major acoustic mismatch between the probe and skin. Ultrasound gel eliminates the air interface and allows efficient transmission of ultrasound into the body.

13. Scattering

When ultrasound interacts with structures smaller than the wavelength or with irregular surfaces, energy is scattered in multiple directions.

Scattering contributes to tissue appearance and is fundamental to Doppler imaging because moving red blood cells scatter ultrasound back toward the transducer.

14. Refraction

Refraction is a change in direction of the ultrasound beam when it crosses an interface obliquely between tissues with different propagation properties.

It can:

- displace structures
- duplicate structures
- create misleading appearances

When anatomy appears inconsistent:

Change the imaging plane before accepting the finding.

15. Absorption and Attenuation

As ultrasound travels through tissue, its intensity progressively decreases. This loss of energy is called **attenuation**.

Attenuation results mainly from:

- absorption
- reflection
- scattering

A crucial relationship is:

Higher-frequency ultrasound undergoes greater attenuation.
This explains why high-frequency ultrasound provides excellent detail but poorer deep penetration.

16. Resolution

Resolution describes the ability to distinguish two nearby structures as separate.

Three forms are particularly important:

Axial resolution — along the ultrasound beam.

Lateral resolution — side by side.

Temporal resolution — changes over time.

17. Axial Resolution

Axial resolution is the ability to distinguish two structures lying one behind another along the direction of the beam.

It depends primarily on **spatial pulse length**. Shorter spatial pulse length improves axial resolution. Higher frequencies generally permit better axial resolution.

PEDIATRIC APPLICATION

Good axial resolution is important when assessing:

- thin septa
- small VSDs
- valve leaflets
- coronary arteries
- PDA morphology

18. Lateral Resolution

Lateral resolution refers to the ability to distinguish two structures lying side by side, perpendicular to the ultrasound beam.

It depends largely on **beam width**. The beam is narrowest near the focal zone. Therefore:

Place the focus at or just below the structure of interest.
Examples:

- **Coronary artery** → focus near aortic root
- **ASD** → focus near atrial septum
- **Pulmonary valve** → focus near RVOT/pulmonary valve
- **PDA** → focus near the ductal region

19. Temporal Resolution

Temporal resolution is the ability to accurately display rapidly changing cardiac events. It depends strongly on **frame rate**.

This is especially important in neonates and infants because their hearts beat rapidly. A neonatal heart rate may be:

140–180/min

A low frame rate can blur:

- valve motion
- septal movement
- ventricular contraction
- short-duration cardiac events

20. What Determines Frame Rate?

Frame rate generally improves when you:

- reduce depth
- narrow sector width
- reduce unnecessary focal zones
- reduce color-box size

It decreases when you:

- increase depth
- widen the sector
- use multiple focal zones
- use a large color Doppler box

PEDIATRIC ECHO PEARL

Small heart + fast heart rate = prioritize temporal resolution.

21. Depth and Pulse Repetition Frequency

The machine must wait for echoes from the deepest region before sending another pulse along the same line.

Therefore:

Greater depth → Longer waiting time → Lower PRF → Lower frame rate

This explains why unnecessary depth is particularly undesirable in neonatal echocardiography.

TECHNIQUE TIP

Use the shallowest depth that completely includes the anatomy of interest.

22. Sector Width

A wide sector displays more anatomy but requires more ultrasound lines to construct each frame.

Therefore:

Wide sector → More anatomy, lower frame rate
Narrow sector → Less anatomy, higher frame rate
A useful approach is:

Survey wide → Interrogate narrow.
First obtain orientation. Then narrow the sector around the structure of interest.

23. Focus

The focal zone is where the ultrasound beam is narrowest and lateral resolution is best. Place the focus at the level of the structure being examined.

Too many focal zones may improve selected spatial detail but reduce frame rate. In a tachycardic neonate, a single appropriately placed focus is often preferable for targeted imaging.

24. Gain

Gain amplifies returning ultrasound signals. It affects **display brightness**. It does not increase the ability of ultrasound to penetrate deeper tissue.

TOO MUCH GAIN

May produce:

- noisy chambers
- apparently thick valve leaflets
- loss of tissue boundaries
- false impression of septal thickening

TOO LITTLE GAIN

May produce:

- poor endocardial definition
- loss of thin structures
- apparent septal dropout

IMPORTANT RULE

If penetration is inadequate, do not simply increase gain—reconsider frequency and acoustic window.

25. Time-Gain Compensation

Deeper echoes are weaker because they have travelled farther and undergone greater attenuation. Time-gain compensation allows different amplification at different depths.

For example, **near field adequate + far field too dark** → adjust deeper TGC rather than making the entire image excessively bright.

26. Dynamic Range

Dynamic range determines how many shades of gray are displayed.

WIDER DYNAMIC RANGE

- more gray shades
- softer image
- lower contrast

NARROWER DYNAMIC RANGE

- fewer gray shades
- greater contrast

The goal is not simply to make the image attractive. The goal is to distinguish:

- blood pool
- endocardium
- myocardium
- valves
- septa
- vessel walls

27. M-Mode Physics

M-mode repeatedly samples one ultrasound line over time. Because only one line is repeatedly processed, it has excellent temporal resolution.

It can be useful for:

- ventricular dimensions
- wall motion
- selected valve motion
- timing relationships

However:

An oblique M-mode cursor produces an oblique measurement.
Good physics cannot compensate for incorrect anatomical alignment.

28. The Doppler Effect

Doppler echocardiography measures changes in ultrasound frequency caused by moving red blood cells.

The Doppler equation is:

$\Delta f = 2f_0 V \cos\theta / c$

where:

- Δf = Doppler frequency shift
- f_0 = transmitted frequency
- V = blood velocity
- θ = angle between beam and blood flow
- c = propagation speed

WHY IS THERE A FACTOR OF "2"?

The **2** is present because **two separate Doppler shifts occur** during a round-trip echo:

1. The first shift happens when the stationary source transmits a wave received by the moving target.
2. The second shift happens when the moving target acts as a moving source, reflecting the wave back to the stationary receiver.

For clinical echocardiography, the most important component is **θ**, the angle between beam and blood flow, because this determines the effect of beam alignment.

When the beam is parallel to blood flow, this gives the optimal Doppler shift. As the angle increases, measured velocity becomes increasingly underestimated. At 90°, there is essentially no useful Doppler shift from motion perpendicular to the beam.

GOLDEN DOPPLER RULE

Doppler wants parallel alignment.

This is particularly important when assessing:

- pulmonary stenosis
- aortic stenosis
- coarctation
- RVOT obstruction
- LVOT obstruction
- VSD gradients
- TR velocity

30. Pulsed-Wave Doppler

PW Doppler transmits pulses and listens for returning signals from a selected location.

Its major advantage is:

Range specificity

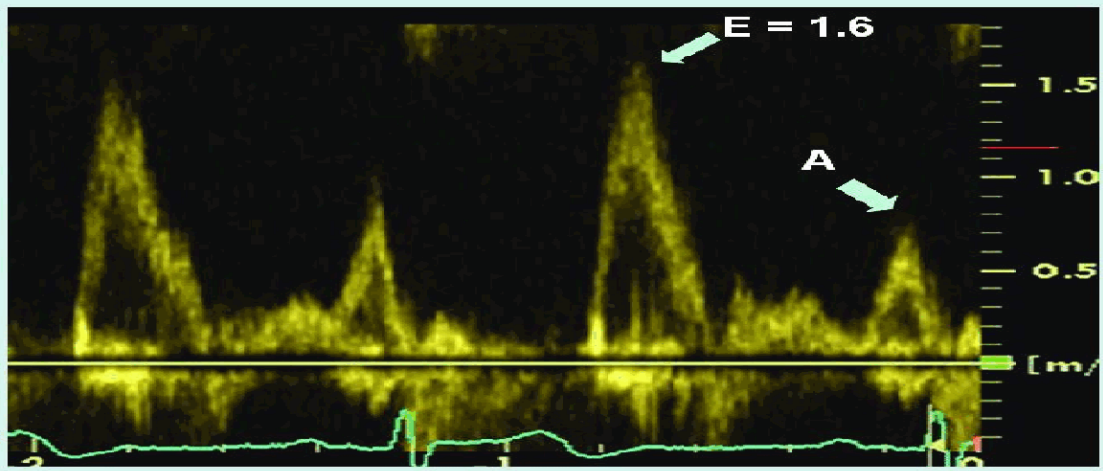
The sample volume can be placed exactly where flow is being assessed.

Common applications:

- mitral inflow
- tricuspid inflow
- pulmonary veins
- LVOT
- RVOT
- branch pulmonary arteries
- descending/abdominal aorta

Its major limitation is:

Aliasing at high velocities.



31. Continuous-Wave Doppler

CW Doppler continuously transmits and receives ultrasound. It can measure very high velocities without the same aliasing limitation as PW Doppler.

Useful for:

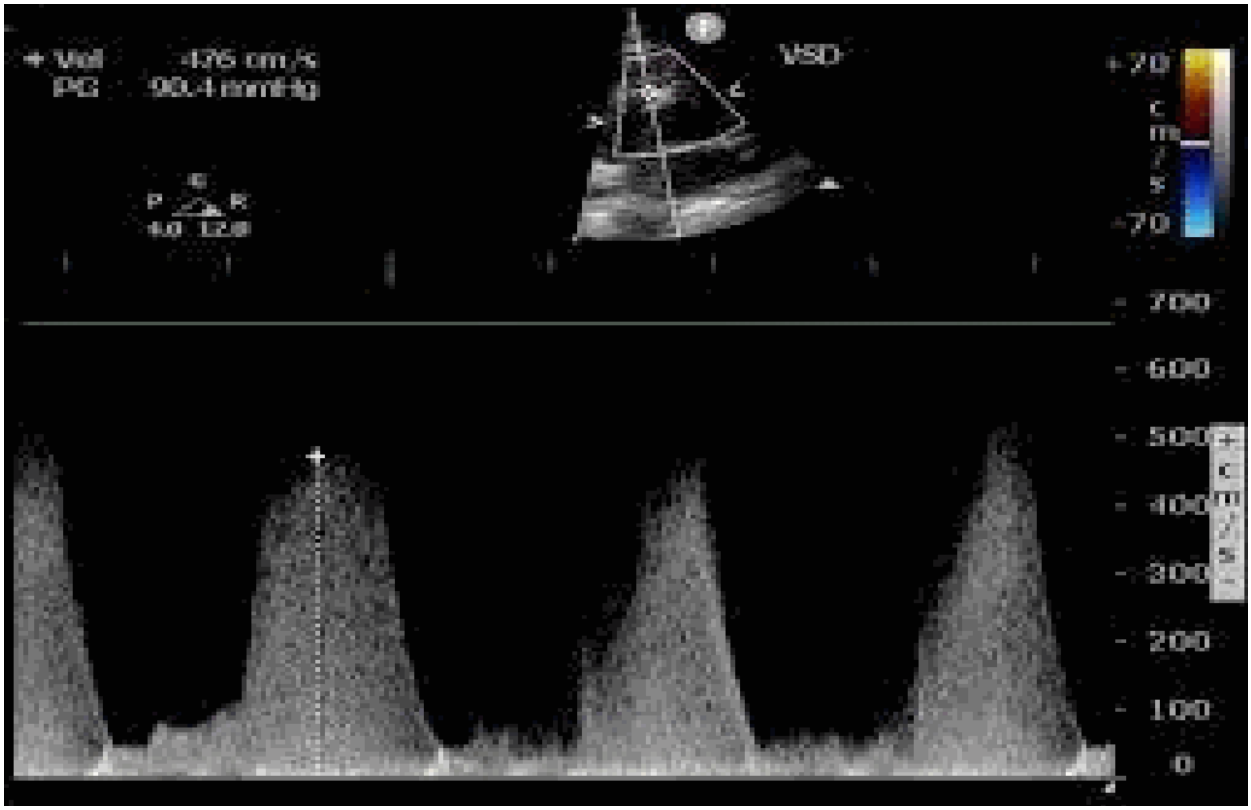
- pulmonary stenosis
- aortic stenosis
- restrictive VSD
- coarctation
- high-velocity regurgitant jets

Its limitation is:

Lack of precise depth localization.

MEMORY RULE

PW = Where? CW = How fast?



32. Pulse Repetition Frequency and the Nyquist Limit

Pulse repetition frequency (PRF) is the number of ultrasound pulses emitted per second.

For PW Doppler, the maximum Doppler shift that can be displayed without aliasing is related to the Nyquist limit:

Nyquist Limit = PRF / 2

If the Doppler shift exceeds the Nyquist limit, **aliasing occurs**.

33. Aliasing

On spectral Doppler, aliasing causes the waveform to wrap around the baseline. On color Doppler, exceeding the selected velocity limit produces abrupt color changes.

Aliasing does **not automatically mean severe disease**.

Its appearance depends on:

- blood velocity
- PRF
- depth
- transmitted frequency
- selected scale

PRACTICAL RESPONSES TO PW ALIASING

Depending on the clinical question:

- increase velocity scale/PRF
- shift the baseline
- reduce depth where feasible
- use a lower transmitted frequency
- switch to CW Doppler for a true high-velocity jet

34. Color Doppler Physics

Color Doppler displays information about blood-flow direction and mean velocity over the 2D image.

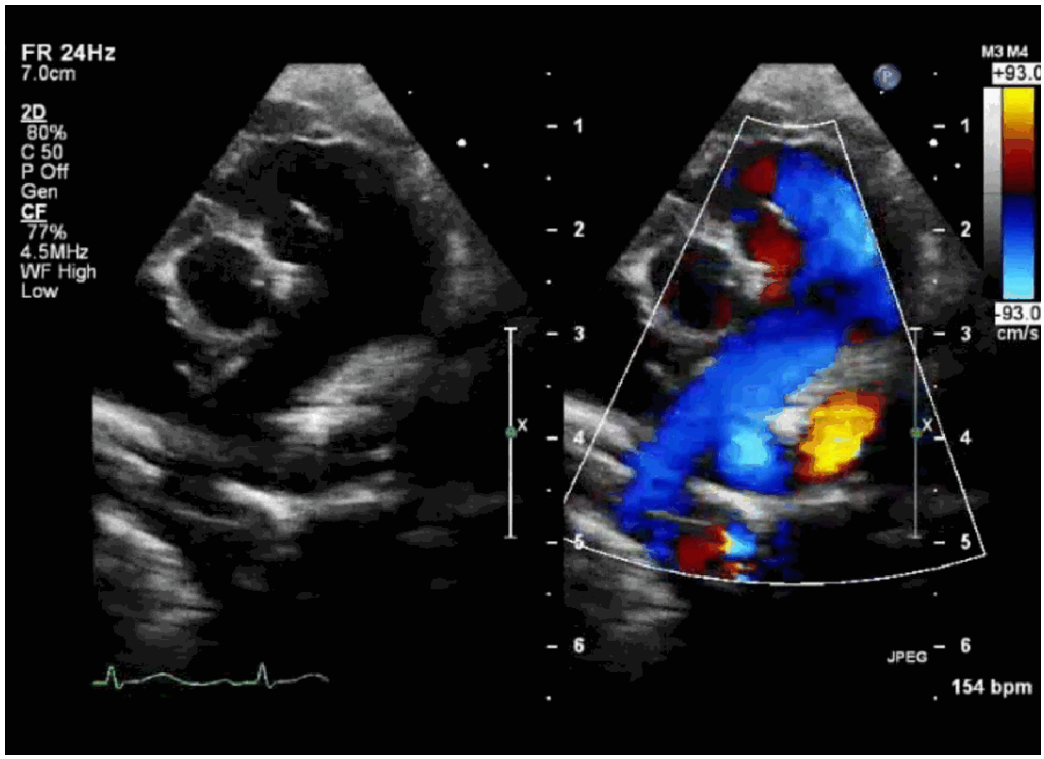
Conventionally:

Red = toward the transducer
Blue = away from the transducer

But:

Red does not mean oxygenated blood.
Blue does not mean deoxygenated blood.

The colors describe direction relative to the transducer.



35. Color Scale

The color scale should be adapted to the expected flow.

LOW-VELOCITY FLOW

Examples:

- pulmonary veins
- systemic veins
- coronary arteries
- some low-velocity shunts

Use a relatively **lower color scale**.

HIGH-VELOCITY FLOW

Examples:

- restrictive VSD
- pulmonary stenosis
- aortic stenosis
- regurgitant jets

A higher scale may be required.

TECHNIQUE TIP

Do not use the same color settings for a coronary artery and a restrictive VSD.

36. Color Box Size

Color Doppler requires additional processing. A large color box reduces frame rate.

Therefore:

Keep the color box as small as necessary to answer the question.
This is especially important in neonates. For coronary imaging, the box may need to include only the aortic root and proximal coronary artery—not the entire heart.

37. Color Gain

Color gain controls amplification of Doppler signals.

TOO LOW

- weak flow disappears
- small shunts may be missed
- coronary flow may not appear

TOO HIGH

- color extends outside vessel boundaries
- noise appears in surrounding tissue
- jets may appear artificially large

A useful practical technique is:

Increase color gain until mild noise appears, then reduce it slightly.

38. The Simplified Bernoulli Equation

One of the most clinically useful equations in echocardiography is:

$\Delta P = 4V^2$

where:

- ΔP = pressure gradient in mmHg
- V = peak velocity in m/s

EXAMPLE

A pulmonary stenosis jet has V = 4 m/s. Therefore:

$\Delta P = 4(4)^2 = 64$ mmHg

39. Why Alignment Matters So Much

Suppose the true velocity is 4 m/s. The calculated gradient is 64 mmHg.

Poor Doppler alignment records only 3 m/s. The calculated gradient becomes 36 mmHg.

A technical acquisition error has therefore produced a major difference in the estimated gradient.

CRITICAL PITFALL

Never grade an obstruction from a poorly aligned Doppler envelope simply because the machine produced a number.

Search for the best-aligned window and obtain the highest clean, reproducible velocity.

40. Physics of Coronary Artery Imaging

Coronary arteries are:

- small
- rapidly moving
- relatively superficial in young children
- associated with relatively low-velocity color flow

Successful imaging therefore often requires:

High frequency + shallow depth + narrow sector + correct focus + small color box + low color scale + optimized gain

This is not a special trick. It is the practical application of ultrasound physics.

41. Physics of Pulmonary Venous Imaging

Pulmonary venous flow may be missed when:

- color scale is too high
- gain is too low
- depth is excessive
- color box is too large
- the imaging plane does not follow the vein

Therefore:

Failure to see color flow is not the same as absence of blood flow.

Optimize the settings and obtain additional views before drawing an anatomical conclusion.

42. Ultrasound Artifacts

Artifacts are image features that do not accurately represent the true anatomy.

Important artifacts include:

- dropout
- reverberation
- acoustic shadowing
- refraction
- mirror-image artifact
- side-lobe artifact
- Doppler aliasing

Recognizing artifacts is particularly important in congenital heart disease because false structures or apparent defects can alter diagnosis.

43. Dropout Artifact

A thin structure may disappear when the beam is unfavorably aligned. This is particularly important in:

- atrial septum
- ventricular septum
- thin valve tissue

Dropout may mimic:

- ASD
- VSD
- tissue discontinuity

CRITICAL PITFALL

Never diagnose a septal defect from dropout in a single plane.

Confirm with:

- another plane
- systematic sweep
- color Doppler
- anatomical consistency

44. Reverberation

Reverberation occurs when sound repeatedly reflects between strong interfaces.

It may produce multiple false echoes that can mimic:

- a membrane
- intracardiac mass
- additional anatomical structure

If an unexpected structure is seen:

Change the transducer position and imaging plane.
True anatomy should remain anatomically consistent.

45. Acoustic Shadowing

A strong reflector may prevent adequate ultrasound transmission to deeper structures. The region behind it appears dark or poorly visualized.

Common causes include:

- prosthetic material
- calcification
- surgical material
- ASD/VSD occluders
- PDA devices

If a device obscures important anatomy:

TECHNIQUE TIP
Change the acoustic window rather than trying to see through the shadow.

46. Physics-Based Troubleshooting

When the image is poor, use a systematic approach rather than changing controls randomly.

- **D — Depth** — Is there unnecessary tissue below the heart?
- **F — Frequency** — Can frequency be increased for better detail, or reduced for penetration?
- **F — Focus** — Is the focal zone at the structure of interest?
- **G — Gain** — Are tissue boundaries adequately displayed?
- **S — Sector** — Can the sector be narrowed?
- **D — Doppler** — Are alignment, scale, gain, sample volume and box size appropriate?

Depth → Frequency → Focus → Gain → Sector → Doppler

47. Common Pediatric Echo Scenarios

Problem	Physics to Consider	Practical Adjustment
Neonatal anatomy looks coarse	Frequency too low	Increase frequency if penetration allows
Far field poorly visualized	Insufficient penetration	Reduce frequency/change window
Neonatal image looks sluggish	Low frame rate	Reduce depth and sector width
Coronary visible in 2D but no color	Scale too high/gain too low	Lower color scale, optimize gain
Pulmonary veins poorly seen	Low-flow sensitivity inadequate	Lower scale, small color box
PS gradient seems unexpectedly low	Poor Doppler alignment	Search for a more parallel window
Apparent ASD in one plane	Dropout	Confirm in subcostal/multiple planes
Device obscures deeper anatomy	Acoustic shadow	Change acoustic window

48. The Major Physics Trade-Offs

Adjustment	Advantage	Trade-Off
Increase frequency	Better spatial resolution	Less penetration
Decrease frequency	Greater penetration	Lower spatial resolution
Reduce depth	Better frame rate	May exclude deeper anatomy
Narrow sector	Better temporal resolution	Smaller field of view
Increase gain	Stronger displayed echoes	More noise
Lower color scale	Better low-flow sensitivity	More aliasing
Increase color scale	Better display of high velocities	Low-velocity flow may disappear
Smaller color box	Better frame rate	Smaller flow region displayed
Correct focus	Better lateral resolution	Requires targeted adjustment
Multiple focal zones	Better detail at several depths	Lower frame rate

49. Ten Physics Rules for Pediatric Echocardiography

Rule 1 — Use the highest frequency that provides adequate penetration.
Rule 2 — Reduce unnecessary depth.
Rule 3 — Narrow the sector when interrogating a specific structure.
Rule 4 — Place the focus near the structure of interest.
Rule 5 — Do not use gain to compensate for inadequate penetration.
Rule 6 — Prioritize frame rate in neonates and tachycardic infants.
Rule 7 — Keep the color box small and match the color scale to expected velocity.
Rule 8 — Align spectral Doppler as parallel to flow as possible.
Rule 9 — Recognize artifacts before diagnosing pathology.
Rule 10 — When an image is inadequate, optimize the physics before declaring the anatomy abnormal or unvisualized.

50. Essential Equations

A pediatric echocardiographer does not need to perform complex calculations during every study, but should understand these relationships:

WAVE EQUATION

$$c = f\lambda$$

ACOUSTIC IMPEDANCE

$$Z = \rho c$$

DOPPLER EQUATION

$$\Delta f = 2f_0 V \cos\theta / c$$

NYQUIST LIMIT

$$\text{Nyquist} = \text{PRF} / 2$$

SIMPLIFIED BERNOULLI EQUATION

$$\Delta P = 4V^2$$

51. Physics-to-Practice Memory Box

REMEMBER: F-D-F-G-S-D

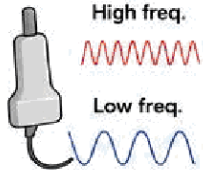
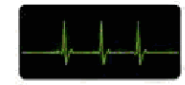
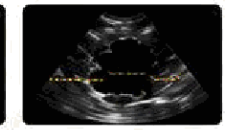
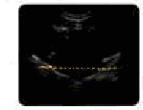
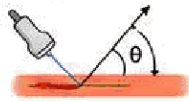
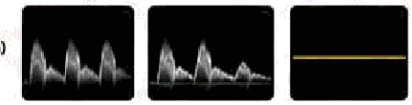
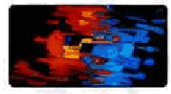
- **F — Frequency** — Highest frequency with adequate penetration.
- **D — Depth** — Make the structure of interest fill the screen.
- **F — Focus** — Place it at the target structure.
- **G — Gain** — Enough to define tissue without excessive noise.
- **S — Sector** — Keep it as narrow as the diagnostic question allows.
- **D — Doppler** — Optimize alignment, scale, gain and sample location.

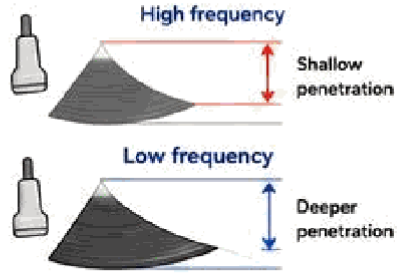
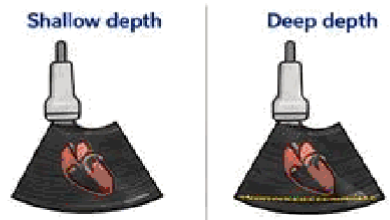
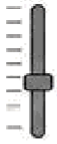
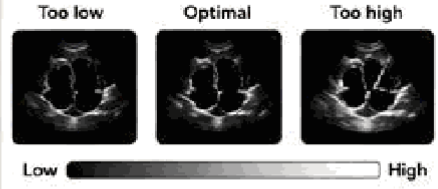
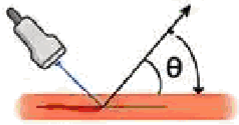
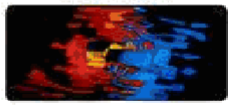
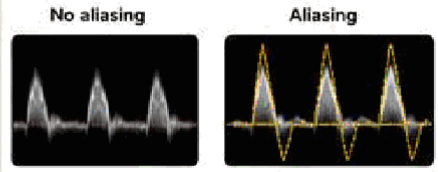
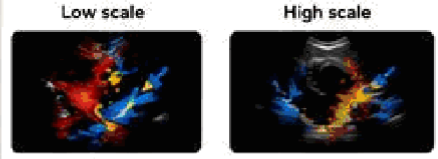
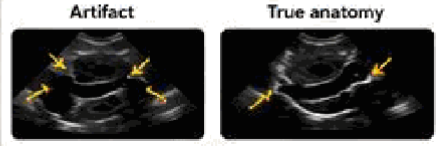
Chapter Key Points

- Ultrasound physics should be learned as a **clinical tool**, not simply as a collection of equations.
- High frequency provides better resolution but less penetration. Low frequency provides greater penetration at the expense of spatial resolution.
- Reducing depth and sector width improves frame rate. Correct focus improves lateral resolution.
- Gain changes displayed signal strength; it does not correct inadequate penetration.
- PW Doppler provides location specificity but is limited by aliasing. CW Doppler measures high velocities but lacks precise range localization.
- Doppler measurements are most accurate when the beam is parallel to flow.
- The Nyquist limit explains aliasing in PW and color Doppler.
- Low-velocity flow such as coronary and pulmonary venous flow requires appropriately sensitive color settings.
- The simplified Bernoulli equation converts measured velocity into an estimated pressure gradient.
- Artifacts such as dropout, reverberation and acoustic shadowing must be recognized before pathology is diagnosed.
- Every machine adjustment involves a trade-off between **resolution, penetration, frame rate, and Doppler sensitivity**.

MASTER PRINCIPLE

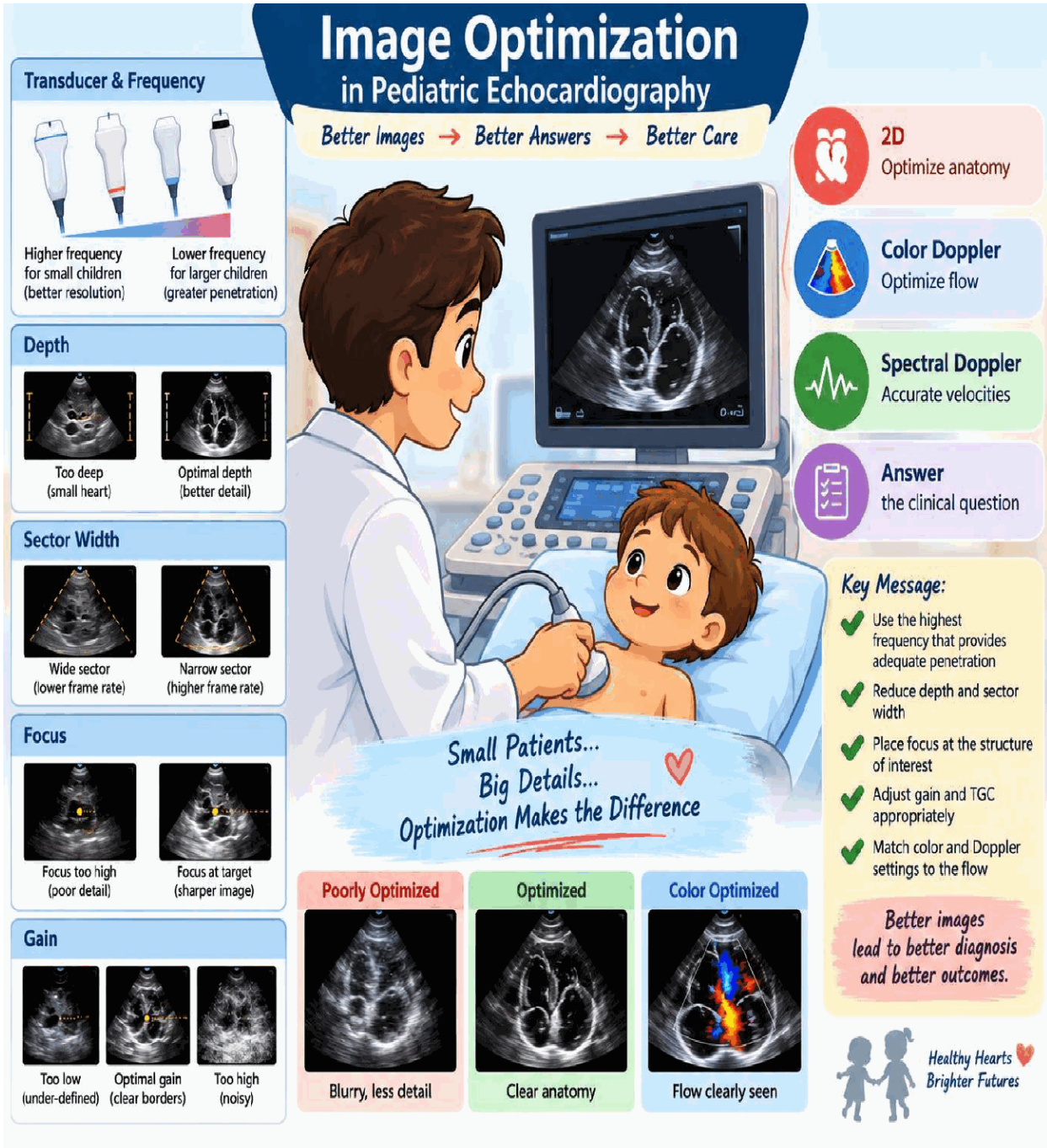
Good echocardiography begins before the diagnosis. Understand the physics → optimize the image → trust the measurement → interpret the physiology.

TOPIC	KEY POINT	PHYSICS EXPLAINED	EFFECT ON IMAGE
Frequency 	• Higher frequency improves resolution but reduces penetration. • Lower frequency does the opposite.	Wavelength (λ) = Speed (c) / Frequency (f) ↑ Frequency → ↓ Wavelength ↑ Frequency → ↑ Resolution ↓ Penetration ↓ Frequency → ↑ Penetration ↓ Resolution	 High frequency Better detail  Low frequency Deeper penetration
Depth 	• Greater depth reduces frame rate. • Use the shallowest depth that includes the heart.	Deeper imaging → longer time for echo return → lower Pulse Repetition Frequency (PRF) → lower frame rate.	 Shallow depth Higher frame rate  Deep depth Lower frame rate
Focus 	• Best lateral resolution is at the focal zone. • Place focus at or just below the structure of interest.	The beam is narrowest at the focus → better ability to distinguish side-by-side structures.	 Focus at target Better detail  Focus too shallow/deep Poor detail
Gain 	• Gain controls displayed brightness. • Too much → noisy. • Too little → poor definition.	Amplifies returning echoes. Does not increase penetration.	 Too low (endocardium not seen)  Optimal  Too high (noisy image)
Doppler Angle 	• Doppler is most accurate when the beam is parallel to flow.	Doppler shift ∝ cos θ θ = 0° → cos θ = 1 (max signal) θ = 60° → cos θ = 0.5 (underestimation) θ = 90° → cos θ = 0 (no signal)	0° Best signal 60° Underestimated 90° No signal 
Nyquist Limit (Aliasing) 	• If velocity exceeds the Nyquist limit, color Doppler or PW Doppler aliases.	Nyquist Limit = PRF / 2 Exceeding this limit → color wrap-around or spectral aliasing.	No aliasing (appropriate scale) Aliasing (scale too low) 
Color Doppler Scale 	• Color scale (velocity range) determines sensitivity to flow.	Low scale → detects low velocity (flow sensitive) but may alias. High scale → shows high velocity but may miss low velocity flow.	Low scale (flow sensitive) High scale (low flow missed) 
Artifacts	• Artifacts can mimic pathology. Recognize and eliminate them before concluding.	Artifacts result from reflection, refraction, scattering or beam-related phenomena.	 Dropout · Reverberation · Acoustic shadow · Mirror image

TOPIC & OPTIMIZATION TIP	CLINICAL EXAMPLE (PEDIATRIC)	VISUAL GUIDE
Frequency  Use the highest frequency that still provides adequate penetration.	• Coronary arteries, AV valves → use high frequency. • Large child or obese patient → use lower frequency for deeper imaging.	
Depth  Reduce depth to just below the area of interest.	Neonate with tachycardia: shallow depth improves visualization of fast cardiac motion.	
Focus  Adjust focus so the target lies at the focal zone.	Pulmonary valve assessment: focus at RVOT/pulmonary valve improves leaflet detail.	
Gain  Increase gain until mild noise appears, then decrease slightly.	Increased gain may create false septal thickening or obscure valve leaflets.	
Doppler Angle 	PS gradient will be underestimated if Doppler beam is not aligned.	
Nyquist Limit (Aliasing) 	Restrictive VSD jet or PS jet may alias if scale is set too low.	
Color Doppler Scale 	Coronary or pulmonary venous flow: use low scale. PS jet: use high scale.	
Artifacts Change imaging plane, transducer position or settings to clarify.	Dropout may mimic ASD/VSD. Reverberation may mimic membranes.	

CHAPTER 3

Image Optimization in Pediatric



Learning Objectives

By the end of this chapter, the learner should be able to:

1. Explain why image optimization is particularly important in neonates, infants, and children.
2. Select an appropriate transducer and imaging frequency for patient size and the structure of interest.
3. Optimize **depth, sector width, focus, gain, TGC, dynamic range, and frame rate**.
4. Optimize color Doppler using appropriate **color-box size, velocity scale, gain, and priority**.
5. Optimize PW and CW Doppler for reliable velocity measurements.

1. Why Image Optimization Matters

Pediatric echocardiography deals with **small structures moving rapidly**.

A coronary artery may measure only a few millimeters. A small VSD can be missed. A pulmonary vein may disappear if the color scale is inappropriate. A neonatal valve can appear thickened because of excessive gain. A stenotic gradient can be underestimated because of poor Doppler alignment.

Therefore, image optimization is not simply about making an image look attractive.

Image optimization is part of the diagnostic process.

A useful principle is:

Do not diagnose from the first image you obtain. Optimize first.

2. The Four Goals of Image Optimization

Every adjustment should serve one or more of four goals:

1. Spatial Resolution

Can I distinguish small adjacent structures?

2. Temporal Resolution

Can I accurately see rapidly moving structures?

3. Penetration

Can ultrasound adequately reach the structure of interest?

4. Flow Sensitivity

Can I detect and accurately measure the blood flow I am looking for?

These goals sometimes compete with each other.

For example:

Higher frequency → better spatial resolution but less penetration
and:

Wider sector → larger field of view but lower frame rate
The skilled echocardiographer understands these trade-offs.

3. Optimize for the Question, Not Just the Patient

There is no single "best" pediatric echo setting.

The optimal setting depends on:

- patient size
- heart rate
- imaging window
- depth of the target
- structure being examined
- expected blood-flow velocity
- clinical question

For example, the settings used to assess a **restrictive VSD** should not be identical to those used to visualize **coronary artery flow**.

PRACTICAL RULE

One patient may require several different optimization strategies during the same examination.

4. The Optimization Sequence

When an image is inadequate, avoid changing multiple controls randomly.

Use a systematic sequence:

- D — Depth
- F — Frequency
- F — Focus
- G — Gain
- S — Sector Width
- F — Frame Rate
- D — Doppler

Or simply:

Depth → Frequency → Focus → Gain → Sector → Frame Rate → Doppler

This sequence can become a routine habit during every pediatric echocardiogram.

5. Transducer Selection

The first optimization decision occurs before the image appears.

Choose the transducer according to:

- patient size
- required penetration
- structure of interest
- acoustic window

Small neonates and infants often permit higher-frequency imaging because their hearts are relatively superficial.

Older or larger children may require lower frequencies for adequate penetration.

GOLDEN RULE

Use the highest frequency that provides adequate penetration to the structure of interest.

6. Frequency Optimization

Frequency determines one of the most important trade-offs in echocardiography.

HIGHER FREQUENCY

Advantages

- shorter wavelength
- better spatial resolution
- better visualization of small structures

Disadvantage

reduced penetration

LOWER FREQUENCY

Advantages

- greater penetration
- improved visualization of deeper structures

Disadvantage

lower spatial resolution

PRACTICAL PEDIATRIC APPLICATIONS

Increase frequency when:

- imaging a premature neonate
- assessing coronary origins
- evaluating valve morphology
- looking for a small VSD
- defining septal margins
- examining a superficial structure

Reduce frequency when:

- imaging a larger child
- the far field is poorly visualized
- the LA or posterior structures are inadequately penetrated
- the aortic arch is difficult to visualize because of depth

CLINICAL PEARL

If the image is bright but the deeper anatomy remains poorly defined, more gain may not solve the problem. Consider lowering the frequency.

7. Depth

Depth determines how much tissue is displayed vertically.

One of the most common beginner errors is using excessive depth.

EXCESSIVE DEPTH

Results in:

- the heart appearing too small
- unnecessary tissue below the heart
- reduced frame rate
- poorer visualization of small structures

APPROPRIATE DEPTH

The structure of interest should occupy a substantial proportion of the display while all necessary anatomy remains visible.

PRACTICAL RULE

Use the shallowest depth that completely includes the anatomy required for the diagnostic question.

8. Why Reducing Depth Improves Frame Rate

The ultrasound machine must wait for returning echoes before sending subsequent pulses.

Greater imaging depth requires more travel time.

Therefore:

Depth ↑ → Pulse travel time ↑ → Frame rate ↓

This becomes especially important in neonates with rapid heart rates.

EXAMPLE

A neonatal parasternal view includes several centimeters of unnecessary tissue below the LV.

Reducing depth will:

- enlarge the heart on the display
- improve temporal resolution
- make valve and septal motion easier to assess

9. Sector Width

The sector determines the width of the field of view.

WIDE SECTOR

Useful for:

- initial orientation
- showing relationships between structures

- complex congenital anatomy

But:

Wide sector → more scan lines → lower frame rate

NARROW SECTOR

Useful for:

- coronary origins
- individual valves
- ventricular function
- VSD interrogation
- RVOT
- LVOT

and improves temporal resolution.

PRACTICAL STRATEGY

Survey wide → Interrogate narrow.

First understand the anatomy.

Then narrow the sector around the structure of interest.

10. Focus

Lateral resolution is best where the ultrasound beam is narrowest—near the focal zone.

Therefore:

Place the focus at or just below the structure of interest.

Examples:

Structure	Suggested Focus
Coronary origin	Aortic root
Atrial septum	IAS
Ventricular septum	IVS
Pulmonary valve	RVOT/PV
Mitral valve	MV leaflet level
PDA	Ductal region
Aortic arch	Arch/isthmus

Multiple focal zones may reduce frame rate.

NEONATAL PEARL

For a rapidly beating neonatal heart, one appropriately positioned focus is often preferable to multiple unnecessary focal zones.

11. Overall Gain

Gain amplifies returning signals and changes the brightness of the displayed image.

It does **not** increase ultrasound penetration.

GAIN TOO HIGH

May cause:

- excessive noise
- loss of blood-tissue distinction
- apparently thickened valves
- blurred endocardial borders
- false appearance of septal thickening

GAIN TOO LOW

May cause:

- poor endocardial definition
- disappearance of thin valve tissue
- apparent septal dropout
- failure to visualize subtle anatomy

PRACTICAL TECHNIQUE

Increase gain until the tissue is clearly visible without filling the blood pool with excessive noise.

12. Time-Gain Compensation

Ultrasound becomes progressively attenuated as it travels deeper.

TGC allows different amounts of gain to be applied at different depths.

For example:

Near field = satisfactory
but

Far field = too dark
Instead of increasing overall gain:

Increase the deeper TGC selectively.
This preserves near-field image quality while improving deeper visualization.

13. Dynamic Range / Compression

Dynamic range controls the range of gray shades displayed.

WIDER DYNAMIC RANGE

- more shades of gray
- softer appearance
- lower contrast

NARROWER DYNAMIC RANGE

- fewer shades
- greater contrast
- sharper black-and-white appearance

Neither extreme is universally correct.

The aim is to clearly distinguish:

- blood pool
- endocardium
- myocardium
- valves
- septa
- vessel walls

Optimize for tissue discrimination, not cosmetic appearance.

14. Frame Rate — Critical in Children

Temporal resolution is especially important in pediatric echocardiography.

Neonates and infants have:

- small hearts
- rapid valve motion
- high heart rates

A low frame rate can make cardiac motion appear blurred.

Frame rate generally improves by:

- reducing depth
- narrowing sector width
- reducing unnecessary focal zones
- reducing color-box size

PEDIATRIC RULE

Small heart + fast heart rate = prioritize frame rate.

15. Zoom

Zoom is useful when assessing small structures such as:

- coronary origins
- aortic valve
- pulmonary valve
- ASD rims
- small VSDs
- PDA
- branch pulmonary arteries

But remember:

Magnification does not create information that was never acquired.

First obtain the best possible original image using appropriate:

- frequency
- depth
- focus
- sector width

Then zoom.

16. Optimize 2D Before Adding Color

A common error is turning on color Doppler before understanding the anatomy.

This can:

- obscure small structures
- lower frame rate
- distract from morphology
- encourage diagnosis based on turbulence alone

The preferred sequence is:

2D → Color → PW/CW → Measurement → Interpretation

First identify the structure.

Then ask a specific flow question.

17. Color Doppler Optimization

Color Doppler is particularly dependent on correct settings.

The major controls are:

1. Color-box size
2. Velocity scale/PRF
3. Color gain
4. Position and steering
5. Wall filter
6. Color priority where available

The settings should be adapted to the expected velocity.

18. Color-Box Size

The color box should cover only the region needed to answer the question.

LARGE COLOR BOX

- more anatomy displayed
- greater processing demand
- lower frame rate

SMALL COLOR BOX

- higher frame rate
- more focused assessment

EXAMPLE

For proximal coronary artery flow:

Do not place color over the entire heart.
Use a small box around:

- aortic root
- coronary ostium
- proximal coronary course

19. Color Velocity Scale

Color scale determines the range of velocities displayed.

The correct setting depends on the expected flow.

LOWER SCALE

Useful for low-velocity flow:

- pulmonary veins
- systemic veins
- coronary arteries
- low-velocity shunts

HIGHER SCALE

Useful for high-velocity flow:

- restrictive VSD
- pulmonary stenosis
- aortic stenosis
- high-velocity regurgitation

GOLDEN RULE

Match the color scale to the expected velocity—not to a fixed machine preset.

20. Color Gain

Color gain determines the amplification of Doppler signals.

TOO LITTLE COLOR GAIN

May cause:

- pulmonary venous flow to disappear
- coronary flow to disappear
- small shunts to be missed

EXCESSIVE COLOR GAIN

May cause:

- color outside vessel boundaries
- excessive noise
- apparent enlargement of jets
- misleading assessment of regurgitation

PRACTICAL TECHNIQUE

Increase color gain until mild background noise appears, then reduce slightly.

21. Wall Filter

Wall filters suppress low-frequency Doppler signals generated by tissue motion.

High wall-filter settings can remove genuine low-velocity blood-flow signals.

This matters when assessing:

- venous flow
- coronary arteries
- slow shunts

For low-flow imaging:

Use an appropriately low wall filter when the machine permits.

22. Color Priority

Color priority determines how the system chooses between displaying gray-scale tissue and color information.

Excessive priority may cause color to overwrite tissue boundaries.

Too little may make low-flow signals difficult to see.

Use it carefully when evaluating small vessels.

23. Spectral Doppler Optimization

A technically poor Doppler tracing can produce a clinically misleading measurement.

Before measuring a velocity, optimize:

- imaging window
- beam alignment
- sample location
- velocity scale
- baseline
- sweep speed
- gain
- sample volume for PW Doppler

24. Doppler Alignment

This is one of the most important principles in echocardiography.

The Doppler equation contains: $\cos\theta$

Measured velocity is most accurate when the beam is parallel to flow.

BEST ALIGNMENT

$\theta \approx 0°$

As the angle increases, velocity is underestimated.

CLINICAL CONSEQUENCE

Poor alignment can underestimate:

- pulmonary stenosis
- aortic stenosis
- coarctation
- VSD gradient
- TR velocity
- RVOT/LVOT obstruction

GOLDEN RULE

Search multiple windows and use the highest clean, reproducible velocity obtained with the best alignment.

25. PW Doppler Optimization

PW Doppler is useful when the location of velocity matters.

Optimize by:

- placing the sample volume precisely
- adjusting sample-volume size
- aligning with flow
- adjusting scale
- increasing sweep speed when detailed waveform analysis is required

Useful for:

- AV valve inflow
- pulmonary veins
- systemic veins
- LVOT
- RVOT
- branch PAs
- descending/abdominal aorta

26. CW Doppler Optimization

CW Doppler is used for high-velocity jets.

Typical applications include:

- pulmonary stenosis
- aortic stenosis
- restrictive VSD
- coarctation
- high-velocity regurgitation

The key issue is not simply placing the cursor across the lesion.

It is obtaining the most parallel alignment possible.

CRITICAL PITFALL

A beautifully traced Doppler envelope can still be wrong if the beam is poorly aligned.

27. Spectral Doppler Gain

Excessive spectral gain produces:

- thick envelopes
- spectral noise
- difficulty identifying the true edge of the waveform

Too little gain may produce:

- incomplete envelopes
- loss of weak signals

Adjust gain until the waveform is clearly defined without excessive background noise.

28. Sweep Speed

Sweep speed determines how much time is displayed horizontally.

A faster sweep speed spreads waveforms apart.

This can improve assessment of:

- valve inflow patterns
- pulmonary venous flow
- tissue Doppler
- timing relationships
- detailed spectral envelopes

The sweep speed should therefore be adapted to the measurement being performed.

29. Optimization for Coronary Arteries

Coronary imaging is an excellent example of physics translated into practice.

Coronary arteries are:

- small
- mobile
- superficial in many children
- associated with relatively low-velocity flow

OPTIMIZE 2D

Use:

- high-frequency transducer
- shallow depth
- zoom
- narrow sector
- focus at aortic root
- appropriate gain
- high frame rate

OPTIMIZE COLOR

Use:

- small color box
- relatively low velocity scale
- appropriate color gain
- low wall filter where available

CORONARY PEARL

Find the coronary origin in high-quality 2D first; use color to confirm flow and course.

30. Optimization for Pulmonary Veins

Pulmonary venous flow may be relatively low velocity and easily lost with inappropriate settings.

Use:

- optimized depth
- appropriate view
- small color box
- lower color scale
- adequate color gain
- low wall filter where appropriate

CRITICAL PITFALL

Failure to display a pulmonary vein with color does not prove that the vein is absent or anomalously connected.

Change the view and optimize before drawing a conclusion.

31. Optimization for ASD

The atrial septum is thin and vulnerable to dropout.

2D OPTIMIZATION

- use the subcostal window whenever feasible
- optimize depth
- focus at IAS
- avoid excessive gain
- sweep through the entire septum

COLOR OPTIMIZATION

- reduce color-box size
- use an appropriate scale for atrial-level flow
- adjust gain
- confirm shunt direction

CRITICAL PITFALL

Never diagnose ASD from apparent septal dropout in a single view.

32. Optimization for VSD

VSD evaluation requires both anatomy and hemodynamics.

2D

- narrow sector around septum
- optimize depth and focus
- use multiple planes
- sweep from base to apex

COLOR

Use color to identify:

- location
- direction
- additional defects
- associated flow abnormalities

SPECTRAL DOPPLER

For restrictive defects, align CW Doppler as parallel to the jet as possible.

The VSD jet direction determines the best Doppler window.

33. Optimization for PDA

The high left parasternal and suprasternal/ductal views can be particularly useful depending on age and anatomy.

Optimize by:

- reducing depth
- focusing on the ductal region
- narrowing the sector
- using a small color box
- adjusting the color scale according to ductal flow velocity

Assess:

- ductal anatomy
- direction of flow
- restriction
- pulmonary artery relationship
- descending aorta

34. Optimization for Coarctation

For suspected coarctation:

2D

Optimize the suprasternal long-axis view to visualize:

- transverse arch
- isthmus
- descending aorta

Use:

- appropriate depth
- narrow sector when focusing on the isthmus
- high frequency when penetration allows

COLOR

Look for:

- focal acceleration
- turbulence
- narrowing

SPECTRAL DOPPLER

Obtain:

- descending aortic Doppler
- abdominal aortic Doppler where useful

Look beyond peak velocity for:

- diastolic continuation
- waveform abnormality
- physiological context

35. Optimization for Pulmonary Stenosis

2D

Optimize:

- RVOT
- pulmonary valve
- annulus
- main pulmonary artery
- branch pulmonary arteries

COLOR

Use an appropriate scale to demonstrate the site of acceleration.

CW DOPPLER

Search for the most parallel beam.

Do not assume one window provides the true maximum velocity.

The highest clean, reproducible, well-aligned velocity is generally the most useful.

36. Optimization for Pericardial Effusion

When evaluating suspected effusion:

- use sufficient depth to include the pericardium
- avoid excessive gain
- assess multiple views
- distinguish fluid from artifact
- examine chamber interaction
- use M-mode/Doppler where clinically relevant

Do not focus only on the amount of fluid.

The important question is:

Is the effusion affecting cardiac filling and hemodynamics?

37. Lesion-Specific Optimization Table

Target	2D Strategy	Color/Doppler Strategy
Coronary arteries	High frequency, shallow depth, zoom, focus at root	Small box, low scale, optimized gain
Pulmonary veins	Targeted venous views	Lower scale, adequate gain, low wall filter
ASD	Subcostal, focus on IAS, multiplane sweep	Small box, appropriate low-flow scale
VSD	Septal sweeps, narrow sector	Color localization + aligned CW
PDA	High left parasternal/ductal view	Small box, scale matched to flow
Coarctation	Optimized suprasternal arch view	Color acceleration + spectral Doppler
Pulmonary stenosis	RVOT/PV optimization	Color localization + best-aligned CW
Neonatal function	Shallow depth, narrow sector	Prioritize high frame rate
Pericardial effusion	Multiple views, adequate depth	Doppler for physiological effect

38. Common Optimization Errors

ERROR 1 — TOO MUCH DEPTH

Result: Small heart, reduced frame rate.

Correction: Reduce depth.

ERROR 2 — SECTOR TOO WIDE

Result: Reduced temporal resolution.

Correction: Narrow the sector after orientation.

ERROR 3 — TOO MANY FOCAL ZONES

Result: Lower frame rate.

Correction: Use one appropriately placed focus when possible.

ERROR 4 — EXCESSIVE GAIN

Result: Noisy chambers and apparently thick structures.

Correction: Reduce gain.

ERROR 5 — LOW GAIN

Result: Poor endocardial definition and possible dropout.

Correction: Increase gain appropriately.

ERROR 6 — LARGE COLOR BOX

Result: Lower frame rate.

Correction: Restrict color to the area of interest.

ERROR 7 — COLOR SCALE TOO HIGH

Result: Low-velocity coronary or venous flow may disappear.

Correction: Lower the scale appropriately.

ERROR 8 — COLOR SCALE TOO LOW

Result: Excessive aliasing in high-velocity jets.

Correction: Increase the scale according to the diagnostic question.

ERROR 9 — POOR DOPPLER ALIGNMENT

Result: Underestimated velocity and gradient.

Correction: Change the window and seek more parallel alignment.

39. The "Before and After" Optimization Method

When teaching pediatric echo, compare images in pairs:

BEFORE OPTIMIZATION

- excessive depth
- wide sector
- inappropriate frequency
- incorrect focus
- poor gain

AFTER OPTIMIZATION

- shallow appropriate depth
- targeted sector
- appropriate frequency
- correct focus
- balanced gain

Ask the trainee:

What changed—and why did the diagnostic information improve?
This develops optimization skills faster than memorizing machine controls.

40. The 10-Second Image Optimization Check

Before recording a clip, ask:

1. Is the depth appropriate?
2. Is this the best frequency?
3. Is the focus at the target?
4. Is the gain appropriate?
5. Can I narrow the sector?

6. Is the frame rate adequate?

Before recording Doppler, additionally ask:

7. Is the color box small enough?
8. Is the velocity scale appropriate?
9. Is the gain appropriate?
10. Is my Doppler beam aligned with flow?

41. Image Optimization Sheet

Control	If Increased	If Decreased	Pediatric Use
Frequency	Resolution ↑, penetration ↓	Penetration ↑, resolution ↓	Use highest frequency with adequate penetration
Depth	Field/depth ↑, frame rate ↓	Frame rate ↑	Keep shallow when possible
Sector width	Field ↑, frame rate ↓	Frame rate ↑	Narrow for targeted imaging
Focal zones	More depths optimized, frame rate ↓	Frame rate ↑	One target focus often ideal
2D gain	Brighter/noisier	Darker	Optimize tissue boundaries
Color-box size	Coverage ↑, frame rate ↓	Frame rate ↑	Keep small
Color scale	High velocities displayed	Low-flow sensitivity ↑	Match expected flow
Color gain	Sensitivity/noise ↑	Weak flow may disappear	Increase then reduce slightly
Spectral gain	Envelope/noise ↑	Weak signals disappear	Define clean envelope

42. Key Clinical Pearls

Pearl 1: Optimize for the structure—not merely for the patient.

Pearl 2: High frequency is valuable only if adequate penetration remains.

Pearl 3: Reducing depth is one of the simplest ways to improve a neonatal image.

Pearl 4: A narrow sector and small color box are powerful tools for improving frame rate.

Pearl 5: Gain improves display brightness, not penetration.

Pearl 6: Low-flow structures require low-flow Doppler settings.

Pearl 7: Doppler velocity is only as reliable as beam alignment.

Pearl 8: Never accept an apparent anatomical defect until technical artifact has been excluded.

Pearl 9: A technically beautiful image is useful only if it answers the clinical question.

Pearl 10: Optimization should become automatic before image acquisition.

Chapter Key Points

- Pediatric image optimization requires continuous adjustment because patient size, heart rate, imaging depth, and diagnostic targets vary widely.
- Use the **highest-frequency transducer that provides adequate penetration**.
- Reduce unnecessary depth and sector width to improve temporal resolution.
- Position the focal zone at or just below the structure of interest.
- Adjust gain to optimize tissue boundaries; do not use gain as a substitute for penetration.
- Prioritize frame rate in neonates and tachycardic infants.
- Optimize 2D anatomy before adding color Doppler.
- Keep the color box small and match the velocity scale to expected flow.
- Low-velocity structures such as coronary arteries and pulmonary veins require appropriately sensitive color settings.
- PW Doppler provides localization; CW Doppler is appropriate for high-velocity interrogation.
- Doppler alignment is critical for reliable velocity and gradient estimation.
- Different lesions require different optimization strategies.
- When anatomy appears abnormal, first ask whether the image has been technically optimized.

MASTER OPTIMIZATION RULE

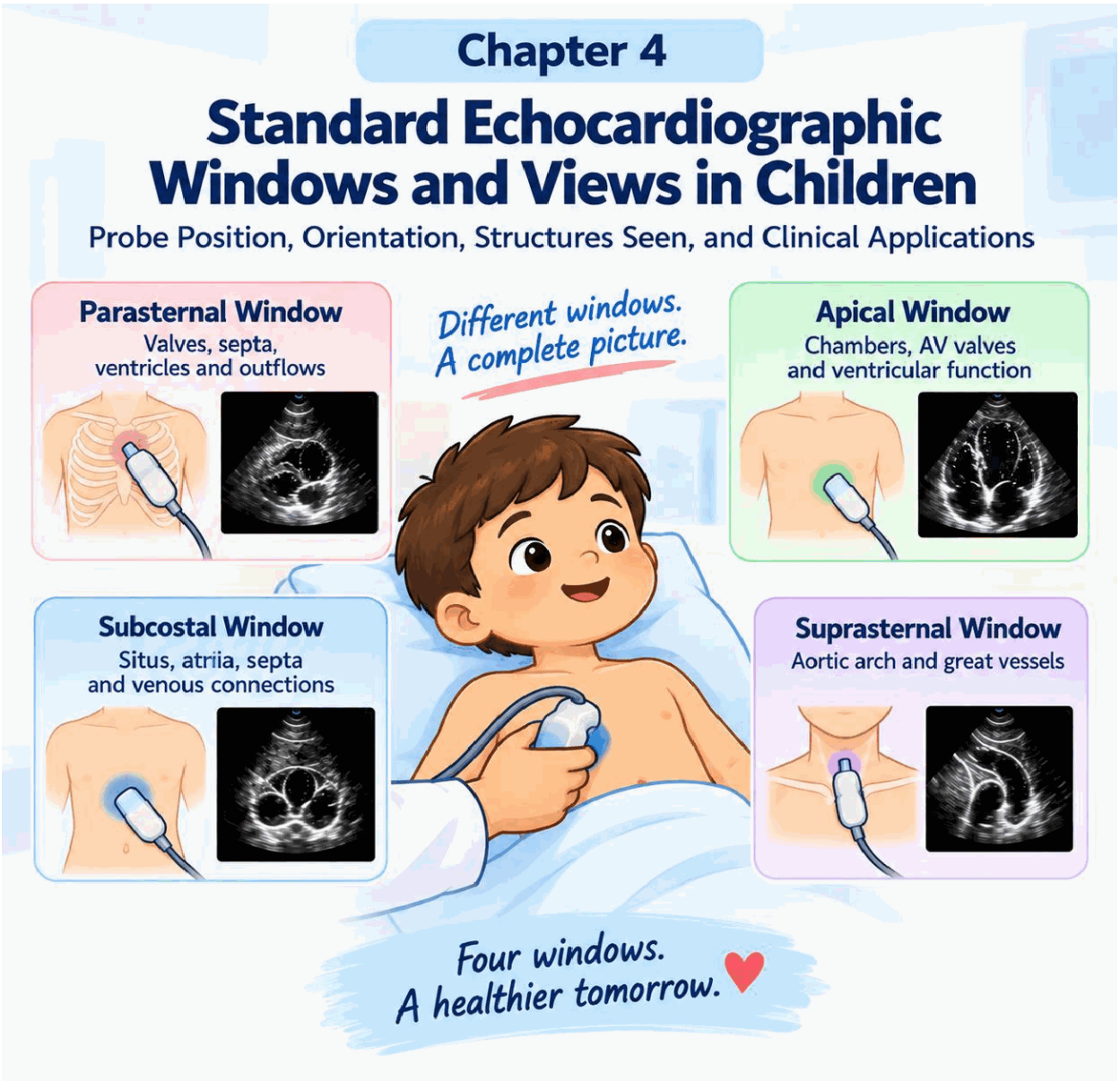
Do not accept the first image. Adjust → Improve → Confirm → Record → Interpret.

The Pediatric Echo Optimization Formula

Best frequency + Minimum necessary depth + Targeted focus + Balanced gain + Narrow sector + Appropriate Doppler = Better diagnostic information.

CHAPTER 4

Standard Echocardiographic Views in Children



Learning Objectives

By the end of this chapter, the learner should be able to:

1. Identify the major transthoracic echocardiographic windows used in children.
2. Position and orient the transducer appropriately for each standard view.
3. Recognize the principal cardiac structures demonstrated in each imaging plane.
4. Perform systematic sweeps rather than relying on isolated images.
5. Select the best view for evaluating specific structures and congenital lesions.
6. Use modified views when standard planes do not adequately demonstrate the anatomy.
7. Integrate 2D, color Doppler, PW Doppler, and CW Doppler appropriately within each window.
8. Recognize common technical pitfalls associated with individual views.
9. Perform a reproducible window-to-window pediatric echocardiographic examination.
10. Understand that congenital anatomy must be reconstructed using multiple complementary views.

1. Why Standard Views Matter

A pediatric echocardiogram should not be a random collection of attractive images.

The standard echocardiographic windows provide a systematic method of examining the heart from different directions. Each window reveals structures that may be poorly seen or misleading from another.

The four principal transthoracic windows are:

1. Subcostal
2. Parasternal
3. Apical
4. Suprasternal

Additional modified views include:

- high left parasternal
- right parasternal
- modified subcostal
- modified apical
- right-sided views in dextrocardia

The central principle is:

One heart → Multiple windows → Multiple planes → One integrated diagnosis

2. Window Versus View

These terms should not be confused.

ACOUSTIC WINDOW

The location on the patient's body from which ultrasound enters.

Examples:

- subcostal
- parasternal
- apical
- suprasternal

ECHOCARDIOGRAPHIC VIEW

The particular imaging plane obtained from that window.

For example, from the **parasternal window**, one may obtain:

- parasternal long-axis view
- parasternal short-axis view
- RV inflow view
- RV outflow view

Therefore:

Window = Where the probe is placed.
View = What imaging plane is produced.

3. The Basic Transducer Movements

A skilled pediatric echocardiographer does not simply place the transducer and hold it still.

Five movements are essential.

Slide

Move the entire probe to another position on the chest.

Rotate

Turn the transducer clockwise or counterclockwise.

Tilt

Move the imaging plane from side to side while maintaining the probe position.

Angle

Direct the beam superiorly, inferiorly, medially, or laterally.

Sweep

Move continuously through adjacent anatomical planes.

ECHO PEARL

Congenital heart disease is three-dimensional. Sweeps often provide more information than perfect still frames.

4. Orientation Before Imaging

Before interpreting any view:

- know where the probe is located
- know where the transducer marker is directed
- understand the laboratory's display convention
- identify structures by morphology

Screen orientation conventions can vary between laboratories and equipment.

Therefore:

Do not identify anatomy simply by where it appears on the monitor.

This is particularly important in:

- dextrocardia
- heterotaxy
- ventricular inversion
- congenitally corrected TGA
- complex single-ventricle anatomy

5. A Practical Scanning Sequence

A useful pediatric sequence is:

Subcostal → Apical → Parasternal → Suprasternal

Some laboratories begin parasternal and use another sequence. The precise order is less important than being **consistent and complete**.

For congenital heart disease, the subcostal window is an excellent starting point because it can establish:

- abdominal situs
- systemic venous anatomy
- atrial arrangement
- AV connections

before assumptions are made about the remainder of the heart.

PART I — SUBCOSTAL WINDOW

6. Why the Subcostal Window Is Important in Children

The subcostal window is particularly valuable in neonates and young children because the liver provides an excellent acoustic window.

It can demonstrate:

- abdominal situs, IVC and aorta, systemic venous connections
- atrial septum, AV connections, ventricular morphology, ventricular septum
- outflow tracts, pulmonary veins in selected planes

PEDIATRIC ECHO PEARL

In an infant with suspected congenital heart disease, never underestimate the subcostal window.

7. Subcostal Abdominal Situs View

PROBE POSITION

Place the transducer just inferior to the xiphisternum and obtain a transverse section through the upper abdomen.

STRUCTURES TO IDENTIFY

- liver, spine, stomach
- inferior vena cava, descending aorta

The first purpose is not to diagnose the intracardiac lesion.

It is to answer: **What is the situs?**

Determine the relationship of:

IVC ↔ Aorta ↔ Spine

Then follow the IVC superiorly toward the atrium.

CLINICAL APPLICATIONS

- situs solitus, situs inversus, heterotaxy
- interrupted IVC, azygos continuation, abnormal systemic venous return



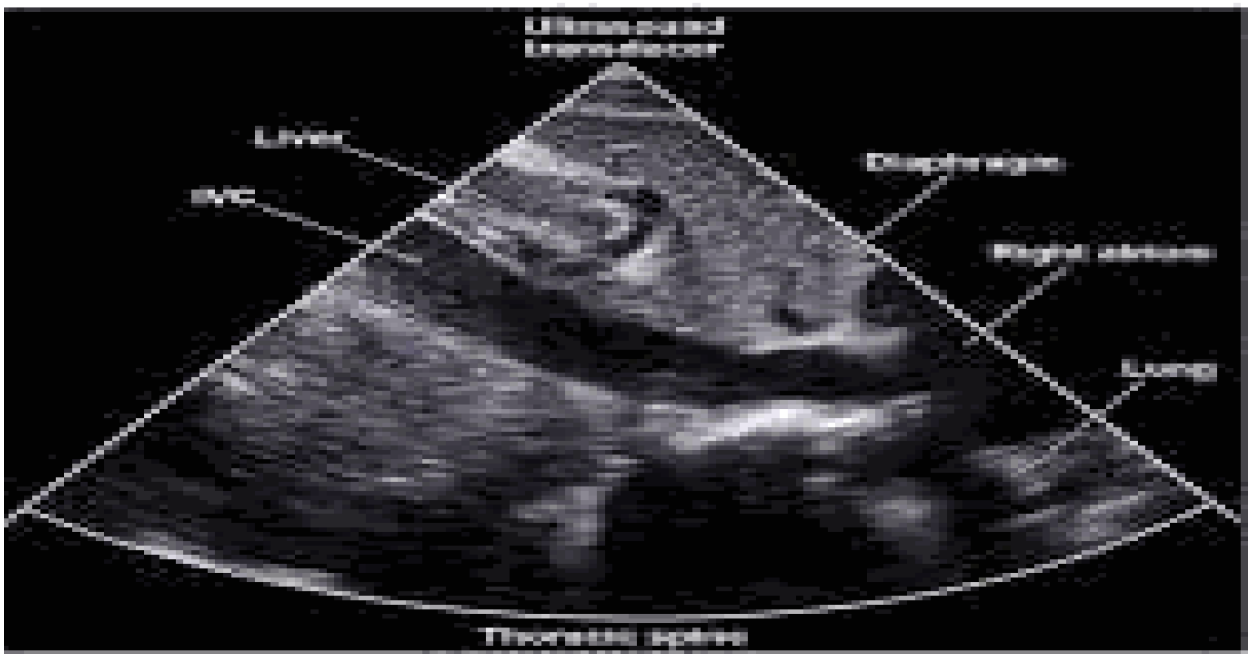
8. Subcostal View of the Atrial Septum

The subcostal window is usually one of the most useful transthoracic approaches for evaluating the atrial septum.

Because the ultrasound beam can often intersect the atrial septum more favorably than in an apical view.

ASSESS

- PFO, secundum ASD
- primum region
- shunt direction with color Doppler

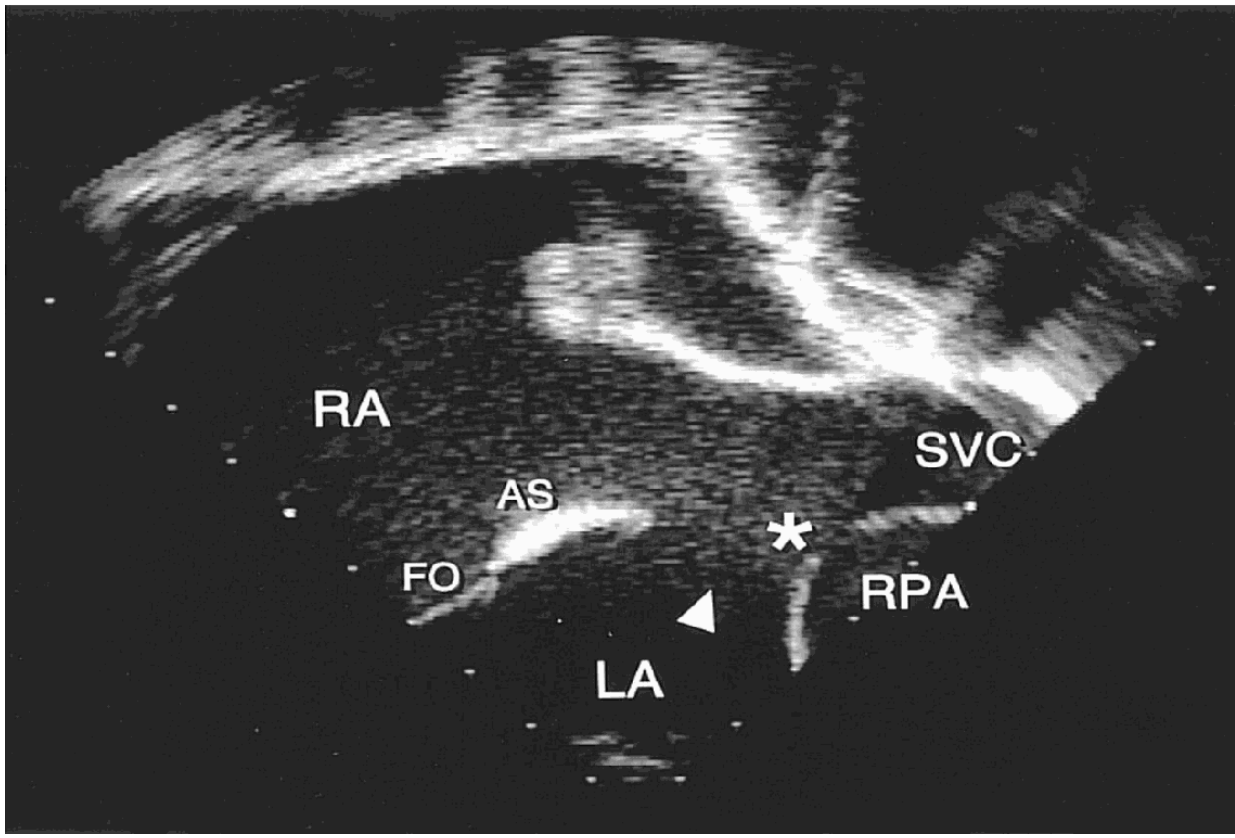


9. Subcostal Bicaval View

Rotate and angle the transducer to display both caval veins entering the right atrium.

Particularly useful for sinus venosus ASD.

When evaluating a suspected sinus venosus defect, do not examine the atrial septum without also examining the pulmonary veins.



10. Subcostal Four-Chamber View

From the subxiphoid position, direct the ultrasound beam superiorly through the liver toward the heart.

STRUCTURES SEEN

- RA, LA, RV, LV, tricuspid valve, mitral valve
- atrial septum, ventricular septum, cardiac crux

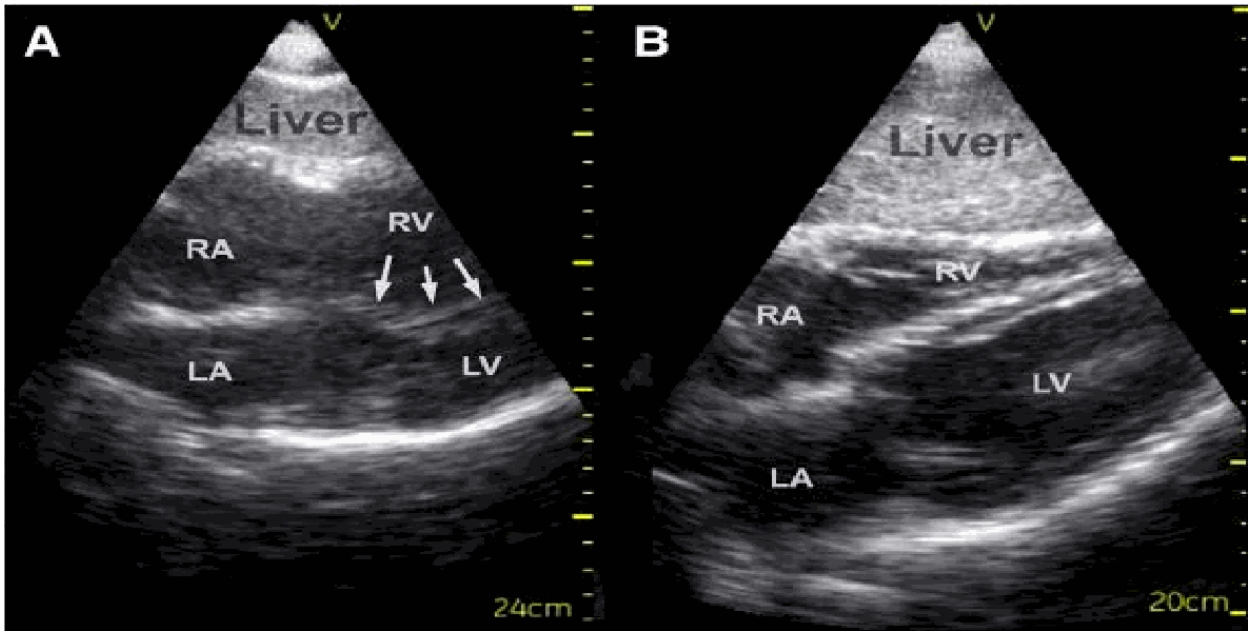
11. Subcostal Outflow Views

Anterior angulation from the subcostal four-chamber plane may demonstrate:

- RVOT, pulmonary valve, main PA
- LVOT, aortic root

These views can be particularly useful in:

- TOF, pulmonary atresia
- critical pulmonary stenosis
- TGA
- DORV
- neonatal outflow obstruction



12. Subcostal Short-Axis Sweep

The probe can be rotated to obtain short-axis imaging and then swept through the heart.

A systematic sweep may demonstrate:

Systemic veins → Atria → Atrial septum → AV valves → Ventricles → Outflow tracts

This can be extremely useful in complex congenital anatomy.

- **PART II — PARASTERNAL WINDOW**

13. Parasternal Long-Axis View

PROBE POSITION

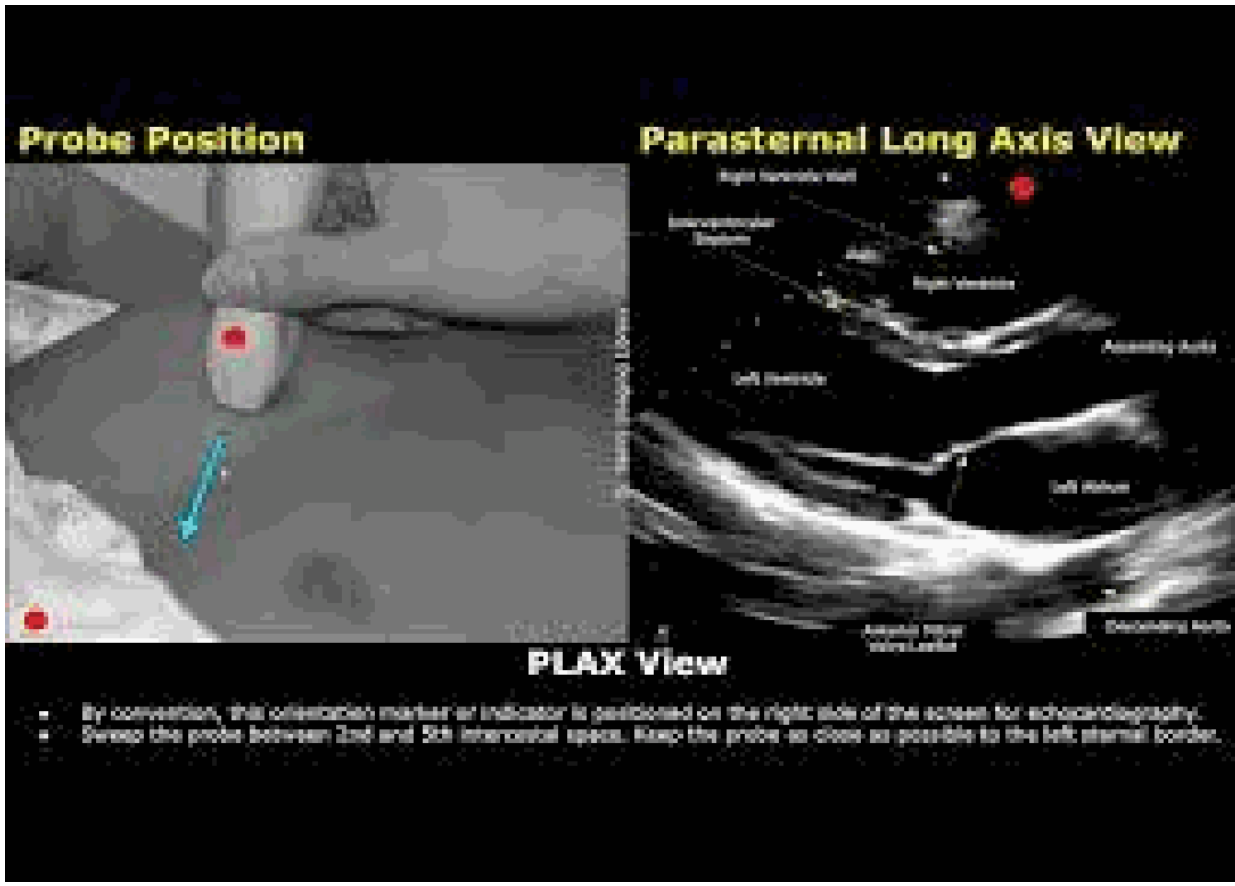
Place the transducer at the left parasternal border, generally around the third to fourth intercostal space with notch towards right shoulder.

Align the imaging plane with the long axis of the LV.

STRUCTURES SEEN

From near field to far field:

- RV, interventricular septum, LV
- LVOT, aortic valve, aortic root
- LA, mitral valve
- descending thoracic aorta in the far field in many patients



14. What to Assess in PLAX

The parasternal long-axis view provides a large amount of information.

Evaluate:

Left Ventricle

- dimensions
- wall thickness
- systolic function

Mitral Valve

- leaflet morphology
- motion
- prolapse
- regurgitation

LVOT

- subaortic region
- obstruction
- membrane

Aortic Valve

- leaflet opening
- thickening
- doming
- regurgitation

Aortic Root

- dimensions
- dilation

Ventricular Septum

- peri-membranous VSD, associated aneurysmal tissue

15. PLAX Clinical Applications

High-yield lesions include:

- VSD, mitral valve disease
- aortic stenosis, subaortic membrane
- aortic regurgitation, aortic root dilation
- hypertrophic cardiomyopathy
- LV dysfunction, pericardial effusion

ECHO PEARL

A good PLAX view should elongate the LV rather than foreshorten it.

16. Parasternal RV Inflow View

From PLAX, angle the transducer toward the right ventricular inflow.

STRUCTURES SEEN

- RA, tricuspid valve
- RV inflow, right ventricular wall

17. Parasternal RV Outflow View

Angle superiorly toward the RVOT.

STRUCTURES SEEN

- RVOT, pulmonary valve, main pulmonary artery
- PDA

Depending on angulation, branch PAs may also be demonstrated.

CLINICAL USES

- TOF, pulmonary stenosis
- RVOT obstruction
- postoperative RVOT assessment
- pulmonary regurgitation
- PDA

18. Parasternal Short-Axis View

From PLAX, rotate the transducer approximately 90° turning the notch towards left shoulder and sweep from the cardiac base toward the apex.

This is one of the most information-rich views in pediatric echocardiography.

The PSAX examination should be thought of as a **continuous sweep**, not one image.

Important levels include:

1. Great artery/aortic valve level
2. Mitral valve level
3. Papillary muscle level
4. Apical ventricular level

19. PSAX — Aortic Valve Level

At the cardiac base, the aortic valve is seen centrally.

STRUCTURES SEEN

- aortic valve, coronary origins where visualized
- RA, tricuspid valve
- RVOT, pulmonary valve
- main PA, branch pulmonary arteries
- PDA

The short-axis view is particularly important for determining:

- tricuspid aortic valve, bicuspid aortic valve

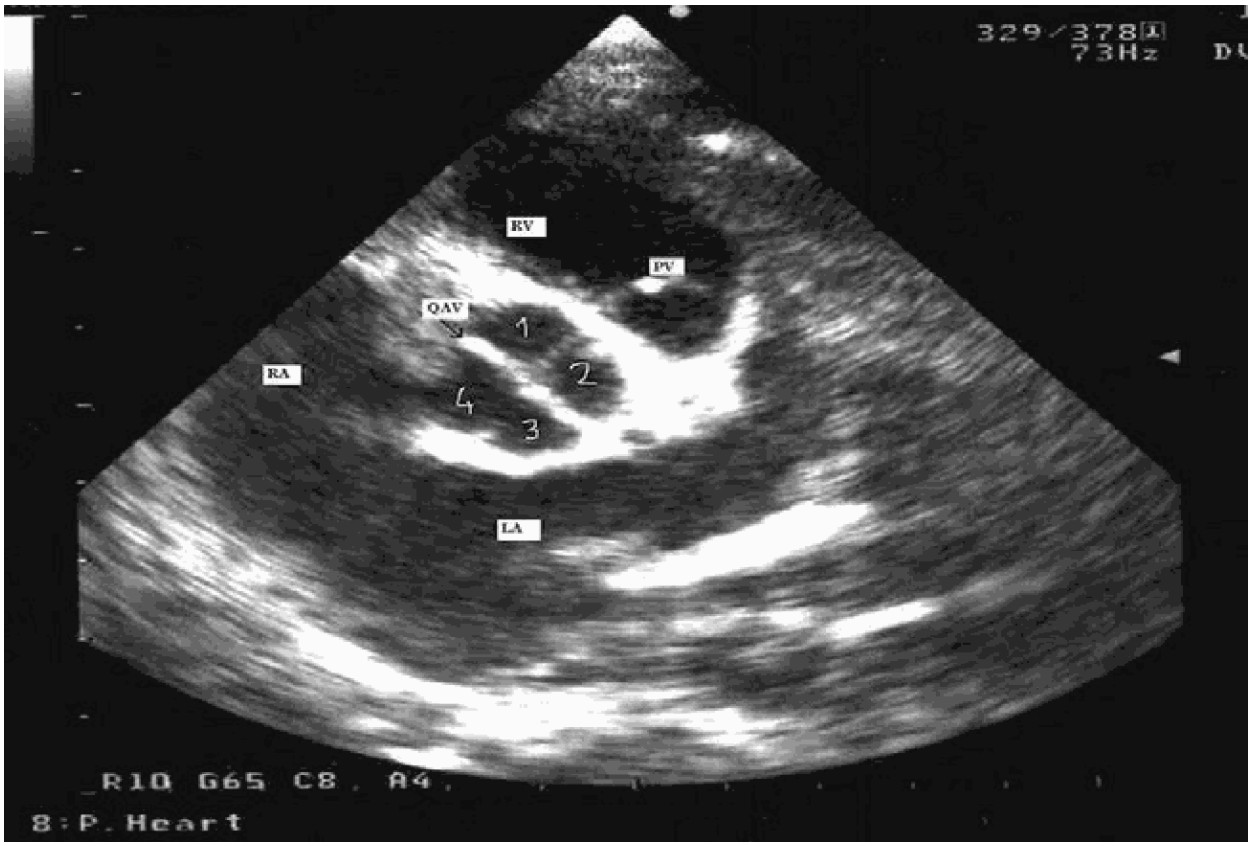
Aortic valve morphology should not be determined from a single frozen frame. Assess leaflet motion throughout systole.

20. Coronary Origins in PSAX

The parasternal short-axis view at the aortic root is a key starting point for visualization of the proximal coronary arteries.

Optimize with:

- higher frequency, shallow depth
- zoom, narrow sector
- small color box, low color scale
- low nequisite limit



21. PSAX — Mitral Valve Level

Sweep inferiorly from the aortic valve level.

The LV becomes circular and the mitral valve is seen in short axis.

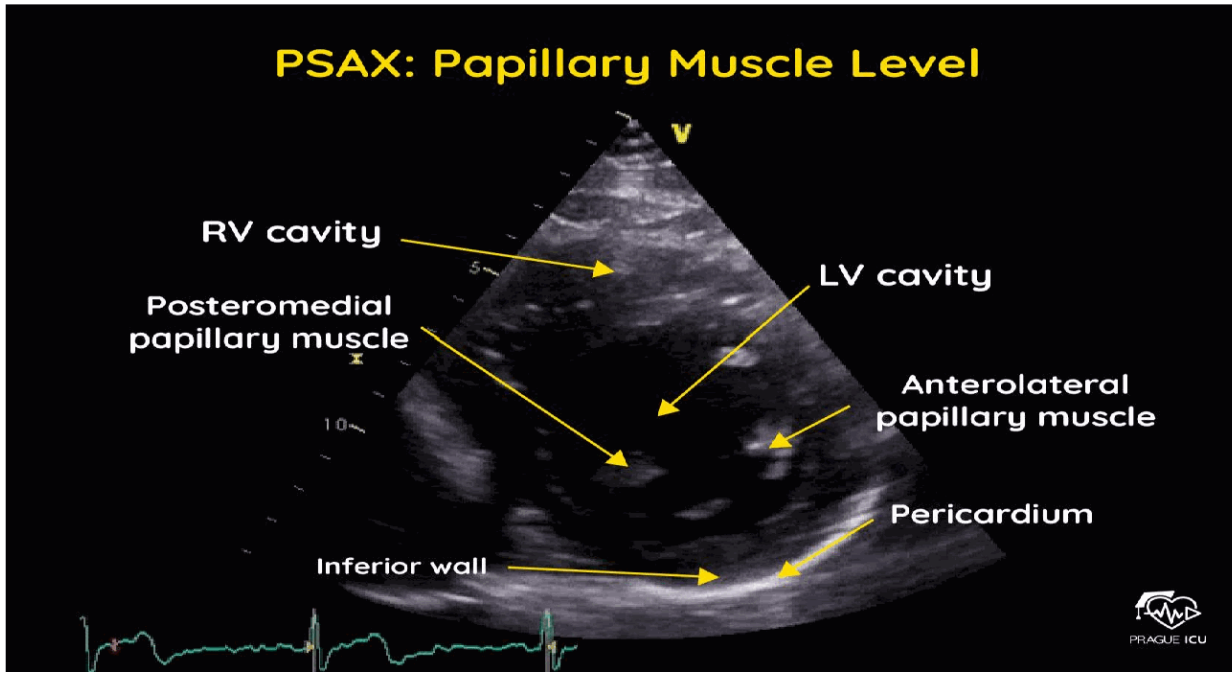
ASSESS

- mitral valve opening, valve orifice
- leaflet morphology, scallops and commissures

22. PSAX — Papillary Muscle Level

Further inferior sweep demonstrates:

- circular LV, papillary muscles
- RV anteriorly, interventricular septum



23. Septal Shape and RV Pressure Loading

Normally, the LV appears approximately circular in short axis.

With significant RV pressure loading, the interventricular septum may flatten.

This can produce a: **D-shaped LV**

24. PSAX — Apical Level

Continue the sweep toward the ventricular apex.

This helps assess:

- apical muscular septum, muscular VSDs

25. High Left Parasternal Window

Moving superiorly along the left parasternal border can provide excellent visualization of the ductal region.

STRUCTURES SEEN

Depending on plane:

- main PA, LPA, RPA
- ductus arteriosus
- descending aorta

PART III — APICAL WINDOW

26. Apical Four-Chamber View

PROBE POSITION

Place the transducer near the cardiac apex.

In infants, the apex may be more medial or superior than expected.

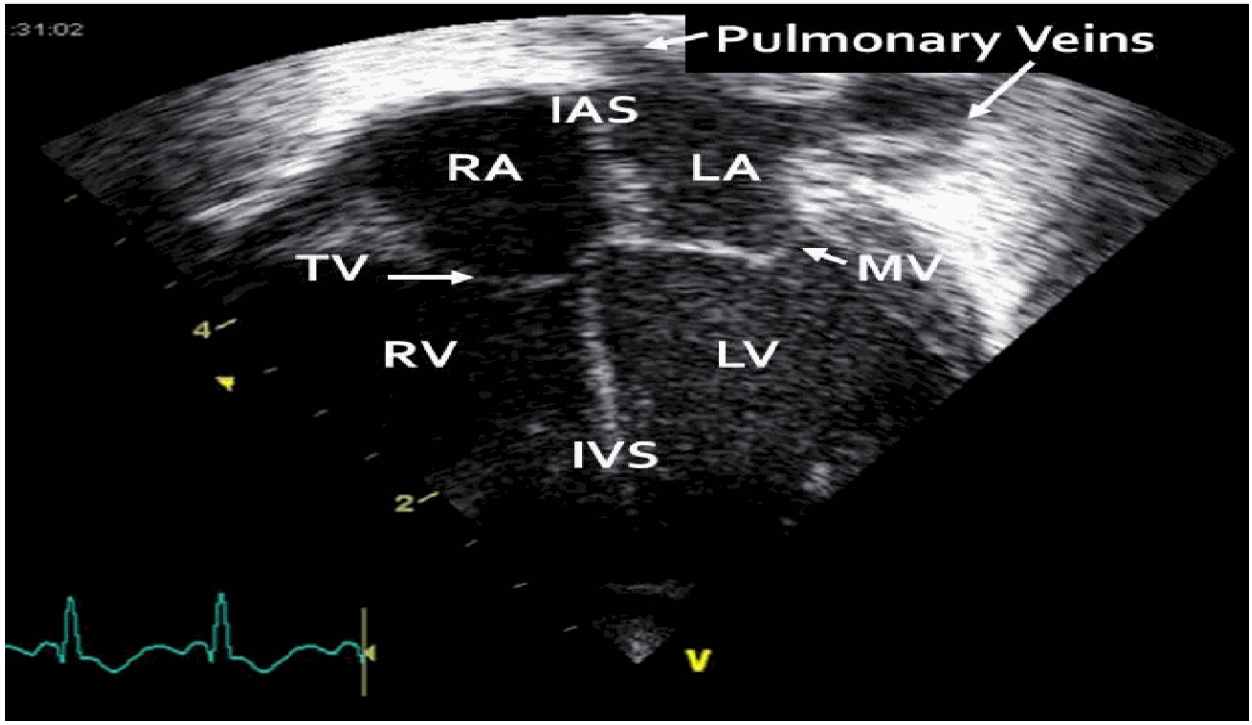
STRUCTURES SEEN

- RA, LA
- RV, LV
- tricuspid valve, mitral valve
- atrial septum, ventricular septum
- cardiac crux, pulmonary veins

A common error is obtaining an apical view in which the true ventricular apex is not included.

Slide the probe until the LV appears elongated with the true apex visible.

Do not accept the first four-chamber image simply because all four chambers are visible.



27. AV Valve Offset

In a normal heart, the tricuspid valve inserts slightly more apically than the mitral valve.

This normal AV valve offset helps identify ventricular morphology.

Loss of normal offset may raise suspicion of:

- AVSD

Marked apical displacement of the tricuspid valve suggests:

- Ebstein anomaly

28. Identifying Morphologic Right Ventricle

- moderator band
- coarse trabeculations
- more apical AV valve insertion

Morphologic Left Ventricle

Look for:

- smoother septal surface
- finer trabeculation
- absence of a moderator band
- more basal AV valve insertion

Name the ventricle by morphology—not by screen position.

However, apparent atrial septal dropout in this view must be interpreted cautiously.

29. Apical Five-Chamber View

From the apical four-chamber view, tilt or angle anteriorly to bring the LVOT and aortic valve into the imaging plane.

The "fifth chamber" is the aortic outflow.

30. Doppler in Apical Five-Chamber View

The apical approach often provides favorable alignment with LV outflow.

But:

Do not assume the apical view always provides the highest aortic velocity.

31. Apical Three-Chamber / Long-Axis View

32. Apical Two-Chamber View

Further rotation can produce a two-chamber view showing principally:

- LA
- LV
- mitral valve

PART IV — SUPRASTERNAL WINDOW

33. Why the Suprasternal Window Is Essential

A study can appear normal from subcostal, parasternal, and apical windows while significant aortic arch disease remains undetected.

The suprasternal window is essential for assessment of:

- aortic arch, arch sidedness
- coarctation, interrupted arch, arch hypoplasia
- head and neck vessels, descending aorta
- pulmonary veins in crab view

34. Suprasternal Long-Axis Aortic Arch View

PROBE POSITION

Place the transducer in the suprasternal notch and direct the beam inferiorly into the mediastinum. Adjust rotation and angulation until the arch is elongated.

STRUCTURES TO DEMONSTRATE

- ascending aorta, transverse arch, head and neck vessels
- aortic isthmus, descending thoracic aorta

COLOR DOPPLER

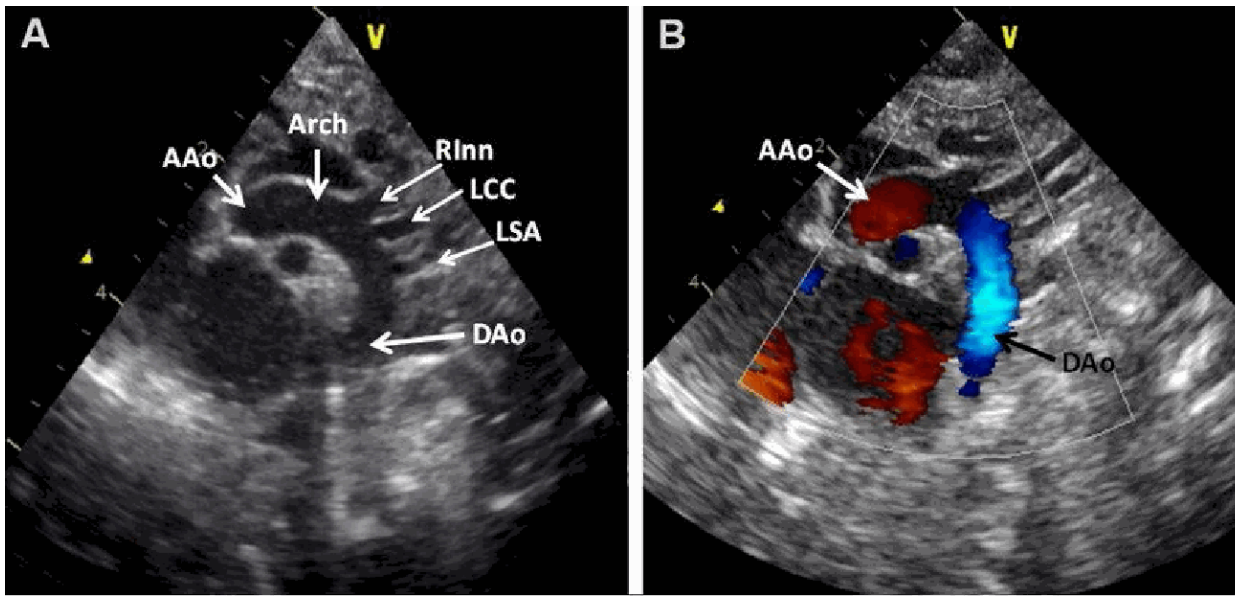
Look for:

- acceleration
- turbulence
- abnormal flow patterns

SPECTRAL DOPPLER

Assess:

- peak velocity
- waveform shape
- diastolic continuation when present



35. Coarctation — Important Echo Clues

Look for a combination of:

- focal narrowing
- small isthmus
- color acceleration, dragger shape envelop
- increased Doppler velocity
- persistent antegrade diastolic flow
- abnormal abdominal aortic Doppler
- associated arch hypoplasia

CRITICAL PITFALL

Do not exclude neonatal coarctation simply because the Doppler gradient is low, particularly while the ductus remains patent.

36. Suprasternal Short-Axis View

Rotation from the arch long-axis plane can provide transverse views of mediastinal structures.

Depending on the level, one may visualize:

- ascending aorta
- SVC, innominate vein
- RPA, LPA, PDA
- arch vessels
- transverse arch

CLINICAL USES

- arch sidedness, branching pattern
- systemic venous anatomy, Crab view, TAPVR
- pulmonary artery relationships, PDA

37. Suprasternal Pulmonary Venous / “Crab” View

A modified suprasternal view can demonstrate pulmonary veins entering the LA.

The veins may create the characteristic appearance sometimes called the **crab view**.

CLINICAL USES

- confirming pulmonary venous return
- suspected PAPVC
- suspected TAPVC
- pulmonary venous obstruction/stenosis

OPTIMIZATION

Use:

- appropriate depth
- small color box
- lower velocity scale
- adequate color gain

RIGHT PARASTERNAL AND MODIFIED VIEWS

38. Right Parasternal Window

The right parasternal window is not required for every structure but can provide important additional information.

It may be useful for:

- SVC–RA region, sinus venosus ASD
- right pulmonary veins
- Doppler alignment with aortic flow

CLINICAL PEARL

When standard views do not answer the question, change the window rather than forcing the diagnosis from an inadequate image.

39. Modified Views Are Part of Congenital Echo

Congenital hearts do not always cooperate with textbook imaging planes.

Modified views may be necessary in:

- dextrocardia
- heterotaxy
- postoperative hearts
- abnormal great artery relationships
- unusual venous anatomy
- chest deformity
- poor acoustic windows

The purpose of a standard protocol is to provide a framework—not to prevent intelligent adaptation.

Standardize the examination, but individualize the imaging.

40. Which View Is Best for Which Structure?

Structure / Lesion	High-Yield View(s)
Abdominal situs	Subcostal abdominal
IVC connection	Subcostal
Atrial septum	Subcostal four-chamber/bicaval
Sinus venosus ASD	Subcostal bicaval + right parasternal
Ventricular septum	Parasternal + subcostal sweeps
AV valves	Apical four-chamber
Mitral valve prolapse	PLAX + apical long-axis
Ebstein anomaly	Apical four-chamber
Aortic valve morphology	PSAX + PLAX
Pulmonary valve	PSAX/RVOT
Coronary origins	PSAX at aortic root
PDA	High left parasternal/ductal view
Pulmonary veins	Subcostal + suprasternal + modified views
Aortic arch	Suprasternal long-axis
Coarctation	Suprasternal + abdominal aortic Doppler
Ventricular function	Apical + parasternal
Pericardial effusion	Multiple windows

41. View-by-View Quick Reference

Window	Key Structures	Major Purpose
Subcostal abdominal	IVC, aorta, liver, spine	Situs
Subcostal 4C	Atria, ventricles, septa, AV valves	Connections/septa
Subcostal bicaval	SVC, IVC, RA, IAS	Venous anatomy/ASD
PLAX	LV, MV, LVOT, AV, LA	Left heart anatomy
PSAX—base	AV, coronaries, RVOT, PV, PAs	Great arteries/outflow
PSAX—ventricles	LV, RV, septum	Function/pressure loading
Apical 4C	Chambers, AV valves, crux	Connections/function
Apical 5C	LVOT, AV	Aortic Doppler
Apical 3C	MV, LV, LVOT, AV	Left heart
Suprasternal LAX	Aortic arch	Arch/coarctation
Suprasternal modified	Pulmonary veins	Venous return
High left parasternal	Ductus, PAs, descending Ao	PDA
Right parasternal	SVC/IAS/RUPV/Ao	Modified congenital assessment

	ECHO WINDOWS	ECHO VIEWS	ABNORMALITIES THAT CAN BE SEEN
1	SUBCOSTAL	Subcostal Transverse View (2D, Color Doppler)	Situs Solitus Situs Inversus Situs Ambiguos
		Sagittal View (2D, M-Mode, Color) Measure IVC Size & Collapsibility Assess Aorta	Interrupted IVC Abdominal CoA
		Bicaval View (2D, Color Doppler)	Sinus Venosus ASD
2	APICAL	Apical 4 Chamber (2D, M-Mode, Color, CW, PW) Assess Atria & Ventricles LA Appendage' AV Valves (TR, MR) E/A Ratio, TAPSI, Visual EF Pulmonary Veins Openings	ASD (Especially Secundum Type) VSD (Muscular, Apical, Perimembranous), AV Canal Defects, Valvular Assessment Chamber Enlargement, Ventricular Dysfunction, Thrombus, Vegetation
		Apical 5 Chamber (2D, Color Doppler, CW)	Aortic Stenosis (Especially Valvular & Sub-valvular) VSD (Especially Perimembranous) Aortic Regurgitation
3	PARASTERNAL	**Parasternal Short Axis (Av Level) (2D Color Doppler, CW)** Assess PA, PV Measure Coronaries Diameter Measure PV Annulus	Valvular Abnormalities (Bicuspid aortic valve, TR, PR, PS) Pulmonary Artery Abnormalities Truncus Arteriosus / Great Vessel Anomalies ASD Rims, PDA Ruptured Sinus of Valsalva Coronaries Dilatation, Coronary Artery Fistula
		Parasternal Short Axis (LV Level) (2D, Color Doppler)	Muscular VSD, LV Dysfunction, D-Shape LV Mitral Valve Leaflets (Prolapse, Flail)
		Parasternal Long Axis (2D, M-Mode, Color Doppler, CW) LV Function – Calculate EF LA/Ao Ratio	Valvular Anomalies (MR, AR) LV Hypertrophy, LV dysfunction Aortic Root Dilation / Dissection Dilated Annulus (Aortic, Mitral) MVP, Flail
4	SUPRASTERNAL	**Suprasternal Notch View (2D, Color Doppler, CW)** SVC Flow & PV Confluence (TAPVC) Crab's View - PVS SVC – PA Flow for Glenn Shunt L-SVC & Innominate Vein	Coarctation of Aorta (Classic Indication) Interrupted Aortic Arch, Aortic Arch Hypoplasia , Patent Ductus Arteriosus Arch Sidedness ,AR, AS AP-Window Defect

42. The Correct Sequence Within Each View

Once a standard view has been obtained:

1. Look → 2. Sweep → 3. Optimize

1. Look — Understand the 2D anatomy.

2. Sweep — Demonstrate the complete structure.

3. Optimize — Adjust depth, frequency, focus, gain, and sector.

4. Color — Assess direction and turbulence.

5. Doppler — Measure flow when clinically relevant.

6. Measure — Perform appropriate dimensions and Z-scores.

7. Record — Store representative stills and loops.

A useful memory rule is:

Find → Sweep → Optimize → Color → Doppler → Measure → Record

43. Common Errors in Standard Views

ERROR 1 — COLLECTING STILL IMAGES WITHOUT SWEEPING

Risk: Missing additional anatomy.

Correction: Record cine sweeps.

ERROR 2 — CALLING AN ASD FROM AN APICAL DROPOUT

Correction: Confirm from the subcostal window with color Doppler.

ERROR 3 — FORESHORTENING THE LV

Correction: Reposition the apical transducer until the true apex is visualized.

ERROR 4 — IDENTIFYING VENTRICLES BY SCREEN POSITION

Correction: Identify morphology.

ERROR 5 — EXAMINING ONLY THE PROXIMAL ARCH

Correction: Demonstrate transverse arch, isthmus, and descending aorta.

ERROR 6 — ASSUMING FOUR PULMONARY VEINS ARE NORMAL WITHOUT DEMONSTRATION

Correction: Actively document venous connections when clinically relevant and obtain modified views if necessary.

ERROR 7 — STOPPING AFTER FINDING THE MAJOR LESION

Correction: Return to the systematic sequence and complete the examination.

44. Lesion-Based Example — Tetralogy of Fallot

A child with suspected TOF should not be diagnosed from a single parasternal image.

Use multiple windows.

Subcostal

Assess:

- Situs, AV connections
- VSD, RVOT, associated anatomy

Parasternal Long Axis

Assess:

- VSD, aortic override
- LV function

Parasternal Short Axis

Assess:

- RVOT, pulmonary valve, pulmonary annulus
- MPA, branch PAs
- coronary origins

Apical

Assess:

- Ventricular function, AV valves
- VSD relationship

Suprasternal

Assess:

- aortic arch, arch sidedness
- PDA, collaterals

This demonstrates an important principle:

A diagnosis is assembled across windows—not discovered in one picture.

45. The Four-Window Memory Map

SUBCOSTAL — START

Think:

Situs + Septa + Systemic veins

PARASTERNAL — STRUCTURE

Think:

Valves + Septum + Outflows + Coronaries

APICAL — CONNECTION & FUNCTION

Think:

AV valves + Chambers + Ventricular function

SUPRASTERNAL — GREAT VESSELS

Think:

Arch + Branches + Descending aorta

A simple memory phrase:

Start → Structure → Function → Finish the Arch

46. Minimum Window Checklist

Before completing a routine pediatric echocardiogram, ask whether you have adequately assessed:

Subcostal

- Situs
- IVC/systemic veins
- Atrial septum
- AV connections
- Ventricular septum

Parasternal

- PLAX
- PSAX sweep
- Mitral/aortic valves
- RVOT/pulmonary valve
- Branch PAs
- Coronary origins where appropriate/feasible

Apical

- Four chamber
- AV valve function
- Ventricular function
- LVOT/aortic flow

Suprasternal

- Aortic arch
- Arch sidedness/branches where required
- Descending aortic flow

Additional Views

- Pulmonary veins adequately assessed
- PDA/ductal region when relevant
- Modified/right parasternal views when required

Chapter Key Points

- The four major pediatric echocardiographic windows are **subcostal, parasternal, apical, and suprasternal**.
- A window is the acoustic access point; a view is the imaging plane obtained from it.
- Probe manipulation requires **sliding, rotating, tilting, angling, and sweeping**.
- Subcostal imaging is particularly important for **situs, venous connections, atrial septum, and AV connections**.
- Parasternal imaging provides detailed assessment of **valves, ventricular septum, outflow tracts, pulmonary arteries, and coronary origins**.
- The apical window is particularly useful for **AV valves, chamber relationships, ventricular morphology, and function**.
- The suprasternal window is essential for **aortic arch anatomy and coarctation assessment**.
- High left parasternal and right parasternal views provide important complementary information.
- Important structures should be assessed in more than one plane whenever feasible.
- A single still frame should never substitute for a systematic sweep.
- Modified views are often necessary in congenital heart disease.
- 2D anatomy should be understood before Doppler information is interpreted.
- A complete study should reconstruct the heart as a connected three-dimensional structure.

MASTER PRINCIPLE

Never let a standard view become a limitation. Use standard windows to build the framework, then sweep, modify, and reposition until the anatomy is understood.

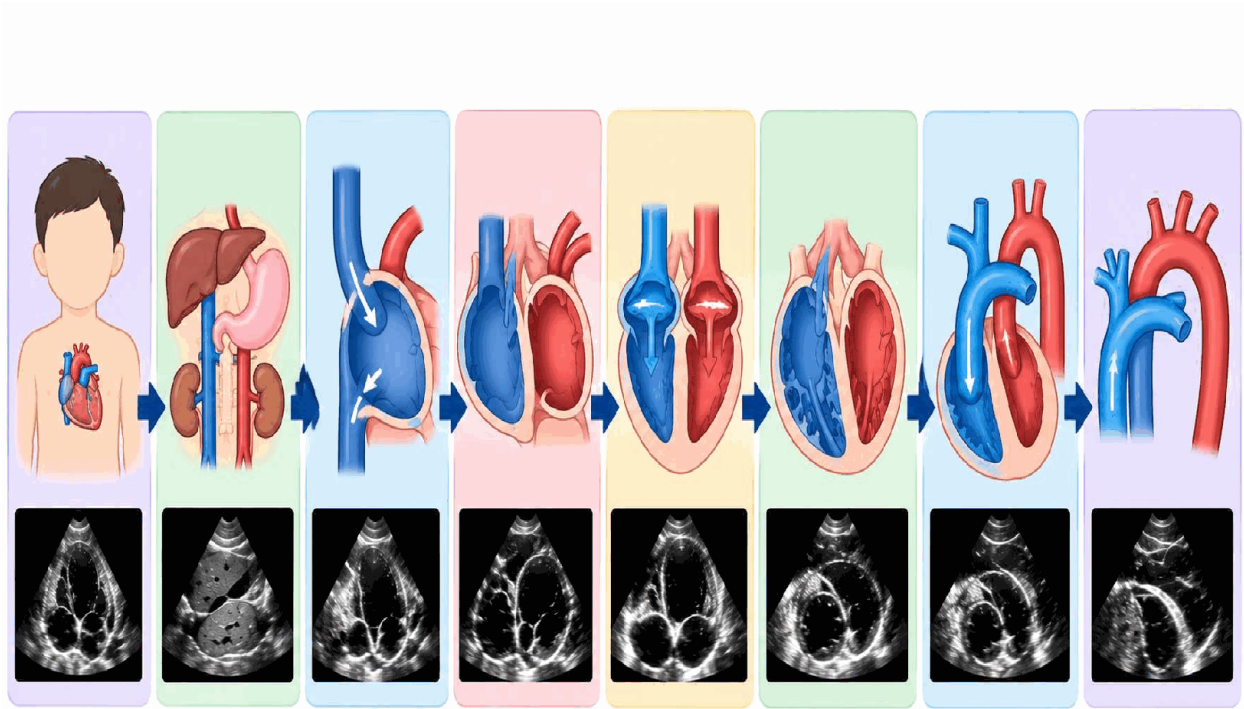
The Pediatric Window Workflow

SUBCOSTAL → PARASTERNAL → APICAL → SUPRASTERNAL

Orient → Define Anatomy → Assess Function → Complete the Great Vessels

CHAPTER 5

Sequential Segmental Analysis



Learning Objectives

By the end of this chapter, the learner should be able to:

1. Explain the purpose of sequential segmental analysis (SSA) in congenital heart disease.
2. Establish cardiac position and abdominal/atrial situs before interpreting intracardiac anatomy.
3. Identify atrial arrangement using morphological features rather than position alone.
4. Identify the morphologic right and left ventricles.
5. Determine atrioventricular (AV) connections.
6. Identify the morphologic aorta and pulmonary artery and determine ventriculoarterial (VA) connections.
7. Describe the spatial relationship of the great arteries without using it as a substitute for defining VA connections.
8. Evaluate systemic and pulmonary venous connections.
9. Apply segmental analysis to complex lesions such as TGA, ccTGA, DORV, heterotaxy, and single-ventricle hearts.
10. Produce a concise segmental description before assigning a diagnostic label.

1. Why Sequential Segmental Analysis?

Congenital heart disease can profoundly alter normal cardiac anatomy.

The atria may not be in their expected positions. The morphologic RV may lie on the left. The aorta may arise from the RV. Both great arteries may arise predominantly from one ventricle. Venous connections may also be abnormal.

For this reason, the echocardiographer should not begin with:

"What congenital lesion does this look like?"

Instead, begin with:

"What are the cardiac segments, and how are they connected?"

This is the essence of sequential segmental analysis.

2. The Central Principle

The heart can be reconstructed as a series of segments and connections:

Veins → Atria → AV Junction → Ventricles → VA Junction → Great Arteries

At every stage, answer two questions:

What is this structure?

and

What does it connect to?

Only after establishing these relationships should the final congenital diagnosis be assigned.

3. Anatomy Before Position

One of the most important principles in congenital echocardiography is:

Identify structures by morphology—not simply by location.

- Do not call a chamber the LV because it lies on the left.
- Do not call a vessel the aorta because it lies posteriorly.
- Do not assume that the right-sided atrium is morphologically the RA.

Complex CHD can violate all these positional expectations.

4. The Sequential Segmental Road Map

A practical pediatric echo sequence is:

STEP 1 — Cardiac position and orientation

↓

STEP 2 — Abdominal/atrial arrangement

↓

STEP 3 — Systemic and pulmonary venous connections

↓

STEP 4 — Atrioventricular connections

↓

STEP 5 — Ventricular morphology

↓

STEP 6 — Ventriculoarterial connections

↓

STEP 7 — Great artery relationship

↓

STEP 8 — Associated intracardiac and extracardiac abnormalities

↓

STEP 9 — Hemodynamics and ventricular function

This transforms a complex congenital heart into a logical sequence.

5. Step 1 — Where Is the Heart?

Before analyzing internal anatomy, establish cardiac position.

Assess:

- position of the heart within the thorax
- direction of the cardiac apex
- orientation of the cardiac base

Common terms include:

Levocardia

Apex directed toward the left.

Dextrocardia

Apex directed toward the right.

Mesocardia

Apex positioned approximately in the midline.

IMPORTANT POINT

Cardiac position does not define situs.
A patient can have dextrocardia with situs solitus, situs inversus, or a more complex arrangement.

6. Step 2 — Begin Below the Heart

For congenital echo, an excellent starting point is the **subcostal abdominal transverse view**.

Identify:

- spine
- descending aorta
- IVC
- liver
- stomach when visible

Then determine the relationship of:

IVC ↔ Aorta ↔ Spine
This provides an important clue to abdominal arrangement and alerts the operator to possible heterotaxy.

7. Situs Solitus

In usual abdominal arrangement:

- liver is predominantly right-sided
- stomach is predominantly left-sided
- IVC is usually right of the spine
- descending aorta is usually left of the spine

The heart most commonly has normal atrial arrangement.

However:

Do not stop at the abdominal image. Trace the venous connections and establish atrial morphology.

8. Situs Inversus

In situs inversus, the abdominal arrangement is approximately the mirror image of usual arrangement.

Typical findings may include:

- stomach on the right
- liver predominantly on the left
- IVC leftward
- aorta rightward

Again, the echocardiographer should describe the actual anatomy rather than relying only on a positional label.

9. Heterotaxy / Isomerism

When the arrangement does not fit the usual or mirror-image pattern, consider **heterotaxy**.

Possible clues include:

- abnormal IVC position
- interrupted IVC
- azygos continuation
- midline liver
- unusual systemic venous anatomy
- abnormal pulmonary venous return
- complex AV junction anatomy
- abnormal great artery relationships

ECHO PEARL

An unusual IVC–aorta relationship should trigger a deliberate search for heterotaxy and abnormal venous connections.

10. Atrial Arrangement

The atria should ideally be identified by **morphology**, particularly the atrial appendages.

The morphologic right and left atrial appendages differ in their anatomy, although direct appendage visualization can be difficult on routine transthoracic echocardiography.

Therefore, in clinical pediatric echo, atrial arrangement is usually established by integrating:

- abdominal situs
- systemic venous connections
- pulmonary venous connections
- atrial anatomy
- appendage morphology when demonstrable

CRITICAL PRINCIPLE

Right-sided does not necessarily mean morphologic right atrium.

11. Step 3 — Systemic Venous Connections

After establishing arrangement, trace systemic venous return.

Identify:

Inferior vena cava

Follow the IVC:

Abdomen → diaphragm → atrium

Do not simply identify an abdominal vessel and assume its destination.

Superior vena cava

Demonstrate:

SVC → atrium

Also consider whether there is:

- persistent left SVC
- bilateral SVCs
- interrupted IVC
- azygos continuation
- abnormal hepatic venous drainage

12. Interrupted IVC

If the expected IVC is not seen entering the atrium:

Do not immediately conclude that it is absent.

Search for:

- interruption of the hepatic segment
- enlarged azygos vein
- azygos continuation to the SVC
- separate hepatic venous drainage to the atrium

This is particularly important in heterotaxy.

13. Persistent Left Superior Vena Cava

A persistent left SVC may drain:

- through a dilated coronary sinus into the RA
- less commonly into the LA or another abnormal connection

A **dilated coronary sinus** should prompt a search for persistent left SVC.

PRACTICAL TECHNIQUE

Use:

- parasternal views
- apical views
- suprasternal views
- color Doppler

to define its course and drainage.

14. Pulmonary Venous Connections

Pulmonary veins deserve the same systematic attention as systemic veins.

Whenever feasible, demonstrate pulmonary veins entering the LA.

Assess:

- right upper pulmonary vein
- right lower pulmonary vein
- left upper pulmonary vein
- left lower pulmonary vein

Not all four veins are equally easy to demonstrate in every child, but the operator should actively establish pulmonary venous connection when congenital disease is suspected.

15. When Pulmonary Veins Are Not Clearly Seen

Do not write:

"Pulmonary veins normal"

unless the connection has been adequately assessed.

If pulmonary venous return is uncertain, use multiple windows:

- subcostal
- apical
- parasternal
- suprasternal
- modified right parasternal views

Apply low-flow color optimization.

ECHO PEARL

Failure to visualize a pulmonary vein is a reason to look harder—not evidence that it is normal.

16. Step 4 — Atrioventricular Connections

Now determine:

Which atrium connects to which ventricle?

This defines the AV connection.

There are several important possibilities.

17. Concordant AV Connection

In an AV concordant connection:

Morphologic RA → Morphologic RV

and

Morphologic LA → Morphologic LV

This is the usual AV connection.

Remember:

Concordance describes connection—not position.

18. Discordant AV Connection

In AV discordance:

Morphologic RA → Morphologic LV
and

Morphologic LA → Morphologic RV
This occurs classically in congenitally corrected transposition when accompanied by VA discordance.

The diagnosis requires correct identification of **ventricular morphology**.

19. Double-Inlet AV Connection

In a double-inlet connection, both atria connect predominantly to one ventricular chamber through either:

- two AV valves, or
- a common AV valve

This forms part of the spectrum of functionally univentricular hearts.

The echocardiographer should determine:

- dominant ventricle
- rudimentary ventricle
- AV valve anatomy
- ventricular communication
- outflow relationships

20. Absent AV Connection

One atrium may have no direct connection to a ventricular chamber.

The classic example is: **Tricuspid atresia**

The morphologic RA has no direct connection to the morphologic RV.

Do not describe this merely as an "absent tricuspid valve."

The AV connection itself is absent.

21. Step 5 — Identify Ventricular Morphology

This is one of the most important skills in congenital echocardiography.

The ventricles must be named according to their **morphological characteristics**, not their location.

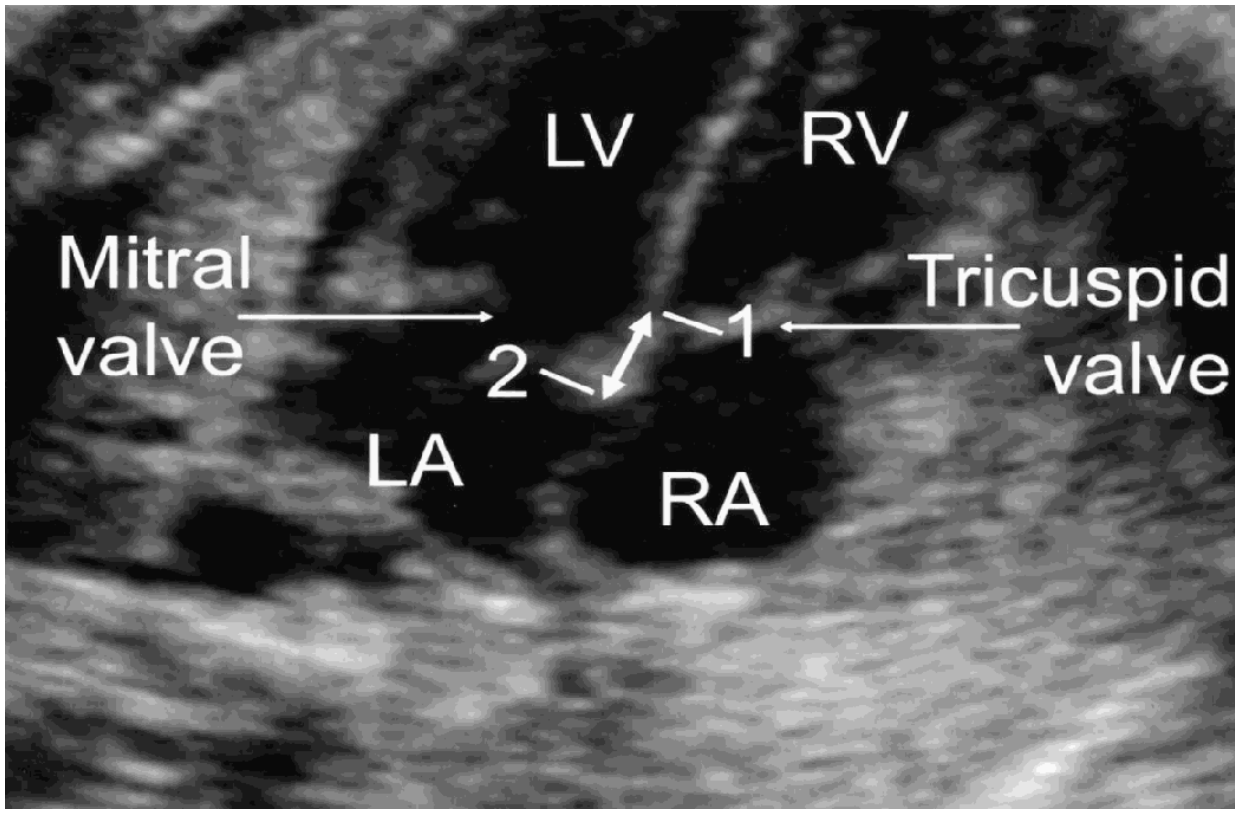
22. Morphologic Right Ventricle

Features favoring a morphologic RV include:

- coarse trabeculations
- moderator band
- more apical insertion of the AV valve
- characteristic irregular internal surface

A useful memory aid:

RV = Rough + Moderator band + Apical AV valve



23. Morphologic Left Ventricle

Features favoring a morphologic LV include:

- relatively smooth septal surface
- finer trabeculations
- absence of a moderator band
- more basal insertion of the AV valve

The LV typically has a more ellipsoidal geometry.

Golden Rule: If the morphologic RV lies on the left side of the screen, it is still the RV.

24. AV Valve Offset

Normally, the tricuspid valve inserts more apically than the mitral valve.

This helps distinguish the ventricles.

However, AV valve insertion can itself be abnormal in congenital disease.

Examples include:

- AVSD

Use several morphological features together rather than relying on one sign.

25. Ventricular Topology

In complex congenital heart disease, ventricular arrangement can be described in terms of topology or handedness.

For practical introductory echocardiography, the learner should first master:

1. identifying morphologic RV and LV,
2. determining their connections,
3. determining which great artery arises from each.

More advanced topology terminology can then be added when required.

26. Step 6 — Ventriculoarterial Connections

Now ask:

Which great artery arises from which ventricle?

Do not identify the arteries solely from anterior/posterior position.

Follow each ventricle through its outflow tract into the great vessel.

27. How to Identify the Aorta

The aorta can be identified by following it into:

- ascending aorta
- aortic arch
- head and neck vessels
- descending aorta

Thus:

Aorta = the great artery that continues into the systemic arterial tree and arch.

28. How to Identify the Pulmonary Artery

The pulmonary trunk is identified by its bifurcation into:

RPA + LPA

Thus:

Pulmonary artery = the great artery that bifurcates into the branch pulmonary arteries.

GOLDEN RULE

Identify great arteries by their branching pattern—not merely by position.

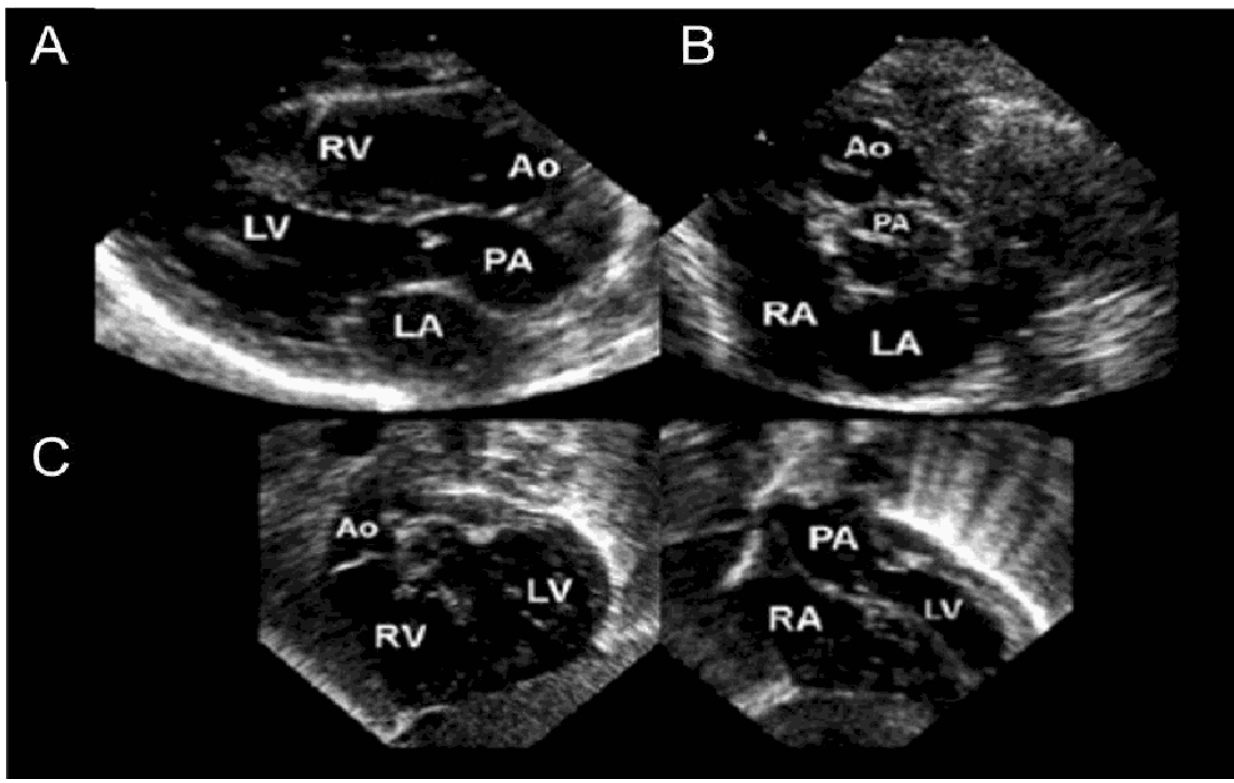
29. Concordant VA Connection

In VA concordance:

Morphologic RV → Pulmonary artery

and

Morphologic LV → Aorta



30. Discordant VA Connection

In VA discordance:

Morphologic RV → Aorta
and

Morphologic LV → Pulmonary artery
This occurs in:

- complete TGA when AV connections are concordant
- ccTGA when AV connections are also discordant

This illustrates why the entire connection sequence matters.

31. Double-Outlet Ventricular Connection

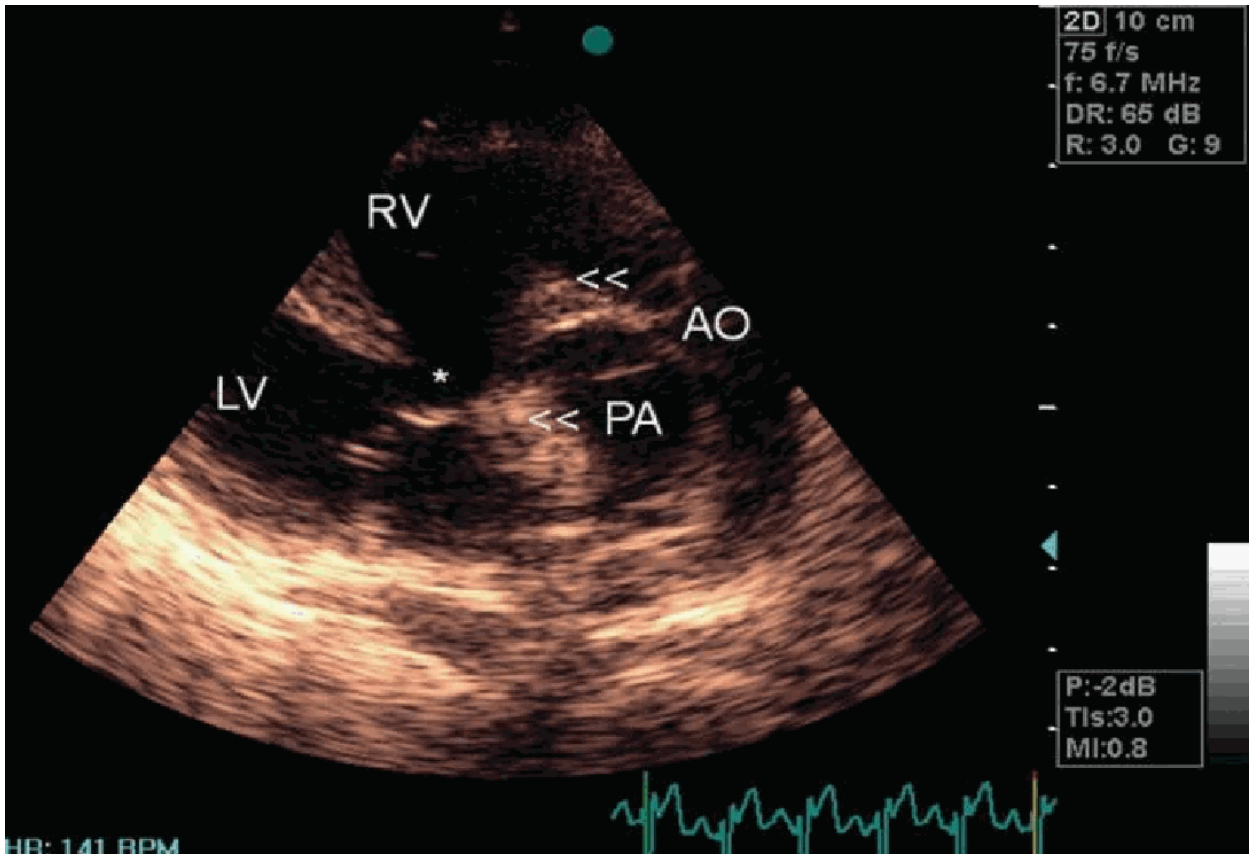
When both great arteries arise predominantly from one ventricle, there is a double-outlet connection.

A common example is: **Double-outlet right ventricle (DORV)**

However, identifying DORV is only the beginning.

The echo must also determine:

- VSD location
- relationship of VSD to great arteries
- great artery relationship
- pulmonary obstruction
- ventricular sizes
- AV valve anatomy
- coronary anatomy where relevant



32. Single-Outlet Connections

In some congenital lesions, only one patent arterial outlet is present.

Examples include:

- pulmonary atresia
- aortic atresia
- common arterial trunk

The echocardiographer should describe the actual ventricular–arterial connection rather than trying to force the anatomy into the usual two-outlet pattern.

33. Step 7 — Great Artery Relationship

After defining VA connections, describe how the great arteries relate spatially.

Possible relationships include:

- usual spiral relationship
- parallel relationship
- aorta anterior and rightward
- aorta anterior and leftward
- side-by-side great arteries

This information is important but should come **after** determining VA connections.

Critical Principle: Great artery position does not define the ventriculoarterial connection.

34. Crossing Versus Parallel Great Arteries

In the normal heart, the great arteries have a crossing relationship.

In classic TGA, they tend to run more parallel.

This can provide an important visual clue.

However: **Parallel vessels suggest TGA but do not establish the diagnosis.**

The diagnosis requires demonstrating:

RV → Aorta
LV → Pulmonary artery

35. Step 8 — Intracardiac Communications

Once the segmental connections are established, systematically evaluate communications.

Atrial Level

- PFO
- ASD
- common atrium
- restrictive atrial communication

Ventricular Level

- VSD
- multiple VSDs
- ventricular communication in univentricular hearts

Great Artery Level

- PDA
- aortopulmonary window
- common arterial trunk

These communications can be crucial for neonatal survival in duct- or mixing-dependent lesions.

36. Step 9 — Obstruction

Systematically search for obstruction along both circulations.

Right-Sided Pathway

Systemic veins → RA → AV valve → RV → RVOT → pulmonary valve → PAs
Left-Sided Pathway

Pulmonary veins → LA → AV valve → LV → LVOT → aortic valve → arch

At each level ask:

Is there narrowing or obstruction?

37. Step 10 — Complete the Physiology

Segmental analysis defines anatomy.

The echocardiogram must then determine what the anatomy is doing physiologically.

Assess:

- shunt direction
- restriction
- pulmonary blood flow
- systemic blood flow
- ventricular pressure
- chamber enlargement
- volume loading
- pressure loading
- AV valve regurgitation
- ventricular function
- ductal dependency

Segmental anatomy tells you what the heart is. Hemodynamics tells you how that heart is functioning.

38. Applying SSA — Normal Heart

A normal segmental sequence can be described as:

Usual atrial arrangement → concordant AV connections → concordant VA connections

Flow pathway:

Systemic veins

↓

Morphologic RA

↓

Morphologic RV

↓

Pulmonary artery

↓

Lungs

↓

Pulmonary veins

↓

Morphologic LA

↓

Morphologic LV

↓

Aorta

This is the reference against which abnormal connections are understood.

39. Applying SSA — Complete TGA

Do not begin with "parallel great arteries."

Instead demonstrate:

Atrial arrangement

Usually usual.

AV connections

Concordant:

RA → RV
LA → LV
VA connections

Discordant:

RV → Aorta
LV → Pulmonary artery
Therefore:

AV concordance + VA discordance = complete TGA
Then assess:

- atrial communication
- VSD
- PDA
- coronary origins
- LVOT obstruction
- arch anatomy

40. Applying SSA — Congenitally Corrected TGA

The sequence is different.

AV Connection

Discordant:

RA → morphologic LV
LA → morphologic RV
VA Connection

Discordant:

Morphologic LV → PA
Morphologic RV → Aorta
Therefore:

AV discordance + VA discordance = double discordance
This results in physiologically "corrected" circulation, although important associated abnormalities may be present.

41. Applying SSA — DORV

First establish:

- atrial arrangement
- AV connection
- ventricular morphology

Then demonstrate that both great arteries arise predominantly from the morphologic RV.

Next ask the crucial anatomical question:

Where is the VSD relative to the great arteries?
Possible relationships include:

- subaortic
- subpulmonary
- doubly committed
- remote/noncommitted

Then assess:

- pulmonary stenosis
- great artery relationship
- ventricular size
- AV valve anatomy
- arch anatomy

42. Applying SSA — Tricuspid Atresia

Do not simply look for a small RV.

Establish:

- atrial arrangement
- absence of direct RA-to-RV connection
- atrial communication
- dominant LV
- hypoplastic/rudimentary RV
- VSD/bulboventricular communication
- VA connection
- pulmonary outflow
- great artery relationship

The diagnosis becomes anatomically coherent when analyzed sequentially.

43. Applying SSA — Single-Ventricle Physiology

In a functionally univentricular heart, do not stop after saying:

"Single ventricle."

Describe:

- atrial arrangement
- systemic venous connections
- pulmonary venous connections
- AV connection
- dominant ventricular morphology
- rudimentary ventricular chamber
- ventricular communication
- VA connections
- pulmonary obstruction
- systemic outflow obstruction
- AV valve regurgitation
- arch anatomy

These details directly influence surgical strategy.

44. Applying SSA — Heterotaxy

Heterotaxy particularly demonstrates the value of sequential analysis.

Do not try to recognize the syndrome from one image.

Instead systematically establish:

Abdominal arrangement

↓

Systemic veins

↓

Pulmonary veins

↓

Atrial arrangement

↓

AV junction

↓

Ventricular morphology

↓

VA connections

↓

Pulmonary outflow

↓

Aortic arch

ECHO PEARL

The more complex the heart looks, the more strictly you should return to the sequence.

45. The Three Connections You Must Never Assume

1. Venous → Atrial Connection

Where do systemic and pulmonary veins drain?

2. Atrial → Ventricular Connection

Which atrium connects to which ventricle?

3. Ventricular → Arterial Connection

Which ventricle gives rise to which great artery?

These three questions alone prevent many major diagnostic errors.

46. The Segmental Analysis Table

Step	Question	What to Establish
1. Position	Where is the heart?	Levocardia/dextrocardia/mesocardia
2. Arrangement	What is the situs?	Usual, mirror-image, heterotaxy
3. Veins	Where does blood enter?	Systemic & pulmonary venous connections
4. AV connection	Which atrium connects to which ventricle?	Concordant, discordant, double inlet, absent
5. Ventricles	Which ventricle is which?	Morphologic RV and LV
6. VA connection	Which artery arises from which ventricle?	Concordant, discordant, double outlet, single outlet
7. Great arteries	How are they related?	Crossing, parallel, anterior/right/left
8. Communications	Where can blood cross?	ASD, VSD, PDA, other
9. Obstruction	Where is flow restricted?	Inflow, outflow, valves, arch
10. Physiology	How is the heart coping?	Pressure, volume, function, flow

47. The "Do Not Assume" Box

Do not assume:

- Right-sided atrium = RA
- Left-sided ventricle = LV
- Posterior great vessel = aorta
- Anterior vessel = pulmonary artery
- Parallel great arteries = TGA
- Small RV = tricuspid atresia
- One large ventricle = complete diagnosis

Instead:

Identify → Trace → Connect → Confirm

48. Common Mistakes

MISTAKE 1 — STARTING WITH A DIAGNOSTIC LABEL

"This looks like DORV."

Better Approach

First determine the connections; then assign the diagnosis.

MISTAKE 2 — IDENTIFYING VENTRICLES BY POSITION

Correction

Use:

- moderator band
- trabeculation
- AV valve insertion
- ventricular morphology

MISTAKE 3 — IDENTIFYING GREAT ARTERIES BY POSITION

Correction

Trace:

Aorta → arch
and

Pulmonary artery → RPA + LPA

MISTAKE 4 — IGNORING PULMONARY VEINS

Correction

Actively establish pulmonary venous drainage, especially in:

- heterotaxy
- ASD
- suspected TAPVR/PAPVR
- unexplained cyanosis

MISTAKE 5 — DESCRIBING TGA ONLY BY PARALLEL ARTERIES

Correction

Demonstrate VA discordance.

MISTAKE 6 — FINDING ONE ABNORMALITY AND STOPPING

Correction

Complete the entire segmental sequence.

49. Practical Scanning Workflow

SUBCOSTAL — ESTABLISH THE BEGINNING

- abdominal arrangement, IVC/aorta
- systemic veins, atria
- AV connections, ventricular morphology

↓

PARASTERNAL — DEFINE THE VENTRICLES AND OUTFLOWS

- Ventricular morphology
- VSD
- VA connections, great arteries
- Valves, RVOT/LVOT
- branch PAs
- coronaries where relevant

↓

APICAL — CONFIRM CONNECTIONS

- AV junction
- ventricular morphology
- AV valves
- ventricular function

↓

SUPRASTERNAL — COMPLETE THE CIRCULATION

Assess:

- arch
- descending aorta
- systemic veins where appropriate
- pulmonary veins where appropriate

50. A Simple Reporting Framework

For complex congenital disease, a structured anatomical description may begin:

Cardiac position: Levocardia.

Atrial arrangement: Usual.

Systemic venous return: SVC and IVC drain to the morphologic RA.

Pulmonary venous return: Pulmonary veins drain to the morphologic LA.

AV connections: Concordant.

Ventricular morphology: Morphologic RV right-sided; morphologic LV left-sided.

VA connections: Discordant, with the aorta arising from the morphologic RV and pulmonary artery from the morphologic LV.

Great artery relationship: Aorta anterior and rightward.

The anatomical conclusion then becomes:

Findings are consistent with complete transposition of the great arteries.
This is much more robust than beginning the report with "TGA."

51. The MASTER Approach Applied to Segmental Analysis

The framework introduced earlier can now be expanded:

M — Morphology

Identify atria, ventricles, valves, and great arteries.

A — Anatomy & Connections

Trace veins → atria → ventricles → arteries.

S — Shunts

Identify ASD, VSD, PDA, and other communications.

T — Turbulence & Pressure

Assess obstruction and pressure gradients.

E — Effect

Determine chamber loading and ventricular function.

R — Relevance

Explain the implications for management, intervention, or surgery.

52. Segmental Analysis Memory Map

For rapid recall, remember:

P–A–V–V–A–G–H
P — Position

Where is the heart?

A — Atrial arrangement

What is the atrial/situs pattern?

V — Veins

Where do systemic and pulmonary veins drain?

V — Ventricles

Which is morphologic RV and LV, and how are they connected to the atria?

A — Arteries

Which great artery arises from which ventricle?

G — Great artery relationship

How are the vessels positioned?

H — Hemodynamics

What are the physiological consequences?

Chapter Key Points

- Sequential segmental analysis is the foundation of echocardiographic assessment of complex congenital heart disease.
- **Morphology and connections are more reliable than position.**
- Begin with cardiac position and abdominal/atrial arrangement.
- Trace systemic and pulmonary venous return.
- Determine AV connections before assuming ventricular identity.
- Identify morphologic RV by features including coarse trabeculation, moderator band, and more apical AV valve insertion.
- Identify the great arteries by their **destinations and branching patterns**.
- Define VA connections before describing great artery position.
- TGA requires demonstration of **VA discordance**, not simply parallel great arteries.
- ccTGA demonstrates **double discordance: AV discordance + VA discordance**.
- DORV requires further description of VSD relationship, outflow obstruction, and great artery anatomy.
- In single-ventricle hearts and heterotaxy, strict sequential analysis becomes even more important.
- After defining anatomy, complete the assessment with shunts, obstruction, hemodynamics, and ventricular function.

MASTER PRINCIPLE

Never diagnose congenital heart disease from where structures appear. Identify what each structure is, trace where it comes from, determine where it goes, and then name the lesion.

The Sequential Segmental Formula

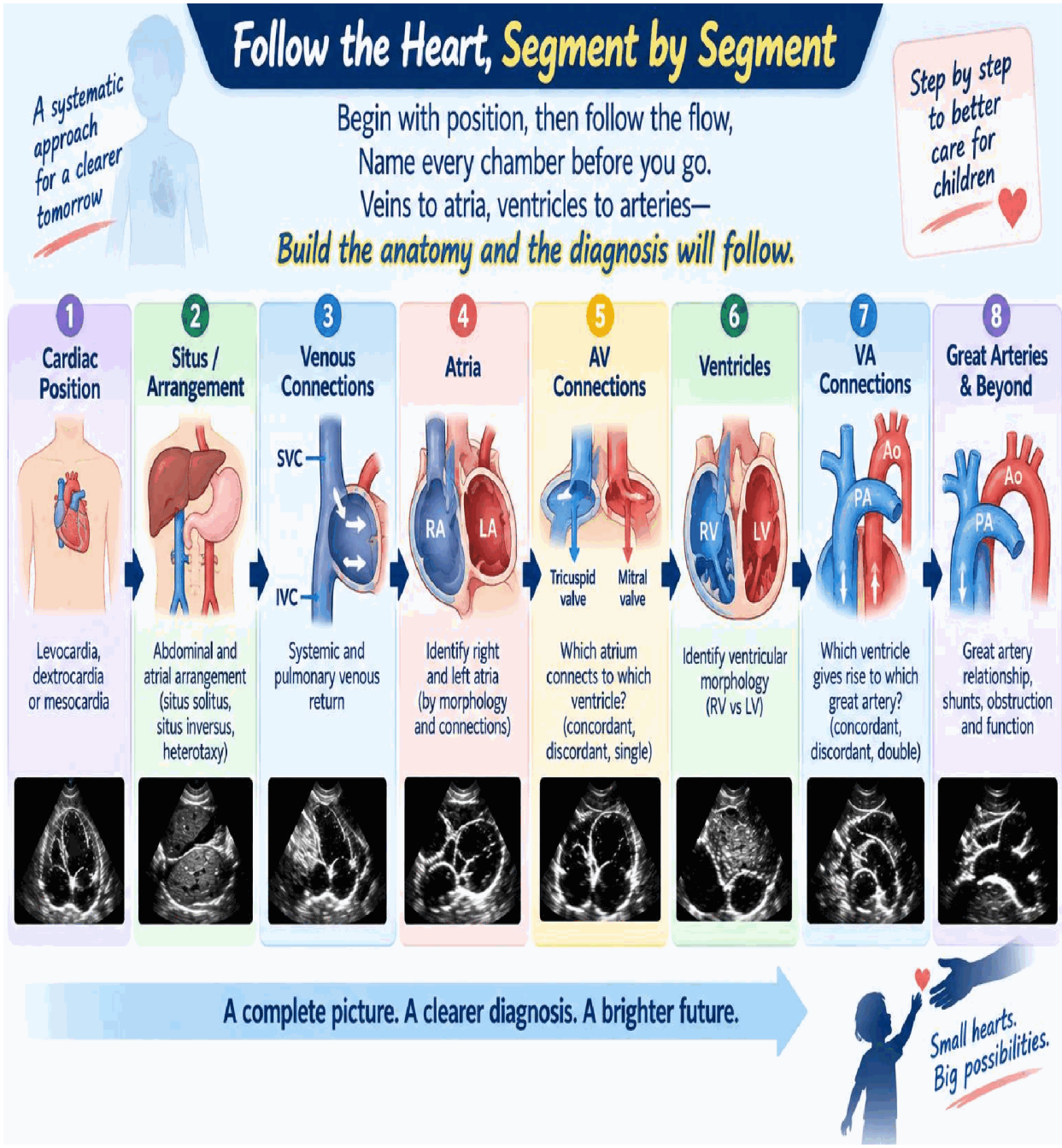
POSITION → ARRANGEMENT → VEINS → ATRIA → AV CONNECTION → VENTRICLES → VA CONNECTION → GREAT ARTERIES → SHUNTS → OBSTRUCTION → PHYSIOLOGY

Or, at its simplest:

Identify → Connect → Trace → Confirm → Diagnose

CHAPTER 6

Cardiac Measurements & Z-Scores



1. Why Measurements Matter in Children

In adults → fixed numbers matter.

In pediatrics → body size matters more than raw numbers.

A 12 mm aortic annulus:

- Normal in a neonate
- Small in a 10-year-old

That is why we use Z-scores.

2. What Is a Z-Score?

Z-score =
How many standard deviations a measurement is from normal for that body surface area (BSA).

Simple Interpretation:

Z-Score	Meaning
0	Perfectly average
+2	Upper limit normal
–2	Lower limit normal
> +2	Dilated
< –2	Small / hypoplastic

3. Body Surface Area (BSA)

Before interpreting measurements:

Calculate BSA.
Always ensure:

- Height is correct
- Weight is recent

Wrong BSA = wrong Z-score.

4. Essential Measurements in Pediatric Echo

These are mandatory in every structured study.

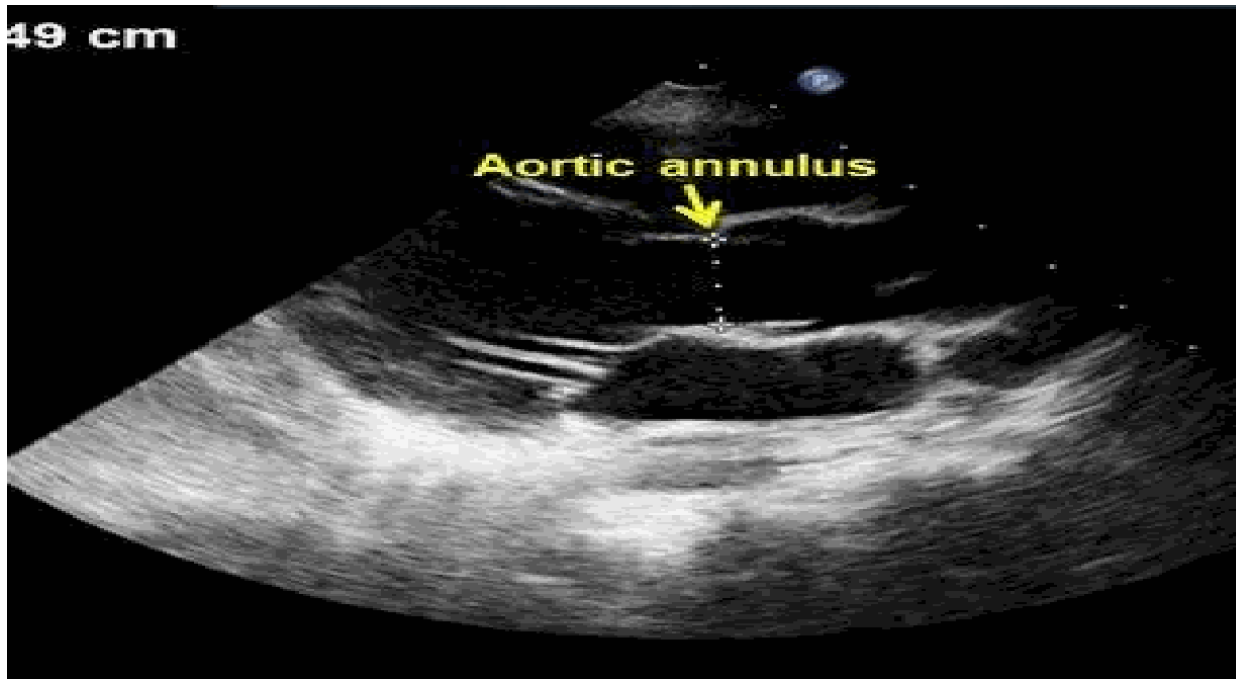
AORTIC ANNULUS

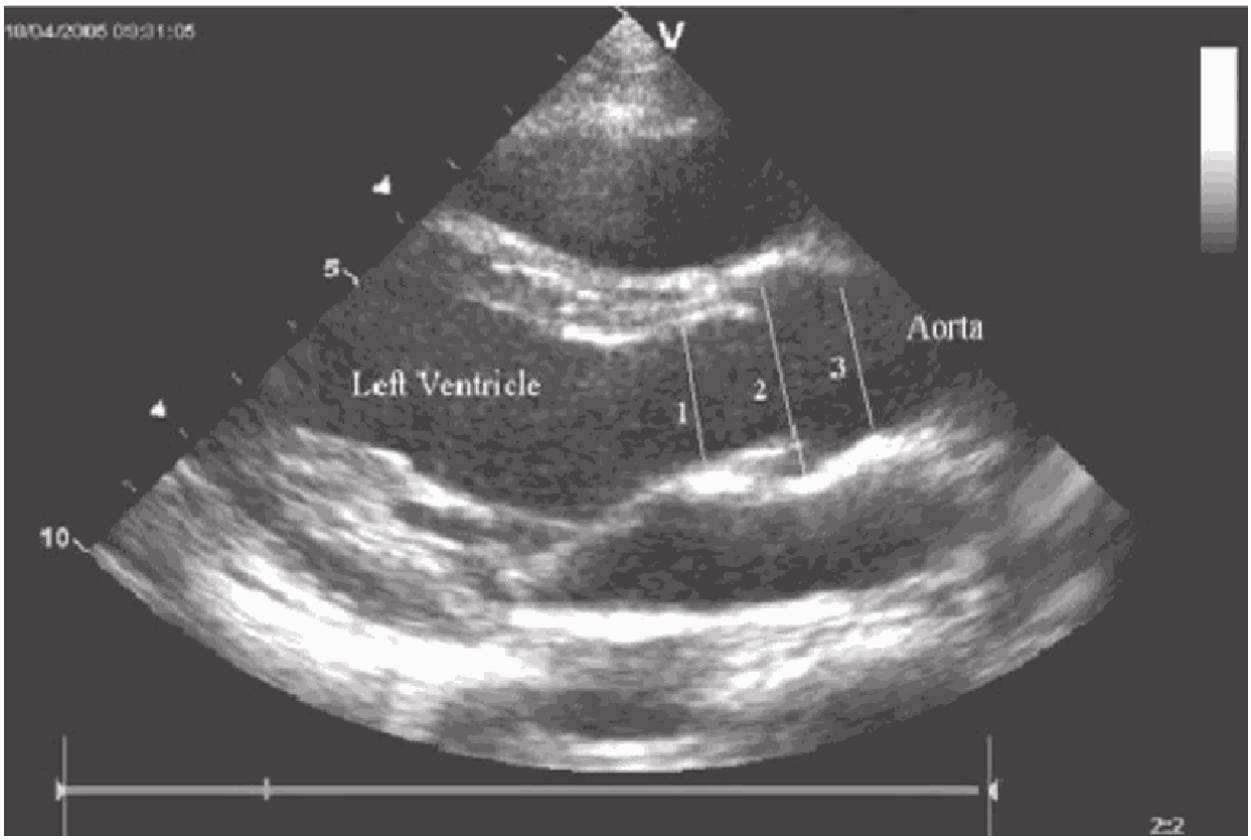
Measure in:

- Parasternal long axis
- Inner edge to inner edge
- Mid-systole

Used for:

- Balloon valvotomy planning
- TAV planning
- Subaortic obstruction evaluation





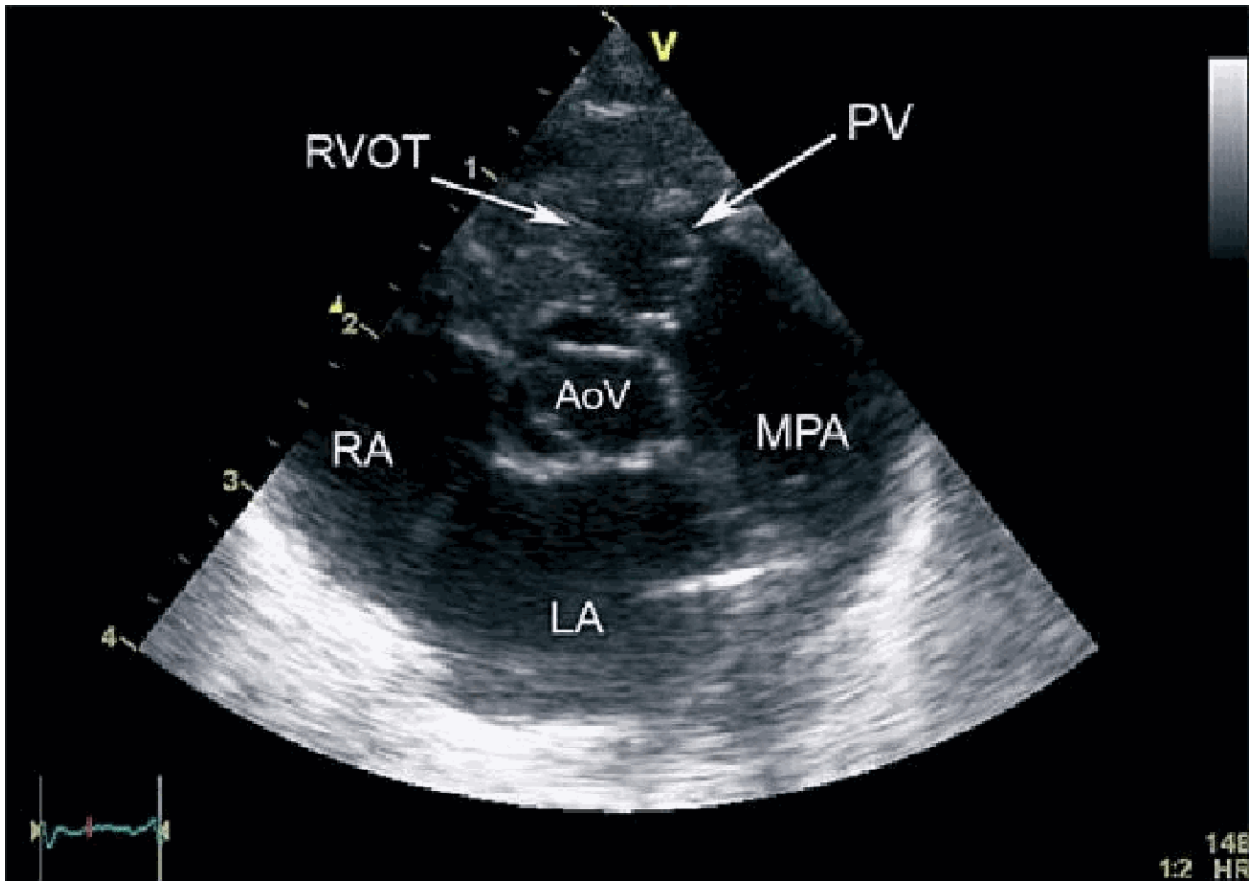
PULMONARY VALVE ANNULUS

Measure in:

- Parasternal short axis
- Systole

Important for:

- TOF repair planning
- Balloon pulmonary valvotomy
- PVI candidacy



MITRAL VALVE ANNULUS

Measure in apical 4 chamber during diastole.

Important in:

- MS
- AVSD
- LV hypoplasia

LV DIMENSIONS

Measure in PLAX:

- LVEDD (end-diastolic diameter)
- LVESD (end-systolic diameter)

Calculate:

- Fractional shortening (FS)
- Ejection fraction (EF)

Normal FS in children:

28–45%

Normal EF in children:

60–80%

AORTIC ROOT & ASCENDING AORTA

Important in:

- Marfan
- Bicuspid AV
- Connective tissue disorders

5. Interpreting Z-Scores Clinically

Example 1:

Pulmonary annulus Z = –3

→ Significant hypoplasia

→ May influence TOF surgical approach

Example 2:

Ascending aorta Z = +3

→ Dilated

→ Follow up required

6. Common Measurement Mistakes

- Measuring at wrong timing (diastole vs systole)
- Outer edge to outer edge
- Oblique plane measurement
- Not zooming
- Forgetting Z-score in report

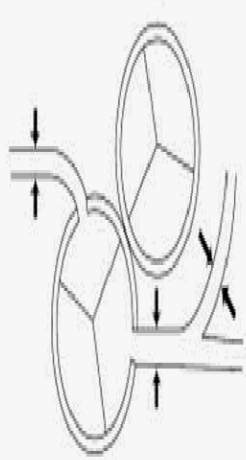
7. What Must Always Have Z-Score?

- Aortic annulus
- Pulmonary annulus
- Aortic root
- LVEDD
- RVOT (in TOF)
- Coronary arteries

8. Normal Echocardiographic Reference Values

The following reference tables provide mean values and prediction limits (2 and 3 standard deviations) indexed to body surface area (BSA), for use when calculating Z-scores.

TWO-DIMENSIONAL ECHOCARDIOGRAPHIC MEASUREMENTS OF MEAN AND PREDICTION LIMITS FOR 2 AND 3 STANDARD DEVIATIONS FOR MAJOR CORONARY ARTERY SEGMENTS*

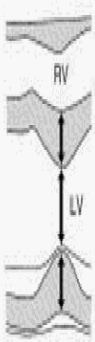


Echo Views	BSA	0.2	0.3	0.4	0.5	0.6	0.7	0.8	1.0	1.2	1.4	1.6	1.8	2.0
LAD	Mean	1.2	1.4	1.6	1.8	1.9	2.0	2.2	2.3	2.5	2.7	2.8	2.9	3.0
	Mean + 2 SD	1.5	1.8	2.1	2.3	2.5	2.7	2.8	3.0	3.3	3.5	3.7	4.0	4.2
	Mean + 3 SD	1.7	2.0	2.3	2.5	2.8	3.0	3.2	3.4	3.8	4.0	4.3	4.5	4.7
RCA	Mean	1.3	1.4	1.6	1.7	1.8	2.0	2.1	2.3	2.5	2.7	2.8	3.0	3.2
	Mean + 2 SD	1.9	2.1	2.3	2.4	2.6	2.7	2.8	3.1	3.4	3.6	3.8	4.0	4.3
	Mean + 3 SD	2.2	2.4	2.6	2.8	3.0	3.1	3.3	3.5	3.8	4.1	4.3	4.5	4.8
LMCA	Mean	1.7	1.9	2.1	2.3	2.4	2.5	2.7	2.9	3.1	3.3	3.4	3.6	3.7
	Mean + 2 SD	2.3	2.6	2.8	3.0	3.3	3.4	3.6	3.9	4.2	4.4	4.6	4.8	5.1
	Mean + 3 SD	2.7	3.0	3.2	3.4	3.7	3.9	4.0	4.3	4.7	4.9	5.2	5.5	5.8

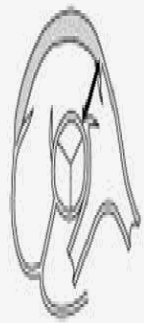
Normal Echocardiographic Values

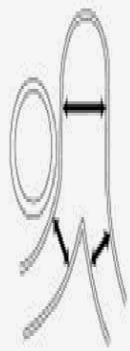
TABLE D-1

TWO-DIMENSIONAL ECHOCARDIOGRAPHY-DERIVED M-MODE MEASUREMENTS OF THE LEFT VENTRICULAR DIMENSION AND WALL THICKNESS: MEAN (-2 SD TO +2 SD)*

Echo Views	BSA	0.2	0.3	0.4	0.5	0.6	0.7	0.8	0.9	1.0	1.2	1.4	1.6	1.8	2.0	2.2
	LV EDD	19.5 (15.5–23.0)	23.0 (19.0–27.0)	26.0 (22.0–30.5)	29.5 (24.5–34.0)	31.5 (27.0–36.5)	33.5 (29.0–38.5)	35.5 (30.5–41.0)	37.5 (32.0–43.0)	39.5 (33.5–45.0)	42.0 (36.0–48.0)	45.0 (38.5–51.0)	47.0 (40.5–54.0)	49.5 (42.5–57.0)	51.5 (44.0–60.0)	53.5 (45.5–62.0)
	IVS (D)	4.5 (3.0–5.5)	5.0 (3.5–6.0)	5.0 (4.0–6.5)	5.5 (4.0–7.0)	6.0 (4.5–7.5)	6.0 (4.5–8.0)	6.5 (4.5–8.5)	7.0 (5.0–9.0)	7.0 (5.0–9.5)	8.0 (5.5–10.0)	8.5 (6.0–11.0)	9.0 (6.0–12.0)	9.5 (6.5–12.5)	10.5 (7.0–13.5)	11.0 (7.5–14.5)
	LVPW (D)	4.0 (3.0–5.0)	4.5 (3.0–5.5)	4.5 (3.5–6.0)	5.0 (3.5–6.5)	5.5 (4.0–7.0)	6.0 (4.0–7.5)	6.0 (4.5–8.0)	6.5 (4.5–8.0)	7.0 (5.0–8.5)	7.5 (5.5–9.5)	8.0 (5.5–10.0)	8.5 (6.0–11.0)	9.0 (6.5–11.5)	9.5 (7.0–12.0)	10.0 (7.5–13.0)
	LV ESD	12.0 (8.0–15.0)	15.0 (11.5–18.0)	17.0 (13.5–20.0)	18.5 (15.0–22.5)	20.0 (16.5–24.5)	21.5 (17.5–26.0)	23.0 (18.5–28.0)	24.0 (19.5–29.0)	25.5 (20.5–31.0)	28.0 (22.0–33.5)	29.5 (23.5–35.5)	31.5 (24.5–37.5)	33.0 (25.5–39.5)	34.5 (26.5–41.5)	36.0 (27.5–43.0)
	IVS (S)	6.5 (5.0–8.0)	7.0 (5.5–9.0)	7.5 (6.0–9.5)	8.0 (6.0–10.0)	8.5 (6.5–10.5)	9.0 (7.0–11.0)	9.5 (7.5–11.5)	9.5 (7.5–12.0)	10.0 (8.0–12.5)	10.5 (8.0–13.5)	11.5 (9.0–14.5)	12.0 (9.0–15.5)	12.5 (9.5–16.5)	13.5 (10.0–18.0)	14.0 (10.0–19.0)
	LVPW (S)	6.5 (5.5–8.0)	7.0 (6.0–8.5)	8.0 (6.5–9.5)	9.0 (7.0–10.5)	9.5 (7.5–11.0)	10.0 (8.0–12.0)	10.5 (8.5–12.5)	11.5 (9.0–13.5)	11.5 (9.0–14.0)	12.5 (10.0–15.0)	13.0 (10.5–16.0)	14.0 (11.0–17.5)	14.5 (11.0–18.5)	15.0 (11.5–19.5)	16.0 (12.0–20.0)

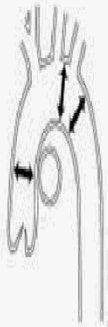
TWO-DIMENSIONAL ECHOCARDIOGRAPHIC MEASUREMENTS OF THE PULMONARY VALVE AND PULMONARY ARTERIES (IN MM): MEAN (-2 SD TO +2 SD)*



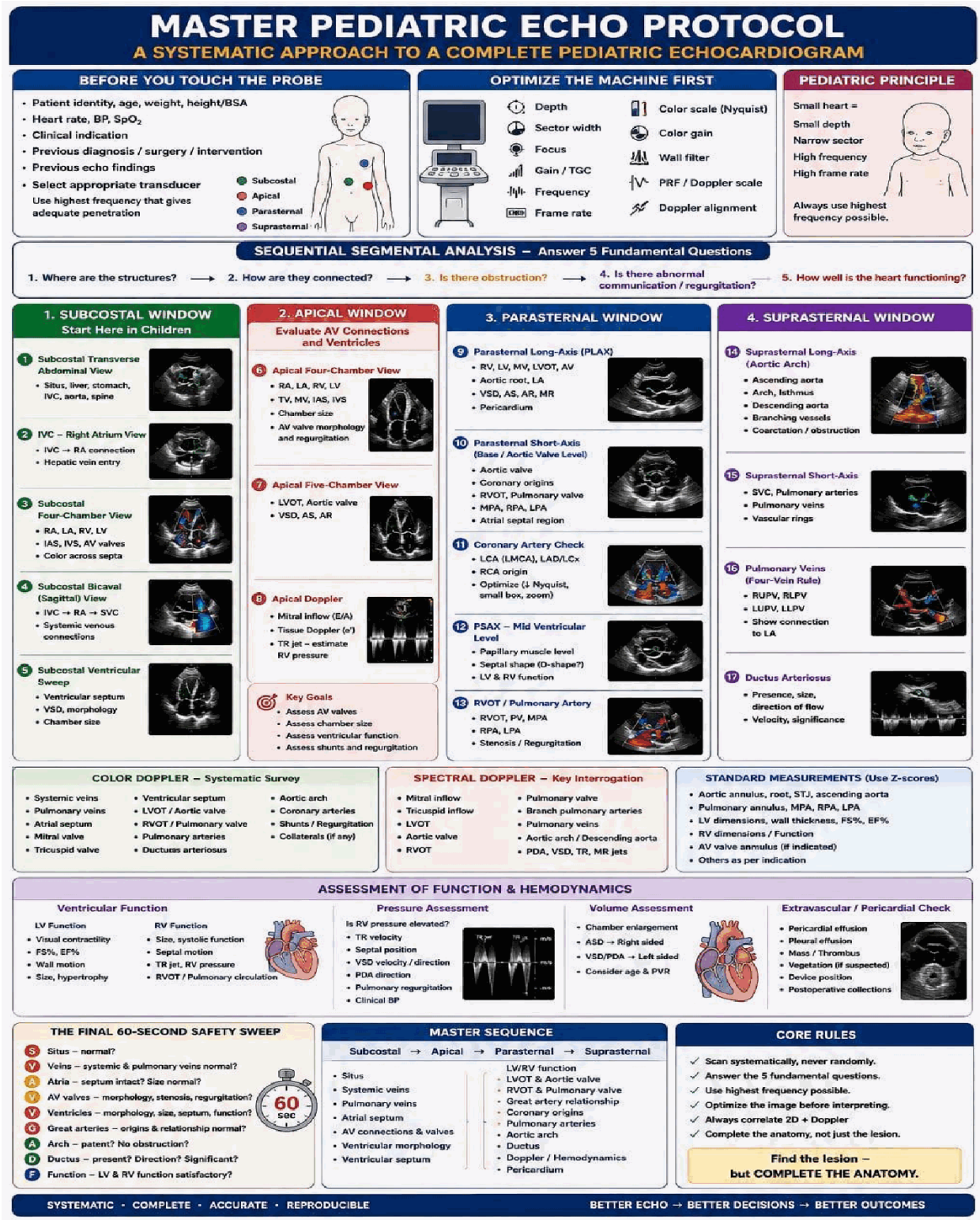


Echo Views	BSA	0.2	0.3	0.4	0.5	0.6	0.7	0.8	0.9	1.0	1.2	1.4	1.6	1.8	2.0	2.2
Pulmonary annulus		8.5 (6.0–10.5)	10.0 (8.0–12.5)	11.5 (9.0–14.0)	13.0 (10.0–16.0)	14.0 (11.0–17.5)	15.5 (11.5–19.0)	16.5 (12.0–20.5)	17.5 (13.0–21.5)	18.5 (13.5–23.0)	20.0 (15.0–25.0)	22.0 (16.0–27.5)	23.5 (17.0–29.0)	25.0 (18.0–30.5)	26.0 (19.0–33.0)	27.0 (19.5–34.0)
Main PA		7.5 (5.0–10.0)	9.0 (6.5–12.0)	10.5 (7.5–14.0)	12.0 (9.0–15.0)	13.0 (9.5–16.5)	14.0 (10.0–17.5)	15.0 (11.0–18.5)	16.0 (11.5–20.0)	17.0 (12.0–21.0)	18.5 (13.5–23.0)	20.0 (14.5–25.5)	21.0 (15.0–28.0)	22.5 (16.0–30.0)	24.0 (16.5–32.0)	25.0 (17.0–33.0)
Right PA		5.0 (3.5–7.0)	6.0 (4.5–8.0)	7.0 (5.0–9.0)	8.0 (5.5–10.0)	9.0 (6.0–11.0)	9.5 (6.5–12.0)	10.0 (7.0–13.0)	10.5 (7.5–13.5)	11.0 (8.0–14.0)	12.5 (9.0–16.0)	13.0 (9.5–17.5)	14.0 (10.0–18.5)	15.0 (10.5–20.0)	15.5 (10.5–21.0)	16.5 (11.0–22.0)
Left PA		4.5 (3.0–6.5)	5.5 (4.0–7.5)	6.5 (4.5–8.5)	7.5 (5.0–9.5)	8.0 (5.5–10.5)	9.0 (6.0–11.0)	9.5 (6.5–12.0)	10.0 (7.0–13.0)	10.5 (7.5–14.0)	11.5 (8.0–15.5)	12.5 (8.5–16.5)	13.5 (9.0–18.0)	14.0 (9.0–19.0)	15.0 (9.5–20.0)	15.5 (10.0–21.0)

TWO-DIMENSIONAL ECHOCARDIOGRAPHIC MEASUREMENTS OF AORTIC ROOT AND AORTA: MEAN (-2 SD TO +2 SD) (IN MM)*

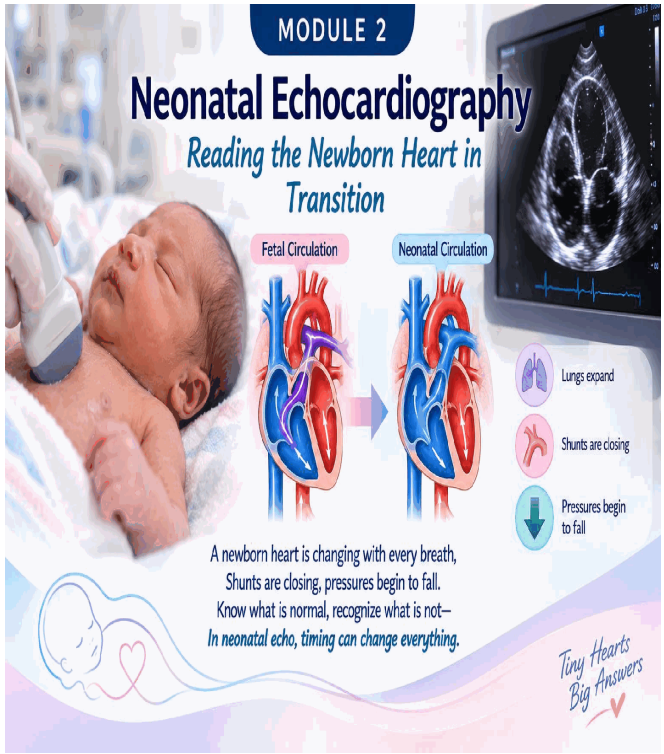
Echo Views	BSA	0.2	0.3	0.4	0.5	0.6	0.7	0.8	0.9	1.0	1.2	1.4	1.6	1.8	2.0	2.2
	Aortic annulus	7.0 (5.5–9.0)	8.5 (7.0–10.0)	10.0 (8.0–12.0)	11.0 (9.0–13.5)	12.0 (10.0–14.5)	13.5 (11.0–15.5)	14.0 (11.5–16.5)	15.0 (12.5–17.5)	15.5 (13.0–18.5)	17.5 (14.5–20.5)	18.5 (15.0–22.0)	20.0 (16.0–23.5)	21.0 (17.0–25.0)	22.0 (18.0–26.0)	23.0 (18.5–27.5)
	Sinus of Valsalva	9.5 (7.0–12.0)	11.5 (9.0–14.0)	13.0 (10.0–16.0)	14.5 (11.5–17.5)	16.0 (13.0–19.5)	17.5 (14.0–21.0)	18.5 (15.5–22.0)	19.5 (15.5–23.5)	20.5 (16–25.0)	22.0 (18.0–27.0)	24.0 (19.0–30.0)	25.5 (20.0–31.5)	27.0 (21.0–33.5)	28.5 (22.0–35.5)	30.5 (23.0–38.5)
	Sinotubular junction	8.0 (6.0–10.0)	10.0 (7.5–12.0)	11.0 (9.0–13.5)	12.5 (10.0–15.0)	14.0 (11.0–16.5)	15.0 (12.0–18.0)	16.0 (12.5–19.0)	16.5 (13.0–20.5)	17.5 (14.0–21.5)	19.5 (15.5–24.0)	21.0 (16.5–26.0)	22.0 (17.0–27.5)	24.0 (18.0–29.0)	25.0 (19.0–31.0)	26.0 (20.0–32.0)
	Ascending aorta	8.0 (5.5–11.0)	10.0 (7.0–13.0)	11.5 (8.5–15.0)	13.0 (10.0–16.0)	14.5 (11.0–17.5)	15.5 (12.0–19.0)	16.5 (13.0–20.5)	17.5 (14.0–21.5)	18.5 (14.5–23.0)	20.5 (15.5–25.5)	22.0 (16.5–27.5)	24.0 (18.0–29.5)	25.5 (19.0–31.0)	26.5 (20.0–33.0)	28.0 (21.0–35.0)
	Transverse aorta	6.5 (4.0–8.5)	8.0 (5.5–10.0)	9.5 (6.0–13.0)	10.5 (8.0–13.0)	11.5 (9.0–14.5)	12.5 (9.5–15.5)	13.0 (10.0–17.0)	14.0 (11.0–18.0)	15.0 (11.5–19.0)	17.0 (12.5–20.5)	18.0 (14.0–22.0)	19.5 (15.0–24.0)	20.5 (15.5–25.5)	21.5 (16.0–27.0)	22.5 (17.0–28.5)
	Aortic isthmus	3.5 (3.0–7.5)	6.5 (4.0–9.0)	7.5 (5.0–10.0)	8.5 (6.0–11.0)	9.5 (6.5–12.5)	10.5 (7.0–13.5)	11.0 (7.5–14.5)	12.0 (8.0–15.5)	12.5 (8.5–16.0)	13.5 (9.5–17.5)	15.0 (10.0–19.5)	16.0 (10.5–21.0)	17.0 (11.0–22.0)	17.5 (11.5–23.5)	18.0 (12.0–25.0)

Summary of Pediatric Echo Protocol



MODULE 2

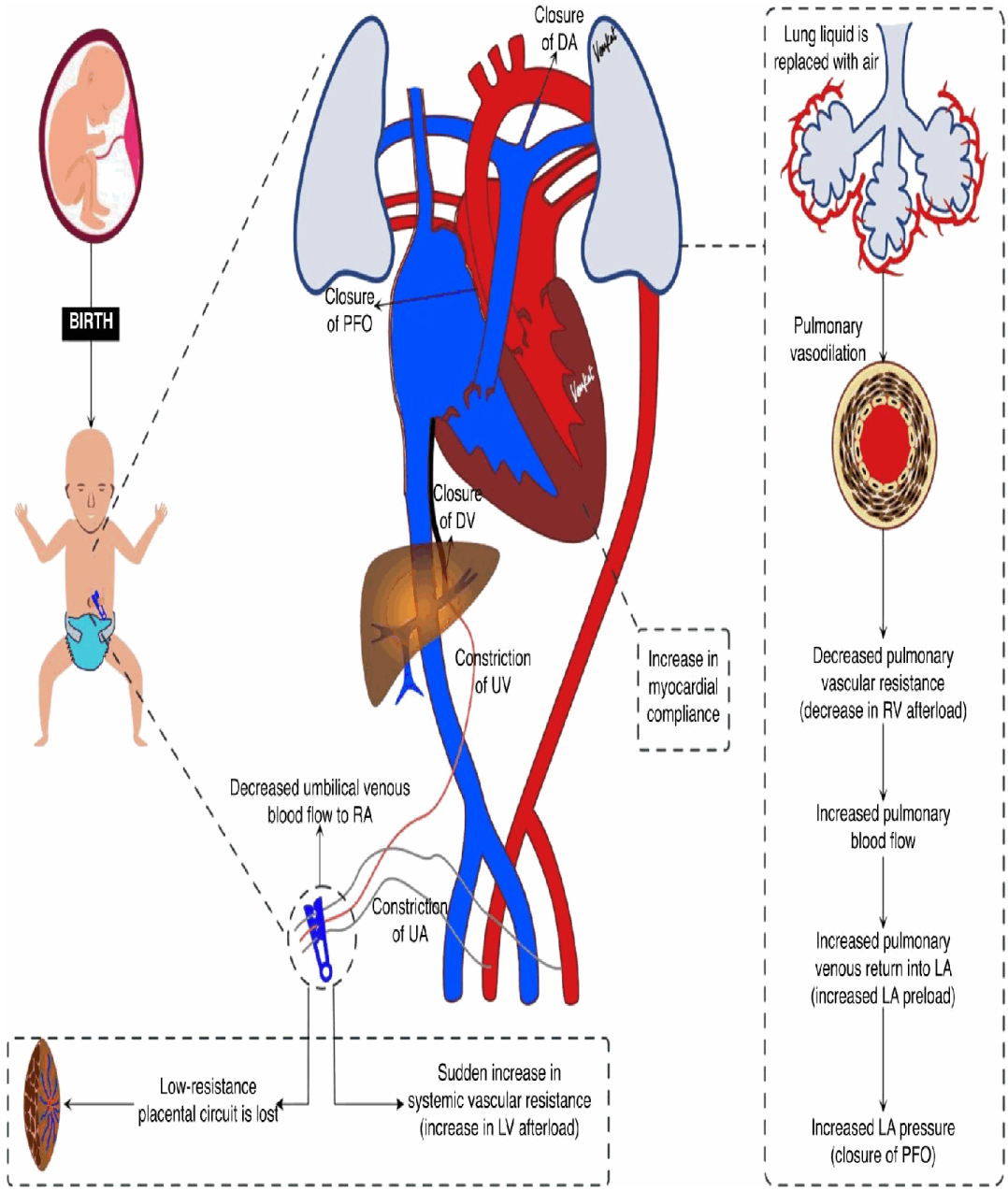
Neonatal Echocardiography



Chapter	Topic
7	The Transitional Neonatal Circulation
8	Normal Neonatal Echo
9	Duct-Dependent Systemic Circulation
10	Duct-Dependent Pulmonary Circulation
11	Persistent Pulmonary Hypertension of the Newborn
12	Neonatal Coarctation of the Aorta
13	Patent Ductus Arteriosus

CHAPTER 7

The Transitional Neonatal Circulation



Understanding the Heart Before Interpreting the Echo

1. The Fetal Circulation

The fetal lungs do not perform gas exchange. Oxygenated blood comes from the placenta.

Three fetal pathways are particularly important:

I. Ductus venosus

Allows a proportion of umbilical venous blood to bypass the liver.

II. Foramen ovale

Allows blood to pass: **RA → LA**

III. Ductus arteriosus

Allows blood from the pulmonary artery to enter the descending aorta:

These pathways are normal and essential before birth.

2. What Happens at Birth?

The circulation changes dramatically when:

The umbilical cord is clamped

Placental circulation is removed.

Systemic vascular resistance rises.

When the newborn breathes lungs expand and oxygenation increases.

Pulmonary vascular resistance falls.

Pulmonary blood flow increases

More blood returns through the pulmonary veins to the LA.

Left atrial pressure rises

The flap of the foramen ovale is pushed toward functional closure.

The ductus begins to constrict

The circulation progressively changes from fetal parallel to normal postnatal serial circulation.

3. Pulmonary Vascular Resistance

Pulmonary vascular resistance is high in fetal life.

After birth it progressively decreases as the lungs:

- expand
- oxygenate
- recruit pulmonary vessels

This changes:

- RV pressure
- PA pressure
- interventricular septal shape
- ductal shunt direction
- atrial shunting
- TR velocity

These changes must be considered when interpreting neonatal Doppler findings.

4. The Patent Foramen Ovale

A PFO is expected in many newborns.

Color Doppler may demonstrate atrial-level flow.

The important questions are not simply:

"Is there a PFO?"

Instead ask:

- What is the direction of flow?
- Is the communication restrictive?
- Is adequate mixing required?
- Is there evidence of elevated right atrial pressure?
- Is the atrial septum abnormal?

5. Atrial Shunt Direction

Depending on transitional physiology, atrial flow may be:

- predominantly left-to-right
- right-to-left
- bidirectional

A small amount of right-to-left flow early after birth does not automatically indicate structural heart disease.

Interpret it alongside:

- clinical oxygenation
- RV pressure
- ductal flow
- septal shape
- pulmonary vascular resistance

6. The Ductus Arteriosus

The ductus is one of the most important structures in neonatal echocardiography.

Always assess:

- presence
- size
- direction of flow
- velocity
- restriction
- relationship to PA and descending aorta

Possible patterns include:

Right-to-left

PA → Aorta
May occur with high pulmonary pressure or when systemic output depends on ductal flow.

Left-to-right

Aorta → PA
Becomes increasingly common as pulmonary vascular resistance falls.

Bidirectional

May occur during early transition or with abnormal pressure relationships.

7. The Ductus Is a Physiological Clue

Do not treat the PDA merely as an anatomical structure.

Its Doppler pattern provides information about:

Pulmonary pressure ↔ Systemic pressure
For example:

Predominantly right-to-left ductal flow

should prompt consideration of:

- high pulmonary vascular resistance
- severe pulmonary hypertension
- duct-dependent systemic circulation

The surrounding anatomy determines which explanation is correct.

8. Interventricular Septal Shape

The interventricular septum provides another useful clue to pressure relationships.

Normal LV

Approximately circular in short axis.

Increased RV Pressure

Septal flattening may produce: **D-shaped LV**

Severe RV Pressure Loading

The septum may bow toward the LV.

9. Estimating Pulmonary Pressure

No single echo measurement should be interpreted in isolation.

Useful clues include:

TR velocity

When an adequate TR jet is present:

$\Delta P = 4V^2$

This can contribute to estimation of RV systolic pressure when RA pressure is considered.

PDA flow

Direction and velocity provide information about PA–aortic pressure relationships.

Septal shape

Provides a qualitative assessment of RV/LV pressure relationship.

Pulmonary artery Doppler

May provide additional physiological information.

Atrial shunting

Right-to-left or bidirectional flow may support elevated right-sided pressure.

Use multiple concordant signs rather than one number.

10. The Transitional RV

The RV is the dominant ventricle during fetal life and initially remains relatively prominent after birth.

Therefore, a newborn may normally have:

- relatively prominent RV dimensions
- increased RV wall thickness compared with an older child

11. The Transitional LV

After birth, increased pulmonary blood flow increases pulmonary venous return to the LA and LV. The LV becomes responsible for the entire systemic output.

In some congenital lesions, this transition reveals previously concealed obstruction.

Examples include: critical aortic stenosis, coarctation

12. Why Duct Closure Can Be Dangerous

In a normal newborn, ductal closure is part of normal transition.

In some congenital lesions, however, the ductus is maintaining life.

Two major physiological groups must be recognized.

Duct-Dependent Systemic Circulation

The PDA helps provide systemic blood flow.

Examples include severe left-sided obstructive lesions such as:

- critical coarctation, interrupted aortic arch, critical aortic stenosis
- HLHS

As the ductus constricts:

Systemic perfusion falls → acidosis → shock

13. Duct-Dependent Pulmonary Circulation

The PDA supplies or supplements pulmonary blood flow.

Examples include:

- pulmonary atresia, critical pulmonary stenosis, severe TOF/RVOT obstruction
- selected complex single-ventricle lesions

As the ductus constricts:

Pulmonary blood flow falls → cyanosis worsens

14. Mixing-Dependent Circulation

Some neonatal lesions depend primarily on adequate mixing between otherwise poorly communicating circulations.

The classic example is: **Transposition of the great arteries**

The echo must urgently assess:

- PFO/ASD, atrial shunt
- PDA, VSD if present

A restrictive atrial communication in TGA can represent a critical physiological problem even when ventricular function appears preserved.

15. Three Questions About Every Neonatal PDA

Whenever a ductus is seen, ask:

1. Which way is blood flowing?
2. Why is it flowing that way?
3. What happens if this ductus closes?

16. Transitional Finding or Congenital Disease?

A neonatal echocardiographer frequently faces this distinction.

Finding	May Be Transitional	May Indicate Pathology
PFO	Common	Restrictive mixing lesion
PDA	Expected early	Duct-dependent circulation/significant PDA
Elevated RV pressure	Early transition	PPHN/CHD
Bidirectional PDA	Early transition	Severe pulmonary/systemic pathology
Septal flattening	Transitional PVR	Significant RV pressure loading
Mild TR	May occur	Structural TV disease/high RV pressure
Right-to-left atrial flow	Transitional	PPHN/obstruction/mixing physiology

The distinction requires:

Age + anatomy + Doppler + physiology + clinical context

17. The Neonatal Echo Mindset

For every newborn, ask:

A — Anatomy

Is the heart structurally normal?

B — Blood flow

Where is blood going?

C — Connections

Are the circulations connected appropriately?

D — Ductus

What role is the PDA playing?

E — Effect

How are the ventricles and organs being perfused?

THE MOST IMPORTANT NEONATAL PRINCIPLE

A technically beautiful echocardiogram is insufficient if it fails to identify the physiology that threatens the newborn.

Therefore:

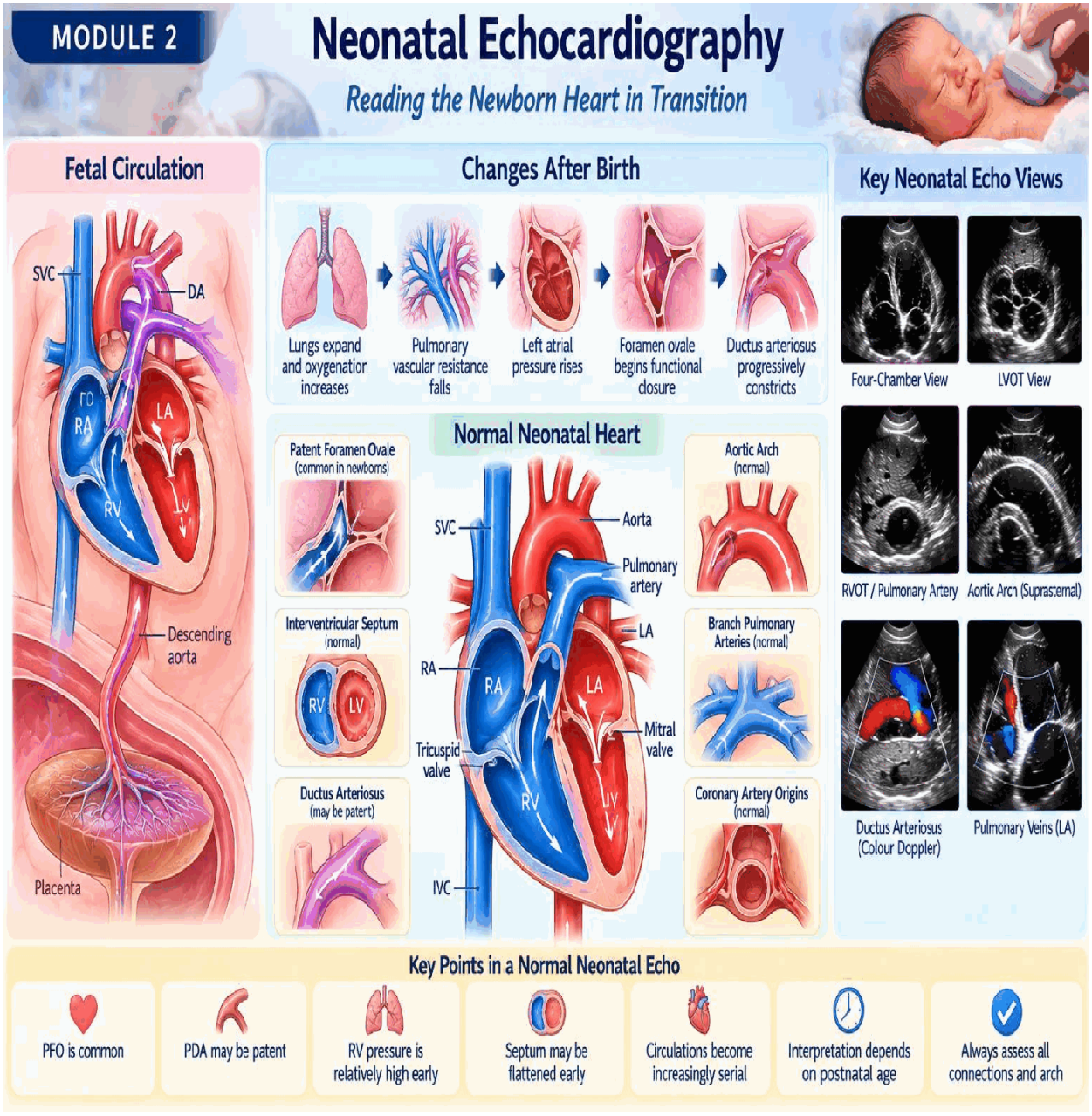
In neonatal echocardiography, anatomy tells you the lesion—but physiology tells you the urgency.

MASTER PRINCIPLE

Never interpret a neonatal echo without asking where the circulation is in its transition from fetal to postnatal life.

CHAPTER 8

Normal Neonatal Echocardiography



Learning Objectives

By the end of this chapter, the learner should be able to:

1. Recognize structurally normal neonatal heart on echo
2. Distinguish normal transitional physiology from congenital pathology
3. Perform a systematic neonatal echocardiography study
4. Assess the PFO and PDA appropriately on echo
5. Interpret neonatal right-sided pressure and septal configuration on echo

1. What Is a "Normal" Neonatal Echo?

A normal neonatal echocardiogram is not necessarily an echocardiogram in which every fetal communication has already disappeared.

During the first hours and days after birth, it may be entirely expected to find:

- a patent foramen ovale
- a patent ductus arteriosus
- relatively prominent right ventricle
- relatively elevated pulmonary artery pressure
- changing atrial and ductal shunt patterns
- transitional septal configuration

Therefore, neonatal echo interpretation requires three pieces of information:

ANATOMY + PHYSIOLOGY + POSTNATAL AGE

A finding that is expected shortly after birth may require a different interpretation if it persists or is associated with abnormal physiology.

2. Start With the Clinical Context

Before placing the probe, know:

- gestational age/postnatal age
- oxygen saturation, respiratory support
- blood pressure and perfusion
- reason for echocardiography
- prenatal cardiac findings if available

In a premature infant, also consider the degree of prematurity and whether the study is being performed for structural assessment, PDA evaluation, pulmonary hypertension, or hemodynamic instability.

NEONATAL ECHO PEARL

Always document postnatal age when interpreting transitional findings.

3. A Systematic Normal Neonatal Study

Use the same anatomical discipline for a healthy-looking newborn as for a baby with suspected congenital heart disease.

A practical sequence is:

Situs → Veins → Atria → AV connections → Ventricles → VA connections → Valves → Septa → Outflows → Pulmonary arteries → Ductus → Pulmonary veins → Aortic arch → Function → Hemodynamics

Do not stop simply because the four-chamber view appears normal.

Several critical neonatal lesions can have a deceptively normal four-chamber appearance.

4. Step 1 — Cardiac Position and Situs

Begin with the subcostal abdominal view.

Establish:

- cardiac position
- apex direction
- abdominal arrangement
- relationship of IVC and aorta to the spine

In the usual arrangement:

- heart is generally left-sided
- apex points leftward
- IVC is usually right of the spine
- descending aorta lies left of the spine

Then trace the IVC toward the heart.

IMPORTANT PRINCIPLE

Do not infer atrial arrangement solely from the direction of the cardiac apex.

5. Step 2 — Systemic Venous Return

Demonstrate the systemic veins entering the appropriate atrium.

Assess:

SVC

Follow the SVC into the right atrium.

IVC

Follow the IVC through the diaphragm into the right atrium.

Hepatic veins

Identify their drainage when clinically relevant.

Also look for indirect clues to venous abnormalities.

For example, a markedly dilated coronary sinus should prompt consideration of a persistent left superior vena cava.

6. Step 3 — Pulmonary Venous Return

Pulmonary venous anatomy deserves deliberate assessment even when the rest of the heart appears normal.

Whenever feasible, demonstrate pulmonary venous return to the left atrium.

Useful windows include:

- subcostal
- apical
- parasternal
- suprasternal
- modified right parasternal views

Color Doppler can assist visualization, but settings must be optimized for relatively low-velocity flow.

CRITICAL PEARL

A normal four-chamber view does not prove normal pulmonary venous connection. If pulmonary venous drainage is uncertain, continue imaging rather than assuming normality.

7. Step 4 — Atrial Anatomy

In the normal neonatal heart:

- systemic venous return reaches the morphologic RA
- pulmonary venous return reaches the morphologic LA
- the atrial septum separates the atria
- the foramen ovale may remain functionally patent

The atria should be assessed for:

- size
- septal anatomy
- PFO
- additional atrial communication
- shunt direction

8. The Patent Foramen Ovale

A PFO is a common normal finding in newborns.

The foramen ovale is not equivalent to a true secundum ASD.

A PFO represents persistence of the fetal communication between the septum primum flap and surrounding atrial septal structures.

On echo, assess:

- flap valve
- size of communication
- color-flow direction
- restriction where clinically relevant

NORMAL TRANSITIONAL FINDING

A small atrial communication through the PFO in an otherwise normal newborn is usually part of postnatal transition rather than congenital septal deficiency.

9. PFO Versus ASD

Feature	PFO	Secundum ASD
Basic mechanism	Incomplete functional/anatomic fusion of fetal flap tissue	True deficiency of atrial septal tissue
Flap valve	Usually present	May be deficient
Neonatal occurrence	Common	Structural lesion
Shunting	Depends on atrial pressure relationship	Usually becomes L → R as PVR falls
Interpretation	Often transitional/physiological	Congenital heart defect

ECHO PEARL

Do not label every neonatal interatrial color jet as an ASD. First identify the anatomy of the atrial septum and flap valve.

10. Atrial Shunting During Transition

Atrial shunt direction depends on the pressure relationship between the two atria.

Flow may be: Left-to-right or Bidirectional

and transient right-to-left components can occur depending on early transitional physiology.

Persistent or clinically significant right-to-left atrial shunting should prompt evaluation of:

- pulmonary hypertension
- right-sided obstruction
- abnormal pulmonary venous return
- ventricular dysfunction
- other congenital heart disease

11. Step 5 — Atrioventricular Connections

In a normal heart:

Morphologic RA → Tricuspid valve → Morphologic RV
and

Morphologic LA → Mitral valve → Morphologic LV
This is: AV concordance

Confirm the connection rather than assuming it from chamber position.

12. Normal AV Valve Appearance

Assess both valves for:

- leaflet morphology, mobility, insertion, opening, regurgitation

The tricuspid valve normally inserts slightly more apically than the mitral valve.

This normal AV valve offset is useful for recognizing ventricular morphology.

13. Physiological Tricuspid Regurgitation

A small degree of TR may be seen in otherwise normal neonates.

It can also provide useful hemodynamic information if the spectral envelope is complete and technically adequate.

However, significant TR should prompt assessment of:

- RV pressure
- tricuspid valve morphology
- Ebstein anomaly
- RV dysfunction
- pulmonary hypertension

IMPORTANT PRINCIPLE

A Doppler jet should always be interpreted with valve anatomy and hemodynamics.

14. Step 6 — Ventricular Morphology

Identify the ventricles by morphology.

Morphologic RV

Features include:

- coarse trabeculations
- moderator band
- more apical tricuspid insertion

Morphologic LV

Features include:

- smoother septal surface
- finer trabeculation
- mitral valve insertion relatively more basal

In the normal neonatal heart, the RV is right-sided and the LV left-sided, but morphology remains the correct basis for identification.

15. The Normal Neonatal Right Ventricle

One of the major differences between neonatal and later pediatric echocardiography is the appearance of the RV.

The fetal RV performs a major share of combined ventricular output.

After birth, pulmonary vascular resistance does not fall instantaneously.

Consequently, early neonatal echo may show:

- relatively prominent RV
- relatively thick RV wall
- elevated RV pressure relative to later infancy
- transitional septal flattening

IMPORTANT PITFALL

Do not apply the appearance of an older child's RV to a newborn without considering transitional physiology.

16. Interventricular Septal Configuration

The parasternal short-axis view is particularly useful.

A normal low-pressure RV allows the LV to maintain a relatively circular short-axis shape.

With increasing RV pressure:

RV pressure rises

↓

Septum flattens

↓

LV becomes increasingly D-shaped

In the early neonatal period, some septal flattening may reflect transitional pulmonary pressure.

Persistent or marked flattening requires explanation.

17. Step 7 — Ventricular Septum

The ventricular septum should be examined systematically in multiple planes.

Assess:

- inlet septum
- perimembranous region
- outlet septum
- muscular septum
- apical septum

Use: 2D + systematic sweeps + color Doppler

A single normal four-chamber image does not exclude a VSD.

ECHO PEARL

Sweep the ventricular septum from base to apex.

18. Step 8 — Ventriculoarterial Connections

In the normal heart:

Morphologic RV → Pulmonary artery

and

Morphologic LV → Aorta

This represents: VA concordance

Trace each vessel rather than identifying it by position alone.

19. Identifying the Normal Great Arteries

Pulmonary Artery

Identify it by demonstrating:

Main PA → Right PA + Left PA

Aorta

Identify it by following:

Ascending aorta → Arch → Descending aorta

The normal great arteries have a crossing rather than parallel relationship.

20. Normal Pulmonary Valve and Pulmonary Arteries

Assess:

- pulmonary valve morphology
- leaflet mobility
- pulmonary annulus
- main PA, right PA, left PA

Color Doppler should show unobstructed antegrade flow.

Spectral Doppler should not show evidence of significant obstruction.

Branch pulmonary artery velocities should be interpreted in the context of age, vessel size, and transitional physiology.

21. Transitional Branch Pulmonary Artery Flow

Some neonates, particularly smaller infants, may demonstrate relatively increased velocities in the branch pulmonary arteries as the pulmonary circulation adapts after birth.

This must be distinguished from fixed anatomical branch pulmonary artery stenosis.

Assess:

- vessel anatomy
- caliber
- location of acceleration
- Doppler pattern
- symmetry
- evolution on follow-up where appropriate

Velocity alone should not replace anatomical assessment.

22. Normal Aortic Valve and LVOT

Assess:

- LVOT
- aortic valve morphology
- valve opening
- annulus
- aortic root
- ascending aorta

The LVOT should be unobstructed.

The aortic valve should open freely without significant stenosis or regurgitation.

In short axis, carefully evaluate valve morphology rather than relying on one frozen frame.

23. Coronary Artery Origins

Whenever technically feasible, particularly in a comprehensive neonatal structural examination, assess the proximal coronary origins.

The parasternal short-axis view at the aortic root is commonly useful.

Optimize using:

- high-frequency imaging
- shallow depth
- narrow sector
- focus at the aortic root
- zoom where appropriate

Demonstrate the anatomical origin in 2D where possible.

IMPORTANT PRINCIPLE

Color near the aortic root does not by itself establish a normal coronary origin.

24. The Normal Neonatal Aortic Arch

The suprasternal window should demonstrate:

- ascending aorta
- transverse arch
- head and neck vessels where feasible
- isthmus
- descending thoracic aorta

Assess:

- continuity
- caliber
- focal narrowing
- flow pattern

Color Doppler should show unobstructed flow without focal pathological acceleration.

25. Never Skip the Arch

A neonate may have:

- apparently normal four chambers, normal ventricular function

and still have important arch obstruction.

Therefore:

No comprehensive neonatal structural echo is complete without assessment of the aortic arch.

This becomes particularly important when there are:

- weak femoral pulses
- upper/lower limb blood pressure differences
- LV dysfunction
- prenatal suspicion of coarctation

26. The Patent Ductus Arteriosus

The ductus arteriosus is commonly patent shortly after birth.

Its presence alone does not indicate disease.

The important assessment includes:

- size
- morphology
- direction of flow
- velocity
- restriction
- physiological effect

Possible flow patterns include: Right-to-left, Bidirectional, Left-to-right

These patterns evolve as pulmonary vascular resistance falls.

27. Ductal Flow in Normal Transition

Early after birth, pulmonary artery pressure may remain relatively high.

Ductal flow can therefore initially be bidirectional or include a right-to-left component.

As pulmonary vascular resistance decreases:

Aortic pressure > Pulmonary artery pressure
and ductal flow typically becomes increasingly:

Aorta → Pulmonary artery
before functional closure.

KEY PRINCIPLE

Ductal direction must always be interpreted according to postnatal age and the rest of the circulation.

28. What Does a PDA Tell You?

The PDA can function as a natural hemodynamic window into the pressure relationship between systemic and pulmonary circulations.

When assessing ductal flow, ask:

1. Which direction is the blood flowing?
2. How restrictive is the ductus?
3. What does this suggest about pulmonary versus systemic pressure?
4. Is the ductus merely transitional—or is the circulation dependent on it?

This last question is crucial.

29. The Normal Neonatal Aortic Isthmus and the Open Ductus

The presence of a PDA can complicate assessment of the aortic isthmus.

In suspected coarctation, an open ductus may partially maintain flow through the region and reduce the apparent Doppler gradient.

Therefore:

A low gradient does not exclude neonatal coarctation.
Assess:

- arch dimensions
- isthmus
- posterior shelf where present
- ductal anatomy
- flow direction
- LV size/function
- serial evolution when clinically required

This is one of the most important neonatal diagnostic traps.

30. Normal Ventricular Function

Both ventricles should be assessed for

- size
- wall motion
- systolic function
- loading conditions

Interpretation must account for the rapidly changing preload and afterload of neonatal transition.

NEONATAL PEARL

Do not interpret ventricular function without asking what loading conditions the ventricle is facing.

31. Estimating RV and Pulmonary Pressure

Several echocardiographic findings can be integrated.

TR Velocity

When a reliable TR jet is present:

$$\Delta P = 4V^2$$

The RV–RA gradient can contribute to estimation of RV systolic pressure.

Interventricular Septum

Assess septal configuration in short axis.

PDA

Assess direction and velocity of ductal flow.

Atrial Shunt

Evaluate direction of flow.

No single measurement should be used alone. Integrate multiple hemodynamic clues.

32. Normal Neonatal Pulmonary Pressure Is Dynamic

Immediately after birth, pulmonary pressure remains relatively high and then falls with transition.

Therefore, findings that may be compatible with early transition include:

- elevated estimated RV pressure
- mild septal flattening
- bidirectional ductal flow
- variable atrial shunting

The interpretation changes when these findings:

- are severe, persist beyond the expected transition
- accompany hypoxemia and ventricular dysfunction

33. Normal Pericardium

The pericardial space should be assessed from multiple views.

There should be no significant pericardial effusion.

A tiny amount of fluid may occasionally be visible, but apparent echo-free spaces should be distinguished from:

- pleural fluid
- normal anatomical spaces
- artifact

Do not diagnose effusion from a single view.

35. Normal Transitional Findings Versus Red Flags

Finding	Can Be Transitional	Red Flag
PFO	Common	Restrictive communication when mixing is essential
PDA	Common early	Duct-dependent circulation or hemodynamically significant shunt
Bidirectional PDA	May occur early	Persistent R→L flow with hypoxemia/abnormal anatomy
Prominent RV	Expected early	Severe dilation or dysfunction
Elevated RV pressure	Expected during transition	Persistent systemic/suprasystemic pressure
Septal flattening	May occur early	Marked/persistent flattening
Small TR jet	May occur	Significant TR/abnormal TV
Interatrial shunting	Common	Persistent significant R→L shunting
Branch PA acceleration	May be transitional	Fixed focal narrowing
Open arch with PDA	May appear reassuring	Coarctation can still be present

Ten Questions Before Calling a Neonatal Echo "Normal"

Before concluding that a neonatal study is normal, ask:

1. Is situs and cardiac position established?
2. Have systemic venous connections been demonstrated?
3. Have pulmonary venous connections been adequately assessed?

4. Are AV connections concordant?
5. Are both ventricles morphologically normal and appropriately sized?
6. Are VA connections concordant?
7. Are the ventricular and atrial septa adequately assessed?
8. Have both outflow tracts and valves been examined?
9. Have the ductus and aortic arch been assessed?
10. Are the physiology and ventricular function appropriate for the baby's postnatal age?

If one of these remains unanswered, the study may be incomplete rather than normal.

37. The "Normal Four-Chamber" Trap

One of the most important lessons in neonatal echocardiography is:

Normal four chambers ≠ Normal heart

Potentially serious lesions may have a relatively reassuring four-chamber appearance, including:

- TGA
- coarctation
- interrupted aortic arch in selected appearances
- anomalous pulmonary venous connections
- coronary abnormalities
- some outflow tract abnormalities

Therefore:

Always complete the connections, great arteries, pulmonary veins, ductus, and arch.

38. The "It Is Just PPHN" Trap

A cyanotic neonate may demonstrate:

- right-to-left atrial shunting
- right-to-left ductal shunting
- elevated RV pressure

These findings can occur with pulmonary hypertension.

But similar physiology may accompany structural congenital heart disease.

Before concluding PPHN, systematically exclude:

- TGA, TAPVR
- pulmonary atresia
- critical pulmonary stenosis
- left-sided obstruction

Master Rule: PPHN is a physiological diagnosis

39. The “Low Gradient Means No Coarctation” Trap

In the presence of an open ductus:

- pressure relationships may be altered
- the ductus may support descending aortic flow
- significant anatomical narrowing may produce little gradient

Therefore:

In neonatal coarctation, anatomy can be more important than the instantaneous gradient.

This concept will be developed in detail in the dedicated coarctation chapter.

41. Echo Pearls for the Normal Neonatal Study

Pearl 1: Normality depends partly on postnatal age.

Pearl 2: A PFO is common; an ASD is a structural tissue deficiency.

Pearl 3: A PDA may be normal, pathological, or essential for survival.

Pearl 4: The neonatal RV may normally appear more prominent than in an older child.

Pearl 5: Early pulmonary pressures may remain relatively high.

Pearl 6: Interpret TR velocity, septal shape, PDA flow, and atrial shunting together.

Pearl 7: Do not call pulmonary venous return normal unless it has been adequately assessed.

Pearl 8: A normal four-chamber view does not exclude critical CHD.

Pearl 9: A low arch gradient with an open PDA does not exclude coarctation.

Pearl 10: Before calling an echo normal, make sure it is complete.

Chapter Key Points

- A normal neonatal echocardiogram must be interpreted according to postnatal age and transitional physiology.
- PFO and PDA are commonly encountered during normal neonatal transition.
- Pulmonary vascular resistance and RV pressure are relatively high after birth and normally decline.
- The neonatal RV may appear relatively prominent compared with the RV of an older child.
- Ductal and atrial shunt direction can change during transition.
- Establish systemic and pulmonary venous connections rather than assuming them.
- Confirm AV and VA concordance.
- Examine the ventricular septum systematically in multiple planes.
- Evaluate both outflow tracts, pulmonary arteries, coronary origins where feasible, and the complete aortic arch.
- The PDA should be interpreted according to its direction, restriction, pressure relationship, and physiological role.
- An open ductus can conceal important aortic arch obstruction.
- Ventricular function must be interpreted in the context of neonatal loading conditions.
- A structurally normal-looking four-chamber view does not exclude critical congenital heart disease.
- A study should not be called normal until the major connections and great vessels have been adequately demonstrated.

MASTER PRINCIPLE

In the newborn, “normal” is dynamic. Know the anatomy, understand the transition, and interpret every shunt in the context of time.

The Normal Neonatal Echo Formula

STRUCTURE + CONNECTIONS + SHUNTS + PRESSURES + FUNCTION + POSTNATAL AGE = MEANINGFUL INTERPRETATION

34. What Should a Normal Neonatal Echo Report Include?

A structured report should document:

Position & Situs

- cardiac position
- abdominal/atrial arrangement

Venous Connections

- systemic venous return, pulmonary venous return

Connections

- AV connection, VA connection

Septa

- atrial septum/PFO
- ventricular septum

Valves

- morphology
- stenosis/regurgitation

Great Vessels

- pulmonary arteries
- ductus arteriosus
- aortic arch

Ventricles

- size
- systolic function

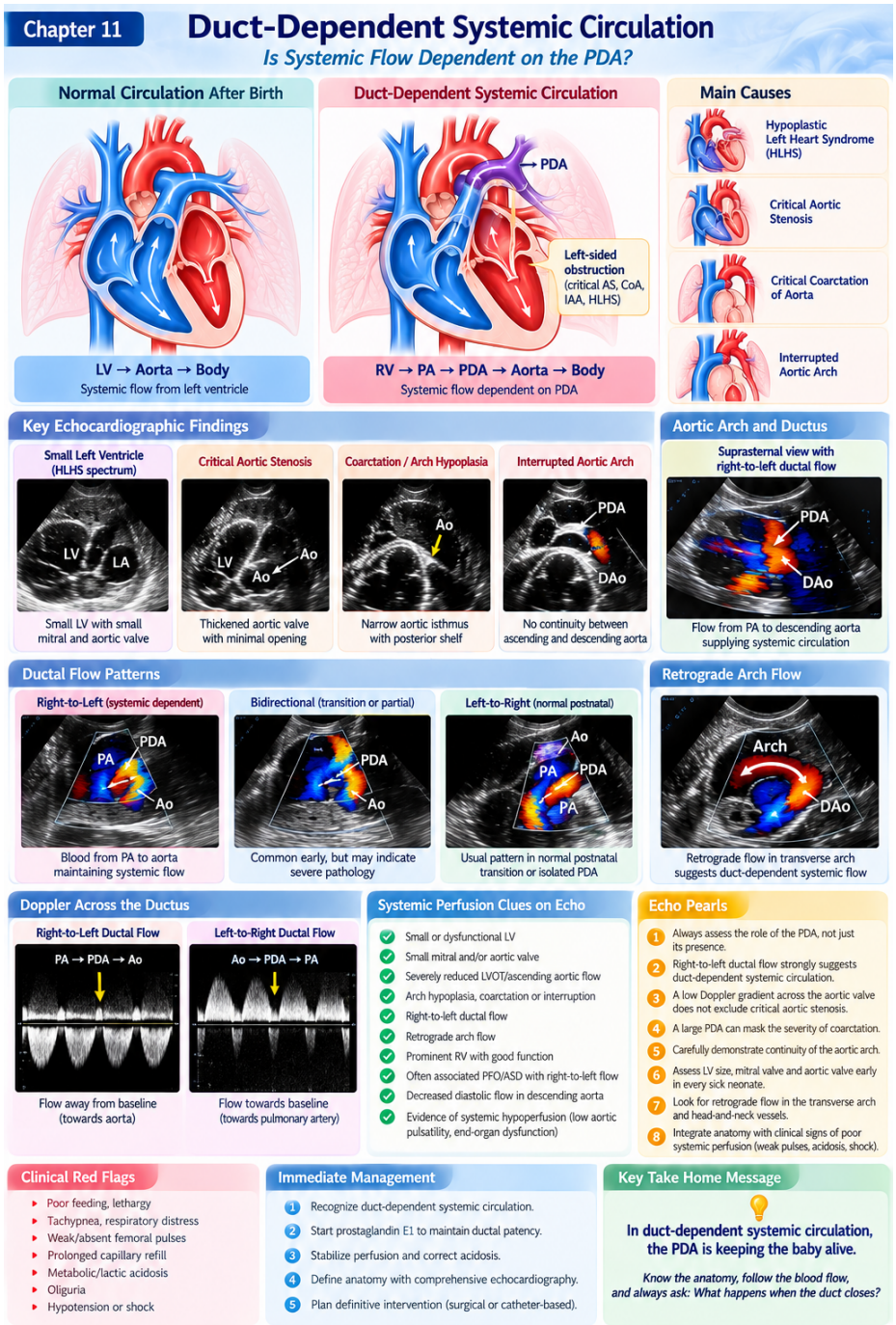
Hemodynamics

- shunt direction
- estimated RV/PA pressure where relevant

Conclusion:

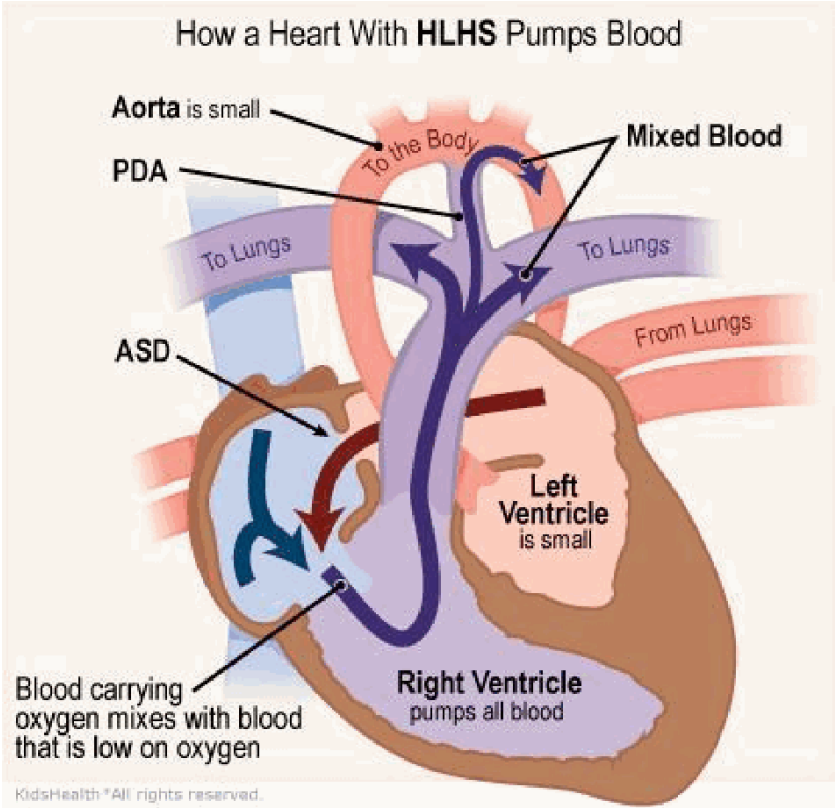
CHAPTER 9

Duct-Dependent Systemic Circulation



Learning Objectives

By the end of this chapter, the reader should be able to recognize duct-dependent systemic circulation on neonatal echocardiography; identify HLHS, critical aortic stenosis, critical coarctation/arch hypoplasia, and interrupted aortic arch; determine whether systemic perfusion is being supplied through the PDA; recognize retrograde flow in the aortic arch or ascending aorta; understand why Doppler gradients can underestimate severe obstruction; and perform a focused emergency neonatal echo when systemic perfusion is poor.



1. The Question That Changes the Scan

In a critically ill neonate, finding a PDA is not enough.

The echocardiographer must answer:

Is the PDA contributing essential blood flow to the systemic circulation?

Normally:

LV → Aortic valve → Ascending aorta → Arch → Descending aorta → Body

In duct-dependent systemic circulation:

RV → Pulmonary artery → PDA → Descending aorta → Body

Depending on the severity of left-sided obstruction, ductal blood may also travel retrogradely through the arch toward the head-and-neck vessels and ascending aorta.

This produces the central physiological concept:

PA → PDA → Aorta = Think systemic ductal dependency

2. Lesions to Look For

The major anatomical substrates are:

Lesion	Key Echo Problem
HLHS	LV/mitral/aortic structures cannot support systemic output
Critical aortic stenosis	Severe obstruction of LV ejection
Critical coarctation	Duct may bypass severe isthmic obstruction
Severe arch hypoplasia	Antegrade systemic flow is inadequate
Interrupted aortic arch	No direct aortic continuity to descending aorta
Borderline left heart	Uncertain ability of LV to support systemic circulation

ECHO PEARL

Do not start with the PDA. Start with the systemic pathway.

Trace:

LV → LVOT → Aortic valve → Ascending Ao → Transverse arch → Isthmus → Descending Ao

3. The Five Echo Questions

A practical neonatal examination can be reduced to five questions.

1. Can the LV generate systemic output?

Assess LV size, systolic function, geometry and endocardial appearance.

2. Can blood enter and leave the LV?

Assess mitral valve, LVOT and aortic valve.

3. Is there an uninterrupted systemic pathway?

Trace the entire aortic arch.

4. Where is descending aortic blood coming from?

Use color Doppler to distinguish antegrade arch flow from ductal supply.

5. What happens if the PDA closes?

This is the final physiological question.

If the answer is **systemic perfusion will deteriorate**, the circulation is duct-dependent.

4. Step 1 — Assess the Left Ventricle

Do not simply report:

"LV small."

Ask:

Is the LV capable of supporting systemic circulation?

Assess:

- LV length and width, LV cavity, systolic function
- mitral valve size, mitral inflow
- LVOT, aortic annulus, ascending aortic dimensions
- presence of endocardial fibroelastosis

A small LV is a **finding**, not a physiological diagnosis. Always interpret LV size together with:

Mitral valve + Aortic valve + LVOT + Ascending aorta + Arch + Ductal flow

5. Step 2 — Examine the Mitral Valve

In a borderline left heart, the mitral valve gives important information about the adequacy of the left-sided circulation.

Evaluate:

Annulus → leaflets → chordae → papillary muscles → inflow

Measure the annulus and use an appropriate pediatric **Z-score** where available.

Look for:

- mitral stenosis, supramitral ring
- hypoplastic annulus
- abnormal papillary muscles, parachute morphology
- abnormal chordal attachments

ECHO PEARL

Do not judge LV adequacy from ventricular dimensions alone. A borderline LV with a severely hypoplastic mitral valve is physiologically very different from a similar-sized LV with satisfactory inflow.

6. Step 3 — Critical Aortic Stenosis

Evaluate the valve in 2D before relying on spectral Doppler.

CRITICAL ECHO PEARL

A low Doppler gradient does NOT exclude critical aortic stenosis.

Gradient is flow-dependent.

If LV output is severely reduced, velocity across a critically stenotic valve may also be low.

Therefore:

Low flow + severe obstruction = deceptively low gradient
Do not equate:

Low gradient = mild stenosis
Instead integrate:

Valve morphology + LV function + antegrade aortic flow + ductal direction

7. Step 4 — Follow the Entire Aortic Arch

The suprasternal examination is mandatory.

Do not stop after seeing an apparently normal ascending aorta.

Trace:

Ascending Ao → proximal transverse arch → distal transverse arch → isthmus → descending Ao
Then identify:

PDA → descending Ao
Use both 2D and color Doppler.

ECHO PEARL
Never diagnose or exclude neonatal coarctation from a single sagittal image. Perform a slow sweep through the entire arch.

8. Coarctation — The Ductus Can Hide the Disease

This is one of the most important neonatal echo traps.

A large PDA may partially bypass the obstructed isthmus and make the coarctation appear less significant.

Therefore, severe coarctation may coexist with:

- relatively low Doppler velocity
- apparently reasonable descending aortic flow
- palpable femoral pulses
- only modest upper–lower limb pressure difference

while the duct remains widely patent.

ECHO PEARL
A low coarctation gradient in a neonate with a large PDA does not exclude critical coarctation. Look at anatomy, not merely velocity.

Assess:

- transverse arch diameter
- isthmus diameter
- posterior shelf
- long-segment narrowing
- ductal insertion
- flow direction
- Doppler pattern distal to the obstruction

9. The Isthmus–PDA Junction

This deserves deliberate imaging.

The region where the ductus joins the descending aorta can be anatomically complex.

A wide ductal ampulla may temporarily maintain lower-body perfusion and obscure the true severity of coarctation.

TECHNIQUE TIP

From the suprasternal notch:

Find arch → narrow sector → optimize depth → identify PDA → sweep through isthmus → add color → obtain PW/CW Doppler

Do not allow a large color box to hide the anatomy.

10. Step 5 — Determine Ductal Flow Direction

This is one of the highest-yield observations in the entire examination.

Normal postnatal tendency

Aorta → PDA → PA

Duct-dependent systemic circulation

PA → PDA → Aorta

The second pattern means the right ventricle is contributing to systemic perfusion.

ECHO PEARL

Right-to-left ductal flow + left-sided obstruction = systemic ductal dependency until proven otherwise.

But direction must always be interpreted with neonatal physiology because pulmonary vascular resistance can also produce right-to-left or bidirectional ductal flow.

Therefore, never diagnose systemic dependency from ductal direction **alone**.

11. Follow the Blood Backward

This is an excellent advanced echo habit.

Place color on the descending aorta.

Ask:

Where did this blood come from?
If it came through the PDA:

Descending Ao ← PDA ← PA ← RV
Then examine the arch.

If flow proceeds from the ductal region backward toward the transverse arch:

Descending Ao → transverse arch → ascending Ao
the systemic circulation is being supplied retrogradely.

ECHO PEARL

Do not simply describe: "Retrograde arch flow present." Interpret it: retrograde arch flow indicates inadequate antegrade contribution from the left ventricle and should trigger immediate evaluation for severe left-sided obstruction.

12. Retrograde Ascending Aortic Flow — A Major Clue to HLHS

In severe HLHS with aortic atresia, the tiny ascending aorta may essentially function as a conduit supplying the coronary arteries.

The neonatal cardiology resource describes an ascending aorta that may measure only **1–2 mm**, with Doppler demonstrating **retrograde flow**; it also emphasizes evaluating RV function, tricuspid regurgitation, atrial restriction, ductal restriction and the aortic isthmus.

The pathway may be conceptualized as:

RV → PA → PDA → Arch → Ascending Ao → Coronary arteries

ECHO PEARL

In a neonate with a tiny LV and aortic atresia: always put color Doppler on the ascending aorta. The direction of flow provides crucial physiological information.

13. HLHS — Do Not Stop After Seeing the Small LV

Once HLHS is recognized, the examination has only begun.

Systematically evaluate:

A. Mitral valve

Stenotic or atretic?

B. Aortic valve

Stenotic or atretic?

C. Ascending aorta

Measure carefully.

D. Aortic arch

Hypoplasia/coarctation?

E. PDA

Size and restriction.

F. Atrial septum

Is pulmonary venous egress unrestricted?

G. RV

Size and systolic function.

H. Tricuspid valve

Severity of regurgitation.

I. Pulmonary veins

Look for consequences of atrial restriction.

The reference describes the classic echo appearance as a diminutive/fibrotic LV, severely hypoplastic aortic valve and diminutive ascending aorta.

14. The Atrial Septum in HLHS

This can determine how sick the baby is.

Pulmonary venous blood enters the LA but cannot proceed normally through the left-sided circulation.

It must therefore cross:

LA → atrial communication → RA

A restrictive atrial communication causes:

LA hypertension → pulmonary venous hypertension → pulmonary edema → severe respiratory compromise

ECHO PEARL

In HLHS, assess two lifelines: the PDA for systemic output and the atrial communication for pulmonary venous decompression.

15. Pulmonary Venous Doppler Can Reveal Atrial Restriction

Do not assess the atrial septum only by measuring its opening.

Interrogate pulmonary venous flow.

Abnormal pulmonary venous Doppler, particularly prominent reversal associated with impaired LA egress, can indicate significant atrial restriction.

ECHO PEARL

Small ASD/PFO + high Doppler velocity + abnormal pulmonary venous Doppler = think restrictive atrial septum.

This can be more physiologically meaningful than defect diameter alone.

16. Interrupted Aortic Arch — Prove Continuity

IAA creates another major imaging trap.

A ductus connecting the pulmonary artery to the descending aorta can be mistaken for continuation of the arch.

Therefore ask:

Can I actually trace the ascending aorta continuously into the descending aorta?

ECHO PEARL

Do not diagnose continuity because two vascular structures appear to meet. Trace the vessel.

Identify:

Ascending aorta → head-and-neck branches → transverse arch → descending aorta

Then separately identify:

PA → PDA → descending aorta

17. Use the Head-and-Neck Vessels as Landmarks

When the arch anatomy is confusing, branch vessels are extremely useful.

Determine:

- where the first branch arises,
- how many branches are seen,
- where continuity is lost,
- where the ductus joins the descending aorta.

ECHO PEARL

In suspected interrupted arch:

Do not merely look for the interruption—map the branches.
This helps define the level of interruption and prevents the PDA from being mistaken for the distal arch.

18. Do Not Forget the Abdominal Aorta

Move to a subcostal sagittal view and obtain PW Doppler from the abdominal descending aorta.

This provides a downstream window into systemic hemodynamics.

Look at:

- systolic velocity
- pulsatility
- diastolic flow
- abnormal continuous forward flow
- flow reversal where relevant
- reduced pulsatility

ECHO PEARL

When the arch image is uncertain, distal systemic Doppler can provide important physiological evidence.
But distal Doppler does **not replace direct arch imaging**.

19. The Borderline Left Heart — Anatomy Plus Physiology

Sometimes the diagnosis is not classic HLHS.

The LV may be moderately small, with combinations of:

- mitral stenosis
- aortic stenosis
- subaortic narrowing
- arch hypoplasia
- coarctation

The important question becomes:

Can this LV ultimately support systemic circulation?

The neonatal reference emphasizes careful assessment of the mitral valve, LV dimensions, subaortic region, aortic arch and isthmus in these patients. It also notes that systolic retrograde arch flow suggests that the LV cannot adequately supply even the upper-body circulation.

ECHO PEARL

Do not make a univentricular-versus-biventricular judgment from LV size alone.

Integrate:

Mitral valve + LV volume + LV function + LVOT + Aortic valve + Arch + Flow direction

20. Ductal Restriction — The Lifeline May Be Closing

Finding a PDA does not mean it is adequate.

Assess:

- diameter, morphology
- color jet, spectral velocity, direction
- systolic/diastolic pattern

A small restrictive PDA in a systemic-dependent circulation can produce rapid deterioration.

ECHO PEARL

“PDA present” is an anatomical statement. “PDA adequate for systemic perfusion” is a physiological conclusion.

The 60-Second Emergency Echo

For the collapsing neonate:

LOOK

LV

↓

Mitral valve

↓

LVOT

↓

Aortic valve

↓

Ascending aorta

↓

Arch + isthmus

↓

PDA

↓

Ductal direction

↓

Retrograde arch flow?

↓

Atrial septum

↓

RV function + TR

Then ask:

Where is the systemic blood coming from?

Critical Pitfalls

Pitfall 1

"PDA is present, therefore systemic circulation is secure."

Correction: Determine whether it is large enough and how blood is flowing through it.

Pitfall 2

"The aortic stenosis gradient is only 25 mmHg."

Correction: Severe LV dysfunction can produce a low gradient across critical obstruction.

Pitfall 3

"There is no significant coarctation gradient."

Correction: A large PDA can bypass/decompress the obstruction.

Pitfall 4

"The descending aorta has flow, so the arch must be intact."

Correction: That blood may be coming through the PDA.

Pitfall 5

"HLHS diagnosed—scan complete."

Correction: Define atrial restriction, PDA, arch, ascending aorta, RV function and TR.

Pitfall 6

"Bidirectional PDA is normal neonatal transition."

Correction: It may be transitional, but in a sick infant it must trigger assessment of both pulmonary pressure and systemic outflow obstruction.

Reporting the Study

Avoid:

"Large PDA with right-to-left flow."

Prefer a physiological report:

Severe left-sided systemic outflow obstruction with predominant right-to-left ductal flow supplying the descending aorta. Retrograde flow is present within the transverse/ascending aorta, consistent with duct-dependent systemic circulation.

For coarctation:

Hypoplastic transverse arch and isthmus with juxtaductal narrowing. A large PDA supplies the descending aorta. Doppler gradient across the coarctation should not be used in isolation to assess severity while the ductus remains widely patent.

This tells the treating team what the echo **means**, not merely what it shows.

MASTER PRINCIPLE

Never simply document that the PDA is open. Determine what the PDA is doing.

If the pathway is:

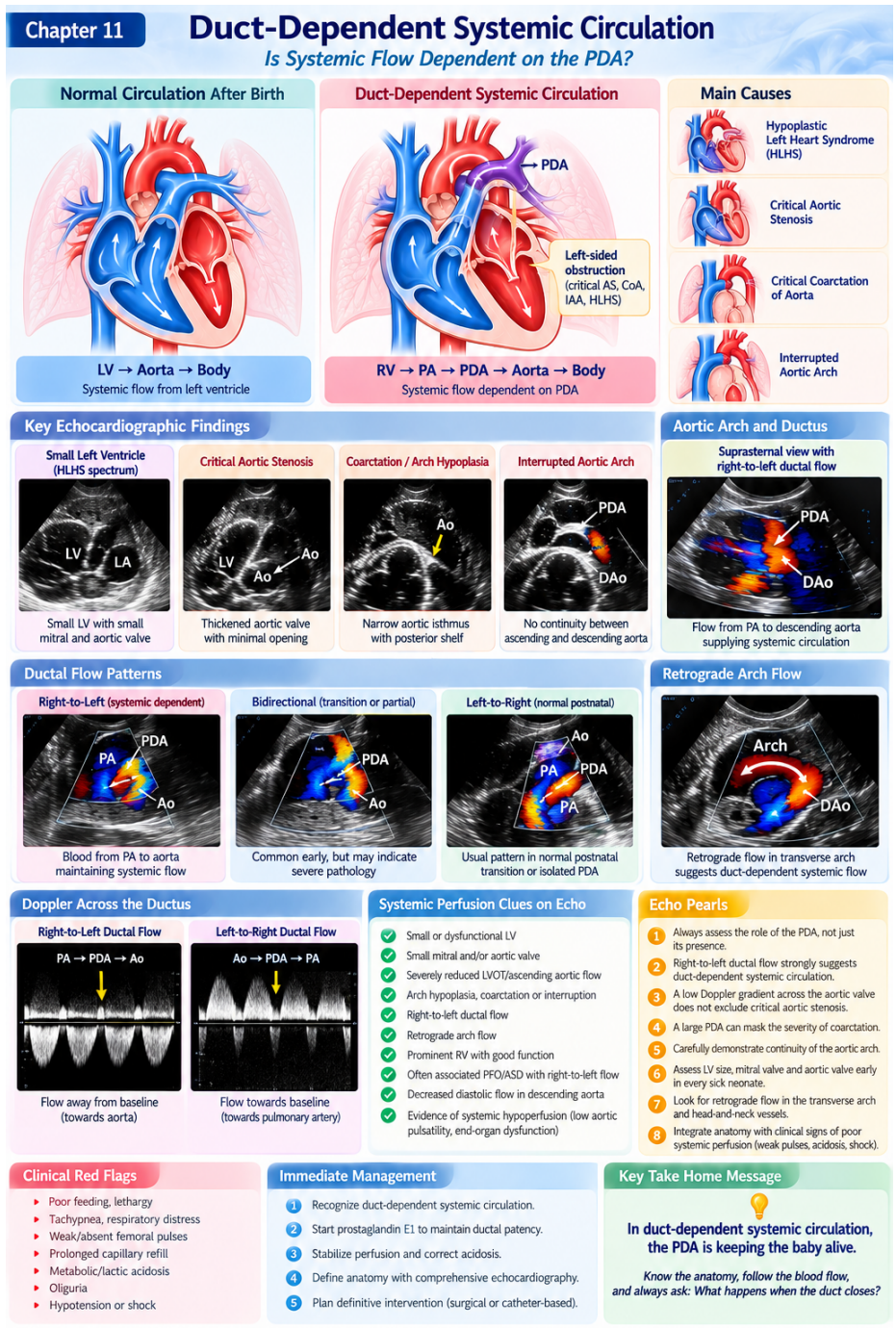
RV → PA → PDA → Aorta → Body

the ductus is functioning as a **systemic lifeline**.

And if the duct closes before the underlying obstruction is recognized, a seemingly stable newborn can rapidly progress to profound systemic hypoperfusion.

Mastery formula:

ANATOMY → FLOW DIRECTION → SYSTEMIC PERFUSION → DUCTAL DEPENDENCY



The Critical Question: Is Systemic Flow Dependent on the PDA?

In the sick neonate, one of the most important questions during echocardiography is not simply

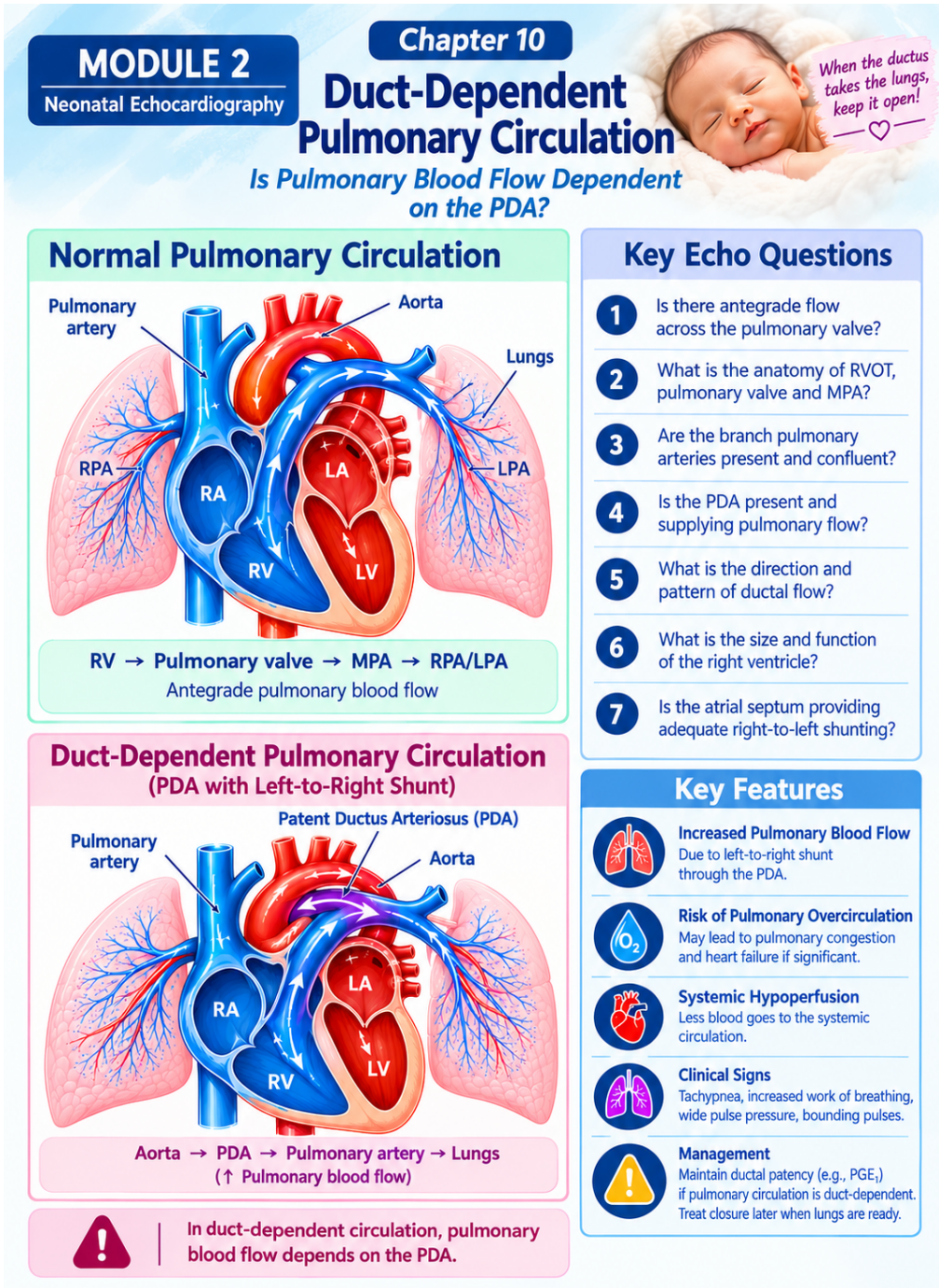
"Is the PDA open?"

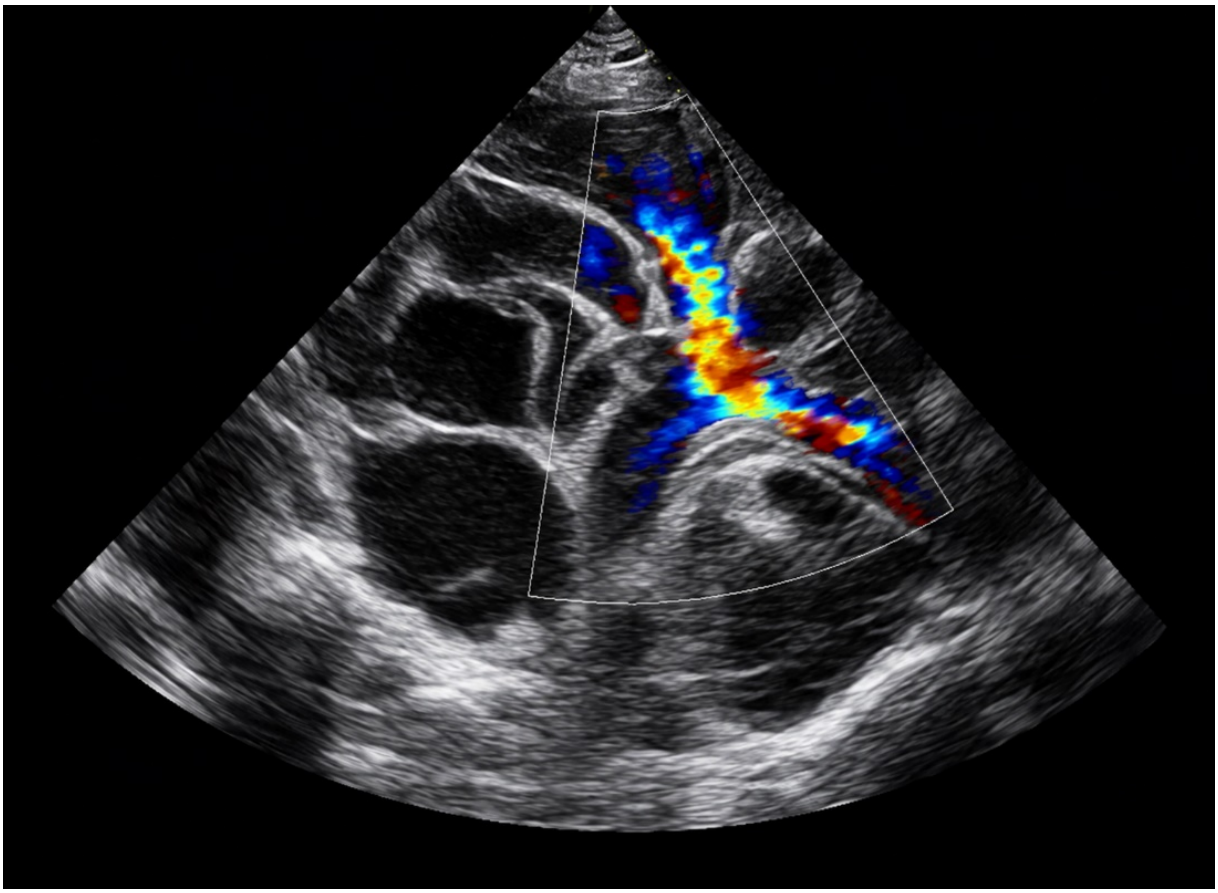
but:

"Is this PDA keeping the systemic circulation alive?"

CHAPTER 10

Duct-Dependent Pulmonary Circulation





Learning Objectives

By the end of this chapter, the learner should be able to:

1. Explain the physiology of duct-dependent pulmonary blood flow.
2. Recognize the major lesions associated with duct-dependent pulmonary circulation.
3. Perform a systematic echocardiographic assessment of a cyanotic neonate with suspected reduced pulmonary blood flow.
4. Determine whether pulmonary blood flow is antegrade from the right ventricle, supplied through the PDA, supplied by aortopulmonary collaterals, or derived from multiple sources.
5. Differentiate pulmonary atresia from critical pulmonary stenosis.

1. The Central Question

In suspected duct-dependent pulmonary circulation, the most useful question is not:

"Is there a PDA?"

The better question is:

"Where is the pulmonary blood flow coming from?"

Normally:

RA → RV → RVOT → Pulmonary valve → MPA → RPA/LPA → Lungs

When the pulmonary outlet is critically obstructed or atretic:

Aorta → PDA → Pulmonary arteries → Lungs

The ductus has changed from a transitional fetal structure into a **lifeline for pulmonary blood flow**.

Therefore, the physiological question becomes:

What will happen to pulmonary blood flow when the ductus constricts?

If pulmonary blood flow falls critically with ductal closure, the circulation is **duct-dependent**.

2. Which Lesions Can Produce Duct-Dependent Pulmonary Blood Flow?

Important neonatal lesions include:

Lesion	Why Pulmonary Flow May Be Duct-Dependent
Pulmonary atresia with intact ventricular septum (PA/IVS)	No effective antegrade RV-to-PA flow
Critical pulmonary stenosis	Antegrade pulmonary flow is inadequate
Severe TOF/RVOT obstruction	Pulmonary output through RVOT may be insufficient
TOF with pulmonary atresia / PA-VSD	No direct RV-to-PA continuity
Tricuspid atresia with severe PS/PA	Pulmonary flow limited by VSD/outflow pathway
Single-ventricle lesions with PS/PA	Effective pulmonary outlet absent or severely restricted
Selected severe Ebstein physiology	Functional or anatomical pulmonary atresia can occur

3. First Principle: Anatomy Before Color

The practical congenital echocardiography reference emphasizes obtaining **2D anatomy before spectral or color Doppler**, so that flow information is interpreted within a defined anatomical framework. This is particularly important in duct-dependent pulmonary circulation because color can be misleading. A beautiful color jet within the pulmonary artery does **not** necessarily mean the RV is supplying it. The blood may have entered through the PDA.

ECHO PEARL

Seeing blood in the pulmonary artery does not prove RV-to-PA continuity.

First establish the anatomy.

Then determine the direction and source of flow.

4. A Practical Scan Strategy

For a beginner, reduce the examination to seven questions:

P — PULMONARY OUTLET

Is the RVOT open?

V — VALVE

Does the pulmonary valve open? Is there antegrade flow?

A — ARTERIES

Are the MPA, RPA, and LPA present and confluent?

D — DUCTUS

Is the PDA present? Where does it originate and insert?

F — FLOW

Which direction is blood travelling?

R — RIGHT HEART

What are the TV, RV size, pressure, and function?

S — SEPTUM

What is happening across the atrial and ventricular septa?

5. Start With a Complete Segmental Examination

Do not begin with the assumption:

"This is pulmonary atresia."

Establish:

Situs → systemic veins → pulmonary veins → atria → AV connections → ventricular morphology → ventricular septum → VA connections → great arteries

Then focus on the pulmonary circulation.

This maintains the same principle used throughout this book:

Never assume anatomy. Demonstrate it.

6. Step 1 — Look at the Four-Chamber View

The four-chamber view often provides the first clue.

Ask:

- Is the RV normal, small, hypertrophied, or dilated?
- Is the RV apex forming?
- Is the tricuspid valve normal, hypoplastic, dysplastic, or absent?
- Is there significant TR?
- Is the ventricular septum intact?
- What is the direction of atrial shunting?

ECHO PEARL

A very small RV immediately changes the diagnostic pathway.

Think particularly of:

PA/IVS → hypoplastic TV → hypoplastic RV → possible coronary sinusoids

But a normal-sized or even enlarged RV does **not** exclude pulmonary atresia.

7. Step 2 — Assess the Tricuspid Valve

This is especially important in PA/IVS.

Assess:

Annulus → leaflets → inflow → regurgitation

Measure the tricuspid annulus carefully and use an appropriate pediatric Z-score.

The apical four-chamber view is an excellent starting point, but confirm anatomy in additional planes.

ECHO PEARL

In PA/IVS, the tricuspid valve is a window into potential RV adequacy.

A very hypoplastic TV generally accompanies a smaller RV.

A relatively well-sized tricuspid valve may accompany a more developed RV.

However:

TV size alone should not determine the treatment pathway.
RV morphology, outflow anatomy, coronary circulation, and function must also be considered.

8. Step 3 — Decide Whether the RV Is Tripartite

For the pediatric cardiology trainee, describe whether the RV has recognizable:

Inlet + trabecular/body + outlet
A severely hypoplastic RV may lack a well-developed body or outlet.

The apical four-chamber view helps assess relative RV size, but the whole RV does not lie in one plane. Sweeping through subcostal and parasternal planes is therefore important. *Moss and Adams* specifically emphasizes multiplane imaging when assessing the inlet, trabecular and outlet portions of the RV.

TECHNIQUE TIP

Do not freeze one apical four-chamber frame and decide:

"RV is small."

Instead:

Find → sweep anteriorly → sweep posteriorly → assess inlet/body/outlet separately.

9. Step 4 — Find the RVOT

This is one of the critical parts of the examination.

Useful views include:

- parasternal short axis
- modified parasternal long axis
- subcostal anterior oblique/RVOT view
- high parasternal views where necessary

Assess:

- whether the RVOT is present
- infundibular caliber
- muscular obstruction
- continuity with the pulmonary valve
- whether atresia is valvar or extends into the infundibulum

ECHO PEARL

Pulmonary atresia is not one anatomy. Define where continuity stops.
This matters particularly in PA/VSD and TOF/PA.

10. Step 5 — Examine the Pulmonary Valve in 2D

Do not start by placing CW Doppler through it.

First ask:

Can I see valve tissue?
Does it move?
Does an orifice exist?
Is the annulus hypoplastic?
Is atresia membranous or muscular?

In critical pulmonary stenosis, the valve may appear severely thickened and barely mobile.

In membranous pulmonary atresia, valve tissue may be present but imperforate.

ECHO PEARL

"I cannot see forward color flow" is not equivalent to "the valve is atretic."

This distinction is one of the most important diagnostic traps in neonatal echocardiography.

11. Pulmonary Atresia Versus Critical Pulmonary Stenosis

These can look remarkably similar.

In critical pulmonary stenosis:

RV → tiny amount of antegrade flow → PA

In pulmonary atresia:

No true antegrade RV → PA flow

However, a large PDA may generate considerable color turbulence within the main PA and make interpretation difficult.

The neonatal cardiology reference specifically notes that forward flow in critical PS may be so small that it becomes difficult to separate from ductal turbulence. A small pulmonary regurgitation jet in the RVOT can demonstrate that the pulmonary valve is patent rather than completely atretic.

HIGH-YIELD ECHO PEARL

Tiny pulmonary regurgitation can prove that a valve you thought was atretic is actually patent.

Look specifically for pulmonary insufficiency with appropriately sensitive color settings.

12. Do Not Let the PDA Fool You

Imagine the following:

Aorta → PDA → MPA

Blood enters the main PA and then spreads toward:

RPA + LPA

Part of that ductal flow can extend toward the pulmonary valve.

On color Doppler this may create the false impression of flow originating across the pulmonary valve.

CRITICAL PITFALL

Ductal flow striking the pulmonary valve can masquerade as antegrade pulmonary-valve flow.

Always establish the direction of flow at the level of the valve itself.

13. Critical Pulmonary Stenosis — What Should You Assess?

The practical echocardiography reference recommends assessing:

- pulmonary valve morphology
- pulmonary annulus/Z-score
- subvalvar region
- RV hypertrophy
- tricuspid regurgitation
- atrial-level shunting
- MPA and branch PAs
- PDA
- RV function

It also notes that neonates at the critical end of the PS spectrum may have right-to-left atrial shunting, suprasystemic RV pressure and PDA-dependent pulmonary blood flow.

ECHO PEARL

Do not call the lesion "critical" based solely on the valve gradient.

Critical physiology means:

The RV cannot provide adequate effective pulmonary blood flow without additional ductal contribution.

14. Doppler Across a Critically Stenotic Pulmonary Valve

Use:

PW Doppler → localize obstruction

followed by:

CW Doppler → measure maximal velocity

March PW through:

RV → infundibulum → valve → MPA

This helps distinguish:

- subvalvar obstruction
- valvar obstruction
- supravalvar obstruction
- combinations of obstruction

ECHO PEARL

A gradient is a consequence of flow as well as obstruction.

Therefore, in a critically compromised neonatal circulation, anatomy and physiology should not be reduced to a single Doppler number.

15. Step 6 — Examine the Main and Branch Pulmonary Arteries

Once pulmonary outflow obstruction is identified, immediately move downstream.

Demonstrate:

MPA → RPA + LPA

Measure:

- pulmonary annulus
- MPA
- proximal RPA
- proximal LPA

and use Z-scores when clinically appropriate.

Questions include:

- Are both branch PAs present?
- Are they confluent?
- Are they hypoplastic?
- Does the PDA enter one branch preferentially?
- Is there narrowing at the ductal insertion?

ECHO PEARL

Do not assume that because you see one pulmonary artery, the pulmonary arteries are confluent.

16. Why the Ductal Insertion Matters

In TOF and pulmonary atresia, abnormalities of the branch pulmonary arteries are common.

Moss and Adams notes that ductal tissue at the LPA insertion can contribute to focal narrowing and that pulmonary arterial anatomy becomes increasingly variable toward the pulmonary-atresia end of the TOF spectrum.

PRACTICAL TEACHING RULE

Whenever you identify a duct-dependent pulmonary circulation:

Find the point where the duct enters the pulmonary arterial tree.

Do not merely measure the middle of the PDA.

Look at its **pulmonary insertion**.

This becomes particularly important if ductal stenting is being considered.

17. Step 7 — Find the PDA Properly

Useful views include:

HIGH LEFT PARASTERNAL

Excellent for:

MPA → RPA → LPA → PDA

A classic view can produce the appearance of a pulmonary arterial **"trifurcation."**

PARASTERNAL SHORT AXIS

Useful for the pulmonary valve, PA bifurcation and ductal region.

SUPRASTERNAL SAGITTAL

Particularly important when the ductus has an unusual or vertical course.

The PDA is normally assessed using several complementary planes rather than a single view.

ECHO PEARL

When pulmonary blood flow depends on the ductus, define the whole duct—not merely the fact that it is open.

18. The "Reverse" or Vertical Ductus

This is particularly relevant in pulmonary atresia.

A ductus may arise from the undersurface of the distal arch and travel in a more vertical or tortuous direction toward the pulmonary arteries.

These ducti can be difficult to appreciate from the standard high parasternal window alone.

ECHO PEARL

If pulmonary atresia is present but you cannot find the ductus: go to the suprasternal sagittal window and search from the distal arch toward the pulmonary arteries.

Do not conclude "no PDA" because the standard ductal view is negative.

19. Step 8 — Determine Ductal Flow Direction

In pulmonary ductal dependency, the physiologically important flow is:

Aorta → PDA → Pulmonary artery

This is conventionally described as **left-to-right ductal flow**.

The PA/IVS echocardiographic approach in the practical congenital echo reference specifically identifies PA/IVS as duct-dependent and expects systemic-to-pulmonary flow through the PDA.

COMPARE THIS WITH SYSTEMIC DUCTAL DEPENDENCY

Pulmonary-dependent circulation:

Aorta → PDA → PA

Systemic-dependent circulation:

PA → PDA → Aorta

This simple contrast is worth memorizing.

20. But Do Not Use Direction Alone

Early neonatal PVR may still be elevated.

Therefore a ductus may show:

- left-to-right flow
- bidirectional flow
- changing systolic/diastolic patterns

depending on pressure relationships and transitional physiology.

ECHO PEARL

Ductal direction tells you about pressure relationships. Anatomy tells you why those relationships exist.

Always integrate:

Ductal flow + RVOT anatomy + pulmonary valve anatomy + oxygenation + postnatal age

21. Ductal Velocity and Restriction

Do not report only:

"PDA present."

Assess:

- diameter
- course
- tortuosity
- site of origin
- pulmonary insertion
- direction
- spectral pattern
- degree of restriction

A restrictive ductus may be inadequate even if technically patent.

MASTER TEACHING POINT

"Open" does not necessarily mean "adequate."

The physiological question is:

Is enough blood reaching the pulmonary arteries through it?

22. Step 9 — Examine the Atrial Septum

Right-sided obstructive lesions frequently require right-to-left atrial decompression.

In PA/IVS:

Systemic veins → RA → cannot exit adequately through RV → PFO/ASD → LA

Thus the interatrial communication becomes an essential part of the circulation.

Moss and Adams emphasizes assessing the interatrial communication from the subcostal window because obligatory right-to-left atrial shunting may be necessary to maintain output.

Assess:

- size
- flap morphology
- direction of flow
- restriction
- RA pressure effect

ECHO PEARL

In right-heart obstruction, the atrial septum is not simply a "PFO check." It is part of the escape pathway for systemic venous return.

23. Right-to-Left Atrial Flow Is a Physiological Clue

A strongly right-to-left atrial shunt should make you ask:

Why can blood not leave the right heart normally?

Possible explanations include:

- pulmonary atresia
- critical pulmonary stenosis
- severe RVOT obstruction
- severe pulmonary hypertension
- tricuspid obstruction
- RV dysfunction

Therefore:

Right-to-left PFO flow is a clue—not the diagnosis.

24. PA/IVS — The Right Ventricle Changes the Entire Management Question

PA/IVS deserves special attention.

The echo examination must define:

TV → RV → RVOT → pulmonary valve → PA → PDA

But also:

RV → coronary sinusoids → coronary arteries

The practical congenital echo text emphasizes assessment of:

- RV size
- whether the RV is tripartite
- TV annular Z-score
- RVOT anatomy
- pulmonary valve
- MPA/branch PAs
- TR
- atrial shunting
- PDA
- coronary sinusoids
- coronary flow
- ventricular function

25. Look for Coronary Sinusoids in PA/IVS

A hypertensive RV may maintain communications with the coronary circulation.

With color Doppler, look within the RV myocardium for unusual channels.

Practical adjustments include:

- high-frequency imaging
- shallow depth
- narrow sector
- targeted zoom
- reduced color scale/Nyquist limit
- careful gain adjustment

ECHO PEARL

Coronary sinusoids on echo should immediately elevate the importance of coronary assessment.

But there is an equally important limitation:

Seeing sinusoids does not by itself prove RV-dependent coronary circulation.

26. The Major Limitation of Echo in PA/IVS

This is critical for trainees.

Echocardiography can identify:

- RV-coronary communications
- proximal coronary origins
- abnormal flow patterns
- regional ventricular dysfunction

But it may not reliably establish the complete coronary arterial anatomy or exclude critical coronary stenosis/atresia. Echo cannot reliably identify all relevant neonatal coronary stenosis or interruption in PA/IVS, and invasive angiographic assessment can therefore be important before RV decompression.

CRITICAL ECHO PEARL

Never report "no RVDCC" simply because you did not see abnormal coronary flow on transthoracic echo.

A better report is:

"Coronary sinusoids are/are not demonstrated; echocardiography cannot fully exclude right-ventricle-dependent coronary circulation."

27. Tetralogy of Fallot With Severe Pulmonary Obstruction

In neonatal TOF, define:

- large anterior malalignment VSD
- aortic override
- degree and level of RVOT obstruction

The pulmonary obstruction may occur at:

- infundibular level
- pulmonary valve
- annulus
- MPA
- branch PA level

Toward the severe end of the spectrum, pulmonary blood flow may become duct-dependent; the extreme form is pulmonary atresia. The practical congenital echo reference explicitly describes this continuum.

ECHO PEARL

In neonatal TOF, do not simply measure the pulmonary-valve gradient. Map every level of the pulmonary outflow.

28. TOF — What the Echo Technician Should Record

Obtain clear imaging of:

VSD → aortic override → infundibulum → pulmonary valve → annulus → MPA → RPA/LPA → PDA

Then document:

- severity and level of RVOTO
- pulmonary annulus
- branch PA dimensions
- PDA
- arch sidedness
- additional VSDs
- coronary anatomy where feasible

The practical echo source also emphasizes careful coronary assessment because an important coronary may cross the RVOT and alter surgical strategy.

29. TOF With Pulmonary Atresia / Pulmonary Atresia With VSD

Now the question changes dramatically.

There is no effective RV-to-PA route.

You must determine:

Where does each lung receive its blood supply?

Possible sources include:

- PDA
- native central pulmonary arteries
- MAPCAs
- combinations of these

In tetralogy with pulmonary atresia the central and segmental PAs may be confluent or nonconfluent and pulmonary blood supply comes from systemic arterial sources.

ECHO PEARL

Pulmonary atresia with VSD is not fully described until the source of pulmonary blood flow has been addressed.

30. Confluent or Nonconfluent Pulmonary Arteries?

This is a major pre-intervention question.

Try to demonstrate:

RPA ↔ central PA confluence ↔ LPA

If the branches cannot be shown connecting, do not assume confluence.

Look individually for:

- RPA
- LPA
- ductal insertion
- possible separate ductal supplies
- systemic collaterals

ECHO PEARL

"Both branch PAs seen" does not equal "branch PAs confluent."

Trace the connection.

31. The Left Atrial Appendage Trap

In TOF with pulmonary atresia and absent central pulmonary arteries, a structure near the expected MPA location can be mistaken for the pulmonary artery.

The practical congenital echo text specifically identifies the **left atrial appendage** as a potential mimic and recommends careful 2D and color assessment.

ECHO PEARL

Before calling a structure the MPA, prove that it connects to the branch pulmonary arteries.

Anatomical continuity beats visual resemblance.

32. MAPCAs — Know the Limit of Echocardiography

Echo may demonstrate:

- large collateral vessels
- flow arising from the descending aorta
- pulmonary arterial hypoplasia
- absence of an obvious ductal supply

But echocardiography may not reliably map:

- the full number of MAPCAs
- their origins
- stenoses
- segmental pulmonary supply
- connections between collateral and native PAs

IMPORTANT REPORTING PRINCIPLE

When anatomy remains incomplete, say so. For example:

"Central pulmonary arteries are severely hypoplastic. Additional systemic-to-pulmonary collateral supply is suspected but cannot be fully delineated by transthoracic echocardiography."

This is better than false precision.

33. Tricuspid Atresia and Single-Ventricle Physiology

When the tricuspid valve is absent, pulmonary blood flow depends on the downstream pathway.

Assess:

RA → atrial communication → LA → dominant LV → VSD/bulboventricular foramen → outlet chamber → pulmonary artery

Then determine:

- pulmonary stenosis or atresia
- VSD/bulboventricular foramen size
- great-artery relationship
- PDA
- branch PA anatomy

Neonatal cardiology references emphasize evaluating the atrial communication, VSD, RV/outlet chamber, great-artery relationship and ductus when tricuspid atresia is present.

34. Functional Pulmonary Atresia

Not every neonate without obvious forward pulmonary flow has anatomical pulmonary-valve atresia.

In severe right-heart dysfunction, including severe forms of Ebstein anomaly, the pulmonary valve may be anatomically patent while the RV is unable to generate effective antegrade flow.

ADVANCED ECHO PEARL

No antegrade pulmonary flow ≠ automatically anatomical pulmonary atresia.

Examine:

- valve tissue
- leaflet motion
- pulmonary regurgitation
- RV pressure
- severe TR
- ductal flow
- serial physiology

This distinction can substantially change management.

35. A Window-Based Practical Protocol

SUBCOSTAL — START WITH PHYSIOLOGY

Assess:

- systemic and pulmonary venous return
- atrial septum and shunt direction
- AV connections
- ventricular morphology
- ventricular septum
- RVOT with anterior angulation

Key question: Is right-to-left atrial flow acting as an obligatory escape pathway?

APICAL — ASSESS THE RIGHT HEART

Assess:

- TV morphology
- TV annulus
- RV size
- RV apex
- TR
- ventricular function

Key question: Is this RV capable of contributing meaningfully to pulmonary blood flow?

PARASTERNAL — DEFINE THE OBSTRUCTION

Assess:

- RVOT
- pulmonary valve
- annulus
- MPA
- branch PAs
- VSD where present
- coronaries where relevant

Use:

2D → Color → PW → CW

Key question: Where exactly does forward pulmonary flow become limited?

HIGH LEFT PARASTERNAL — FIND THE DUCTUS

Look for:

RPA — LPA — PDA

Assess:

- PDA course
- insertion
- direction
- restriction

Key question: How much of pulmonary flow is coming from the systemic circulation?

SUPRASTERNAL — COMPLETE THE MAP

Assess:

- aortic arch
- ductal origin
- vertical/reverse ductus
- branch PA anatomy where accessible
- systemic collaterals where visible

Key question: Is an unusual ductal or collateral pathway being missed?

36. Color Doppler Settings Matter

This chapter is one where poor settings can directly create the wrong diagnosis.

For tiny pulmonary-valve flow, coronary sinusoids, or slow ductal flow:

Lower the color velocity scale when necessary.

For high-velocity stenotic jets:

Increase the scale appropriately to reduce excessive aliasing.

Keep:

- depth shallow
- sector narrow
- focus near the structure
- color box small

37. The Most Important Diagnostic Traps

Trap 1 — "NO ANTEGRADE COLOR FLOW = PULMONARY ATRESIA."

Correction: Search carefully for tiny valve opening or pulmonary regurgitation.

Trap 2 — DUCTAL FLOW MISTAKEN FOR PULMONARY-VALVE FLOW

Correction: Determine flow direction directly across the valve.

Trap 3 — "PDA PRESENT = PULMONARY CIRCULATION SECURE."

Correction: Assess ductal restriction, tortuosity and pulmonary insertion.

Trap 4 — BRANCH PAS SEEN = CONFLUENT PAS

Correction: Demonstrate the actual confluence.

Trap 5 — NORMAL-SIZED RV EXCLUDES PA/IVS

Correction: PA/IVS has a wide RV-size spectrum.

Trap 6 — NO SINUSOID SEEN = NO RVDCC

Correction: Echo cannot reliably exclude all clinically important coronary abnormalities.

Trap 7 — A SINGLE RVOT GRADIENT DEFINES SEVERITY

Correction: Combine anatomy, antegrade flow, ductal contribution and oxygenation.

Trap 8 — FAILURE TO FIND A STANDARD PDA = NO PDA

Correction: Search for vertical/reverse ductal anatomy from the suprasternal window.

Trap 9 — FINDING THE PRIMARY LESION ENDS THE STUDY

Correction: Complete the pulmonary arterial anatomy, ductal anatomy and associated lesions.

38. Beginner Versus Trainee Expectations

ECHO TECHNICIAN / BEGINNER SHOULD RELIABLY ESTABLISH

- pulmonary valve opens or does not visibly open
- presence of antegrade pulmonary flow
- PDA presence and flow direction
- MPA, RPA and LPA visualization
- RV size
- TV morphology
- atrial shunt direction
- VSD present or absent
- ventricular function
- obvious associated anomalies

PAEDIATRIC CARDIOLOGY TRAINEE SHOULD ADDITIONALLY DEFINE

- precise level(s) of RVOTO
- valvar versus muscular pulmonary atresia
- TV and pulmonary annular Z-scores
- tripartite versus bipartite/severely hypoplastic RV
- branch PA confluence and ductal insertion
- possible RVDCC/coronary sinusoids
- pulmonary blood-flow sources in PA/VSD
- limitations requiring CT/catheterization
- physiological significance and urgency

39. The 60-Second Emergency Echo

For the severely cyanotic neonate:

Four chambers — RV small, normal or large?

↓

Atrial septum — Right-to-left escape?

↓

Tricuspid valve — Normal, hypoplastic or absent?

↓

Ventricular septum — Intact or VSD?

↓

RVOT — Open, critically narrowed or atretic?

↓

Pulmonary valve — Does it open? Any antegrade flow or PR?

↓

MPA + RPA + LPA — Present? Confluent?

↓

PDA — Where is it? Which direction is flow?

↓

Coronaries if PA/IVS — Sinusoids? Abnormal proximal flow?

Final question: Where are the lungs getting their blood?

40. A Simple Diagnostic Algorithm

Neonate with cyanosis + reduced pulmonary flow

Is there antegrade flow across the pulmonary valve?

YES — Is it adequate?

Adequate → pulmonary circulation probably not duct-dependent.

Tiny/inadequate → think critical pulmonary stenosis or severe RVOTO.

Then assess:

PDA contribution + atrial R →L shunting + RV pressure/function
NO — Is the ventricular septum intact?

YES — Think: PA/IVS. Assess:

TV → RV → sinusoids/coronaries → atrial communication → PDA
NO — Large VSD present — Think: TOF/PA or another PA/VSD anatomy. Assess:

Central PAs → confluence → PDA → MAPCAs → arch

41. How to Describe Duct Dependency in the Report

Avoid:

"PDA present."

A stronger conclusion is:

"Pulmonary blood flow is predominantly supplied through the patent ductus arteriosus, with limited/no effective antegrade flow across the pulmonary outflow."

For critical PS:

"Severe valvar pulmonary stenosis with markedly reduced antegrade pulmonary blood flow and significant systemic-to-pulmonary ductal contribution, consistent with duct-dependent pulmonary circulation."

For PA/IVS:

"Pulmonary atresia with intact ventricular septum. Pulmonary blood flow is supplied through the PDA. The RV is hypoplastic with [description] and the tricuspid annulus measures [value/Z-score]. Coronary sinusoids are [seen/not demonstrated]; RVDCC cannot be excluded by echocardiography alone."

For TOF/PA:

"Pulmonary atresia with large malalignment VSD and overriding aorta. Confluent/hypoplastic branch pulmonary arteries are supplied through [PDA identified]. Additional systemic-to-pulmonary collateral vessels cannot be fully excluded/delineated by echocardiography."

42. Echo Pearls — The Page to Remember

Pearl 1: The first question is not "Is there a PDA?" It is "Where is pulmonary blood flow coming from?"

Pearl 2: Aorta → PDA → PA = think pulmonary ductal dependency.

Pearl 3: Blood inside the MPA does not prove blood crossed the pulmonary valve.

Pearl 4: Tiny pulmonary regurgitation may distinguish critical PS from true pulmonary atresia.

Pearl 5: In PA/IVS, assess TV size, RV morphology and coronary sinusoids—not merely the pulmonary valve.

Pearl 6: Failure to see coronary sinusoids does not exclude RVDCC.

Pearl 7: In pulmonary atresia with VSD, define the source of blood flow to each lung.

Pearl 8: Both PAs being visible does not prove that they are confluent.

Pearl 9: A vertical/reverse ductus may require suprasternal imaging.

Pearl 10: A patent PDA can still be too restrictive to provide adequate pulmonary flow.

Pearl 11: Right-to-left atrial shunting in critical right-heart obstruction may be an essential decompression pathway.

Pearl 12: When anatomy remains uncertain, state the uncertainty rather than inventing anatomical completeness.

43. Chapter Key Points

Duct-dependent pulmonary circulation occurs when antegrade blood flow from the ventricle to the pulmonary arteries is absent or insufficient and pulmonary blood flow therefore depends substantially on the PDA.

The most important lesions are **PA/IVS, critical pulmonary stenosis, severe TOF/RVOT obstruction, TOF with pulmonary atresia/PA-VSD, and selected single-ventricle or tricuspid-atresia anatomies**.

The echocardiographic examination should reconstruct:

RV → RVOT → Pulmonary valve → MPA → Branch PAs
and then separately reconstruct:

Aorta → PDA → Pulmonary arteries
The echocardiographer must determine which pathway is actually providing pulmonary blood flow.

In PA/IVS, assessment must extend beyond the pulmonary valve to the:

Tricuspid valve → RV morphology → coronary sinusoids → atrial communication
because these findings influence subsequent management. Echocardiography remains central to defining anatomy but has limitations in excluding clinically important right-ventricle-dependent coronary circulation.

In TOF/PA or PA/VSD, define the native pulmonary arteries and their confluence, the ductal supply and the possibility of collateral pulmonary blood flow.

MASTER PRINCIPLE

In suspected duct-dependent pulmonary circulation, do not merely find the obstruction. Follow the blood all the way to the lungs.
The diagnostic sequence is:

ANATOMY — Where is the obstruction?

↓

ANTEGRADE FLOW — How much blood can cross the RVOT?

↓

PULMONARY ARTERIES — Are they present, adequate and confluent?

↓

DUCTUS — Does systemic blood enter the pulmonary arteries through the PDA?

↓

ALTERNATIVE SOURCES — Are collaterals present or suspected?

↓

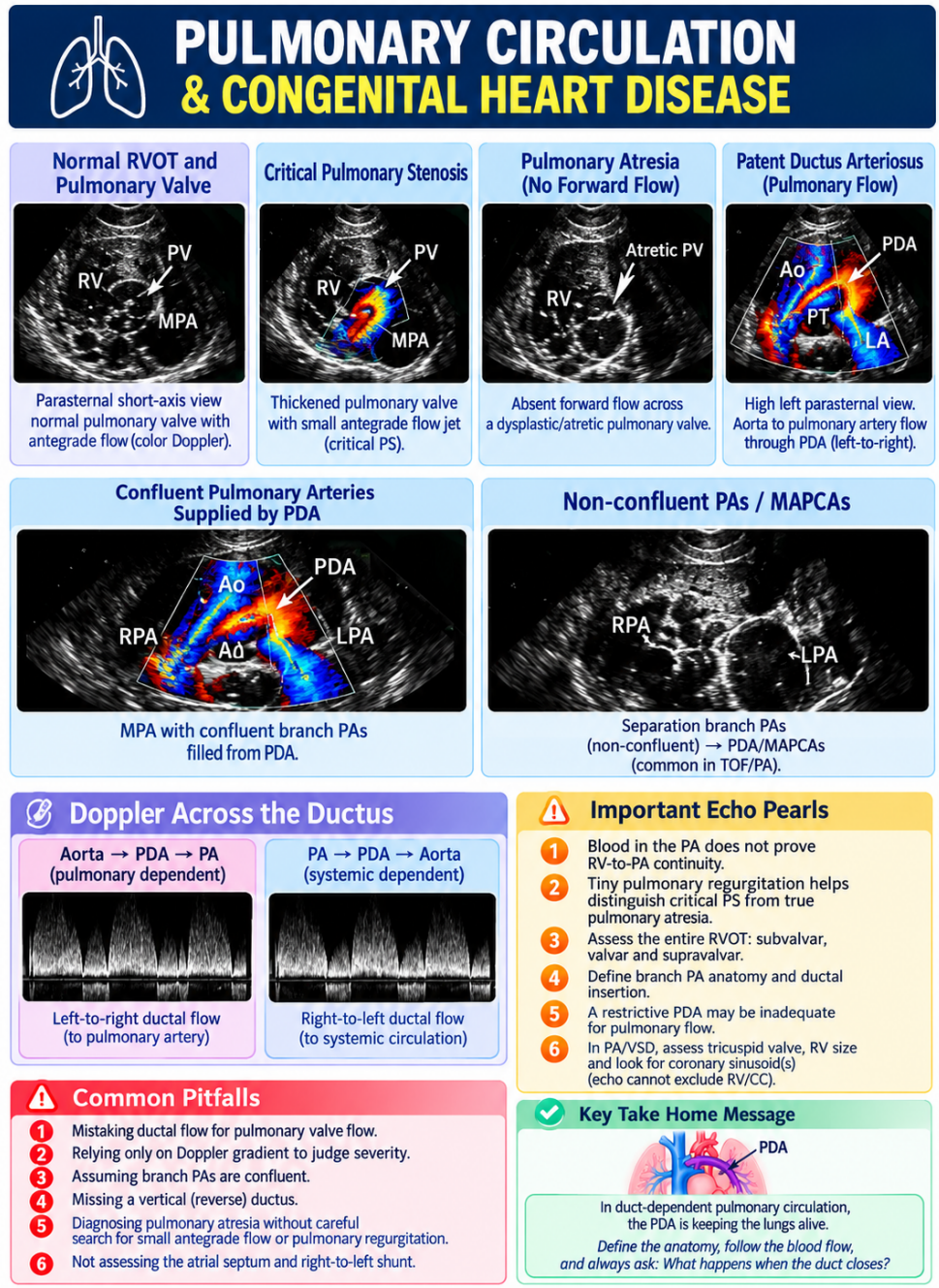
PHYSIOLOGY — What happens when the ductus closes?

Mastering Formula

RV → RVOT → PV → PA → PDA → FLOW DIRECTION → SOURCE OF PULMONARY BLOOD FLOW → URGENCY

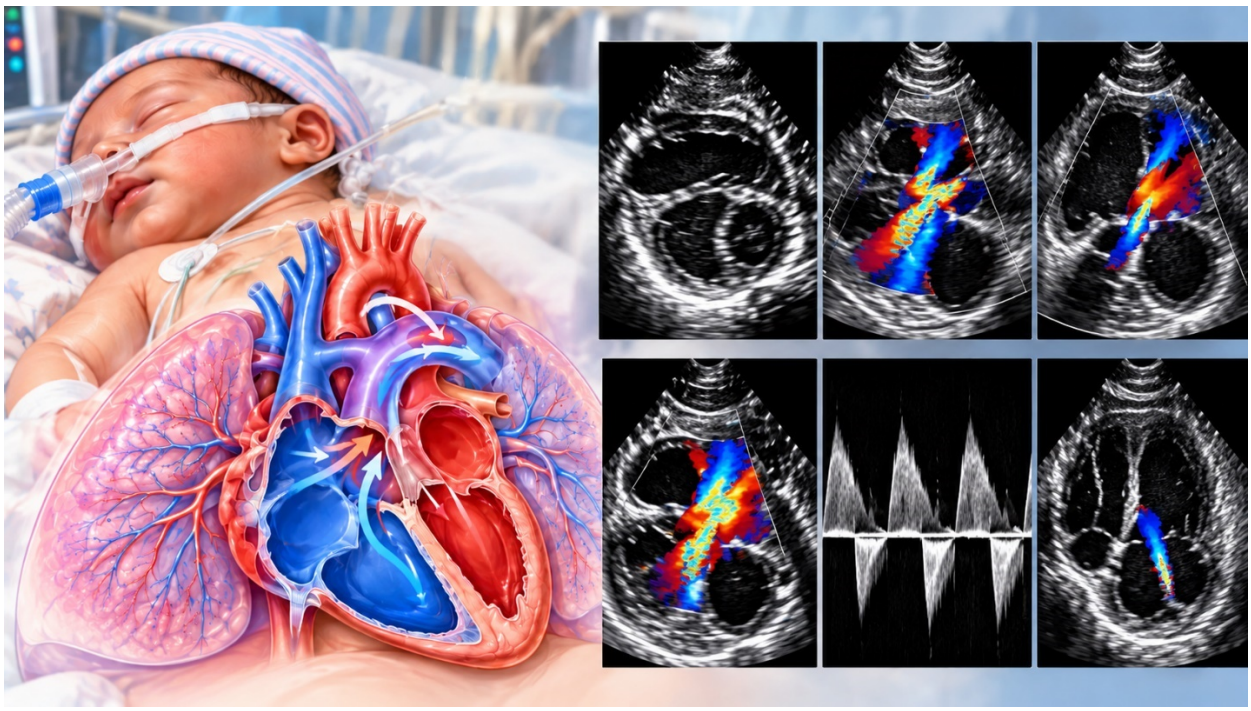
And the single most important question for the learner is:

"Where are the lungs getting their blood?"



CHAPTER 11

Persistent Pulmonary Hypertension of the Newborn



PPHN — Is This Abnormal Transition or Structural Heart Disease

When the Pulmonary Pressure Fails to Fall

Learning Objectives

By the end of this chapter, the learner should be able to:

1. Explain the physiological basis of persistent pulmonary hypertension of the newborn (PPHN).
2. Understand why PPHN is primarily a **physiological diagnosis rather than a structural cardiac diagnosis**.
3. Perform a systematic echocardiographic assessment in a neonate with suspected pulmonary hypertension.
4. Exclude critical congenital heart disease before attributing hypoxemia to PPHN.
5. Estimate pulmonary and right ventricular pressure using multiple echocardiographic clues.
6. Interpret tricuspid regurgitation velocity correctly.
7. Assess interventricular septal configuration as a marker of RV pressure loading.
8. Interpret PDA and atrial-level shunt direction.
9. Recognize obstructed pulmonary venous return as an important PPHN mimic.
10. Use serial echocardiography to assess changing neonatal physiology.

1. What Is PPHN?

During normal transition after birth:

Lung expansion + increasing oxygenation

↓

Pulmonary vascular resistance falls

↓

Pulmonary blood flow increases

↓

Pulmonary venous return to the LA increases

↓

Right-sided pressures fall

↓

Fetal right-to-left shunting disappears

PPHN occurs when this normal fall in pulmonary vascular resistance is inadequate or delayed.

The result is persistence of a fetal-type pressure relationship:

Pulmonary vascular resistance remains abnormally high relative to systemic vascular resistance.

Blood may therefore bypass the lungs through:

PFO: RA → LA

and/or

PDA: PA → Aorta

producing systemic hypoxemia.

2. The Most Important Concept

PPHN is not diagnosed simply because the estimated pulmonary pressure is high.

Pulmonary pressures are normally elevated immediately after birth and fall progressively during neonatal transition.

Therefore:

High pulmonary pressure alone ≠ PPHN

The echocardiographer must integrate:

postnatal age + oxygenation + shunt direction + RV pressure + ventricular function + lung disease + structural anatomy

ECHO PEARL

PPHN is abnormal persistence of high pulmonary vascular resistance—not merely the presence of a high neonatal RV pressure.

3. The First Echo Question Is Not "How High Is the PA Pressure?"

The first question is: **Is the heart structurally normal?**

Before diagnosing PPHN, systematically exclude congenital lesions capable of producing similar:

- cyanosis
- right-to-left atrial shunting
- right-to-left ductal flow
- elevated RV pressure
- RV enlargement
- ventricular dysfunction

Particularly important lesions include:

- TGA
- obstructed TAPVR
- pulmonary atresia
- critical pulmonary stenosis
- severe TOF/RVOT obstruction
- Ebstein anomaly
- critical left-sided obstruction
- aortic arch obstruction
- pulmonary venous obstruction

GOLDEN RULE

PPHN should never be used as a shortcut around complete anatomical assessment.

4. The "It Is Just PPHN" Trap

A cyanotic neonate may show:

right-to-left PFO flow + right-to-left PDA flow + high RV pressure

This certainly fits PPHN physiology.

But the same pattern can accompany important congenital heart disease.

A particularly dangerous mimic is **obstructed total anomalous pulmonary venous return**. The neonatal cardiology reference specifically stresses urgent echocardiographic assessment of pulmonary venous anatomy when PPHN is suspected because obstructed anomalous pulmonary venous return may closely mimic it.

CRITICAL ECHO PEARL

Before calling a neonate PPHN, demonstrate pulmonary venous return.

Do not write:

"Pulmonary veins appear probably normal."

Whenever technically possible, show their entry into the LA and use color/PW Doppler when obstruction is suspected.

5. What Is Echo Actually Trying to Answer?

A useful PPHN echo should answer five questions:

1. Is there structural CHD?
2. How high is RV/pulmonary pressure relative to systemic pressure?
3. Where is right-to-left shunting occurring?
4. How is the RV coping with the pressure load?
5. How is the LV being affected by ventricular interaction and reduced pulmonary venous return?

A study that reports only:

"Severe pulmonary hypertension" is incomplete.

6. A Practical PPHN Echo Sequence

Use:

ANATOMY → PRESSURE → SHUNTS → RV → LV → PERFUSION

Or remember:

A–P–S–R–L–P

A — Anatomy

Exclude CHD.

P — Pressure

Estimate RV/PA pressure.

S — Shunts

PFO and PDA direction.

R — Right ventricle

Size and function.

L — Left ventricle

Filling and function.

P — Perfusion physiology

Does the echo explain the hypoxemia and circulatory state?

7. Step 1 — Establish Normal Anatomy

Even in an unstable neonate, establish as rapidly as possible normal segmental anatomy.

ECHO PEARL

A normal four-chamber view does not exclude the important PPHN mimics.

TGA, obstructed pulmonary venous return and arch disease may all require views beyond the four-chamber plane.

8. Step 2 — Look at the Right Ventricle

The RV is central to PPHN.

Assess:

- RV size
- wall thickness
- systolic function
- RV pressure loading
- RV volume loading
- TR severity
- septal interaction
- RA size

The RV may be:

- relatively normal early in disease
- hypertrophied
- dilated
- pressure-loaded
- or dysfunctional

ECHO PEARL

Pulmonary hypertension severity and RV failure are not the same thing.

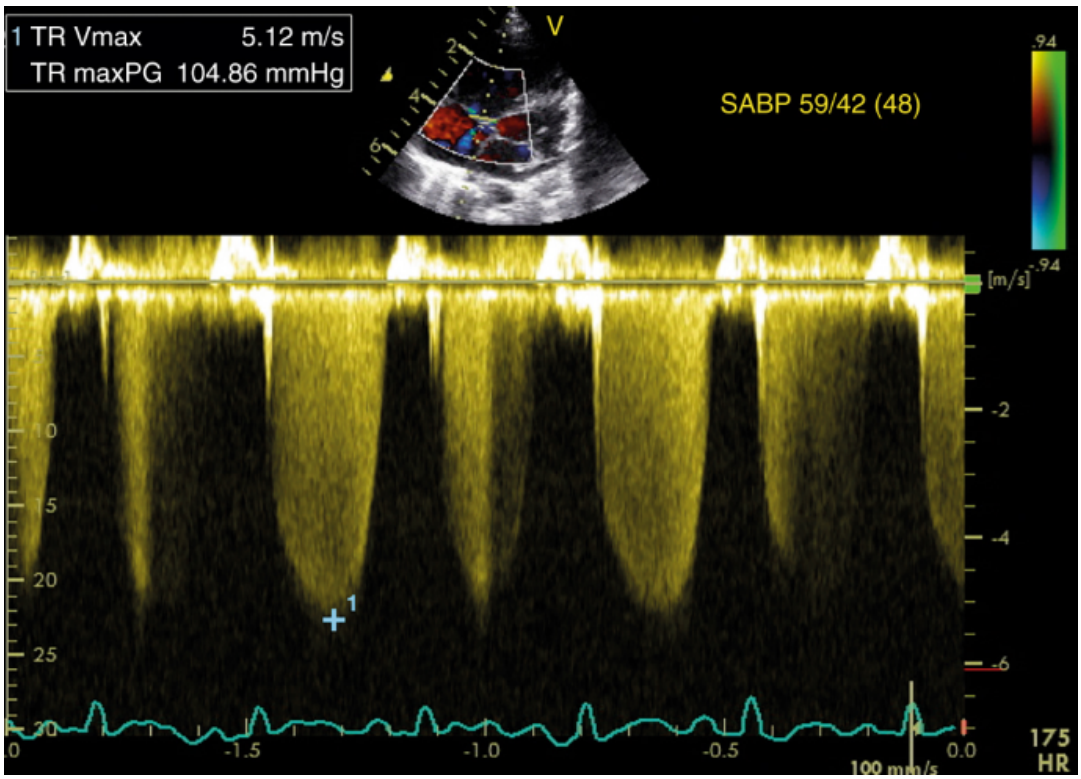
A baby can have very high pulmonary pressure with preserved RV performance.

Another baby can have lower measured pressure because the RV is failing and cannot generate a high pressure.

Therefore:

Always report pressure and function separately.

9. Step 3 — Use Tricuspid Regurgitation



When a complete TR spectral envelope is available:

$\Delta P = 4V^2$

where:

- V = peak TR velocity
- ΔP = RV–RA systolic pressure difference

This can contribute to estimation of RV systolic pressure.

But there are three important conditions.

The TR jet should be:

- adequately aligned
- complete
- and representative of peak velocity

ECHO PEARL

A faint, incomplete TR envelope should not be forced into a pressure estimate.

If the jet is poor: report that RV pressure cannot be reliably quantified from TR and use other physiological clues.

10. Do Not Underestimate Pressure Because the TR Jet Is Poor

This is a common beginner error.

A baby with severe PPHN may have:

- no measurable TR
- trivial TR
- or an incomplete spectral envelope

This does **not** mean pulmonary pressure is normal.

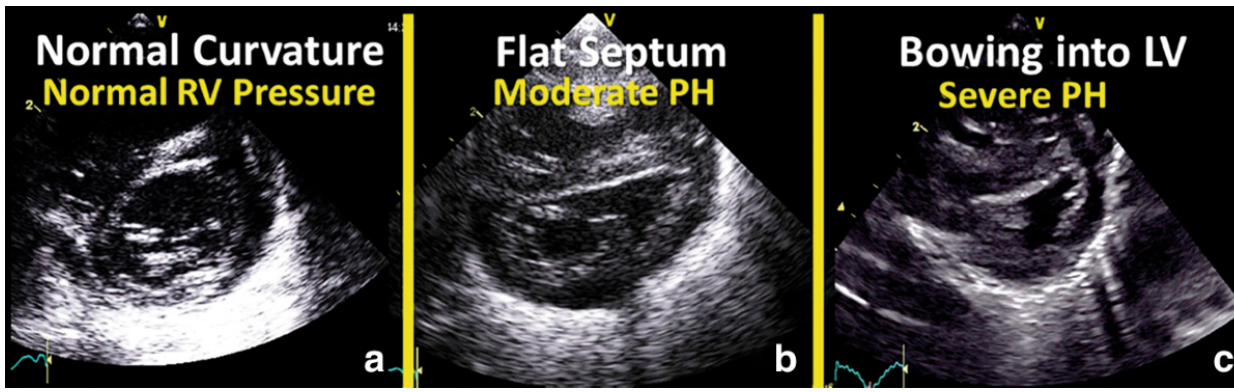
Instead assess:

- septal configuration
- PDA flow
- atrial flow
- RV morphology
- RV function

MASTER PRINCIPLE

No TR jet ≠ no pulmonary hypertension.

11. The Interventricular Septum — Your Pressure Gauge



The parasternal short-axis view is particularly useful.

Lower RV pressure

LV remains approximately circular.

Increasing RV pressure

The interventricular septum progressively flattens.

Severe pressure loading

The LV becomes: **D-shaped**

and with extreme RV pressure, the septum may bow toward the LV.

ECHO PEARL

Always assess septal shape in short axis—not from the apical four-chamber view alone.

12. When Does Septal Flattening Occur?

Look at the timing. This improves interpretation.

Predominantly systolic flattening

Suggests **RV pressure loading**.

Persistent flattening extending through the cardiac cycle

May indicate severe RV loading and altered ventricular interaction.

Predominantly diastolic flattening

May have a greater contribution from RV volume loading.

PRACTICAL TEACHING TIP

Do not simply report:

"Septum flattened."

Describe **when**, **how much**, and **what the RV is doing**.

13. The PDA Is a Natural Pressure Comparator

The ductus connects:

Pulmonary artery ↔ Aorta

Therefore its Doppler pattern provides information about the relative pulmonary and systemic arterial pressures.

Possible patterns are:

Aorta → PA

Left-to-right ductal flow.

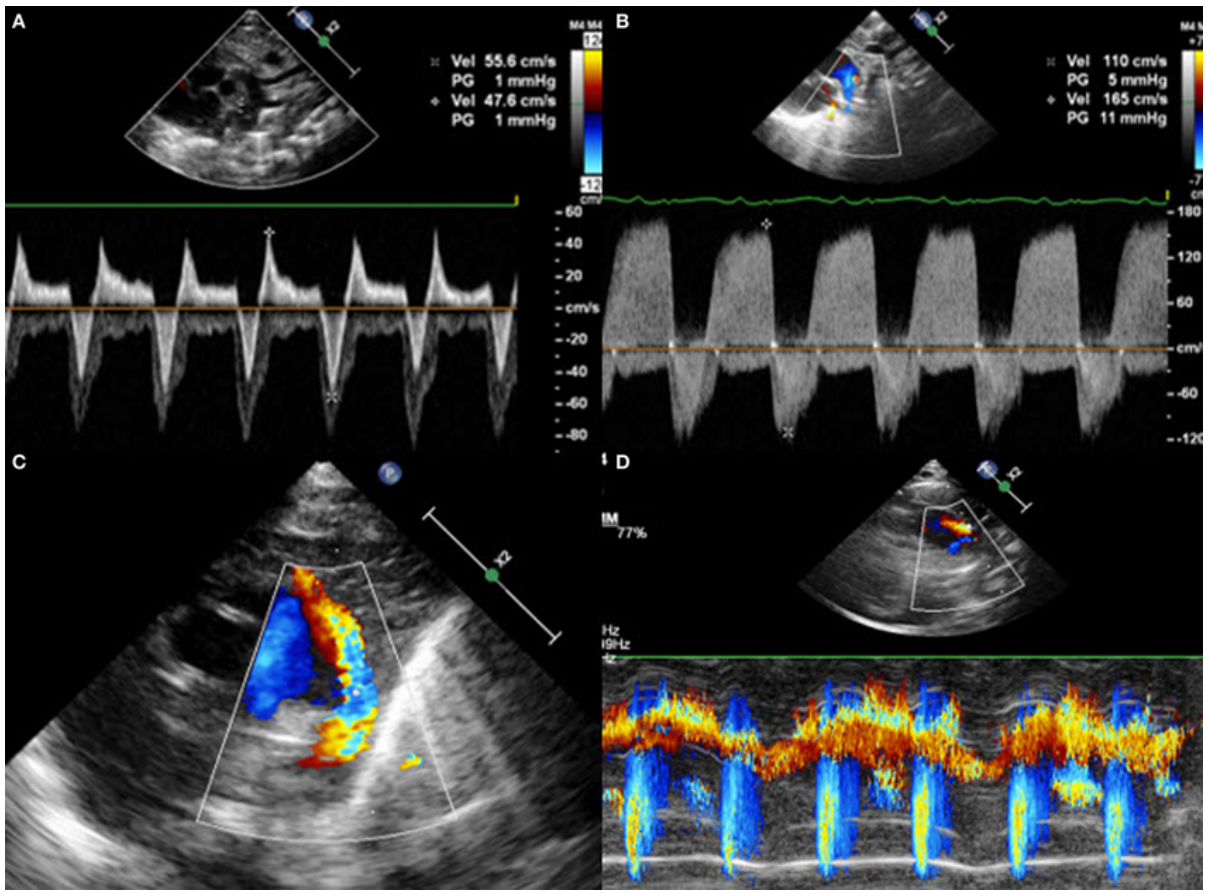
PA → Aorta

Right-to-left ductal flow.

Bidirectional

Pressure relationship changes during the cardiac cycle.

14. PDA Flow in PPHN



With significant pulmonary hypertension:

PA pressure approaches or exceeds aortic pressure
Therefore ductal flow may become:

bidirectional
or

predominantly right-to-left
In severe PPHN:

PA → PDA → descending aorta
contributes directly to systemic desaturation.

ECHO PEARL

Right-to-left ductal flow tells you that pulmonary pressure is competing with or exceeding systemic pressure—but it does not tell you why.

You must still exclude:

- duct-dependent systemic circulation
- abnormal pulmonary venous return
- structural right/left heart obstruction

15. Right-to-Left PDA: PPHN or Systemic Ductal Dependency?

This is one of the most important distinctions in neonatal echo.

PPHN

High PVR → PA pressure high → PA → PDA → Aorta

The systemic pathway from:

LV → aorta → arch

should remain anatomically adequate.

Duct-dependent systemic circulation

Left-heart obstruction → systemic circulation requires PA → PDA → Aorta

Examples include:

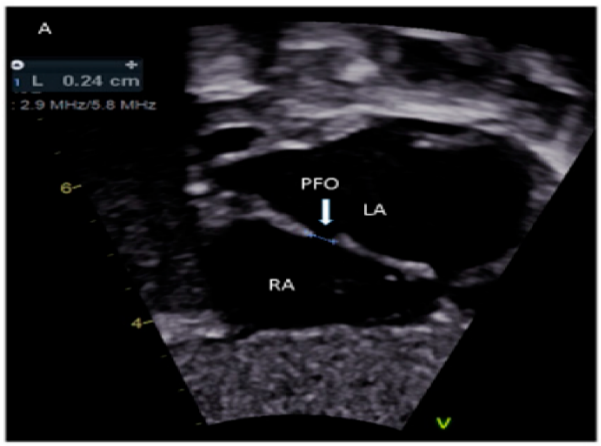
- HLHS
- critical AS
- coarctation
- interrupted arch

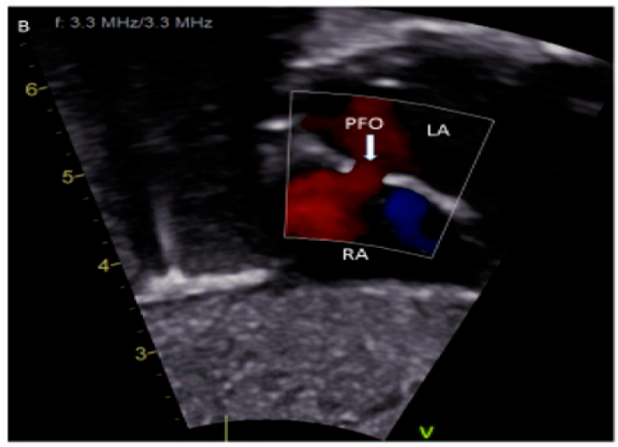
ECHO PEARL

Never interpret right-to-left ductal flow without examining the left heart and entire aortic arch.

16. Atrial-Level Shunting

Assess the PFO/ASD from the subcostal window.





Possible flow:

- left-to-right
- bidirectional
- right-to-left

In PPHN, elevated RA pressure may produce:

RA → LA

This allows desaturated systemic venous blood to bypass the lungs.

ECHO PEARL

Atrial right-to-left flow explains hypoxemia, but does not establish the cause of the high right-sided pressure.

17. PDA and PFO Tell Different Parts of the Story

These two shunts should not be considered interchangeable.

PDA

Reflects principally: **PA pressure versus aortic pressure**

PFO

Reflects principally: **RA pressure versus LA pressure**

Therefore a neonate may show different combinations.

For example:

R→L PDA + L→R PFO
is physiologically possible. So is:

R→L PDA + R→L PFO

ADVANCED ECHO PEARL

Do not expect atrial and ductal shunts always to move in the same direction.
Interpret each according to its own pressure relationship.

18. The Preductal–Postductal Concept

With significant right-to-left ductal shunting:

PA → PDA → descending aorta
desaturated blood preferentially enters the descending aorta.

Therefore lower-body oxygen saturation may be lower than right upper-limb saturation.

This creates a:

preductal–postductal saturation difference
However, the size of this difference depends on anatomy, shunting pattern and mixing.

ECHO PEARL

Absence of a large saturation difference does not exclude PPHN.
Echo remains necessary to understand actual shunt physiology.

19. Step 4 — Assess RV Function

This is one of the most clinically important parts of the examination.

Do not stop after estimating RV pressure.

Assess:

- qualitative RV systolic function
- longitudinal contraction
- free-wall movement
- chamber dilation
- TR
- septal displacement
- interaction with LV filling

Where locally validated and technically reliable, quantitative measurements may supplement visual assessment.

ECHO PEARL

The question is not only "How high is the RV pressure?" but also "Can the RV tolerate it?"

21. The Small LV in Severe PPHN

A small-looking LV in a sick neonate does not always mean structural left-heart hypoplasia.

Reduced pulmonary blood flow produces:

less pulmonary venous return → less LA filling → less LV preload
while septal displacement can further reduce LV filling.

ECHO PEARL

A small underfilled LV is different from an anatomically hypoplastic LV.
Look at:

- mitral valve size
- aortic valve size
- LV morphology
- arch anatomy
- serial change with improvement in pulmonary circulation

22. LV Function Also Matters

Evaluate:

- LV systolic function
- chamber filling
- mitral inflow qualitatively where useful
- systemic output
- interaction with RV

Associated conditions such as:

- perinatal asphyxia
- sepsis
- myocardial ischemia

may produce biventricular dysfunction in addition to pulmonary hypertension.

The neonatal cardiology source notes that perinatal asphyxia can be associated with ventricular dysfunction and that pulmonary hypertension can exacerbate TR and hypoxemia through right-to-left shunting.

23. Pulmonary Artery Doppler

Pulmonary arterial Doppler can provide additional physiological information.

Assess the RVOT/MPA waveform together with:

- TR
- septal shape
- PDA
- ventricular function

The principle is:

Do not use a single pulmonary artery Doppler parameter in isolation.
For the beginner, the key skill is recognizing that abbreviated or abnormal pulmonary ejection patterns may support elevated pulmonary vascular load, but should remain part of a multiparametric assessment.

24. The Multiparametric Approach

A strong PPHN study integrates:

Echo Feature	What It Helps Assess
TR velocity	RV–RA pressure gradient
Septal shape	RV/LV pressure relationship
PDA direction	PA–aortic pressure relationship
PFO direction	RA–LA pressure relationship
RV size	Pressure/volume response
RV systolic function	RV adaptation/failure
LV filling	Ventricular interaction
Pulmonary veins	Exclude obstruction
Structural survey	Exclude CHD

GOLDEN RULE

Never diagnose or grade PPHN from one Doppler number.

25. Mild, Moderate or Severe?

A rigid echo-only grading system can be misleading.

Instead describe the actual physiology.

EXAMPLE: LESS SEVERE PHENOTYPE

- elevated RV pressure
- mild septal flattening
- preserved RV function
- mainly L → R ductal flow
- minimal R → L atrial shunting

MORE SEVERE PHYSIOLOGY

- RV pressure approaching systemic
- marked septal flattening
- bidirectional PDA
- significant R → L atrial flow
- preserved or beginning RV dysfunction

VERY SEVERE PHYSIOLOGY

- systemic/suprasystemic RV pressure pattern
- predominantly R → L ductal shunt
- marked septal displacement
- RV dilation/dysfunction
- impaired LV filling/output

ECHO PEARL

Describe the components rather than hiding complex physiology behind one label such as "severe PH."

26. PPHN Is Dynamic

Pulmonary vascular resistance can change rapidly with:

- oxygenation
- ventilation
- lung recruitment
- acidosis
- agitation
- systemic blood pressure
- underlying disease

Therefore one echo represents: **one physiological moment**, not a permanent state.

ECHO PEARL

Serial echocardiography is often more informative than arguing over a single pressure estimate.

Compare:

Pressure → shunts → RV function → LV filling

over time.

27. The Pulmonary Veins Must Be Seen

This point deserves repetition.

Obstructed pulmonary venous return can produce:

- pulmonary hypertension
- cyanosis
- respiratory distress
- right-heart pressure loading

and may easily be mistaken for PPHN.

Examine:

- right upper pulmonary vein
- right lower pulmonary vein where possible
- left upper pulmonary vein
- left lower pulmonary vein where possible
- LA connection
- confluence if anomalous anatomy suspected
- color flow
- spectral Doppler where obstruction is suspected

CRITICAL PEARL

"PPHN" plus pulmonary edema or unexplained severe respiratory compromise should make pulmonary venous obstruction a priority diagnosis to exclude.

28. Obstructed TAPVR — The Dangerous Mimic

An infant with obstructed TAPVR may look clinically and hemodynamically similar to severe PPHN.

Therefore:

Do not rely on pulmonary pressure findings to distinguish them.

Look for:

- failure to demonstrate pulmonary veins entering LA
- anomalous venous confluence
- abnormal vertical vein/systemic connection
- venous turbulence
- restrictive Doppler pattern
- obligatory atrial-level shunting

The neonatal cardiology reference specifically recommends urgent echo in babies thought to have PPHN to exclude obstructed anomalous pulmonary venous connection.

29. Other Structural Mimics

TGA

Can present with severe cyanosis and high pulmonary pressure.

Confirm: **VA connections**

Do not identify great arteries merely by position.

Pulmonary atresia / critical pulmonary stenosis

Can produce:

- right-to-left atrial shunting
- hypoxemia
- abnormal ductal flow

Examine pulmonary valve and RVOT.

Ebstein anomaly

Severe TR and functional pulmonary atresia may resemble severe PPHN physiology.

Examine TV morphology carefully.

Left-heart obstruction

Right-to-left ductal flow may reflect a systemic ductal lifeline rather than PPHN.

Always assess:

LV → aortic valve → arch

30. PPHN Versus Duct-Dependent Pulmonary Circulation

These can both produce cyanosis, but the ductus performs opposite physiological roles.

PPHN

High pulmonary pressure may drive:

PA → PDA → Aorta

Duct-dependent pulmonary circulation

Systemic arterial blood supplies pulmonary arteries:

Aorta → PDA → PA

ECHO PEARL

Follow the arrow. The direction of ductal blood flow fundamentally changes the physiology.

31. PPHN Versus Normal Transition

Finding	Normal Early Transition	PPHN Concern
RV pressure	Relatively elevated	Persistent systemic/suprasystemic pattern
RV prominence	Common	Marked dilation/dysfunction
Septum	Mild/transient flattening	Marked persistent flattening
PDA	May be bidirectional early	Significant persistent R→L component
PFO	Variable shunting	Persistent significant R→L flow
Oxygenation	Improves with transition	Persistent/labile hypoxemia

Window-by-Window PPHN Protocol

SUBCOSTAL

Start here. Assess:

- situs
- systemic veins
- pulmonary veins
- atrial septum
- direction of PFO flow
- ventricular septum
- outflow connections

Key question: Is the anatomy normal, and is systemic venous blood escaping right-to-left across the atrial septum?

APICAL FOUR-CHAMBER

Assess:

- RA size
- RV size
- TV morphology
- TR
- RV function
- LV filling
- LV function

Obtain CW through the TR jet when adequately aligned.

Key question: How high is RV pressure, and how well is the RV coping?

PARASTERNAL LONG AXIS

Assess:

- LV size
- LV systolic function
- septal position
- aortic valve
- LVOT

Key question: Is LV filling/output being compromised?

PARASTERNAL SHORT AXIS

This is the essential septal-pressure view. Assess:

- RV size
- septal configuration
- LV circularity/D-shape
- RVOT
- pulmonary valve
- MPA

Key question: What does septal geometry tell me about RV pressure?

HIGH LEFT PARASTERNAL / DUCTAL VIEW

Assess:

- PDA
- Direction, velocity/pattern
- MPA and branch PAs

Key question: Is PA pressure approaching or exceeding systemic pressure?

SUPRASTERNAL

Assess:

- aortic arch
- descending aorta
- ductus where necessary
- pulmonary venous structures where relevant

Key question: Am I missing systemic obstruction or abnormal venous anatomy?

The 60-Second PPHN Echo

For a rapidly deteriorating hypoxemic neonate:

1. Four chambers — Normal connections?

↓

2. Pulmonary veins — Do they enter LA?

↓

3. Great arteries — Normal VA connections?

↓

4. Pulmonary valve/RVOT — Patent?

↓

5. Aortic arch — Unobstructed?

↓

6. TR jet — RV pressure estimate possible?

↓

7. PSAX septum — Circular or D-shaped LV?

↓

8. PDA — L→R, bidirectional or R→L?

↓

9. PFO — Which direction?

↓

10. RV/LV function — Preserved or failing?

Then answer: Is this truly PPHN—and how is the heart coping?

Common Diagnostic Pitfalls

Pitfall 1 — CALLING ANY NEONATAL PULMONARY HYPERTENSION PPHN

Correction: Integrate postnatal age and clinical state.

Pitfall 2 — DIAGNOSING PPHN FROM A TR VELOCITY ALONE

Correction: Use multiple concordant signs.

Pitfall 3 — ASSUMING ABSENT TR MEANS NORMAL PA PRESSURE

Correction: Use septum, PDA, PFO and RV morphology.

Pitfall 4 — SEEING RIGHT-TO-LEFT PDA FLOW AND IMMEDIATELY CALLING PPHN

Correction: Exclude duct-dependent systemic circulation.

Pitfall 5 — SEEING RIGHT-TO-LEFT PFO FLOW AND CALLING PPHN

Correction: Exclude RVOT obstruction, pulmonary atresia, abnormal venous return and other CHD.

Pitfall 6 — FORGETTING PULMONARY VEINS

Correction: Specifically exclude obstructed TAPVR.

Pitfall 7 — REPORTING PRESSURE WITHOUT RV FUNCTION

Correction: Pressure describes afterload; function describes the ventricular response.

Pitfall 8 — IGNORING LV UNDERFILLING

Correction: Assess ventricular interaction and systemic output.

Pitfall 9 — USING ONE ECHO AS A FIXED DIAGNOSIS

Correction: PPHN physiology is dynamic. Serial assessment may be necessary.

36. Echo Pearls — The Page to Remember

Pearl 1: PPHN is a physiological diagnosis; first prove that the heart is structurally appropriate.

Pearl 2: High RV pressure shortly after birth can be transitional—context and persistence matter.

Pearl 3: No TR jet does not mean no pulmonary hypertension.

Pearl 4: Use TR + septal shape + PDA + PFO + RV function together.

Pearl 5: A D-shaped LV is a visual clue to significant RV loading.

Pearl 6: Right-to-left PDA flow indicates PA pressure competing with systemic pressure, but not its cause.

Pearl 7: Right-to-left atrial flow reflects the RA–LA pressure relationship, not specifically PA pressure.

Pearl 8: Ductal and atrial shunts do not have to point in the same direction.

Pearl 9: Pressure and ventricular function must be reported separately.

Pearl 10: A failing RV may generate a lower-than-expected pressure—do not mistake this for improvement.

Pearl 11: A small LV may be underfilled rather than truly hypoplastic.

Pearl 12: Never diagnose PPHN without deliberately assessing pulmonary venous return.

Chapter Key Points

PPHN represents failure or delay of the normal postnatal fall in pulmonary vascular resistance.

The echocardiographic diagnosis should never be based on one finding.

A useful assessment integrates:

STRUCTURE + RV PRESSURE + SEPTAL SHAPE + PDA + PFO + RV FUNCTION + LV FILLING

The first responsibility of echocardiography is to exclude structural congenital heart disease.

The most important PPHN mimic to actively exclude is **obstructed pulmonary venous return**, particularly obstructed TAPVR.

TR Doppler can estimate RV pressure when the jet is complete and well aligned, but lack of a usable TR jet does not exclude pulmonary hypertension.

Parasternal short-axis septal configuration provides an important visual assessment of RV pressure loading.

Ductal flow reflects the relationship between pulmonary and systemic arterial pressures.

Atrial shunting reflects the relationship between right and left atrial pressures.

RV function determines how well the heart is tolerating increased pulmonary vascular load.

LV filling may fall because of reduced pulmonary venous return and adverse ventricular interaction.

PPHN is dynamic, so serial echocardiography should focus on changes in **pressure, shunt direction and ventricular performance**, not merely repeat a diagnostic label.

MASTER PRINCIPLE

In PPHN, do not chase a pressure number. Reconstruct the physiology.

The PPHN Echo Formula

EXCLUDE CHD → ESTIMATE PRESSURE → FOLLOW THE SHUNTS → ASSESS THE RV → ASSESS THE LV → REASSESS OVER TIME

And the question that should remain in the echocardiographer's mind throughout the study is:

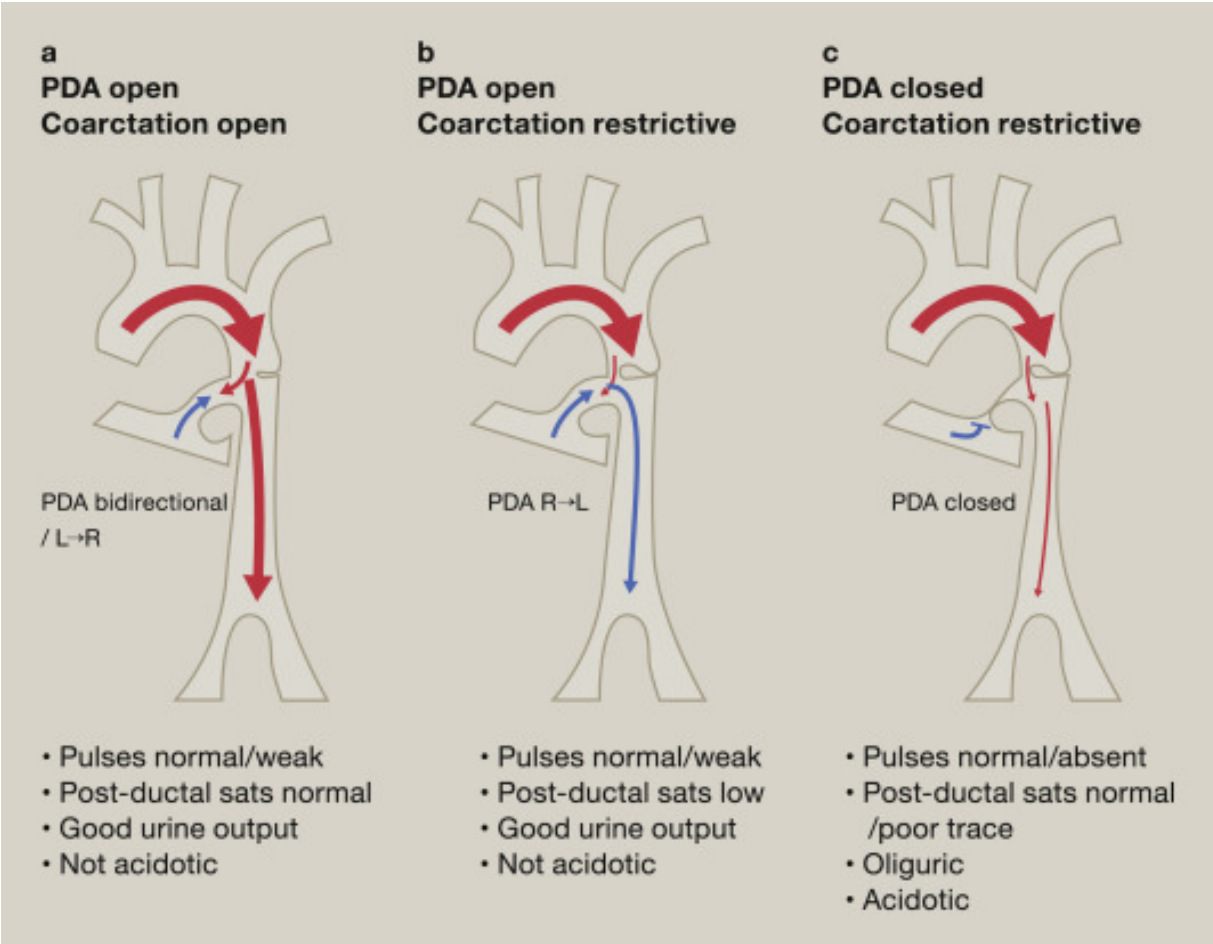
"Is this truly abnormal pulmonary vascular transition—and how is it affecting the whole circulation?"

CHAPTER 12

Neonatal Coarctation of the Aorta

The Lesion the Ductus Can Hide

When a Narrow Arch Looks Deceptively Open



Learning Objectives

By the end of this chapter, the learner should be able to:

1. Explain why neonatal coarctation may be difficult to diagnose while the PDA remains patent.
2. Perform a systematic echocardiographic assessment of the entire neonatal aortic arch.
3. Identify the proximal and distal transverse arch, isthmus, ductal insertion and descending aorta.
4. Recognize a discrete posterior shelf and diffuse arch hypoplasia.
5. Distinguish anatomical severity from Doppler gradient.
6. Interpret ductal flow in relation to systemic perfusion.
7. Recognize retrograde transverse-arch or descending-aortic supply from the PDA.
8. Use color and spectral Doppler appropriately across the arch.
9. Recognize abnormal abdominal aortic Doppler as a downstream clue to obstruction.
10. Differentiate between pre-ductal and post-ductal coarctation.

1. Why Neonatal Coarctation Is Different

In the older infant or child, significant coarctation may produce a clear focal narrowing with accelerated Doppler flow and a measurable pressure gradient.

The neonate is different.

The **ductus arteriosus may still be widely patent**, providing an alternative route for blood to reach the descending aorta. A severe anatomical obstruction may therefore initially produce surprisingly little Doppler acceleration.

Therefore:

Low gradient does not exclude severe neonatal coarctation.
This is the core message of the chapter.

2. The Anatomy You Must Understand

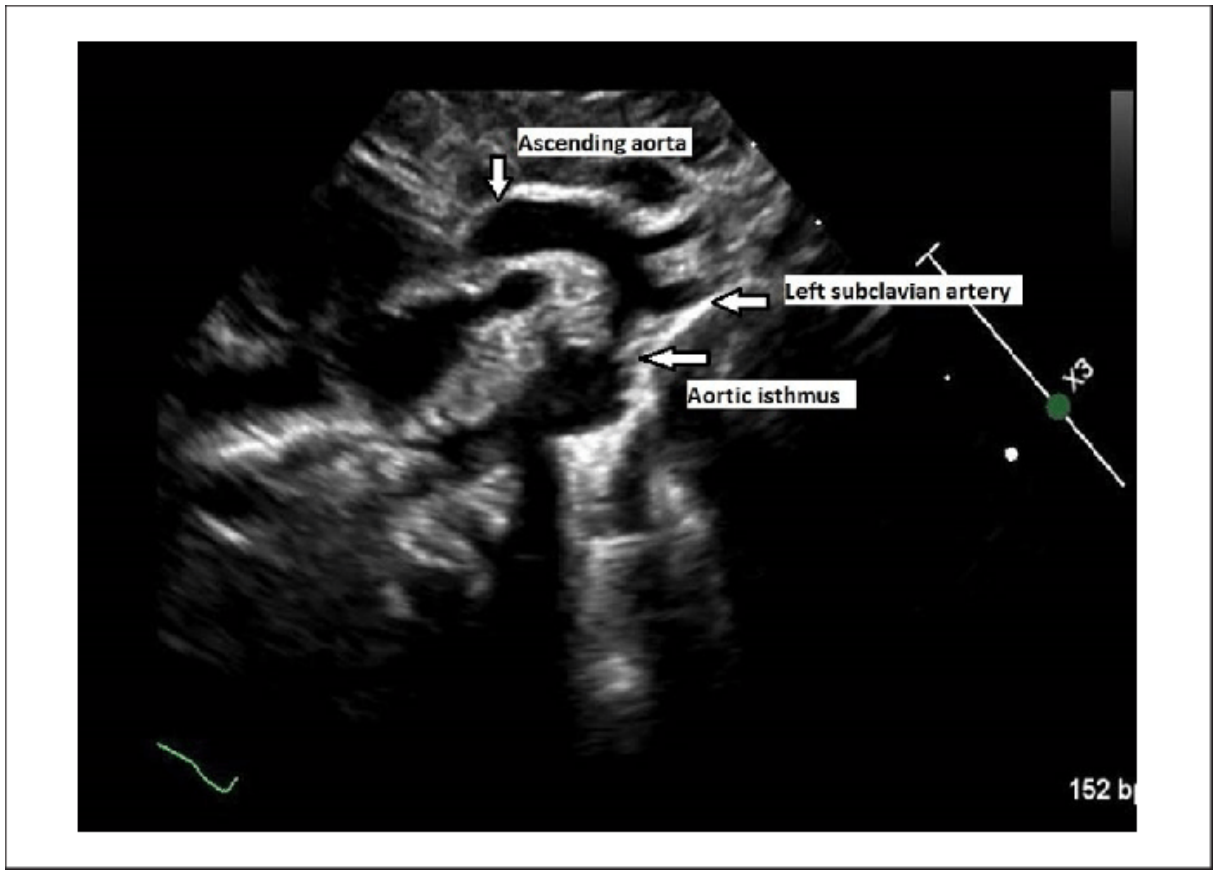
The neonatal arch should be reconstructed systematically:

Ascending aorta → proximal transverse arch → distal transverse arch → isthmus → descending aorta
The ductus joins the descending aortic region near the isthmus.

Moss and Adams describes typical coarctation as localized narrowing just beyond the left subclavian artery, often with a posterior shelf projecting from the posterior aortic wall toward the ductal region. Associated isthmic/transverse-arch hypoplasia may coexist.

Important anatomical landmarks

- Ascending aorta
- Brachiocephalic/innominate artery
- Left common carotid artery
- Left subclavian artery
- Proximal transverse arch
- Distal transverse arch
- Aortic isthmus, Descending thoracic aorta
- Ductal insertion



ECHO PEARL

Do not simply "see an arch." Identify its segments.

3. Where Should the Arch Be Measured?

- **proximal transverse arch** between the innominate and left carotid arteries
- **distal transverse arch** between the left carotid and left subclavian arteries
- **aortic isthmus** distal to the left subclavian artery

Measurements should be interpreted using appropriate pediatric reference data and Z-scores rather than arbitrary absolute values.

TECHNIQUE TIP

For reproducibility:

Measure → record exact level → use the same normative dataset → interpret with anatomy

Do not compare a distal transverse-arch measurement with normative data intended for the isthmus.

4. The Central Echo Question

Do not begin with:

"What is the coarctation gradient?"

Begin with:

"Is there an adequate uninterrupted anatomical pathway from the LV to the descending aorta?"

Then ask:

1. Is the LV supplying the ascending aorta adequately?
2. Is the transverse arch of adequate caliber?
3. Is the isthmus narrow?
4. Is there a posterior shelf?
5. What is the PDA doing?
6. Where is the descending aortic blood actually coming from?

5. The Ductus–Coarctation Relationship

The fetal circulation explains the diagnostic difficulty.

Before birth, much of lower-body flow reaches the descending aorta through the ductus rather than through the isthmus. The neonatal cardiology reference therefore notes why fetal and early postnatal coarctation can be difficult to identify: the isthmus normally carries relatively limited fetal flow and lies very close to the ductal insertion.

After birth:

PVR falls

↓

LV output increases

↓

Ductus begins to constrict

↓

More systemic flow must pass through the arch/isthmus

If significant coarctation is present, this normal transition can suddenly unmask the obstruction.

ECHO PEARL

The ductus can temporarily convert a critical lesion into a deceptively mild-looking echo.

6. How a Large PDA Masks Coarctation

Imagine severe juxtaductal narrowing.

Without a PDA:

LV → arch → narrow isthmus → descending Ao

There may be a clear velocity increase.

With a wide-open PDA:

RV → PA → PDA → descending Ao

The descending aorta can still receive substantial blood despite severe isthmic obstruction.

Therefore the baby may temporarily have:

- reasonable distal flow
- palpable femoral pulses
- modest blood-pressure difference
- relatively low Doppler gradient

CRITICAL ECHO PEARL

A large PDA decreases the diagnostic value of the instantaneous coarctation gradient.

7. The Suprasternal Long-Axis View — Your Main View

This is the key window.

Aim to demonstrate:

Ascending Ao → entire transverse arch → isthmus → descending Ao

Optimize:

- shallow enough depth, narrow sector
- high-frequency transducer where feasible
- focal zone at the arch
- grayscale before color

BEGIN WITH 2D

Look for:

- focal narrowing
- diffuse arch hypoplasia
- posterior shelf
- abnormal arch angulation
- ductal ampulla
- poststenotic enlargement
- continuity of the entire arch

ECHO PEARL

A beautiful color arch with poor 2D anatomy is not an adequate coarctation study.

8. Never Diagnose From One Frozen Arch Image

The aortic arch is three-dimensional and curved.

A single sagittal slice can:

- exaggerate narrowing
- miss the narrowest segment
- or make ductal tissue look like normal aortic continuity

PRACTICAL TEACHING TECHNIQUE

From the suprasternal notch:

Find → sweep right-to-left → sweep anterior-to-posterior → identify branches → find isthmus → identify PDA

This deliberate sweep is more reliable than repeatedly trying to reproduce one textbook image.

ECHO PEARL

Coarctation is a sweep diagnosis, not a screenshot diagnosis.

9. The Posterior Shelf

A discrete coarctation often appears as an echogenic ridge projecting from the posterior wall of the aorta near the isthmus.

However:

Not every neonatal coarctation has an obvious shelf.

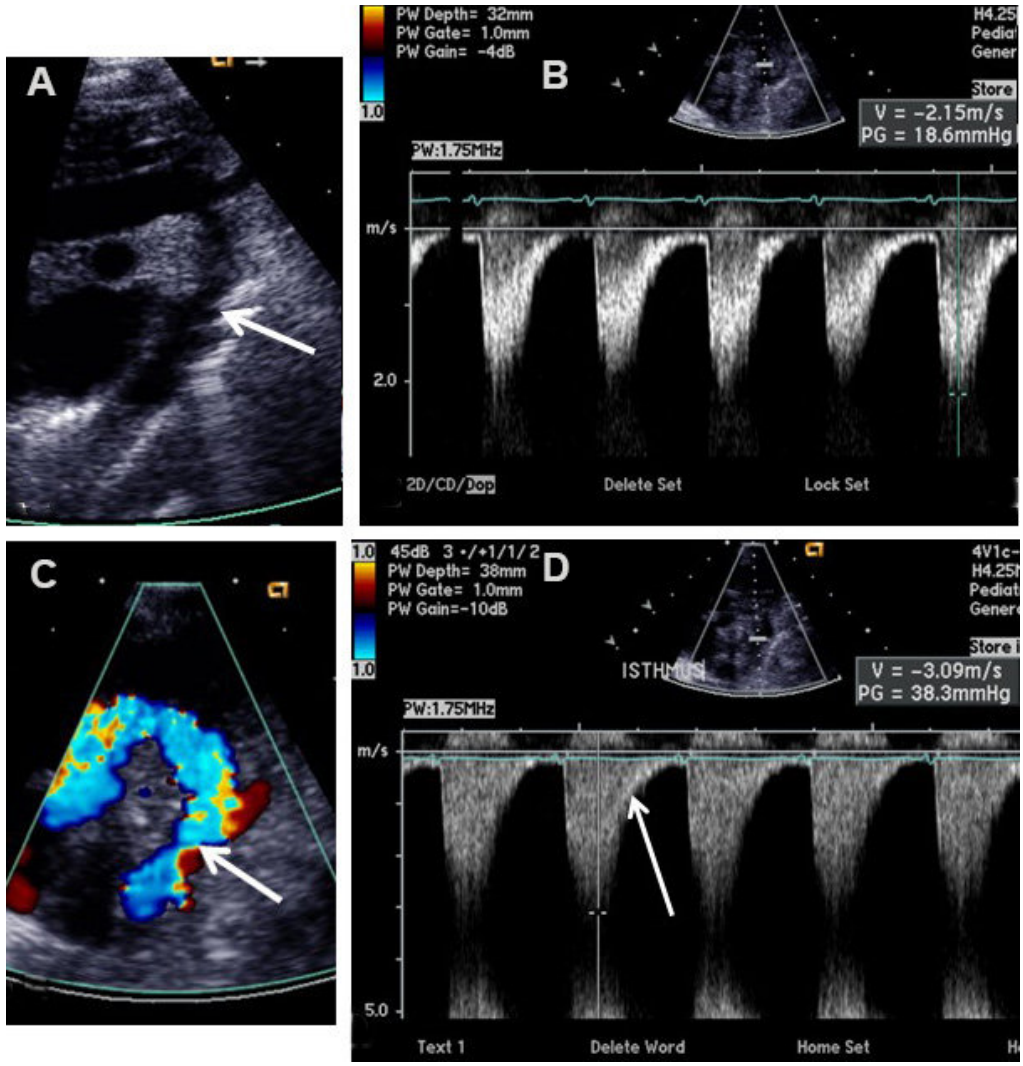
Some babies have predominantly:

- distal transverse-arch hypoplasia
- long-segment narrowing
- diffuse isthmic hypoplasia

rather than a dramatic discrete membrane-like narrowing.

ECHO PEARL

Absence of a visible shelf does not exclude coarctation.



10. Discrete Coarctation Versus Arch Hypoplasia

These often coexist but should be described separately.

Discrete coarctation

Usually:

- focal juxtaductal narrowing
- posterior shelf
- abrupt caliber change

Arch hypoplasia

More diffuse reduction in caliber involving:

- proximal transverse arch
- distal transverse arch
- isthmus
- or combinations

BETTER REPORTING

Avoid:

"Coarctation present."

Prefer:

"Hypoplastic distal transverse arch and isthmus with additional discrete juxtaductal posterior shelf."

That tells the surgeon much more.

11. Identify the Head-and-Neck Branches

The branch vessels provide important anatomical landmarks.

Identify sequentially:

Brachiocephalic → LCCA → LSCA

Then determine:

Where is the narrowing relative to the left subclavian artery?

This helps distinguish:

- distal transverse-arch narrowing
- isthmic narrowing
- discrete juxtaductal coarctation

ECHO PEARL

If you cannot identify the arch branches, be cautious about declaring the arch anatomy completely defined.

12. Look Carefully at the Isthmus–Ductus Junction

This is where the diagnosis is often won or lost.

The ductal ampulla may blend into the descending aortic region.

A wide ductus may create the impression that the isthmus is wider than it really is.

Therefore inspect:

- proximal isthmus
- distal isthmus
- posterior wall
- ductal insertion
- true descending aorta

TECHNIQUE TIP

Use grayscale first.

Then place a **small color box** over the region.

A large color box can obscure the vessel walls and hide subtle narrowing.

13. The "Wide Open Arch" Illusion

One of the most dangerous errors occurs when the ductal ampulla is mistaken for the aortic isthmus.

You may see a large vascular channel entering the descending aorta and conclude:

"The arch looks wide."

But the structure may actually be:

PDA → descending aorta
while the true isthmus is substantially smaller.

ECHO PEARL
Always separate the true aortic isthmus from the ductal ampulla.
Trace each structure back to its origin.

14. Color Doppler — Use It to Map, Not to Diagnose Alone

Color helps identify:

- the site of acceleration
- flow through the transverse arch
- flow through the isthmus
- ductal direction
- retrograde arch flow
- disturbed distal flow

But color settings matter.

Excessively high velocity scale may miss low-velocity ductal or retrograde flow.

Excessively low scale may produce excessive aliasing that hides anatomical localization.

PRACTICAL RULE

2D anatomy → targeted color → PW localization → CW quantification
Not the reverse.

15. Spectral Doppler Across Coarctation

Once the anatomical narrowing is identified, interrogate with PW and CW Doppler.

A significant coarctation may show:

- systolic acceleration
- prolonged systolic flow
- persistent antegrade flow into diastole
- reduced return toward baseline

Moss and Adams describes the classic severe-coarctation spectral pattern as showing persistent diastolic flow across the obstruction.

ECHO PEARL

Look at the shape of the Doppler envelope, not only the peak number.

16. Why the Bernoulli Gradient Can Mislead

The simplified Bernoulli equation:

$$\Delta P = 4V^2$$

works best when assumptions regarding proximal velocity and flow are reasonable.

In neonatal coarctation those assumptions may fail because of:

- large PDA
- diffuse arch hypoplasia
- low cardiac output
- multiple levels of obstruction and abnormal proximal velocity

ECHO PEARL

Do not convert every neonatal coarctation into a single gradient.

17. The Most Important Low-Gradient Scenarios

Scenario 1 — Large PDA

Ductal blood bypasses/decompresses the obstruction.

Scenario 2 — Severe LV dysfunction

There is insufficient flow to generate high velocity.

Scenario 3 — Diffuse arch hypoplasia

No single discrete jet produces a dramatic peak gradient.

Scenario 4 — Near-critical obstruction with distal ductal supply

Descending aortic flow is maintained from the pulmonary artery.

MASTER RULE

Low flow + severe obstruction = potentially low Doppler gradient.

18. Determine the Direction of Ductal Flow

This is fundamental.

Normal postnatal tendency

Aorta → PDA → PA

Significant systemic ductal dependence

PA → PDA → descending aorta

If descending aortic perfusion depends substantially on the RV through the PDA, the lesion is physiologically critical.

ECHO PEARL

Coarctation severity is not just "how narrow is it?" It is "how dependent is systemic perfusion on the ductus?"

19. Bidirectional Ductal Flow Requires Thought

A ductus may show:

- right-to-left flow in part of systole
- left-to-right flow at another phase
- fully bidirectional flow

This can occur because pulmonary and systemic pressures are dynamically related during neonatal transition.

Therefore:

Do not label bidirectional flow as "normal transition" automatically.
In a baby with suspicious arch anatomy, bidirectional or right-to-left ductal flow should increase attention to systemic outflow obstruction.

20. Retrograde Transverse-Arch Flow

This is a major red flag.

If the descending aorta is supplied by the PDA, blood may travel:

PA → PDA → descending Ao
and then:

descending/ductal region → transverse arch
in a retrograde direction.

This indicates that normal LV antegrade systemic output through the arch is inadequate.

Severe arch obstruction can show retrograde filling of the arch, while ductal supply may cause the measured coarctation gradient to be underestimated.

CRITICAL ECHO PEARL

Retrograde arch flow is a physiological finding, not just a Doppler curiosity.
It should trigger urgent assessment of:

- LV adequacy
- aortic valve
- entire arch and ductal dependency

21. Follow the Blood Backward

This practical maneuver is extremely useful.

Place color over the descending aorta.

Ask:

Where is the blood entering this vessel from?
If it originates through the PDA:

Descending Ao ← PDA ← PA ← RV
Then trace the aortic arch upward.

If blood is moving from the ductal region toward the transverse arch:

Systemic circulation is being supported retrogradely.

ECHO PEARL
Do not assume that flow in the descending aorta proves adequate LV-to-aorta continuity.

22. The Abdominal Aorta — A Downstream Clue

Do not finish the coarctation study at the suprasternal notch.

Obtain PW Doppler from the abdominal descending aorta.

In severe obstruction, the waveform may show:

- reduced systolic pulsatility
- blunted systolic peak
- delayed systolic upstroke
- persistent forward diastolic flow

The practical echocardiography reference illustrates markedly reduced pulsatility in severe coarctation and recommends spectral assessment through the descending and abdominal aorta.

ECHO PEARL
The abdominal aorta can tell you that something upstream is wrong—even when the suprasternal image is difficult.

23. But Abdominal Doppler Is Not the Diagnosis

An abnormal abdominal aortic waveform can support the suspicion of arch obstruction.

However it can be affected by:

- low cardiac output
- large PDA
- other systemic outflow lesions
- severe ventricular dysfunction

Therefore:

Abdominal Doppler supports the diagnosis; it does not replace direct arch imaging.

24. Assess the Left Ventricle

Every neonatal coarctation study must assess the LV.

Look for:

- LV size
- systolic function
- hypertrophy
- dilation
- endocardial appearance
- mitral valve
- LVOT
- aortic valve

In a critically ill neonate, LV dysfunction may result from excessive afterload and systemic hypoperfusion.

ECHO PEARL

A sick LV can make a severe coarctation look less impressive by Doppler because it generates less flow.

25. Examine the Aortic Valve

Coarctation is often part of a broader left-sided phenotype.

Assess:

- number/morphology of cusps
- annulus
- opening
- stenosis
- regurgitation

The bicuspid/unicuspid aortic valve and Shone-type multiple left-heart obstruction are among important associations of left-heart disease.

ECHO PEARL

After finding coarctation, do not stop at the arch. Search the rest of the left heart.

26. Examine the Mitral Valve and LV Inflow

Assess:

- annulus
- leaflet morphology
- papillary muscles
- chordal anatomy
- inflow obstruction

Especially look for combinations suggesting:

multilevel left-heart obstruction

such as mitral obstruction + LVOT/aortic obstruction + coarctation.

27. Look for Associated Intracardiac Lesions

The comprehensive study should also identify:

- VSD
- ASD/PFO
- PDA
- bicuspid aortic valve
- other LVOT obstruction
- arch branching abnormalities

Associated shunts can materially alter:

- ventricular loading
- pulmonary pressure
- Doppler gradients
- clinical presentation

ECHO PEARL

Coarctation physiology is influenced by the rest of the heart.

28. The VSD Can Change the Picture

A sizeable VSD may allow pressure equalization or alter ventricular output distribution.

Therefore a baby with:

- coarctation
- VSD
- large PDA

may not display the classic "isolated coarctation" physiology.

This reinforces the core rule:

Reconstruct the entire circulation before interpreting the arch gradient.

29. The Collapsing Neonate

A baby with critical coarctation may deteriorate as the ductus closes.

The sequence is:

Ductal constriction

↓

Descending aortic perfusion falls

↓

LV afterload rises

↓

Systemic perfusion deteriorates

↓

Renal/mesenteric perfusion falls

↓

Lactic acidosis develops

↓

LV dysfunction/shock

At that stage, the baby may be labelled:

"Sepsis" — "Metabolic acidosis" — "Poor feeding" — "Respiratory distress"

when the fundamental problem is critical systemic outflow obstruction.

30. Do Not Be Reassured by Femoral Pulses Early

While the ductus is open, lower-body perfusion can be maintained.

Thus the neonate may still have palpable femoral pulses despite important coarctation.

CLINICAL–ECHO PEARL

Normal femoral pulses on Day 1 do not exclude critical coarctation.

The same logic applies to a modest arm–leg pressure difference while the PDA remains open.

Serial Echocardiography — Sometimes the Correct Answer Is "Not Yet Certain"

This is a particularly important concept for trainees.

When there is:

- suspicious isthmic anatomy
- arch disproportion
- large PDA
- uncertain posterior shelf
- no convincing gradient

the appropriate conclusion may be:

"Coarctation cannot be excluded while the ductus remains widely patent."

This is better than falsely declaring the arch normal.

PRACTICAL TEACHING PRINCIPLE

Diagnostic uncertainty should be documented—not hidden.

Serial imaging as the ductus changes can clarify anatomy and physiology.

33. When Repeat Echo Is Most Informative

Serial reassessment should focus on what changes as ductal flow decreases:

Anatomy

Does the isthmus appear increasingly narrow?

Doppler

Does arch velocity increase?

Diastolic flow

Does persistent forward flow become more apparent?

Distal circulation

Does abdominal aortic pulsatility worsen?

LV

Does afterload-related dysfunction appear?

Clinical findings

Do pulses, perfusion or pressure differences change?

The serial study should answer a new physiological question rather than simply repeat the first examination.

34. Practical Window-by-Window Protocol

SUBCOSTAL

Assess:

- situs
- systemic venous return
- atrial septum
- ventricular balance
- LV function
- abdominal aortic Doppler

Key question: Is systemic output reaching the lower body normally?

APICAL

Assess:

- LV and RV relative size
- ventricular function
- AV valves
- VSD
- inflow abnormalities

Key question: Is there broader left-heart disease?

PARASTERNAL LONG AXIS

Assess:

- mitral valve
- LV
- LVOT
- aortic valve
- ascending aorta
- ventricular function

Key question: Can the LV generate adequate systemic output?

PARASTERNAL SHORT AXIS

Assess:

- aortic valve morphology
- ventricular function
- associated VSD
- RV pressure if relevant

SUPRASTERNAL LONG AXIS

This is the central coarctation view. Assess:

Ascending Ao → proximal transverse arch → distal transverse arch → isthmus → descending Ao → PDA

Measure relevant arch segments. Add: color, PW, CW.

Key question: Is this anatomically and physiologically an adequate systemic pathway?

SUPRASTERNAL SHORT AXIS

Assess:

- arch sidedness
- branch pattern
- descending aorta
- branch pulmonary arteries where useful

The 60-Second Coarctation Screen

For a sick neonate with poor systemic perfusion:

1. LV — Normal size and function?

↓

2. Aortic valve — Adequate opening?

↓

3. Ascending aorta — Normal caliber?

↓

4. Transverse arch — Proximal and distal segments adequate?

↓

5. Isthmus — Narrow?

↓

6. Posterior shelf — Present?

↓

7. PDA — Large, restrictive, direction?

↓

8. Descending aorta — Antegrade from LV or supplied by ductus?

↓

9. Abdominal Doppler — Normal pulsatility?

10. What happens if the ductus closes?

36. The Most Important Diagnostic Traps

Trap 1 — "GRADIENT IS ONLY 10–15 MMHG."

Correction: Judge anatomy and physiology, not gradient alone. A large PDA may bypass/decompress the obstruction.

Trap 2 — "THERE IS GOOD FLOW IN THE DESCENDING AORTA."

Correction: Determine its source and direction. That flow may originate from the PDA.

Trap 3 — "THE ARCH LOOKS OPEN ON COLOR."

Correction: Examine grayscale anatomy and measure the arch. Color can fill a narrow vessel.

Trap 4 — "NO OBVIOUS POSTERIOR SHELF."

Correction: Assess diffuse transverse/isthmic hypoplasia. Not all neonatal coarctation is discrete.

Trap 5 — "FEMORAL PULSES ARE PRESENT."

Correction: The PDA may still be supporting lower-body flow.

Trap 6 — "PRENATAL SUSPICION, BUT POSTNATAL GRADIENT IS LOW."

Correction: The ductus may still be masking the lesion.

Trap 7 — MEASURING THE WRONG STRUCTURE

Correction: Trace the true arch and PDA separately. The ductal ampulla can be mistaken for the isthmus.

Trap 8 — STOPPING AFTER THE ARCH DIAGNOSIS

Correction: Associated left-heart abnormalities may alter management.

Trap 9 — POOR SUPRASTERNAL IMAGE INTERPRETED AS NORMAL

Correction: "Not visualized adequately" is not equivalent to "normal."

37. Beginner Versus Trainee Expectations

Echo Technician / Beginner	Paediatric Cardiology Trainee
Obtain a clear suprasternal arch view	Reconstruct full arch anatomy
Identify PDA	Determine ductal physiological role
Recognize obvious narrowing	Distinguish focal vs diffuse disease
Record arch Doppler	Interpret low-gradient physiology
Obtain abdominal aortic Doppler	Integrate distal waveform with anatomy
Assess LV function	Assess systemic dependency
Identify associated VSD	Define complete left-heart phenotype
Recognize uncertainty	Determine need for serial reassessment/additional imaging

38. A Practical Reporting Framework

Avoid:

"Mild coarctation, peak gradient 12 mmHg."

in a newborn with a large PDA.

That statement may seriously underestimate the lesion.

A better report is:

"The distal transverse arch and isthmus are hypoplastic with focal juxtaductal narrowing/posterior shelf. A large PDA remains patent and contributes to descending aortic flow. The Doppler gradient across the suspected coarctation is low but is not a reliable marker of anatomical severity in the presence of significant ductal flow."

If uncertain:

"The isthmus is small with suspicious juxtaductal morphology; however, the PDA remains widely patent and coarctation severity cannot be reliably determined at present. Serial assessment following change in ductal patency is recommended according to the clinical context."

If duct-dependent:

"Right-to-left ductal flow supplies the descending aorta with retrograde flow within the transverse arch, consistent with duct-dependent systemic perfusion."

39. Echo Pearls — The Page to Remember

Pearl 1: Low gradient does not exclude neonatal coarctation.

Pearl 2: The PDA can bypass the obstruction and hide its physiological severity.

Pearl 3: Measure the arch segment by segment—not as one structure.

Pearl 4: Separate the true isthmus from the ductal ampulla.

Pearl 5: A posterior shelf is helpful when present, but its absence does not exclude coarctation.

Pearl 6: Color shows flow; grayscale defines anatomy.

Pearl 7: Persistent diastolic flow across the coarctation supports significant obstruction.

Pearl 8: Retrograde transverse-arch flow is a major sign of inadequate antegrade systemic output.

Pearl 9: Flow in the descending aorta may be ductal rather than LV-derived.

Pearl 10: Abdominal aortic Doppler is an important downstream clue.

Pearl 11: LV dysfunction may lower the gradient rather than make the lesion less severe.

Pearl 12: When a large PDA prevents certainty, serial echo is part of the diagnosis—not a failure of diagnosis.

Chapter Key Points

Neonatal coarctation is one of the most important **duct-masked lesions** in congenital echocardiography.

The arch should be reconstructed from:

Ascending Ao → proximal transverse arch → distal transverse arch → isthmus → descending Ao
and the PDA should be identified separately.

The echocardiographic assessment must integrate:

2D anatomy + arch dimensions + posterior shelf + color flow + spectral Doppler + PDA direction + LV function + distal systemic flow
A large PDA can maintain descending aortic perfusion and markedly reduce the apparent gradient across a critical coarctation. This is why anatomical narrowing and ductal physiology may be more informative than the instantaneous Doppler gradient in the early neonatal period.

The suprasternal long-axis view is central, but the study should also assess the abdominal aortic waveform, LV, mitral valve, aortic valve and associated intracardiac abnormalities.

When anatomy is suspicious but the ductus remains widely patent, an appropriate conclusion may be:

"Coarctation cannot yet be excluded."

MASTER PRINCIPLE
In neonatal coarctation, never ask only "What is the gradient?" Ask "What is the anatomy, where is the descending aortic blood coming from, and what will happen when the ductus closes?"
The Neonatal Coarctation Formula

ARCH ANATOMY → ISTHMUS → DUCTAL INSERTION → FLOW DIRECTION → DISTAL PERFUSION → LV FUNCTION → SERIAL CHANGE
And the one sentence every learner should remember is:

"A patent ductus can make a critical coarctation look deceptively mild."

Pre-Ductal, Post-Ductal and Juxtaductal Coarctation

The terms **pre-ductal** and **post-ductal coarctation** describe the position of the aortic narrowing **relative to the insertion of the ductus arteriosus**. They are useful for understanding neonatal physiology, although modern congenital cardiology more often describes the lesion anatomically as **juxtaductal coarctation with or without arch hypoplasia**.

PRE-DUCTAL COARCTATION

The narrowing lies **proximal to the ductal insertion**, usually around the distal transverse arch/isthmus.

Conceptually:

Ascending aorta → arch → COARCTATION → ductal insertion → descending aorta
Because the obstruction is before the ductus, the descending aorta can receive blood from:

RV → pulmonary artery → PDA → descending aorta
So the PDA may temporarily bypass the obstruction.

This is why a neonate with severe pre-ductal coarctation may initially have reasonable femoral pulses and relatively preserved lower-body perfusion while the ductus remains open.

As the ductus constricts:

ductal flow falls → descending aortic flow falls → systemic perfusion deteriorates → acidosis/shock
This is the classic **duct-dependent systemic circulation** pattern.

ECHO PEARL

In pre-ductal/juxtaductal neonatal coarctation, the PDA can hide the severity of the obstruction.
The Doppler gradient across the coarctation may therefore be low despite severe anatomical narrowing.

POST-DUCTAL COARCTATION

The narrowing lies **distal to the ductal insertion**.

Conceptually:

Ascending aorta → arch → ductal insertion → COARCTATION → descending aorta
Here, ductal blood enters the aorta **before** the obstruction, so the PDA cannot effectively bypass the distal narrowing.

This pattern is classically associated with later presentation, when collateral circulation has had time to develop.

Typical consequences in older children or adults include:

- upper-body hypertension
- weak/delayed femoral pulses
- arm–leg blood pressure gradient
- collateral arterial enlargement
- rib notching in longstanding disease

ECHO PEARL

Post-ductal physiology depends less on the PDA and more on the severity of the fixed obstruction and development of collaterals.

JUXTADUCTAL COARCTATION

In reality, many neonatal coarctations are best described as **juxtaductal**: the narrowing is at or very near the site where the ductus inserts into the descending aorta.

This is the most useful concept for neonatal echo because the anatomy often involves:

- distal transverse arch hypoplasia
- isthmic hypoplasia
- posterior shelf
- ductal tissue extending into the aortic wall

So the older "pre-ductal vs post-ductal" classification is often too simplistic.

A practical way to remember it is:

Type	Narrowing Relative to PDA	PDA Can Bypass?	Typical Physiology
Pre-ductal	Before ductal insertion	Usually yes	Neonatal, duct-dependent systemic flow
Juxtaductal	At ductal insertion	Often partly	Most common neonatal pattern
Post-ductal	Beyond ductal insertion	Usually no	Later presentation, collaterals may develop

IMPORTANT NEONATAL ECHO DISTINCTION

For a newborn, do not stop at saying **"pre-ductal coarctation."** Instead describe the actual anatomy:

"Hypoplastic distal transverse arch and isthmus with discrete juxtaductal narrowing adjacent to the ductal insertion."

Then describe the physiology:

"The PDA supplies the descending aorta with right-to-left ductal flow."

That gives much more useful information than the historical label alone.

MASTER CONCEPT

Pre-ductal = the ductus can rescue the lower body.

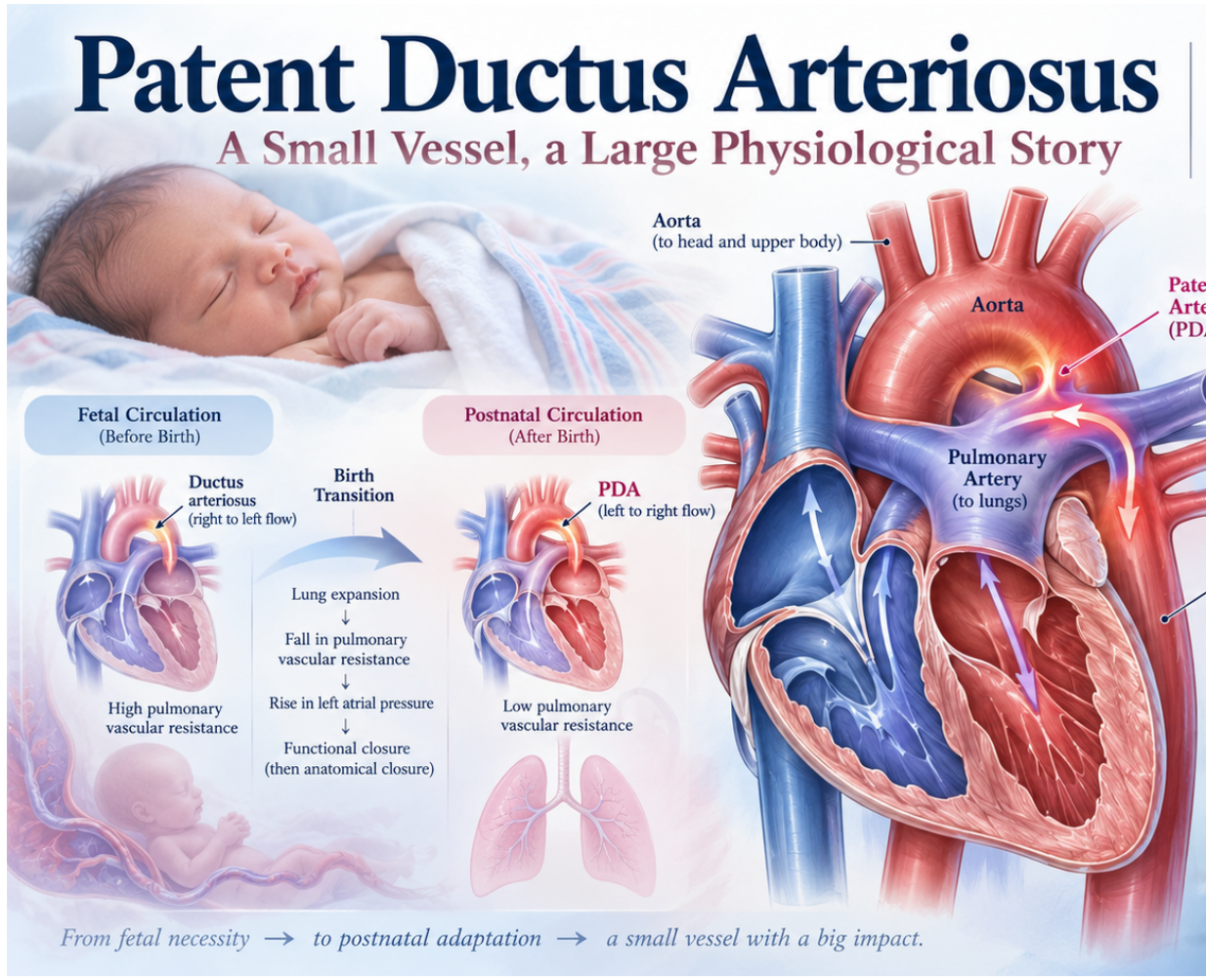
Post-ductal = the obstruction lies beyond the ductal rescue route.

Juxtaductal = the neonatal reality in many cases.

CHAPTER 13

Patent Ductus Arteriosus

A Small Vessel, a Large Physiological Story



Learning Objectives

By the end of this chapter, the learner should be able to:

1. Describe the anatomical relationship of the ductus arteriosus to the pulmonary artery and descending aorta.
2. Explain how ductal physiology changes during normal neonatal transition.
3. Obtain appropriate echocardiographic views for PDA assessment.
4. Identify the PDA using 2D and color Doppler without mistaking adjacent vessels for the ductus.
5. Determine the **direction of ductal flow** and relate it to pulmonary and systemic pressure.
6. Recognize restrictive, nonrestrictive, left-to-right, right-to-left, and bidirectional ductal flow patterns.
7. Assess PDA morphology and its relationship to the branch pulmonary arteries and aortic isthmus.
8. Evaluate the hemodynamic consequences of left-to-right PDA shunting.
9. Assess LA and LV volume loading associated with a significant PDA.
10. Use descending and abdominal aortic Doppler as supplementary markers of systemic runoff.

1. Why the PDA Deserves More Than a Yes/No Diagnosis

A report stating:

"PDA present."

is incomplete.

The clinically useful echocardiographic questions are:

1. How large is the ductus anatomically?
2. Which direction is blood flowing?
3. How restrictive is it?
4. What does the Doppler pattern tell us about pulmonary and systemic pressures?
5. What effect is the shunt having on the heart and systemic circulation?
6. Is the ductus harmful—or is it keeping the baby alive?

That last distinction is especially important in neonatal echocardiography.

2. Normal Ductal Anatomy

The ductus arteriosus provides a vascular connection between the pulmonary and systemic circulations.

In the usual anatomy, it connects the region of the:

Left pulmonary artery / pulmonary arterial side
to the:

Descending aorta near the aortic isthmus
During fetal life the ductus is essential because pulmonary vascular resistance is high and the lungs are not responsible for gas exchange.

Fetal ductal flow is predominantly:

Pulmonary artery → Ductus → Descending aorta
After birth, the pressure relationship changes.

3. What Happens to Ductal Flow After Birth?

With the first breaths:

Lungs expand

↓

Pulmonary vascular resistance falls

↓

Pulmonary artery pressure falls

↓

Systemic vascular resistance rises after placental separation

↓

Pressure gradient across the ductus reverses

The ductal flow therefore progressively changes toward:

Aorta → PDA → Pulmonary artery

before functional closure.

Early ductal flow can be **right-to-left, bidirectional, or left-to-right depending on the stage of transition**, so postnatal age must always be considered.

ECHO PEARL

The direction of PDA flow is physiology displayed in real time.

Never interpret ductal color without knowing the baby's age and surrounding anatomy.

4. The Three Fundamental Ductal Flow Patterns

A. Left-to-Right

Aorta → PDA → Pulmonary artery

Usually indicates: **Systemic pressure > pulmonary artery pressure**

This becomes the typical PDA pattern as neonatal PVR falls.

B. Right-to-Left

Pulmonary artery → PDA → Aorta

Consider:

- high pulmonary vascular resistance
- pulmonary hypertension
- normal very early transition in selected newborns
- duct-dependent systemic circulation

ECHO PEARL

Right-to-left PDA flow is not a diagnosis. It is a pressure relationship that requires explanation.

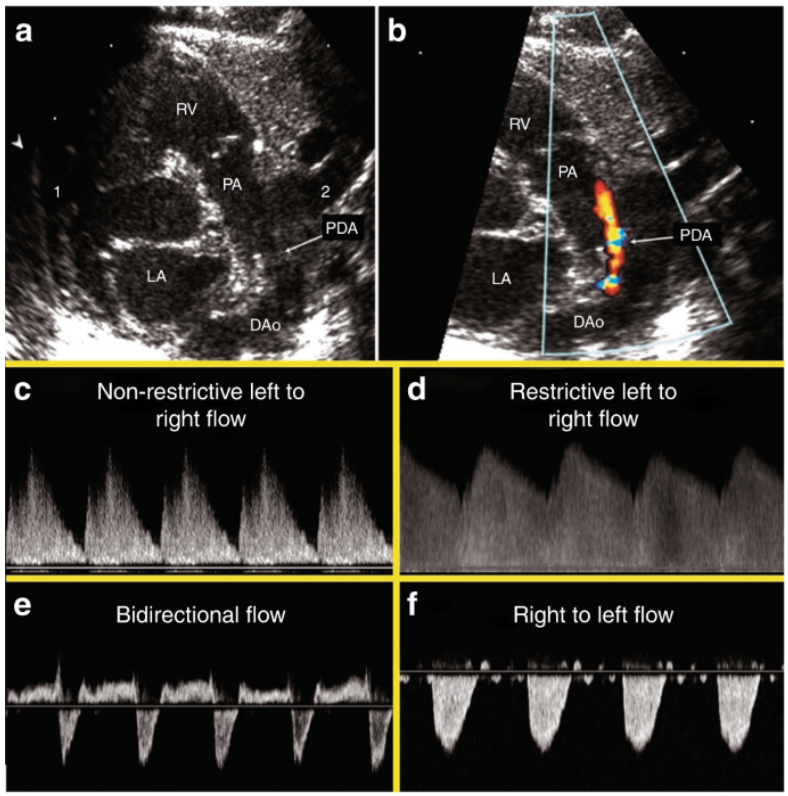
C. Bidirectional

Flow changes direction during the cardiac cycle.

This may occur:

- during normal early transition
- when PA and aortic pressures are similar
- with pulmonary hypertension
- in complex duct-dependent congenital heart disease

Again: **Interpret the pattern in context.**



5. Before Studying the PDA — Know the Clinical Context

Especially in a neonate, document:

- gestational age
- birth weight
- current weight
- postnatal age
- oxygen requirement
- respiratory support
- blood pressure
- systemic perfusion
- reason for the echo
- suspected structural CHD

In a premature infant, the same anatomical PDA may have very different physiological consequences depending on:

- pulmonary vascular resistance
- lung disease
- systemic vascular resistance
- ventricular function
- shunt magnitude

NEONATAL ECHO PEARL

A PDA should never be interpreted independently of the baby's circulation.

6. The Best Echocardiographic Windows

HIGH LEFT PARASTERNAL / DUCTAL VIEW

Often one of the most useful views. It may demonstrate:

Main PA → LPA → PDA → descending aorta

PARASTERNAL SHORT AXIS

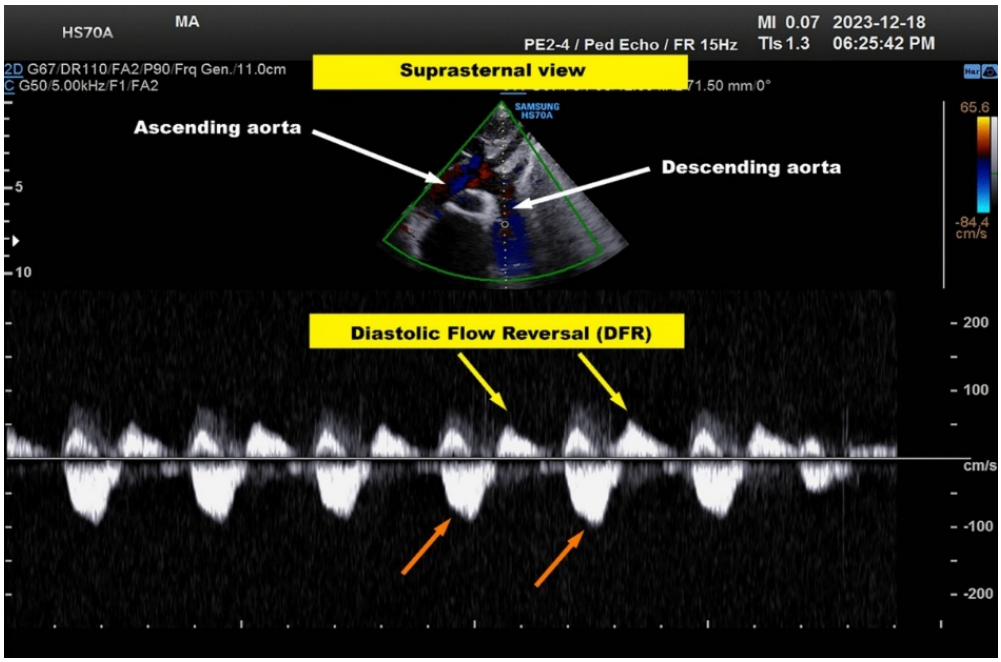
At the level of the great arteries, sweep toward:

- pulmonary valve
- MPA, RPA
- LPA
- ductal region

SUPRASTERNAL VIEWS

Useful for:

- ductal origin
- descending aorta
- unusual ductal course
- arch anatomy
- relationship to coarctation



7. A Beginner's PDA Imaging Sequence

Use:

2D → COLOR → DIRECTION → SPECTRAL DOPPLER → MEASURE → EFFECT

Step 1 — Find the pulmonary arteries.
Step 2 — Identify the descending aorta.
Step 3 — Demonstrate a vascular connection between them.
Step 4 — Add color Doppler.

Step 5 — Establish flow direction.
Step 6 — Add spectral Doppler.
Step 7 — Assess the cardiac and systemic consequences.

8. Do Not Start With Color Alone

Color Doppler makes a PDA easy to notice. But it can also create errors.

Before interpreting the jet:

- know where the MPA is
- identify the LPA
- identify the descending aorta
- define the vessel walls
- understand the imaging plane

Then use color.

ECHO PEARL

Color should prove that blood flows through an anatomically identified ductus—not create the anatomy for you.

This follows the general principle established earlier in the textbook: **2D anatomy first.**

9. Optimize the Ductal Image

For a small or restrictive PDA:

Reduce depth

Make the ductal region occupy more of the screen.

Increase frequency where penetration allows

Improves visualization of small vessel walls.

Narrow the sector

Improves temporal resolution.

Place the focus at the PDA

Improves lateral resolution.

Use a small color box

Preserves frame rate.

Match the color scale to expected velocity

Too high: small/slow ductal flow may disappear.

Too low: a high-velocity restrictive PDA becomes excessively aliased.

10. Confirm That It Really Is a PDA

A common beginner problem is seeing color near the pulmonary artery and calling it a PDA.

Before doing so, demonstrate:

Aortic side ↔ ductal channel ↔ pulmonary arterial side

Potential mimics or confusing structures include:

- LPA flow
- adjacent veins
- collateral vessels
- turbulent branch PA flow
- unusual great-artery anatomy

ECHO PEARL

A color jet near the LPA is not automatically a PDA. Trace the vessel.

11. Assess the Anatomy of the PDA

Do not describe only "small" or "large."

Look at:

- aortic end
- pulmonary end
- narrowest portion
- length
- overall shape
- tortuosity
- relation to LPA
- relation to descending aorta
- relation to aortic isthmus

This becomes especially important before catheter intervention.

TECHNIQUE TIP

Sweep through the whole PDA rather than measuring a single attractive frame.

The narrowest internal diameter should not be assumed to occur at the center of the ductus.

12. Where Should the PDA Be Measured?

Measurement should be:

- obtained in an optimized plane
- perpendicular to the ductal walls as much as feasible
- taken at the relevant narrowest anatomical portion
- accompanied by a clear description of where the measurement was obtained

Avoid oblique measurement.

ECHO PEARL

An oblique PDA measurement can make the duct look larger than it is.

For premature infants, local neonatal protocols may use additional standardized measurements and indexing. These should be applied consistently rather than mixing reference systems.

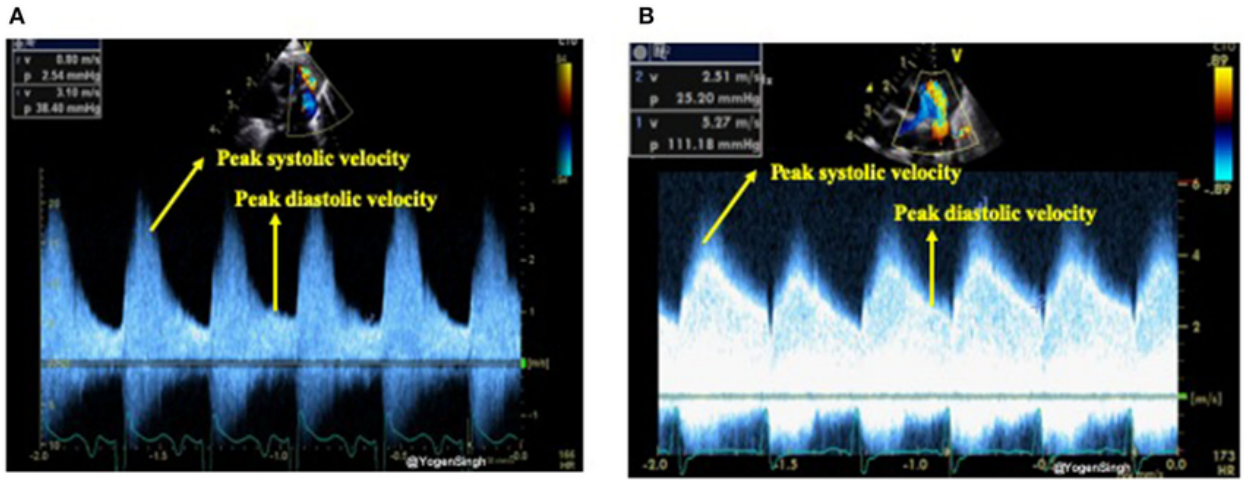
13. Spectral Doppler — What Does the Waveform Tell You?

Spectral Doppler does more than provide velocity.

Look at:

- direction
- peak velocity
- systolic pattern
- diastolic pattern
- continuity throughout the cardiac cycle

The waveform provides information about the pressure difference between the aorta and pulmonary artery.



14. Restrictive PDA

When the pressure difference between the systemic and pulmonary circulations is large and the ductus is narrow, velocity across the PDA may be high.

Conceptually:

Small ductus + large pressure difference → high-velocity jet

ECHO PEARL

High PDA velocity can mean restriction—not necessarily a large shunt.

This is analogous to other restrictive communications.

Do not equate: **High velocity = large PDA.**

15. Nonrestrictive PDA

A large ductus may allow substantial pressure transmission between the aorta and pulmonary artery.

The ductal velocity may therefore be lower than expected.

This becomes especially important when pulmonary artery pressure is elevated.

CRITICAL PEARL

Low ductal velocity can represent a large, nonrestrictive PDA with high pulmonary pressure.

Therefore: **Velocity alone cannot define ductal importance.**

16. PDA as a Window Into Pulmonary Pressure

The ductus directly connects:

Aorta ↔ Pulmonary artery

Therefore Doppler provides clues about their relative pressures.

Large systemic-to-pulmonary gradient

Usually produces a faster left-to-right jet.

Similar systemic and pulmonary pressures

Produces a lower velocity gradient.

Pulmonary pressure exceeding systemic pressure

May produce right-to-left flow.

The PDA is a **natural hemodynamic window into the PA–aortic pressure relationship**.

ECHO PEARL

Do not merely report ductal direction. Ask what the direction tells you about pressure.

17. But PDA Doppler Is Not the Entire Pulmonary Pressure Assessment

Do not use one ductal waveform alone.

Integrate:

- TR velocity
- septal configuration
- RV morphology
- PDA flow
- atrial shunting
- pulmonary artery Doppler
- clinical oxygenation

This is especially important in PPHN.

18. Does the PDA Cause Significant Pulmonary Overcirculation?

Once left-to-right flow is established, the next question is:

How much physiological effect is this shunt producing?

The sequence is:

Aorta → PDA → pulmonary arteries

↓

Pulmonary blood flow increases

↓

Pulmonary venous return increases

↓

LA preload increases

↓

LV preload increases

Therefore significant PDA shunting may produce:

- LA enlargement
- LV enlargement
- increased LV stroke volume
- pulmonary overcirculation

19. Look at the Left Atrium

LA enlargement is an important clue to significant pulmonary recirculation.

Assess the LA in:

- parasternal long axis
- apical views
- other appropriate planes

But:

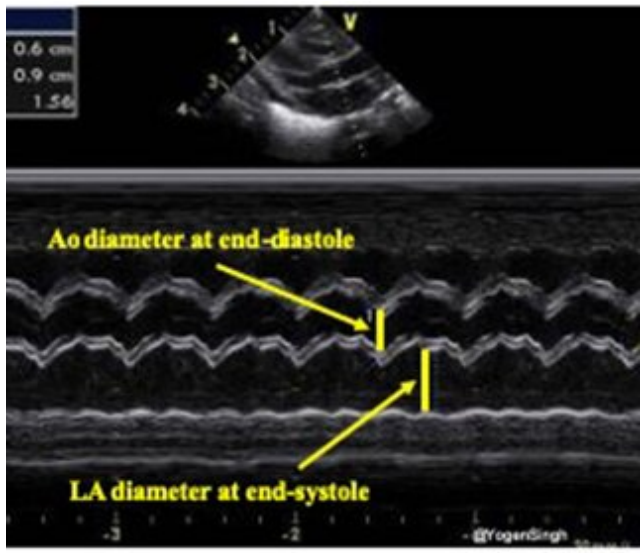
ECHO PEARL

Do not diagnose hemodynamic significance from LA size alone.

LA dimensions are influenced by:

- body size
- mitral valve disease
- atrial communications
- loading conditions
- ventricular function

Interpret the LA as part of the whole circulation.



Left atrium (LA) / Aorta (Ao) ratio measurement

20. Look at the Left Ventricle

In a significant left-to-right PDA shunt:

Pulmonary flow ↑ → pulmonary venous return ↑ → LV preload ↑

The LV may therefore become:

- dilated
- hyperdynamic
- volume loaded

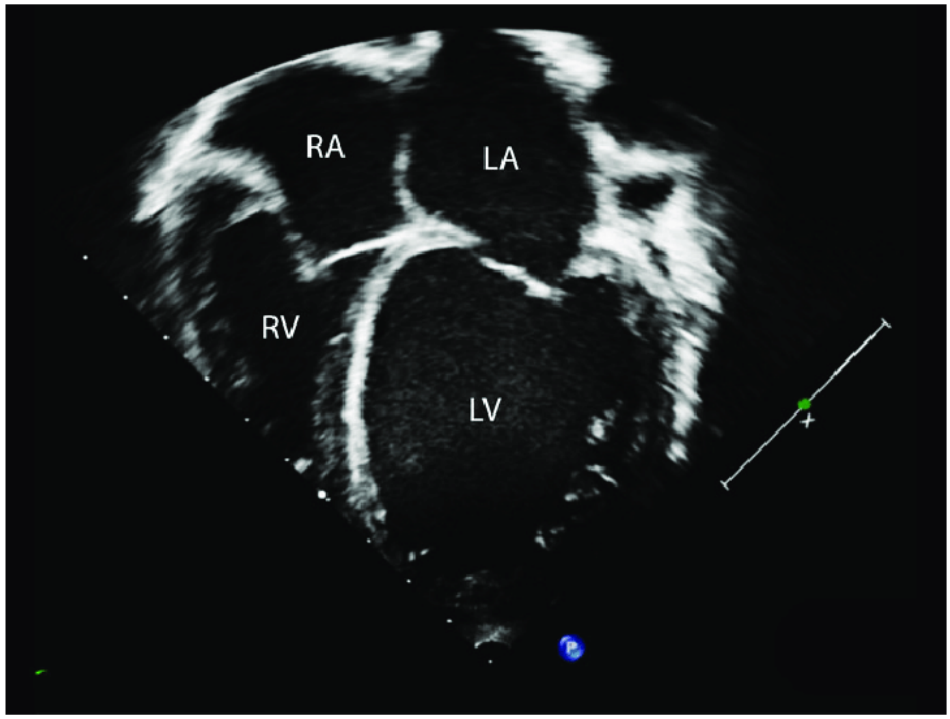
Assess:

- LV end-diastolic dimensions
- systolic function
- volume-loading pattern
- mitral inflow where relevant

Use pediatric reference values and Z-scores where appropriate.

ECHO PEARL

The ductus tells you the shunt exists. The LV tells you how much work that shunt is creating.



21. The Descending Aorta — Look for Systemic Runoff

A large left-to-right PDA allows systemic arterial blood to escape into the pulmonary circulation during diastole.

This can reduce diastolic flow to the lower-body circulation.

Obtain Doppler from the:

- descending thoracic aorta
- abdominal aorta when clinically relevant

Look for abnormal diastolic patterns, including reduced or reversed diastolic flow in appropriate clinical contexts.

ECHO PEARL

A significant PDA affects not only the lungs—it can steal diastolic systemic blood flow.

22. The Premature Infant — Anatomy Is Only Half the Story

PDA assessment in premature infants requires special caution.

A premature infant may have:

- a sizeable PDA
- immature myocardium
- respiratory disease
- changing PVR
- variable systemic vascular resistance
- limited ability to tolerate pulmonary overcirculation

Therefore the central question should not simply be:

"How many millimeters is the PDA?"

It should be:

"What is this PDA doing to this infant?"

23. Hemodynamically Significant PDA — Think in Domains

Instead of relying on a single cutoff, assess several domains.

Domain 1 — Ductal Anatomy

- diameter
- shape
- restriction

Domain 2 — Ductal Flow

- left-to-right/bidirectional/right-to-left
- velocity
- spectral pattern

Domain 3 — Pulmonary Overcirculation

- LA enlargement
- LV enlargement
- increased pulmonary venous return where apparent
- increased pulmonary flow

Domain 4 — Systemic Steal

- descending/abdominal aortic diastolic flow abnormalities
- evidence of compromised systemic perfusion

Domain 5 — Ventricular Response

- LV volume load
- ventricular function
- RV pressure

Domain 6 — Clinical Context

- respiratory support
- oxygen requirement
- blood pressure
- perfusion
- feeding/growth where relevant

MASTER PEARL

Hemodynamic significance is an integrated diagnosis—not a millimeter measurement.

24. A Note About Numerical PDA Thresholds

Different neonatal programs and published protocols use combinations of:

- absolute ductal diameter
- indexed ductal size
- LA-related measurements
- LV output
- systemic Doppler patterns
- ductal flow patterns

For this introductory chapter, the safer teaching principle is:

Use your validated institutional or neonatal protocol for numerical thresholds, and interpret those measurements within the complete hemodynamic picture.

This avoids teaching a single number as universally diagnostic.

25. The PDA Can Be Beneficial

This is one of the most important concepts in congenital cardiology.

A PDA is not always a lesion to close.

In some newborns: **The PDA is maintaining life.**

Two major situations exist.

26. Duct-Dependent Pulmonary Circulation

Here the PDA provides pulmonary blood flow.

Pathway:

Aorta → PDA → Pulmonary arteries → Lungs

Examples include:

- pulmonary atresia
- critical pulmonary stenosis
- severe TOF/RVOT obstruction
- selected single-ventricle lesions

CRITICAL PEARL

Never recommend PDA closure until duct-dependent pulmonary circulation has been excluded.

27. Duct-Dependent Systemic Circulation

Here the PDA provides systemic blood flow.

Pathway:

RV → PA → PDA → Descending aorta

Examples include:

- critical coarctation
- interrupted aortic arch
- critical aortic stenosis
- HLHS spectrum

CRITICAL PEARL

Right-to-left ductal flow in a sick neonate should trigger immediate evaluation of the left heart and aortic arch.

28. PDA in Coarctation — A Major Diagnostic Trap

A large PDA can bypass or decompress significant neonatal coarctation.

Therefore:

- distal aortic flow may look acceptable
- the Doppler gradient may be low
- femoral pulses may still be present

An open PDA can reduce the apparent coarctation gradient; assess arch dimensions, the isthmus, ductal anatomy, flow direction, and LV function rather than relying on gradient alone.

ECHO PEARL

Before calling a PDA "isolated," look carefully at the aortic arch.

29. PDA and Pulmonary Hypertension

When PVR remains elevated:

- ductal velocity may fall
- shunting may become bidirectional
- a right-to-left component may appear

In severe pulmonary hypertension:

PA → PDA → Aorta

may occur.

Do not diagnose PPHN from this alone.

Evaluate:

- structural anatomy
- pulmonary venous return
- RV pressure
- septal shape
- TR velocity
- aortic arch
- ventricular function

ECHO PEARL

The PDA shows the pressure relationship. It does not tell you the cause of that pressure relationship.

30. PDA and the Left Pulmonary Artery

The pulmonary end of the ductus lies near the LPA.

Carefully assess:

- LPA origin
- proximal LPA caliber
- ductal insertion
- surrounding flow disturbance

This is particularly important:

- before intervention
- in unusual ductal anatomy
- in congenital lesions dependent on ductal pulmonary flow

Do not allow color turbulence from the PDA to obscure LPA anatomy.

31. PDA and the Aortic Isthmus

The aortic end lies near an anatomically important region.

Always inspect:

Distal transverse arch → isthmus → ductal insertion → descending aorta

This is especially important in neonates because a large ductal ampulla can confuse the assessment of true isthmic caliber.

ECHO PEARL

The PDA and aortic isthmus should be understood as neighboring structures—not interpreted independently.

32. Common PDA Echo Patterns

Echo Pattern	Likely Interpretation
High-velocity continuous L→R	Restrictive PDA with substantial pressure difference
Lower-velocity L→R	Larger/nonrestrictive duct or elevated PA pressure—interpret context
Bidirectional	Similar PA/systemic pressures, early transition, PH, or complex physiology
Predominantly R→L	High pulmonary pressure or duct-dependent systemic circulation
LA/LV enlargement + L→R PDA	Significant pulmonary recirculation likely
Abnormal descending aortic diastolic flow	Systemic runoff may be important
PDA + retrograde arch flow	Consider duct-dependent systemic circulation
PDA supplying PAs with absent/critical RVOT flow	Duct-dependent pulmonary circulation

33. The 60-Second PDA Assessment

When you identify a PDA, immediately answer:

1. Anatomy — Can I see the entire ductus?

2. Size — Where is the narrowest part?

3. Direction — Aorta → PA or PA → Aorta?

4. Restriction — What does the spectral waveform suggest?

5. Pulmonary pressure — Does the PDA suggest high PA pressure?

6. Heart effect — Are the LA/LV volume loaded?

7. Systemic effect — Is there diastolic runoff?

8. Dependency — Would closing the PDA compromise pulmonary or systemic flow?

34. A Window-Based PDA Protocol

PARASTERNAL SHORT AXIS

Look for: MPA, RPA, LPA, ductal region, pulmonary valve.

Ask: **Is there abnormal color flow entering the PA near the LPA?**

HIGH LEFT PARASTERNAL

Optimize the classic ductal region. Demonstrate:

PDA + LPA + descending aorta

Ask: **Can I define the complete ductal anatomy?**

SUPRASTERNAL

Assess: aortic end, arch, descending aorta, ductal relationship, unusual ductal orientation.

Ask: **Is there associated arch obstruction or unusual ductal anatomy?**

PARASTERNAL LONG AXIS / APICAL

Assess consequences: LA size, LV size, LV function.

Ask: **What is the volume effect of the shunt?**

SUBCOSTAL / ABDOMINAL AORTA

Use Doppler when clinically relevant.

Ask: **Is there evidence of systemic runoff?**

36. Beginner Versus Trainee Expectations

Echo Technician / Beginner	Paediatric Cardiology Trainee
Identify PDA	Define complete ductal anatomy
Obtain ductal view	Identify aortic and pulmonary ends
Determine flow direction	Interpret pressure relationships
Obtain Doppler waveform	Assess restriction/nonrestriction
Measure PDA carefully	Integrate measurements with physiology
Assess LA/LV size	Determine hemodynamic consequences
Examine arch	Recognize duct dependency
Record representative images	Assess suitability for intervention where appropriate

35. Common Diagnostic Pitfalls

Pitfall 1 — PDA PRESENT = DISEASE

Correction: A PDA may be part of normal early neonatal transition. Consider postnatal age and physiology.

Pitfall 2 — HIGH VELOCITY = LARGE PDA

Correction: The opposite may be true. High velocity often reflects a restrictive ductus and large pressure difference.

Pitfall 3 — LOW VELOCITY = INSIGNIFICANT PDA

Correction: Low velocity may occur with a large nonrestrictive ductus or high PA pressure.

Pitfall 4 — RED MEANS SYSTEMIC AND BLUE MEANS PULMONARY

Correction: Color indicates direction relative to the probe, not oxygen content.

Pitfall 5 — ONE MEASUREMENT DEFINES SIGNIFICANCE

Correction: Integrate anatomy, Doppler, chamber loading and systemic flow.

Pitfall 6 — LEFT-TO-RIGHT FLOW MEANS SAFE TO CLOSE

Correction: Complete structural assessment first, especially in complex CHD.

Pitfall 7 — RIGHT-TO-LEFT FLOW AUTOMATICALLY MEANS PPHN

Correction: Exclude critical systemic outflow obstruction.

Pitfall 8 — PDA FOUND, ARCH IGNORED

Correction: Always examine the neonatal arch.

Pitfall 9 — LARGE COLOR JET = ANATOMICALLY LARGE PDA

Correction: Color jet dimensions depend heavily on velocity scale, gain, imaging plane. Measure anatomy in 2D.

Pitfall 10 — REPORTING THE LESION BUT NOT ITS EFFECT

Correction: "PDA 2 mm" alone does not tell the clinician whether it matters.

38. Echo Pearls — The Page to Remember

Pearl 1: Do not report "PDA present." Describe what it is doing.

Pearl 2: Aorta → PDA → PA = left-to-right shunting.

Pearl 3: PA → PDA → aorta requires explanation.

Pearl 4: High ductal velocity often means restriction—not a large shunt.

Pearl 5: Low velocity may mean high PA pressure or a nonrestrictive PDA.

Pearl 6: PDA significance is not defined by diameter alone.

Pearl 7: Look at the LA and LV to understand pulmonary recirculation.

Pearl 8: Look at the descending/abdominal aorta to understand systemic runoff.

Pearl 9: In premature infants, assess hemodynamic significance—not simply ductal patency.

Pearl 10: Right-to-left ductal flow is a physiological clue, not a diagnosis of PPHN.

Pearl 11: Before considering PDA closure, exclude duct-dependent systemic and pulmonary circulations.

Pearl 12: Always examine the neonatal aortic arch when assessing a PDA.

Chapter Key Points

The ductus arteriosus is both an **anatomical structure and a physiological window**.

Its significance changes dramatically from fetal life through neonatal transition and later infancy.

Every PDA assessment should include:

SIZE + MORPHOLOGY + DIRECTION + VELOCITY + RESTRICTION + CARDIAC EFFECT + SYSTEMIC EFFECT + DEPENDENCY

Left-to-right ductal shunting increases pulmonary blood flow and may produce LA/LV volume loading.

A large shunt can also cause systemic diastolic runoff.

Ductal Doppler provides useful information about the pressure relationship between the pulmonary and systemic circulations but should never be interpreted alone.

In the premature infant, no single ductal measurement should substitute for integrated hemodynamic assessment.

Most importantly, a PDA is not always pathological. In selected congenital heart lesions it may be essential for **pulmonary or systemic survival**.

The important questions are not merely whether the ductus is present, but **which direction blood is flowing, how restrictive it is, what the pressure relationship is, and whether the circulation depends upon it.**

MASTER PRINCIPLE

Never ask only, "How big is the PDA?" Ask, "What is the PDA doing to the circulation?"

The PDA Echo Formula

FIND → DEFINE → FOLLOW THE FLOW → INTERPRET PRESSURE → ASSESS VOLUME LOAD → ASSESS SYSTEMIC RUNOFF → EXCLUDE DUCTAL DEPENDENCY

And the single most important question remains:

"What will happen to this circulation if the ductus closes?"

MODULE 3

Cyanotic Congenital Heart Disease

Topics
Cyanosis — Echo Algorithm
Transposition of the Great Arteries (TGA)
Total Anomalous Pulmonary Venous Return (TAPVR)
Tetralogy of Fallot (TOF)
Pulmonary Atresia with VSD (PA-VSD)
TOF with Absent Pulmonary Valve (TOF-APV)
Pulmonary Atresia with Intact Septum (PA-IVS)
Critical Pulmonary Stenosis
Double Outlet Right Ventricle (DORV)
Tricuspid Atresia
Truncus Arteriosus
Single Ventricle Physiology
Glenn & Fontan — Echo Essentials

1. Cyanosis — Echo Algorithm

Not all cyanosis is cardiac.

Before echo, always check:

OXYGEN SATURATION PATTERN

Pre-ductal (Right Hand) and Post-ductal (Foot)

Pattern	Suggestion
Both low	Mixing lesion
Lower body lower	Duct-dependent systemic
Both very low (<75%)	Severe cyanotic CHD

LOOK AT THE 4-CHAMBER VIEW

Ask 4 quick questions:

1. Is one ventricle very small?
2. Is RV dominant?
3. Is there AV valve abnormality?
4. Are all pulmonary veins draining normally into LA?

IS PULMONARY BLOOD FLOW INCREASED OR DECREASED?

Look at: pulmonary valve, pulmonary artery size, Doppler flow, chest X-ray (if available).

Decreased Pulmonary Flow (Oligemic Lungs) — Think:

- Tetralogy of Fallot
- Pulmonary atresia
- Severe pulmonary stenosis

Increased Pulmonary Flow (Plethoric Lungs) — Think:

- Transposition of the great arteries
- Total anomalous pulmonary venous return
- Truncus arteriosus

ASSESS GREAT ARTERIES

Are they crossing normally, parallel, or single outflow?

Parallel vessels → suspect Transposition of the great arteries
Single outflow → suspect truncus or pulmonary atresia

CHECK PDA DIRECTION

Color Doppler tells physiology:

PDA Flow	Meaning
Aorta → PA	Duct-dependent pulmonary
PA → Aorta	Duct-dependent systemic
Bidirectional	Transitional circulation

This single step often solves the case.

ASSESS ATRIAL SEPTUM

In critical cyanosis, a restrictive atrial septum is dangerous—especially in Transposition of the great arteries.

If restrictive → emergency balloon atrial septostomy.

LOOK FOR TOTAL MIXING LESIONS

If saturation equal everywhere + severe cyanosis, consider Total anomalous pulmonary venous return.

Clues:

- Dilated RA/RV
- Small LA
- Pulmonary venous confluence behind LA

RAPID ECHO ALGORITHM SUMMARY

1. Shock? → Yes → Think left-sided obstruction
2. Cyanosis without shock? → Check pulmonary blood flow
3. Decreased pulmonary flow? → RVOT obstruction
4. Increased pulmonary flow? → Mixing lesion (TGA, TAPVR, truncus)
5. Parallel great arteries? → TGA
6. Single ventricle physiology? → Complex CHD

HIGH-YIELD ECHO PATTERNS

Pattern 1 — Small LV + Large RV + Retrograde descending aorta → Duct-dependent systemic

Pattern 2 — No antegrade pulmonary flow + PDA supplying PA → Duct-dependent pulmonary

Pattern 3 — Parallel great arteries + severe cyanosis → TGA

Pattern 4 — Dilated right heart + unobstructed pulmonary arteries + severe hypoxia → Obstructed TAPVR

WHEN TO START PROSTAGLANDIN IMMEDIATELY

Start PGE1 if:

- Severe cyanosis
- Suspected duct dependency
- Uncertain anatomy
- Deteriorating neonate

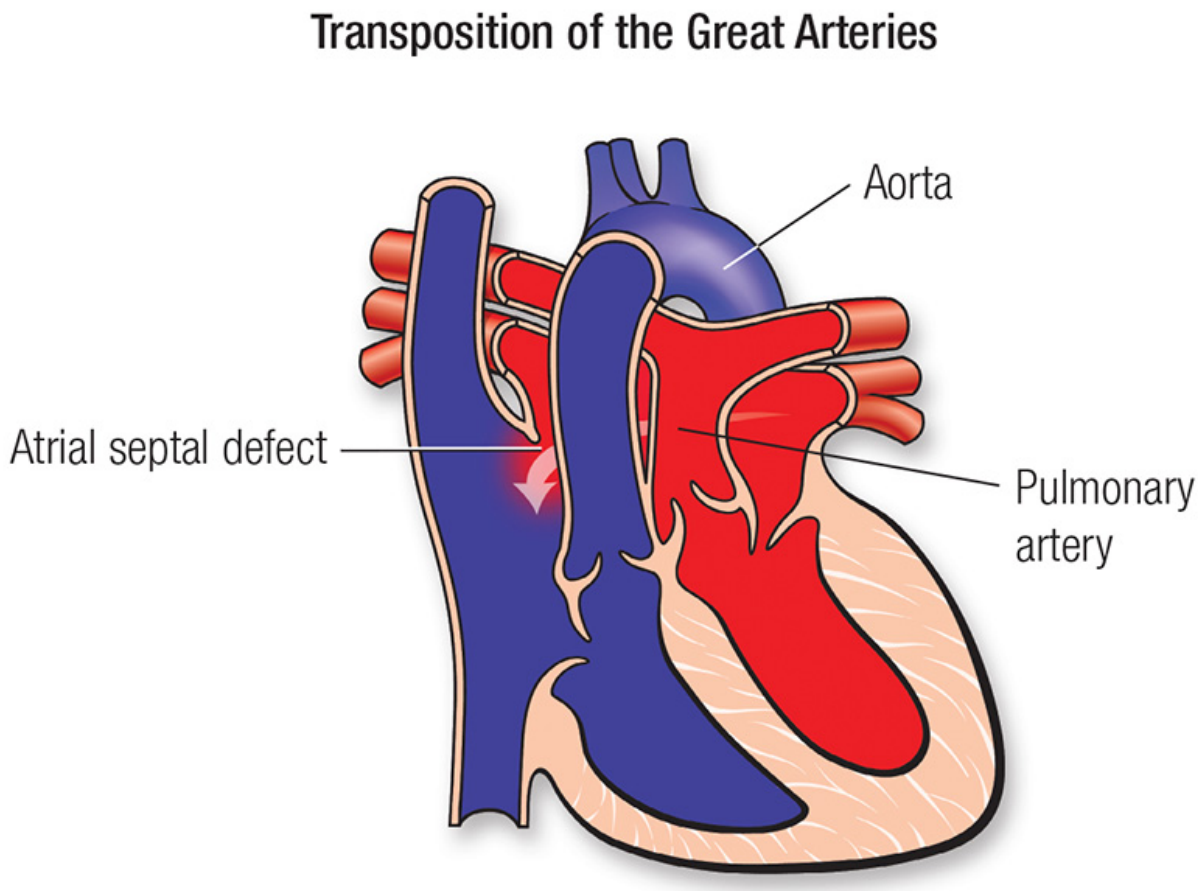
Transposition of the Great Arteries (TGA)

WHAT IS TGA?

Transposition of the great arteries is a congenital heart defect where:

- Aorta arises from the Right Ventricle (RV)
- Pulmonary artery arises from the Left Ventricle (LV)

Instead of crossing, the great arteries run parallel.



TYPES OF TGA

1. D-TGA (Complete TGA) — Most common type. Dextro-Transposition of the great arteries

- Parallel circulation
- Severe early cyanosis
- Requires arterial switch surgery

2. L-TGA (Congenitally Corrected) — Levo-Transposition of the great arteries

- AV and VA discordance
- Less severe cyanosis

- Different physiology

Neonatal cyanosis usually D-TGA.

D-TGA Subtypes:

I. TGA with IVS
II. TGA, VSD
III. TGA, VSD, PS

WHY IT IS DANGEROUS

Two separate circulations form:

- Body → RA → RV → Aorta → Body
- Lungs → LA → LV → PA → Lungs

Survival depends on mixing. No oxygen mixing unless there is: ASD, VSD, or PDA.

1. Echo Diagnosis — Stepwise Approach

STEP 1: APICAL 4-CHAMBER VIEW

Usually looks normal. That's the trap. Both ventricles: normal size, normal AV connections.

TGA is NOT a 4-chamber problem—it is a great artery problem.

STEP 2: PARASTERNAL SHORT AXIS (GREAT ARTERIES)

Normal heart: pulmonary artery wraps around aorta.

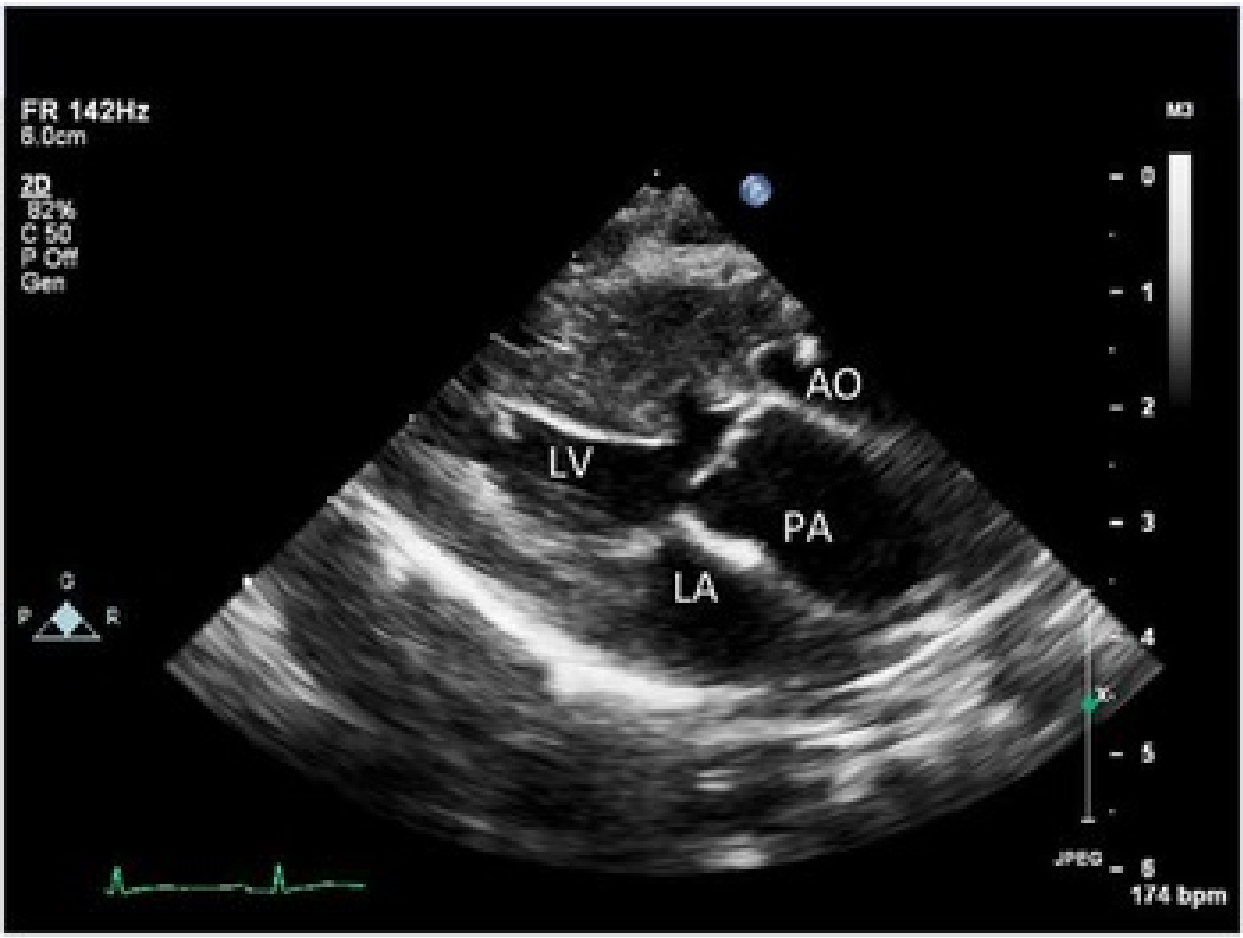
In TGA: great arteries are parallel; aorta is anterior and rightward.

This is the key diagnostic sign.

STEP 3: FOLLOW THE VESSELS

Always trace: RV → which vessel? LV → which vessel?

If RV leads to aorta → TGA confirmed.



STEP 4: ASSESS INTERATRIAL SEPTUM

Critical step. Look for: Adequate ASD, Restrictive septum, Bowing pattern.

If restrictive → emergency balloon atrial septostomy.

STEP 5: ASSESS PDA

Duct is often bidirectional and important for mixing. Prostaglandin maintains ductal flow.

STEP 6: ASSESS CORONARIES AND LV

ECHO CLUES THAT MUST ALERT YOU

- Parallel great arteries
- Aorta anterior
- RV giving rise to aorta
- Severe cyanosis with minimal murmur
- Normal 4-chamber but abnormal outflows

2. Diagnostic Traps

TRAP 1: LOOKING ONLY AT 4-CHAMBER VIEW

4-chamber often appears normal. Always examine outflow tracts.

TRAP 2: MISSING RESTRICTIVE ATRIAL SEPTUM

Baby may deteriorate suddenly. Check: septal thickness, Doppler across ASD, LA pressure signs.

TRAP 3: CONFUSING WITH PERSISTENT PULMONARY HYPERTENSION (PPHN)

PPHN: normal anatomy, high pulmonary pressures.

TGA: abnormal vessel connections, parallel great arteries.

Anatomy always wins over pressure diagnosis.

Transposition of the Great Arteries

- **Simple TGA** → ASO (1–3 wks)
- **TGA + Other simple defects (such as PDA, VSD, dynamic or mild PS)** → ASO (1–3 wks) + Repair of other defects
- **TGA + VSD + Severe PS** → Shunt operation (±) (early in life) → (1) VSD-AO tunnel + Rastelli (>1–2 yr), or (2) REV procedure (>6 mo), or (3) Nikaidoh operation (>1 yr)
- **TGA + Large VSD + Subaortic stenosis** → (Initial BT shunt [±]) → Damus-Kaye-Stansel + VSD closure + RV-PA connection (1–2 yr)

Total Anomalous Pulmonary Venous Return (TAPVR)

WHAT IS TAPVR?

Total anomalous pulmonary venous return is a congenital defect where all pulmonary veins drain into the right atrium (or systemic veins) instead of the left atrium.

The left atrium receives no direct pulmonary venous blood.

Survival requires an ASD (for mixing).

BASIC PHYSIOLOGY

Normal: Lungs → Pulmonary veins → LA → LV → Body

In TAPVR: Lungs → Confluence → Systemic veins → RA → RV

So: oxygenated blood returns to right heart, complete mixing occurs, systemic oxygen saturation depends on mixing efficiency.

TYPES OF TAPVR

Supracardiac (Most Common)

Pulmonary veins → Confluence → Vertical vein → SVC

Look for: vertical vein on left side, dilated SVC.

Cardiac

Pulmonary veins → Coronary sinus → RA

Clue: dilated coronary sinus.

Infracardiac (Often Obstructed)

Pulmonary veins → Portal vein / IVC

Often severe obstruction. Clue: severe pulmonary hypertension, sick neonate.

Mixed Type

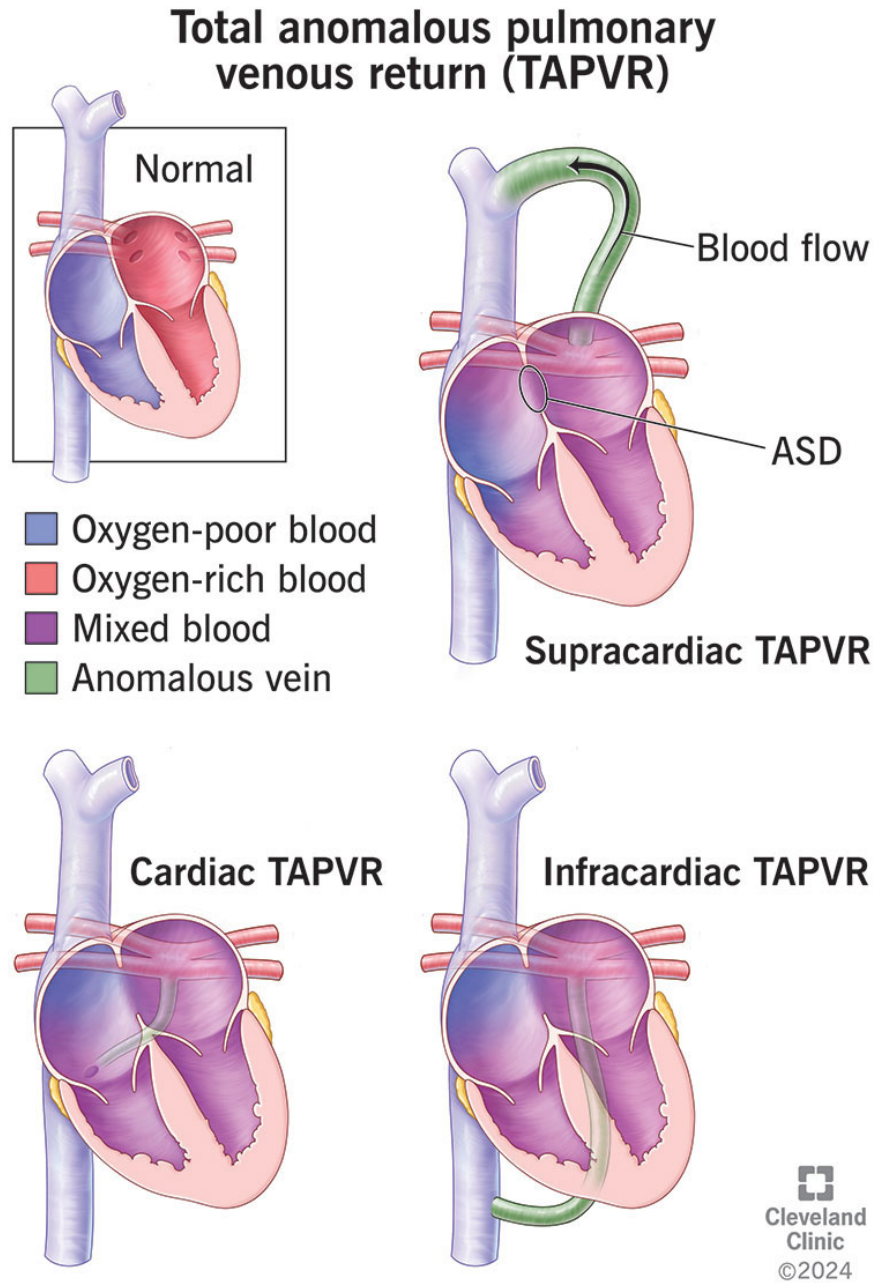
Combination of above pathways.

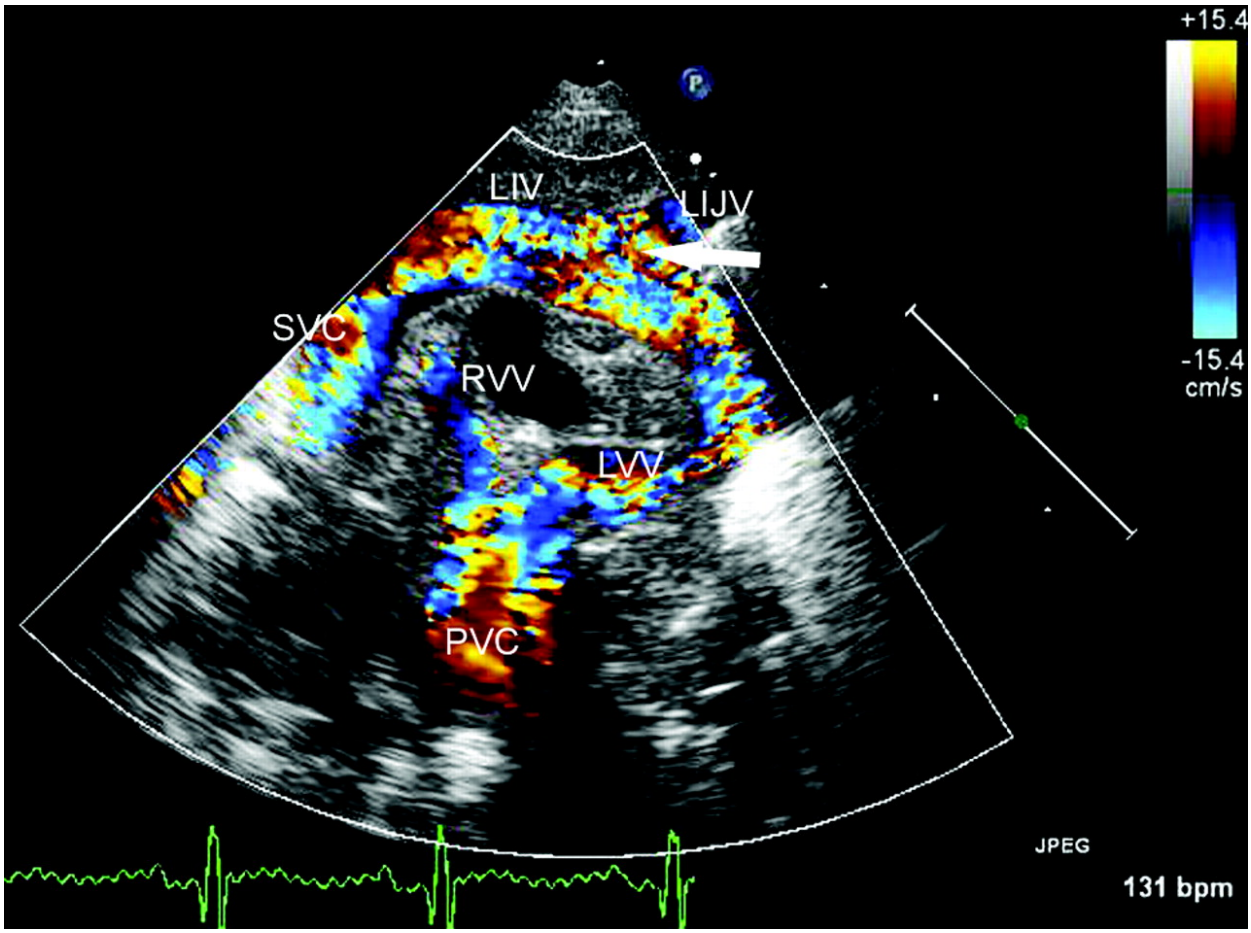
UNOBSTRUCTED TAPVR

- Moderate cyanosis
- Tachypnea
- Increased pulmonary blood flow
- RA/RV enlargement

OBSTRUCTED TAPVR (EMERGENCY)

- Severe cyanosis
- Respiratory distress
- Metabolic acidosis
- Pulmonary edema



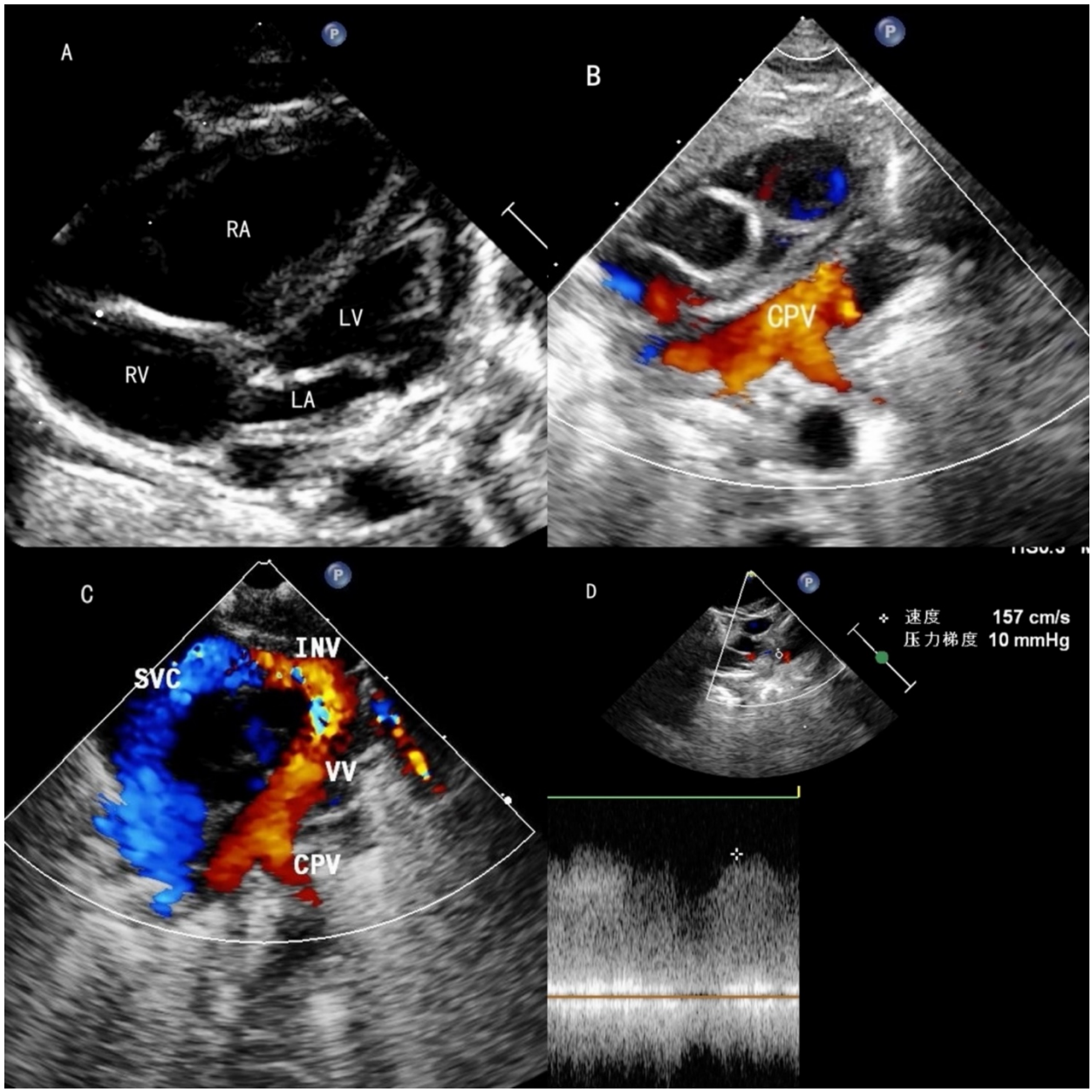


1. Echo Approach — Stepwise

STEP 1: LOOK AT 4-CHAMBER VIEW

Key findings: Dilated RA, Dilated RV, Small LA, No pulmonary veins entering LA.

Small LA is a major clue.



STEP 2: SEARCH FOR PULMONARY VEINS

Normal echo should show 4 pulmonary veins entering LA.

In TAPVR: you will NOT see them entering LA. Instead, look posterior to LA for pulmonary venous confluence.

STEP 3: IDENTIFY DRAINAGE PATHWAY

Trace confluence: upward → supracardiac; to coronary sinus → cardiac; downward through diaphragm → infracardiac.

Use color Doppler to confirm flow direction.

STEP 4: LOOK FOR OBSTRUCTION

Signs: turbulent flow in vertical vein, continuous high-velocity Doppler, pulmonary hypertension, flattened interventricular septum.

Obstructed TAPVR is surgical emergency.

KEY ECHO CLUES YOU MUST NOT MISS

- No pulmonary veins into LA
- Small LA
- Dilated right heart
- Dilated SVC (supracardiac type)
- Dilated coronary sinus (cardiac type)
- Vertical vein visible

2. Diagnostic Traps

TRAP 1: DIAGNOSING PPHN INSTEAD

PPHN: normal pulmonary venous drainage. TAPVR: abnormal venous anatomy.

Always confirm pulmonary veins enter LA.

TRAP 2: MISSING INFRACARDIAC OBSTRUCTION

Baby may look like severe lung disease. Clues: severe hypoxia, pulmonary edema, small LA, no visible pulmonary veins in LA.

TRAP 3: NOT LOOKING BEHIND LA

Pulmonary venous confluence is usually posterior. Use subcostal and suprasternal views.

CLINICAL PATTERNS

Pattern 1 — Cyanotic neonate + dilated right heart + small LA → Think TAPVR.

Pattern 2 — Severe distress + pulmonary edema + cyanosis → Think obstructed infracardiac TAPVR.

MANAGEMENT

Definitive treatment: surgical repair. In obstructed cases: emergency surgery required.

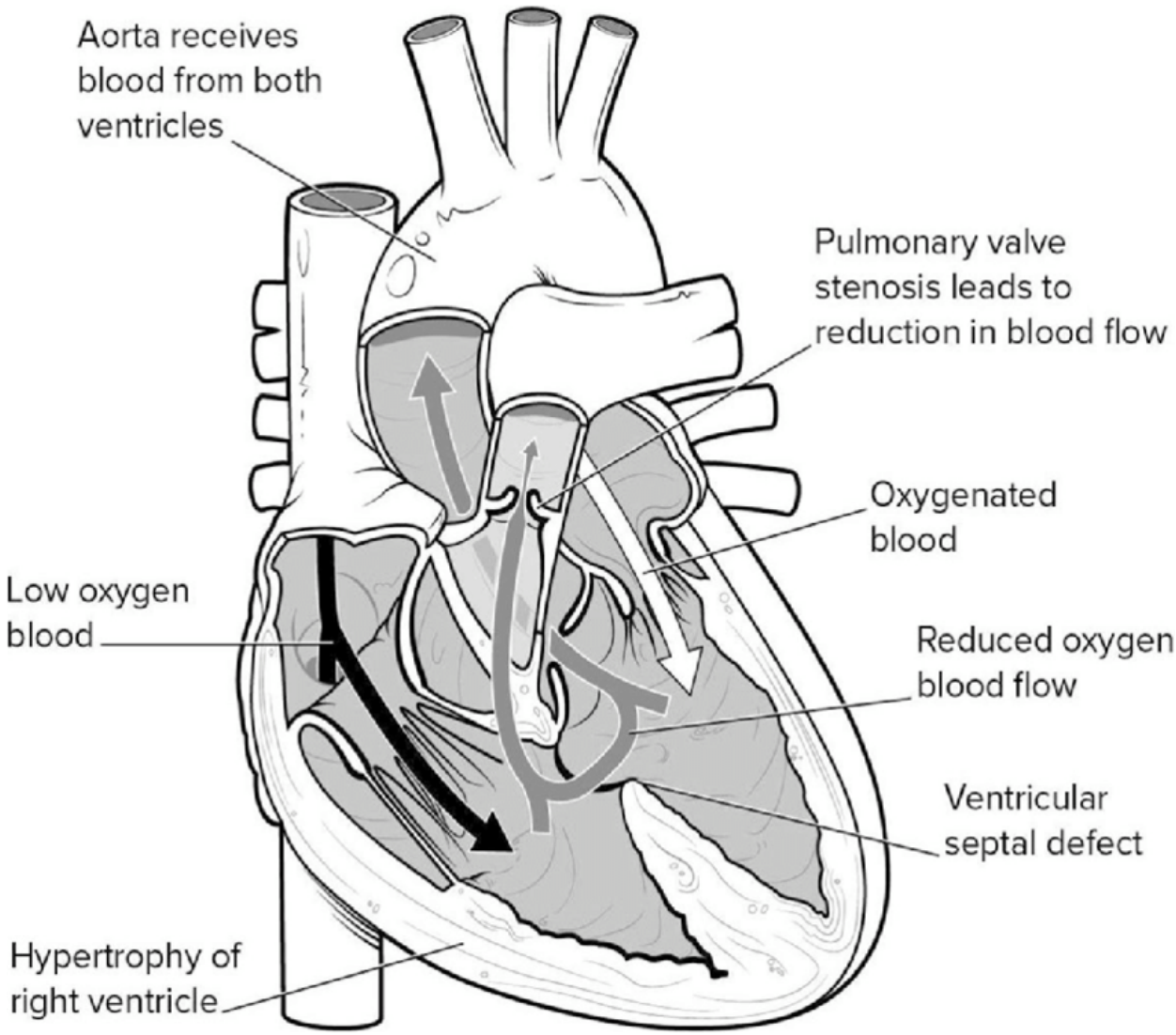
No role for prostaglandin (unless duct helping mixing slightly).

Tetralogy of Fallot (TOF)

WHAT IS TETRALOGY OF FALLOT?

Tetralogy of Fallot consists of four components:

1. Ventricular Septal Defect (VSD)
2. Overriding Aorta
3. Right Ventricular Outflow Tract (RVOT) Obstruction
4. Right Ventricular Hypertrophy



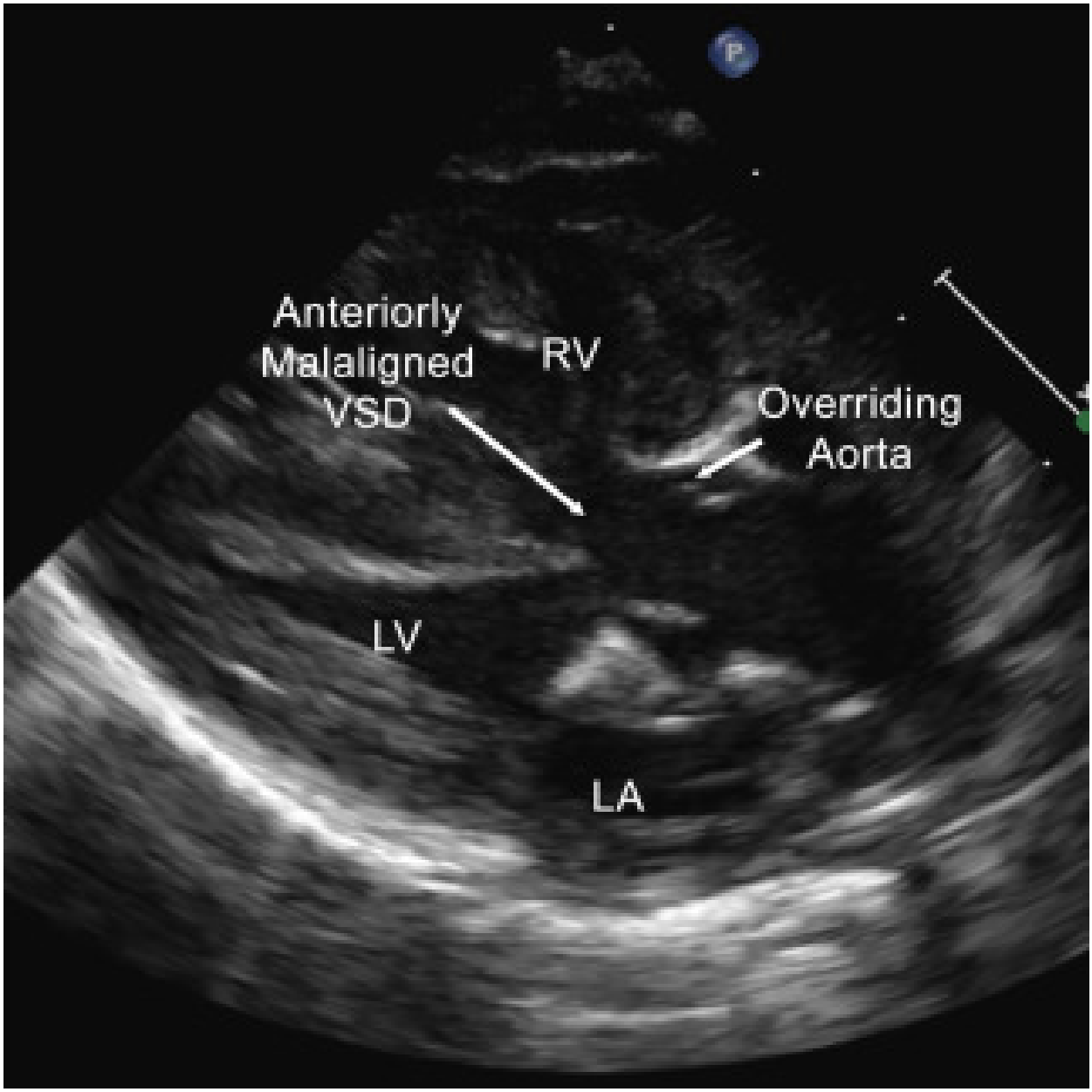
WHY NEONATAL PRESENTATION VARIES

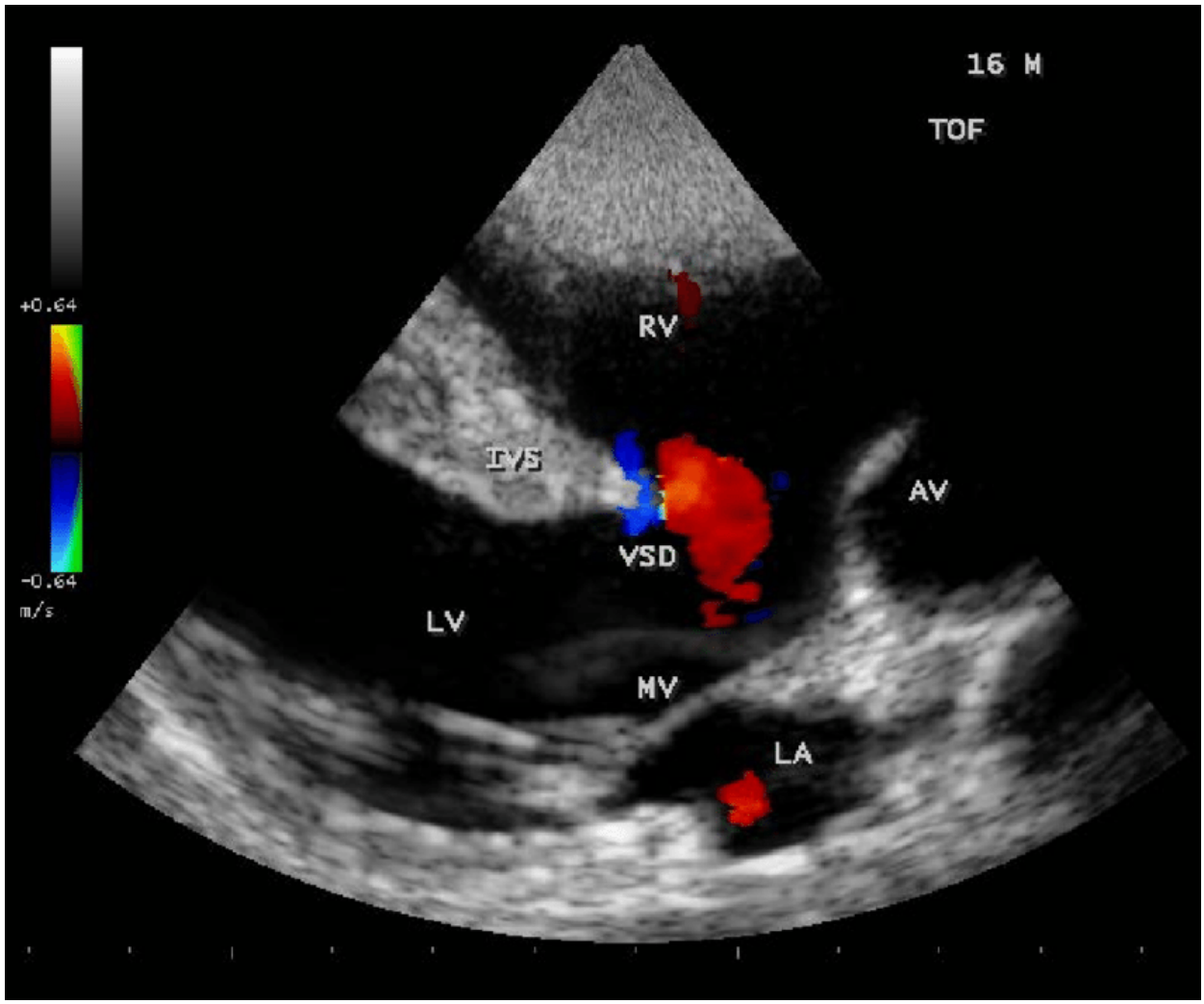
Severity depends mainly on degree of RVOT obstruction:

- Mild obstruction → pink baby (acyanotic)
- Severe obstruction → cyanotic neonate
- Critical obstruction → duct-dependent pulmonary flow

1. Echo Diagnosis

STEP 1: PARASTERNAL LONG AXIS VIEW





Look for: large malalignment VSD, aorta overriding septum, anterior deviation of infundibular septum.

Overriding aorta is hallmark.

STEP 2: PARASTERNAL SHORT AXIS — RVOT

Evaluate: pulmonary valve size, infundibular narrowing, main pulmonary artery size. Color Doppler shows turbulent flow.

STEP 3: MEASURE RVOT GRADIENT

Use CW Doppler: high gradient = significant obstruction.

BUT in very severe obstruction → low forward flow → gradient may be misleadingly low.

Always assess anatomy.

STEP 4: ASSESS PULMONARY ARTERIES

Look for hypoplastic branch pulmonary arteries; confluent vs discontinuous branches. Important for surgical planning.

STEP 5: EVALUATE PDA

If severe RVOT obstruction: pulmonary flow may depend on duct, PDA supplies pulmonary arteries, flow direction Aorta → PA.

This becomes duct-dependent pulmonary circulation.

KEY ECHO FINDINGS

- Large VSD
- Overriding aorta
- RVOT narrowing
- Turbulent pulmonary flow
- Small pulmonary annulus

RV hypertrophy may not be obvious in early neonates.

SEVERITY CLASSIFICATION (ECHO-BASED)

Mild TOF — Good antegrade pulmonary flow, mild cyanosis.

Moderate TOF — Significant RVOT gradient, intermittent desaturation.

"Pink TOF" — Minimal obstruction, increased pulmonary flow.

Critical TOF — Severe obstruction or pulmonary atresia, duct-dependent pulmonary flow.

2. Neonatal Diagnostic Traps

TRAP 1: MISSING TOF WHEN CYANOSIS IS MILD

Mild obstruction/evolving TOF may show only a murmur with minimal desaturation. Always evaluate RVOT carefully.

TRAP 2: CONFUSING WITH PULMONARY STENOSIS + VSD

TOF has malalignment VSD and overriding aorta. Isolated PS + VSD does not.

TRAP 3: ASSUMING LOW GRADIENT = MILD DISEASE

If pulmonary valve is almost atretic, flow is minimal and gradient may appear low. Anatomy is more important than gradient.

Differential Diagnosis in Cyanotic Neonate

Condition	Pulmonary Flow	Great Arteries
TOF	Decreased	Normal relationship
TGA	Increased	Parallel
TAPVR	Increased	Normal
Pulmonary Atresia	Absent	Normal

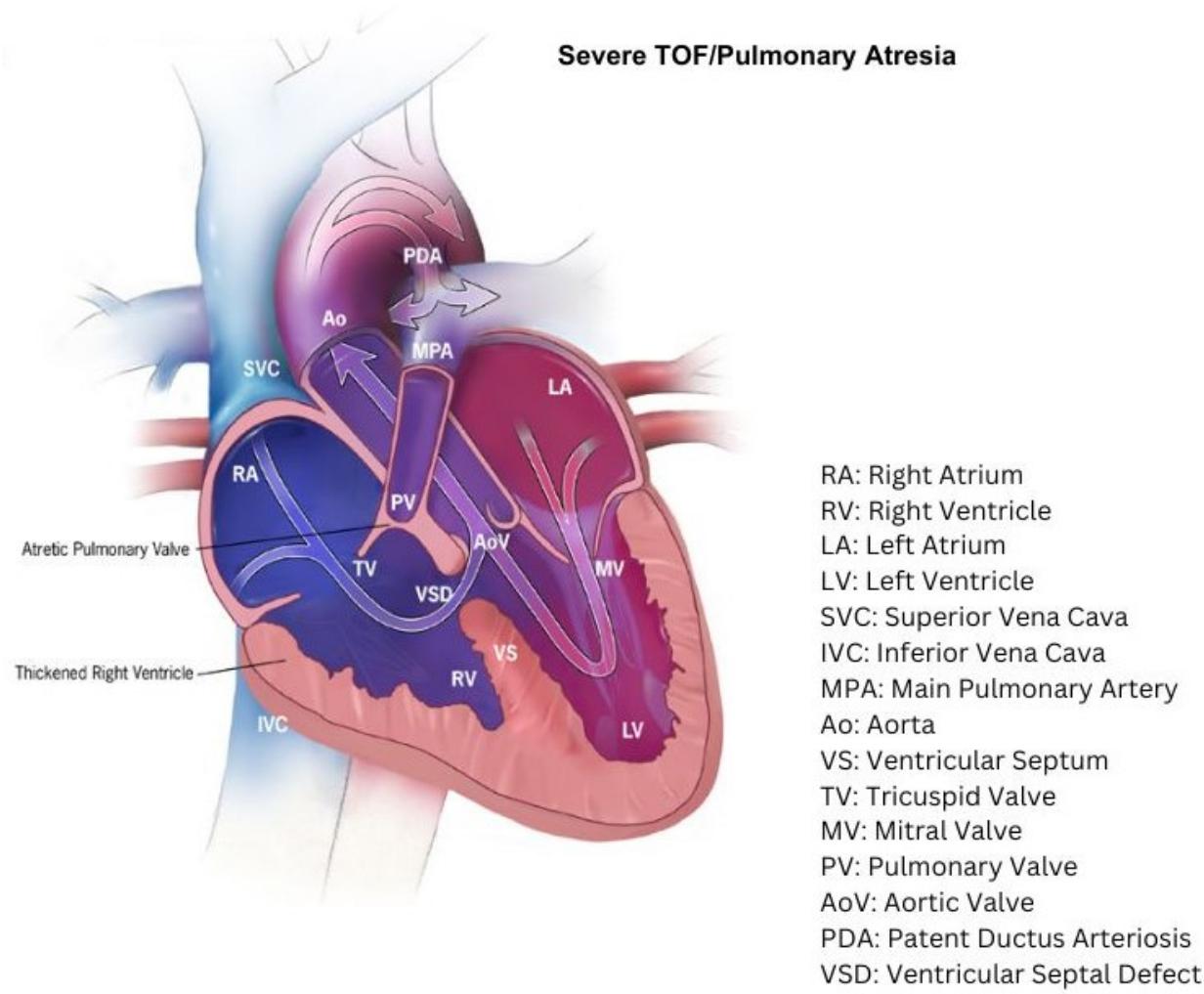
TOF usually shows decreased pulmonary blood flow.

Pulmonary Atresia with VSD (PA-VSD)

Pulmonary atresia with ventricular septal defect is a severe form of conotruncal defect characterized by:

1. Large VSD
2. Complete atresia of the pulmonary valve
3. No antegrade flow from RV to pulmonary artery
4. Pulmonary blood supply from ductus or collaterals

It is often considered the extreme end of Tetralogy of Fallot.



CORE PHYSIOLOGY

Because the pulmonary valve is completely atretic: RV cannot pump blood into pulmonary arteries; pulmonary circulation depends on PDA or Major Aortopulmonary Collateral Arteries (MAPCAs).

Without duct → severe cyanosis. This is typically duct-dependent pulmonary circulation.

Neonate presents with: severe cyanosis, oxygen saturation often <75%, minimal murmur (no forward pulmonary flow), worsening after duct closure.

If PDA closes → rapid desaturation.

1. Echo Diagnosis — Stepwise Approach

STEP 1: PARASTERNAL LONG AXIS

Findings: large malalignment VSD, overriding aorta, no visible pulmonary valve opening. Aorta usually arises predominantly from RV.

STEP 2: RVOT EVALUATION

Key diagnostic point: no antegrade flow across pulmonary valve, no color Doppler jet into pulmonary artery, pulmonary valve appears imperforate.

Absence of flow confirms atresia.

STEP 3: ASSESS MAIN PULMONARY ARTERY

Two possibilities: (A) well-formed pulmonary arteries supplied by PDA; (B) hypoplastic or discontinuous pulmonary arteries supplied by MAPCAs.

Important for surgical planning.

STEP 4: LOOK FOR PDA

Color Doppler shows Aorta → Pulmonary artery flow. This confirms duct-dependent pulmonary circulation.

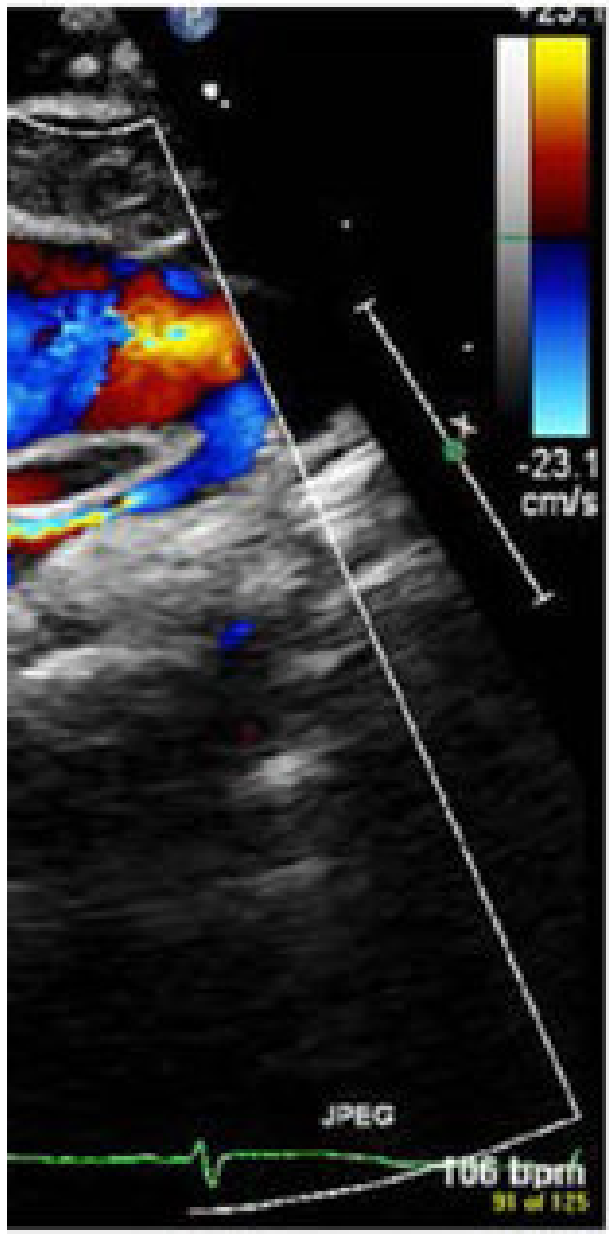
Start prostaglandin immediately if duct closing.

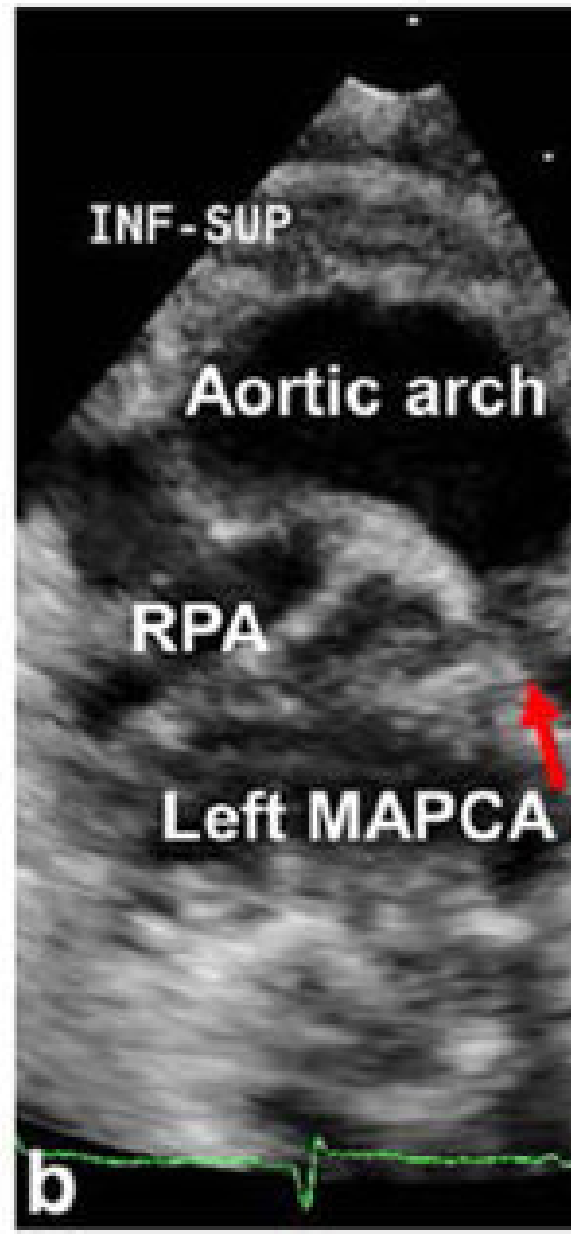
STEP 5: SEARCH FOR MAPCAs

MAPCAs arise from descending aorta or subclavian arteries.

Echo clues: abnormal vessels from aorta, continuous turbulent flow, small or absent central pulmonary arteries.

Often confirmed by CT or cath.





KEY ECHO FEATURES SUMMARY

- Large VSD
- Overriding aorta
- Absent pulmonary valve flow
- RV hypertrophy
- Ductal or collateral pulmonary supply

DIFFERENTIATE FROM SIMILAR CONDITIONS

Pulmonary atresia with intact ventricular septum — No VSD, RV usually small, different surgical pathway.

Severe Tetralogy of Fallot — In TOF, some antegrade pulmonary flow exists; in PA-VSD, no antegrade flow at all.

This distinction is critical.

2. Neonatal Diagnostic Traps

TRAP 1: MISTAKING FOR SEVERE TOF

Always confirm: is there ANY antegrade pulmonary flow? If none → pulmonary atresia.

TRAP 2: MISSING MAPCAs

If pulmonary arteries are small, search for collateral vessels carefully.

TRAP 3: ASSUMING GRADIENT MEASUREMENT HELPS

There is no gradient if no flow. Anatomy is diagnosis.

CLASSIC NEONATAL PATTERN

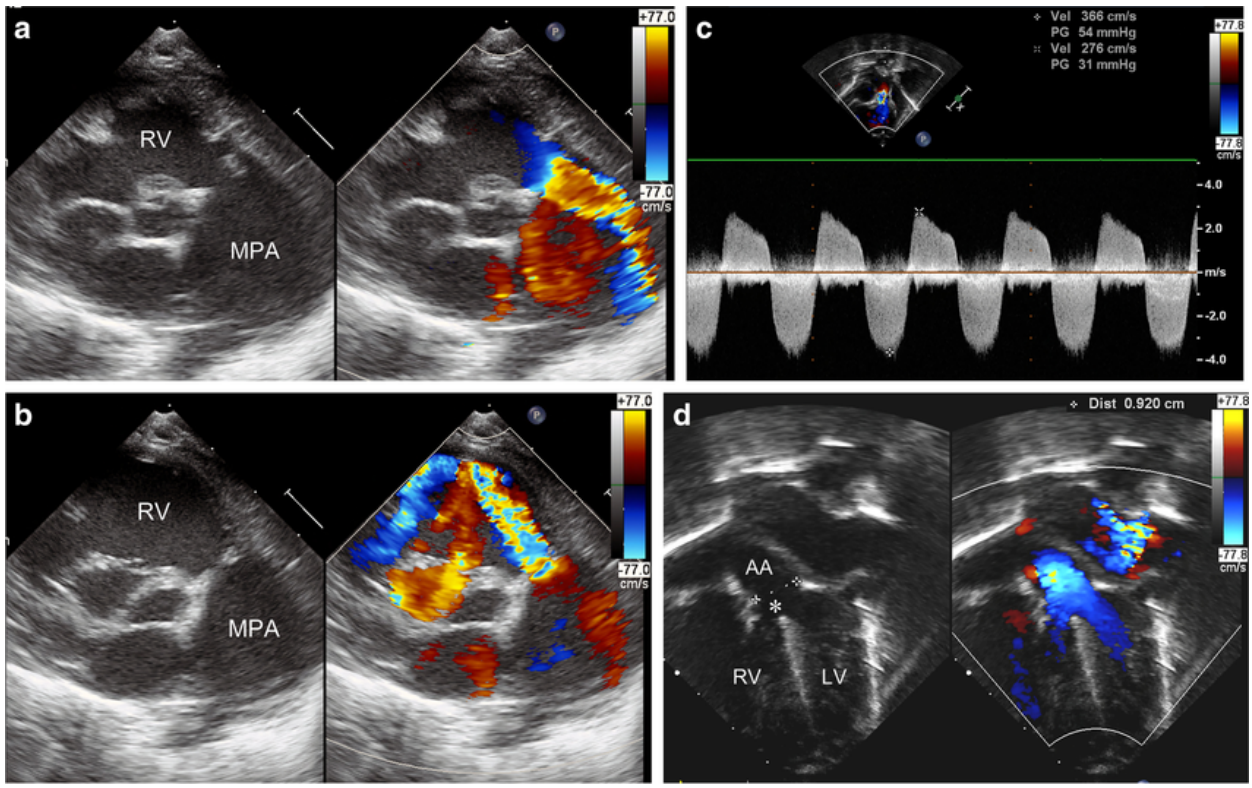
Severe cyanosis + large VSD + overriding aorta + no pulmonary valve flow + PDA supplying lungs → Think: Pulmonary Atresia with VSD.

Tetralogy of Fallot with Absent Pulmonary Valve (TOF-APV)

Tetralogy of Fallot with absent pulmonary valve is a rare variant of Tetralogy of Fallot characterized by:

1. Large malalignment VSD
2. Overriding aorta
3. Rudimentary or absent pulmonary valve
4. Massive pulmonary regurgitation
5. Severely dilated pulmonary arteries

Unlike classic TOF, the main problem is pulmonary artery dilation and airway compression, not just RVOT obstruction.



CORE PATHOPHYSIOLOGY

Because the pulmonary valve is absent or dysplastic: severe pulmonary regurgitation occurs, blood flows back into RV during diastole, pulmonary arteries progressively dilate, enlarged PAs compress bronchi.

Respiratory distress may be more prominent than cyanosis.

CLINICAL PRESENTATION IN NEONATES

- Respiratory distress
- Tachypnea
- Stridor or wheeze
- Hyperinflated lungs on X-ray
- Variable cyanosis

Key feature: airway compression symptoms.

1. Echo Diagnosis — Stepwise Approach

STEP 1: IDENTIFY TOF ANATOMY

Parasternal long axis: large malalignment VSD, overriding aorta. Classic TOF features must be present.

STEP 2: EVALUATE PULMONARY VALVE

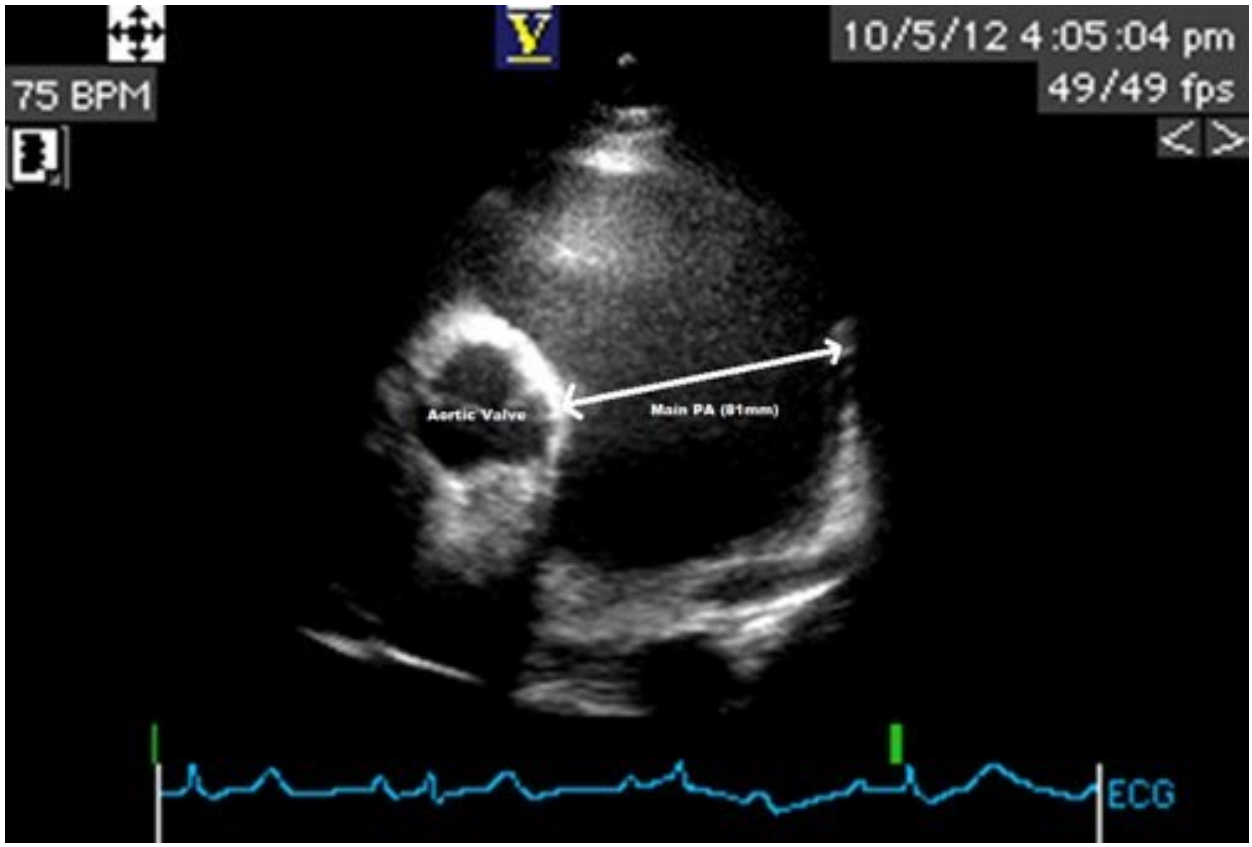
Findings: rudimentary valve tissue, no effective leaflet coaptation, severe pulmonary regurgitation.

Color Doppler: to-and-fro flow across RVOT, large diastolic regurgitant jet.

STEP 3: ASSESS PULMONARY ARTERIES

Characteristic finding: massive dilation of main and branch pulmonary arteries. This is the hallmark of TOF-APV.

Measure main PA diameter and branch PA diameters. They are often aneurysmal.



STEP 4: RV ASSESSMENT

RV may show volume overload (from regurgitation), hypertrophy, dilatation.

Unlike classic TOF, RV obstruction may be mild.

HOW IT DIFFERS FROM CLASSIC TOF

Feature	Classic TOF	TOF-APV
Pulmonary valve	Stenotic	Absent/dysplastic
Pulmonary arteries	Small	Massively dilated
Main issue	RVOT obstruction	Severe regurgitation
Symptoms	Cyanosis	Respiratory distress

IMPORTANT ASSOCIATIONS

TOF-APV is frequently associated with 22q11.2 deletion syndrome. Always consider genetic evaluation.

2. Diagnostic Traps

TRAP 1: THINKING ALL TOF HAVE SMALL PULMONARY ARTERIES

In TOF-APV: pulmonary arteries are massively dilated. This is opposite of classic TOF.

TRAP 2: MISSING SEVERE PULMONARY REGURGITATION

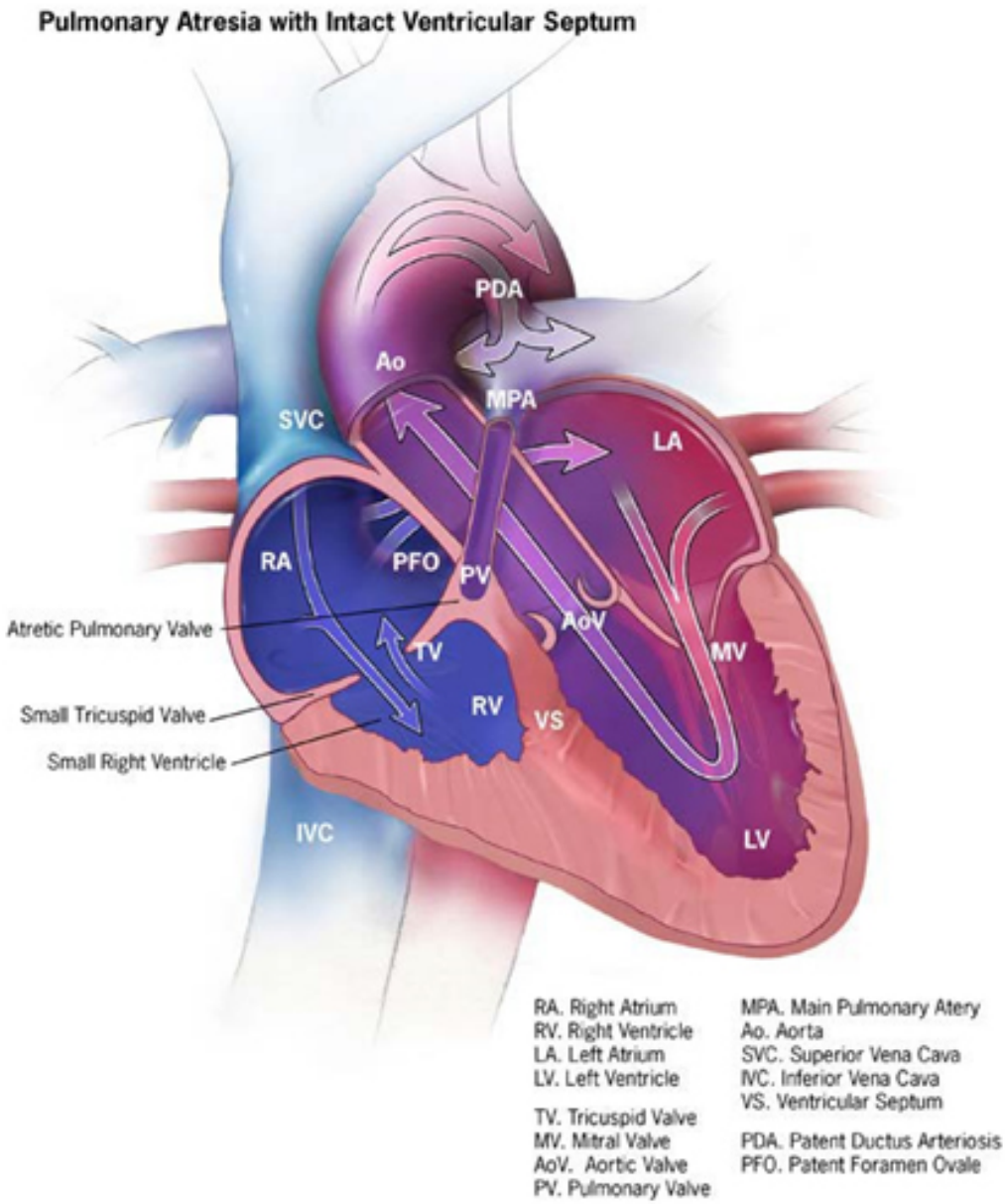
Look for: wide regurgitant jet, short pressure half-time, to-and-fro Doppler flow.

Pulmonary Atresia with Intact Septum (PA-IVS)

Pulmonary atresia with intact ventricular septum is a critical congenital heart defect characterized by:

1. Complete atresia of the pulmonary valve
2. No VSD (ventricular septum intact)
3. No antegrade flow from RV to pulmonary artery
4. Variable degrees of RV hypoplasia

This is a duct-dependent pulmonary circulation lesion.



Because the pulmonary valve is completely atretic: RV cannot eject blood into pulmonary artery; pulmonary blood flow depends on PDA (Aorta → PA).

Without duct → severe cyanosis.

Unlike PA-VSD: there is no VSD to decompress RV, RV pressure becomes suprasystemic.

CLINICAL PRESENTATION

- Severe cyanosis at birth
- Oxygen saturation 60–75%
- Minimal murmur (no antegrade flow)
- Rapid desaturation when duct closes

If PDA closes → life-threatening hypoxemia.

1. Echo Diagnosis — Stepwise Approach

STEP 1: CONFIRM INTACT VENTRICULAR SEPTUM

This is crucial. No VSD, intact septum on multiple views.

This differentiates from Pulmonary atresia with ventricular septal defect.

STEP 2: EVALUATE PULMONARY VALVE

Findings: imperforate pulmonary valve, no color Doppler flow, no CW Doppler jet.

Absence of antegrade pulmonary flow confirms atresia.

STEP 3: ASSESS RIGHT VENTRICLE

RV can be small and hypoplastic, hypertrophied with thick walls, or severely dysfunctional.

Tricuspid valve often small. Measure: tricuspid annulus Z-score, RV cavity size. These determine surgical strategy.

STEP 4: LOOK FOR CORONARY SINUSOIDS (CRITICAL!)

High RV pressure may create RV → coronary artery connections (sinusoids).

Echo clues: color Doppler flow from RV into myocardium, abnormal coronary filling.

If coronary circulation depends on RV pressure → high surgical risk.

STEP 5: ASSESS PDA

Color Doppler shows Aorta → Pulmonary artery flow. Confirms duct-dependent pulmonary circulation.

Start prostaglandin immediately.

SEVERITY SPECTRUM

PA-IVS has 3 broad anatomical patterns:

1. Mild RV Hypoplasia — RV can potentially grow; candidate for biventricular repair.

2. Moderate RV Hypoplasia — May require staged repair.

3. Severe RV Hypoplasia — Single ventricle pathway.

Outcome depends on RV size and coronary anatomy.

KEY ECHO FINDINGS SUMMARY

- Intact ventricular septum
- Atretic pulmonary valve
- Hypoplastic RV
- Small tricuspid valve
- Duct-dependent pulmonary flow
- Possible coronary sinusoids

2. Diagnostic Traps

TRAP 1: MISSING TINY VSD

Carefully exclude small VSD. If VSD present → diagnosis changes to PA-VSD.

TRAP 2: CONFUSING WITH CRITICAL PULMONARY STENOSIS

In critical PS: some antegrade flow exists. In PA-IVS: no antegrade flow at all.

TRAP 3: IGNORING CORONARY ABNORMALITIES

Coronary dependence dramatically changes management. Always evaluate coronaries.

IMMEDIATE MANAGEMENT

1. Start Prostaglandin E1
2. Stabilize oxygenation
3. Detailed echo assessment
4. Refer to tertiary cardiac center

Definitive strategy depends on: RV size, tricuspid size, coronary anatomy.

Critical Pulmonary Stenosis (PS)

Critical PS in the neonate = severe obstruction at the pulmonary valve → RV cannot eject effectively → Pulmonary blood flow becomes duct-dependent.

This is NOT just "severe PS"—it behaves like functional pulmonary atresia.

PATHOPHYSIOLOGY (WHY THE BABY GETS SICK)

- Severely thickened, doming pulmonary valve
- Tiny valve opening
- RV pressure becomes systemic or suprasystemic
- RV fails to deliver blood to lungs
- Pulmonary flow depends on PDA

When PDA closes → profound cyanosis.

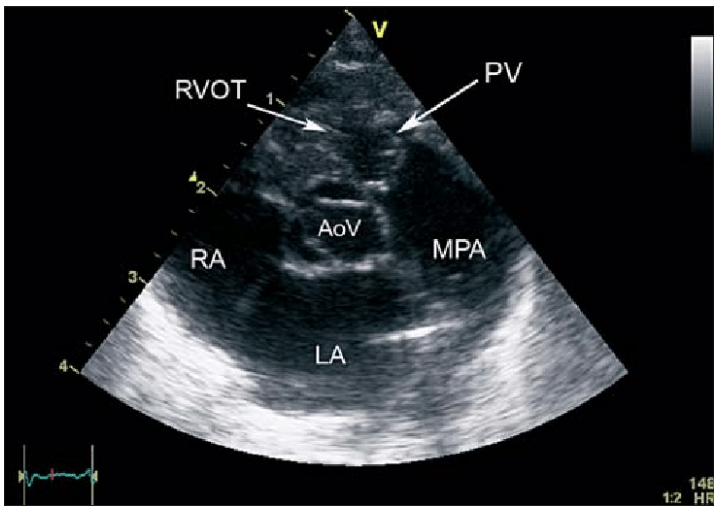
ECHO GOALS

When you suspect critical PS:

1. Confirm valve obstruction
2. Assess RV size & function
3. Confirm duct-dependent pulmonary flow
4. Exclude pulmonary atresia
5. Evaluate TR severity

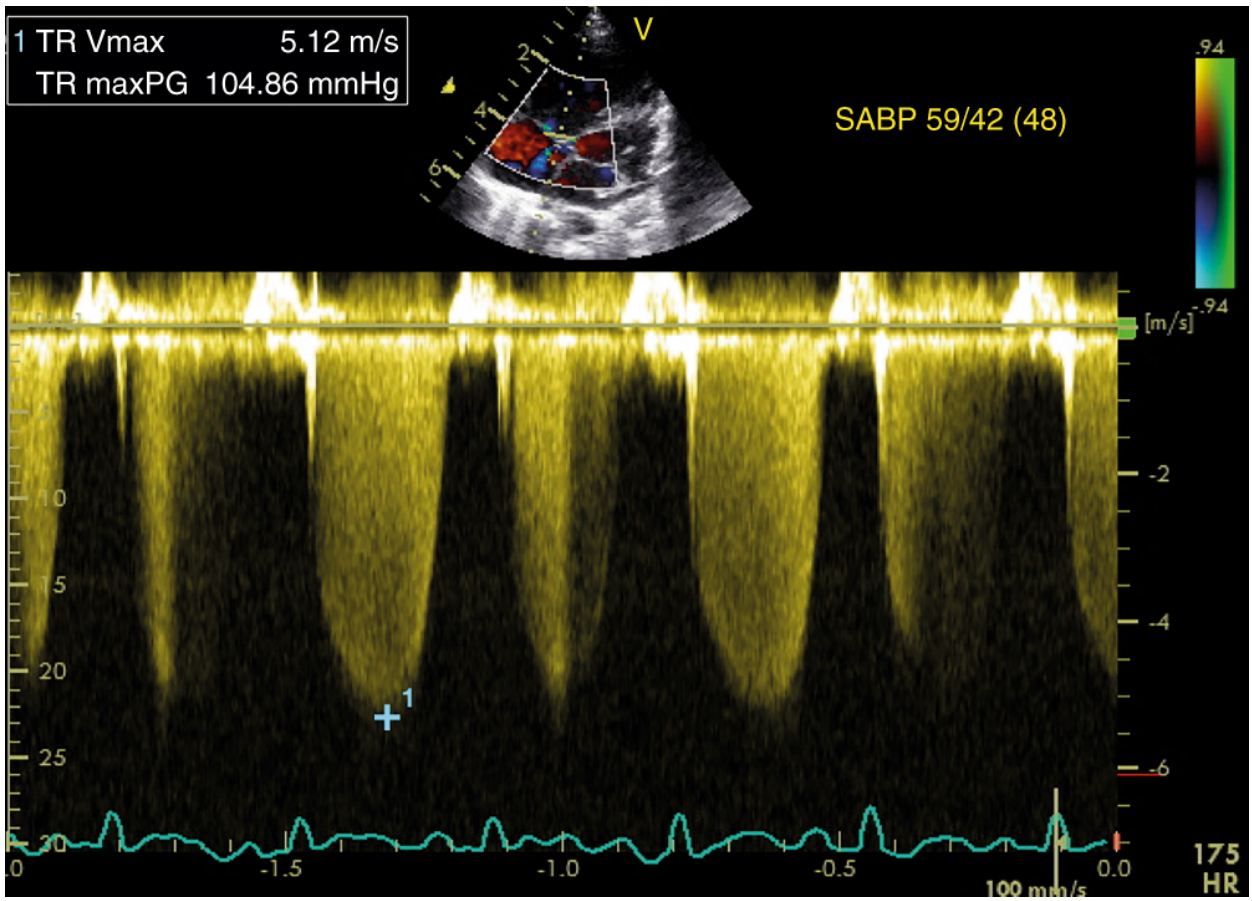
KEY ECHO VIEWS — PARASTERNAL SHORT AXIS (GREAT ARTERIES)

Look for: thickened leaflets, systolic doming, tiny central opening, post-stenotic PA dilation.



RV INFLOW VIEW

Assess: RV hypertrophy, RV dilation, tricuspid regurgitation (usually severe).



SUPRASTERNAL / HIGH PARASTERNAL (DUCT)

Confirm: ductal shunt, pulmonary arteries filling from aorta.

DOPPLER FINDINGS — ACROSS PULMONARY VALVE

Peak gradient may be very high, BUT in failing RV → gradient may be deceptively low.

Low gradient + sick cyanotic baby = critical obstruction

Tricuspid Regurgitation — TR jet estimates RV pressure. Usually RV pressure ≈ systemic.

How to Differentiate from Pulmonary Atresia

Feature	Critical PS	Pulmonary Atresia
Valve opening	Tiny opening present	No opening
Doppler flow	Minimal antegrade flow	No antegrade flow
Color Doppler	Thin jet across valve	No jet
RV	Usually adequate size	May be hypoplastic

MANAGEMENT IMPLICATION

Echo diagnosis → Immediate: start PGE1, urgent balloon pulmonary valvotomy, assess RV adequacy before intervention.

Double Outlet Right Ventricle (DORV)

WHAT IS DORV?

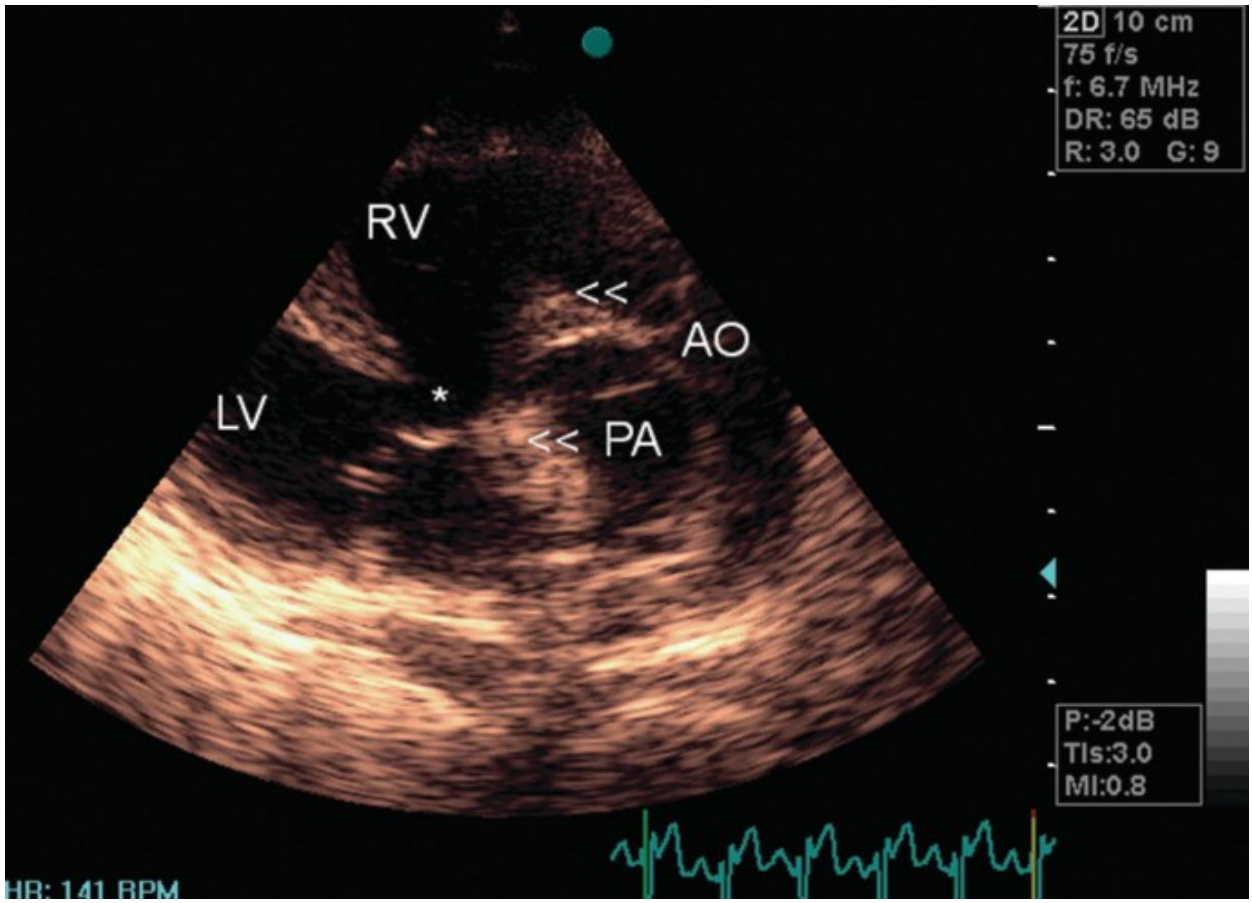
Definition: both great arteries arise predominantly (>50%) from the right ventricle.

Normal heart → Aorta from LV, PA from RV. DORV → Aorta + PA arise from RV.

Systemic output depends on a VSD.

STEP 1 IN ECHO: CONFIRM THE DEFINITION — PARASTERNAL LONG AXIS

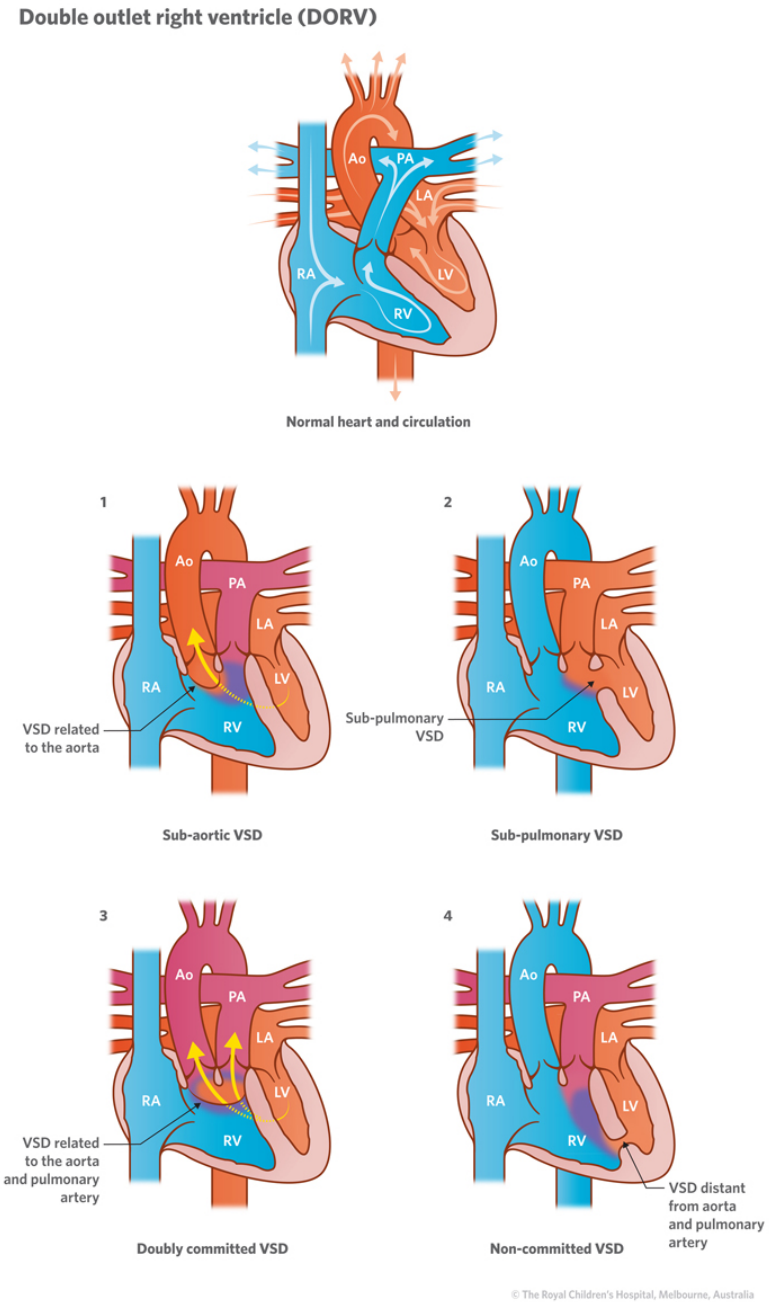
Look for: aorta not connected to LV, both great arteries arising from RV, VSD, aortomitral discontinuity.



TYPES OF DORV (ECHO-BASED CLASSIFICATION)

1. Subaortic VSD (Most common)

- VSD located beneath aorta
- LV blood preferentially enters aorta
- Physiology resembles VSD ± PS
- If PS present → behaves like TOF



2. Subpulmonary VSD (Taussig–Bing)

- VSD beneath pulmonary artery

- LV blood goes to PA
- Physiology resembles TGA
- Severe early cyanosis

High surgical urgency.

3. Doubly Committed VSD

- VSD related to both semilunar valves
- Often associated with outflow obstruction

4. Non-Committed (Remote) VSD

- VSD far from both great arteries
- Complex intracardiac routing
- Often requires complex repair

ESSENTIAL ECHO CHECKLIST

When you diagnose DORV, always answer:

1. VSD size?
2. VSD location?
3. Is there pulmonary stenosis?
4. Aortic override percentage?
5. LV size adequate?
6. RV size and function?
7. Any arch obstruction?
8. Coronary anatomy (if possible)?

NEONATAL PRESENTATION PATTERNS

Cyanotic neonate? Think: subpulmonary VSD, DORV + severe PS.

Heart failure at 4–8 weeks? Think: subaortic VSD without PS.

1. Diagnostic Traps

Mistaking DORV for TOF → In TOF, aorta overrides but still connected to LV; in DORV, both arteries primarily from RV.

Missing subpulmonary VSD → baby may look like TGA.

Not assessing arch → coarctation may coexist.

How to Differentiate From TOF

Feature	DORV	TOF
Aortic connection	>50% from RV	Partial override
LV outlet	Only via VSD	Direct to aorta

Feature	DORV	TOF
Great artery relation	Variable	Usually normal
VSD	Essential for output	Malalignment type

Tricuspid Atresia

Tricuspid atresia is a congenital defect where:

- The tricuspid valve is completely absent
- There is no direct connection between RA and RV
- The RV is hypoplastic

This is a single ventricle physiology lesion.

CORE PATHOPHYSIOLOGY

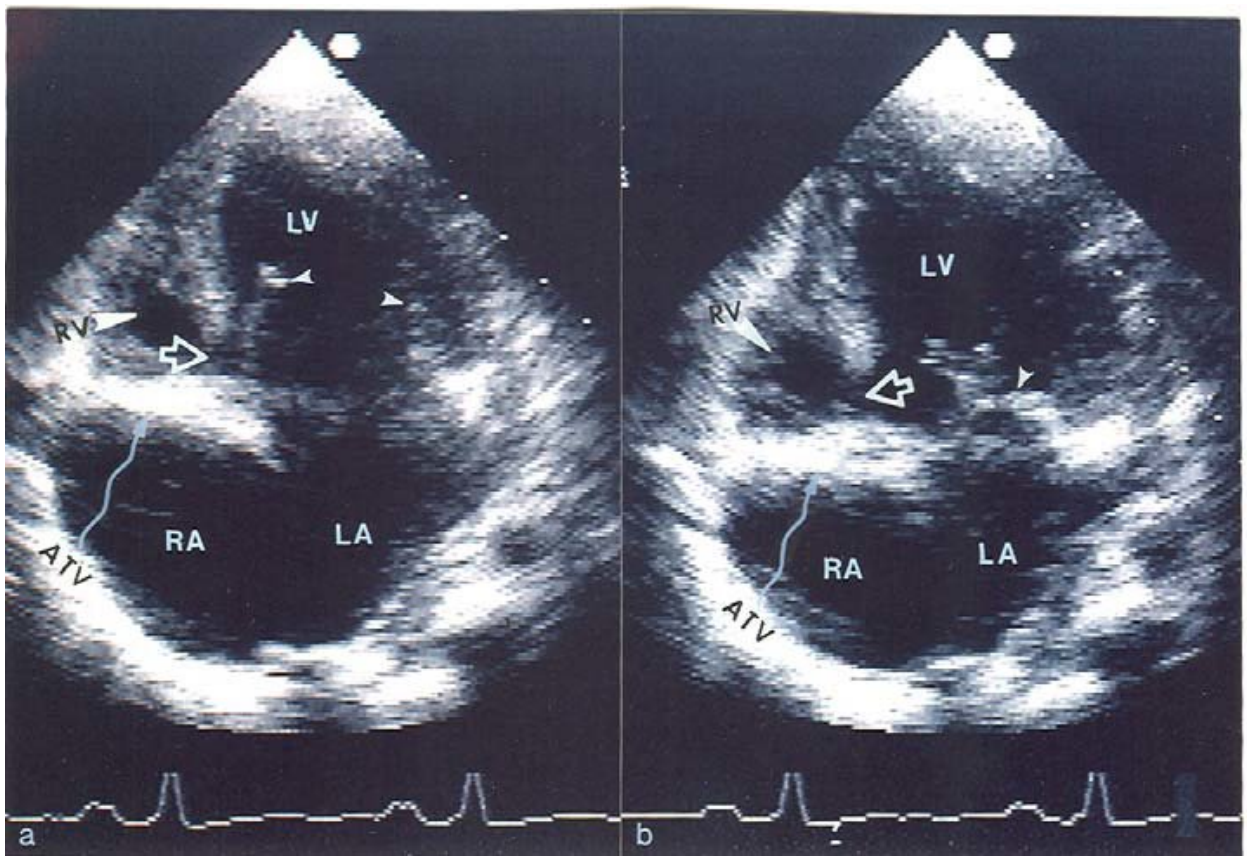
Because RA cannot empty into RV: RA → shunts across ASD to LA; LA → LV → systemic circulation.

Pulmonary blood flow depends on: VSD (if present), PDA, presence/absence of pulmonary stenosis.

ECHO GOAL: CONFIRM NO RA–RV CONNECTION — APICAL 4 CHAMBER VIEW

Look for: no tricuspid valve opening, echogenic plate at tricuspid position, small hypoplastic RV, large LV.

Key sign: No color Doppler flow from RA to RV.



STEP 2: IDENTIFY VENTRICULO-ARTERIAL RELATIONSHIP

Most common type: normally related great arteries.

But may be associated with: Transposition of great arteries, DORV.

Always define: (1) where does the aorta arise? (2) where does the PA arise?

STEP 3: ASSESS PULMONARY BLOOD FLOW

This determines neonatal presentation.

A. With Pulmonary Stenosis (Common) — Restricted pulmonary flow, severe cyanosis, duct-dependent pulmonary circulation.

B. Without Pulmonary Stenosis — Large VSD, excess pulmonary flow, heart failure at 4–8 weeks.

Echo Classification (Clinically Useful)

Type	Great Arteries	Pulmonary Flow
Type I	Normally related	± PS
Type II	TGA	Usually high pulmonary flow
Type III	Other variants	Variable

HEMODYNAMIC PATTERN

RA pressure elevated; large ASD (mandatory for survival); LV = systemic ventricle; RV = hypoplastic.

Important: if ASD is restrictive → emergency septostomy.

KEY ECHO CHECKLIST

When diagnosing tricuspid atresia, always document:

- Type of great artery relationship
- VSD presence and size, ASD size
- Pulmonary stenosis severity
- PDA presence

NEONATAL PRESENTATION PATTERNS

Presentation	Echo Physiology
Severe cyanosis day 1	PS + duct-dependent flow
Cyanosis after duct closes	Inadequate pulmonary flow
CHF at 1 month	No PS + large VSD

1. Diagnostic Traps

Confusing with pulmonary atresia with intact septum → in PA-IVS, tricuspid valve exists (often small).

Missing restrictive ASD → baby deteriorates rapidly.

Not assessing pulmonary outflow obstruction.

CLASSIC ECHO PATTERN

No tricuspid valve, no RA–RV flow, small RV, large LV, mandatory ASD, VSD determines pulmonary flow.

Think: Tricuspid Atresia.

Truncus Arteriosus

Truncus arteriosus (Common Arterial Trunk) is a rare cyanotic congenital heart defect in which a single arterial trunk arises from the heart and supplies the systemic, pulmonary, and coronary circulations.

Instead of separate aorta and pulmonary artery, there is:

- One common arterial trunk
- One truncal valve
- A large outlet (malalignment) VSD
- Variable pulmonary artery anatomy

It accounts for approximately **1–2% of congenital heart disease**.

EMBRYOLOGY

Normally, the truncus arteriosus divides into the aorta and pulmonary artery.

Failure of truncoconal septation results in: single arterial trunk, single semilunar valve, outlet VSD.

PATHOPHYSIOLOGY

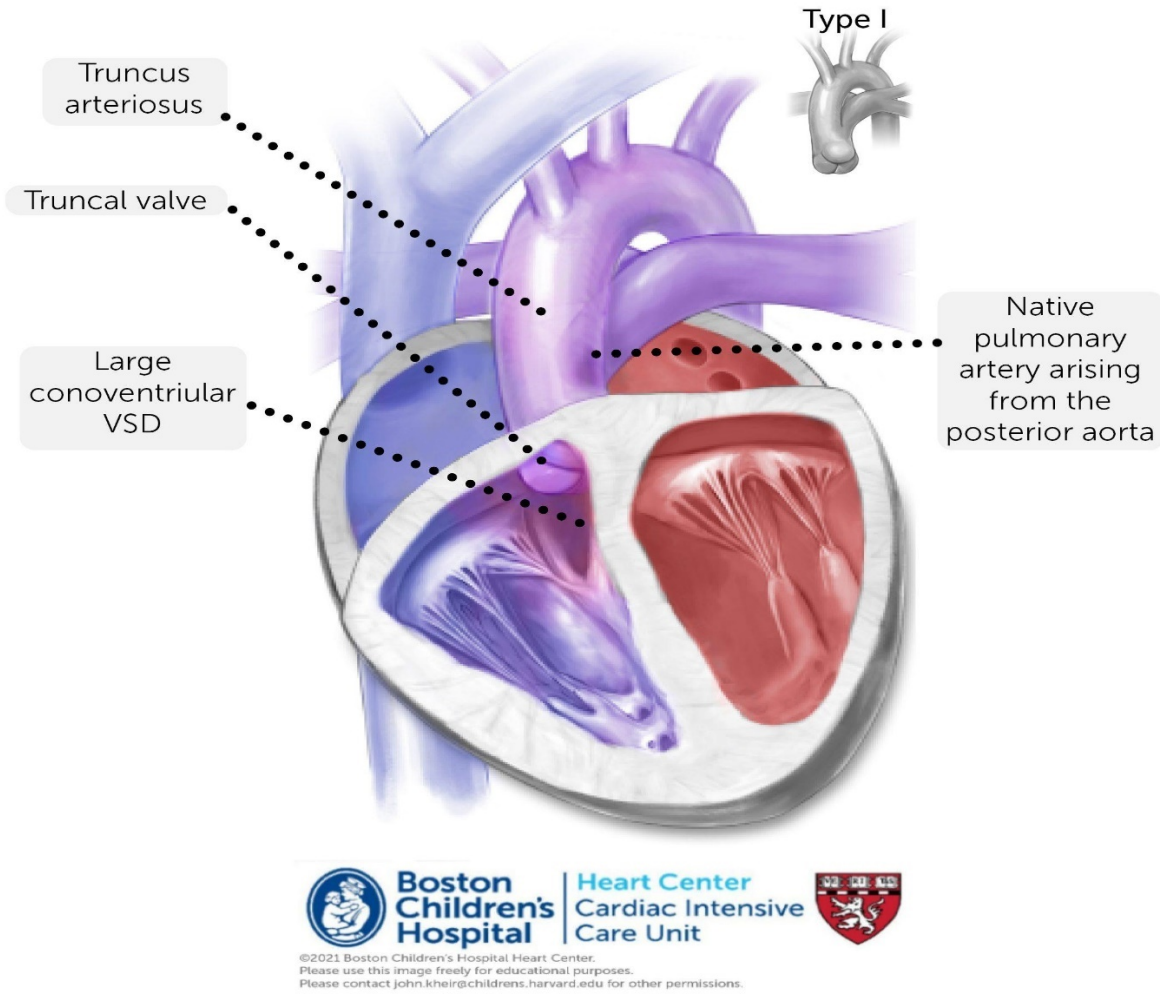
Both ventricles eject into one common artery. Consequences: complete mixing of oxygenated and deoxygenated blood, increased pulmonary blood flow, congestive heart failure, pulmonary hypertension, mild to moderate cyanosis.

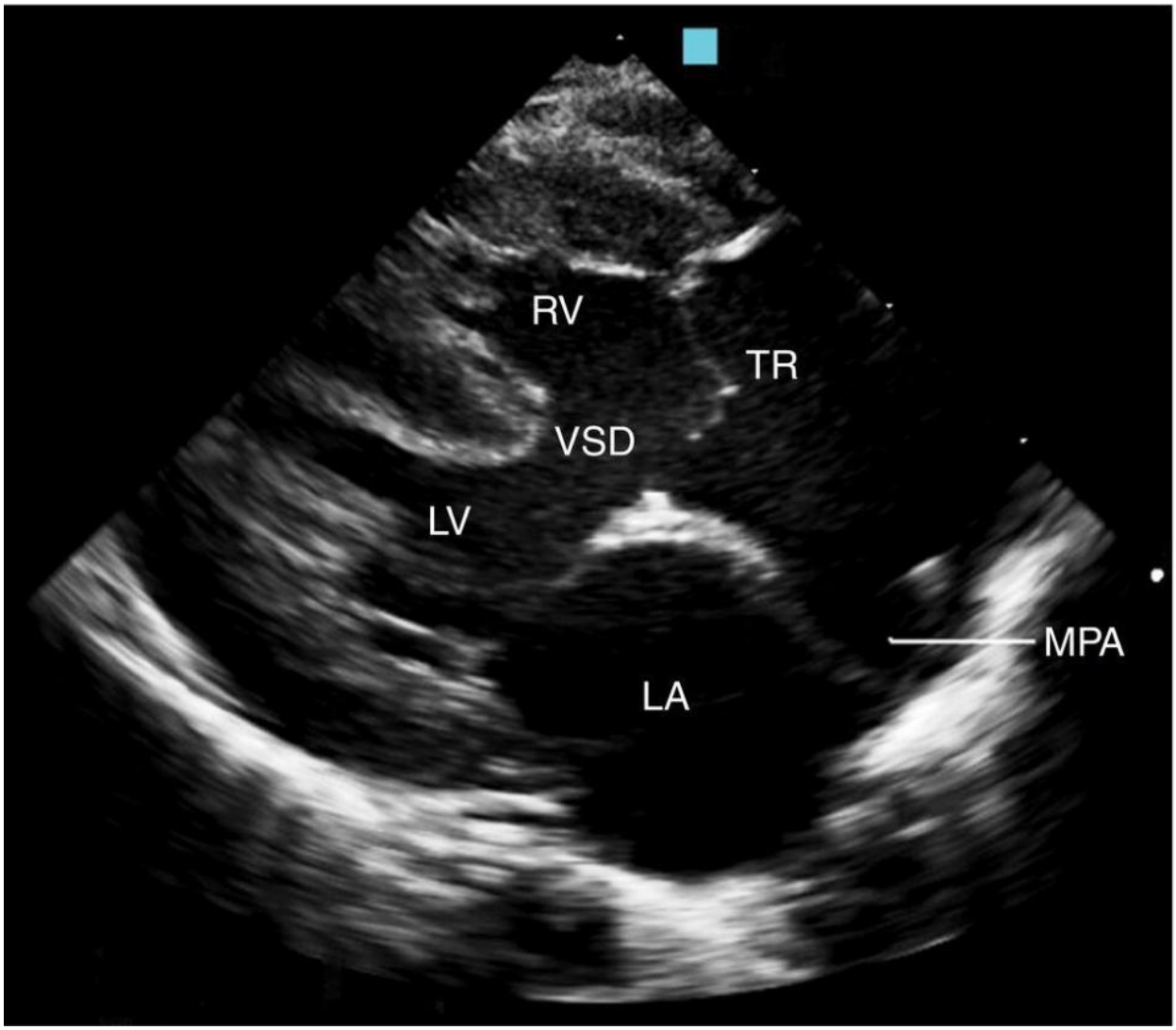
Without repair: progressive pulmonary vascular disease develops early.

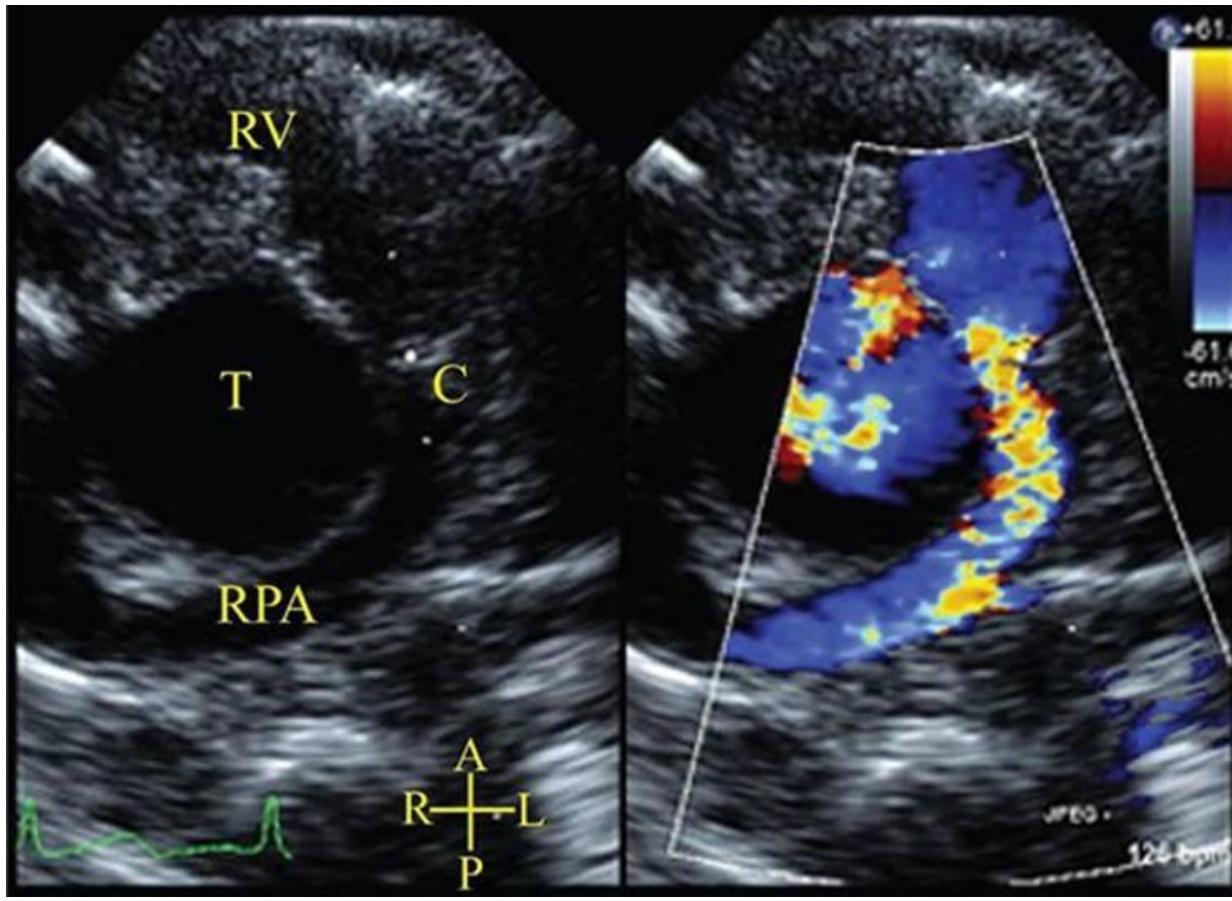
1. Collett and Edwards Classification

TYPE I (MOST COMMON)

A short main pulmonary artery arises from the truncal artery and divides into the right and left pulmonary arteries.

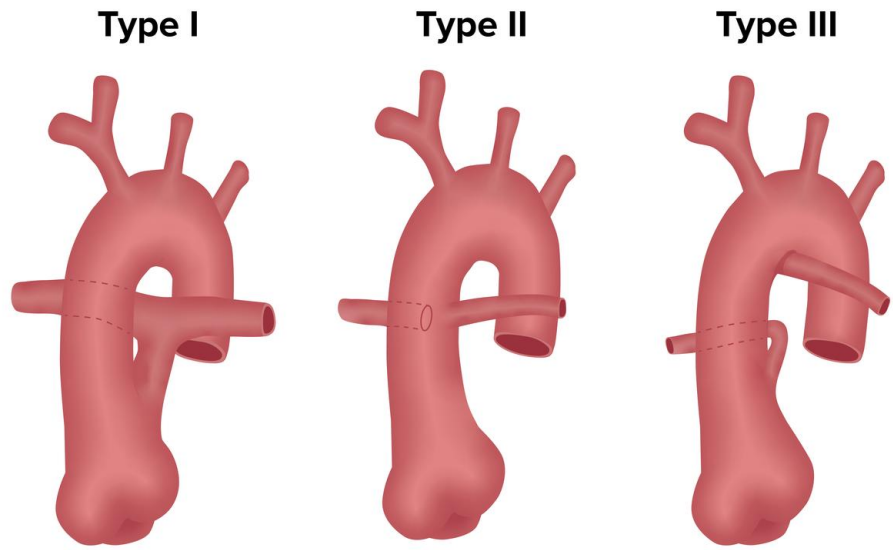

Type I
Truncus arteriosus
Truncal valve
Large conoventriular VSD
Native pulmonary artery arising from the posterior aorta
Boston Children's Hospital
Heart Center
Cardiac Intensive Care Unit
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RV
TR
VSD
LV
MPA
LA



TYPE II

Right and left pulmonary arteries arise separately from the posterior aspect of the truncus.



TYPE III

Right and left pulmonary arteries arise separately from the lateral sides of the truncus.

TYPE IV

Historically described but **no longer considered truncus arteriosus**. It is now classified as **Pulmonary Atresia with VSD**.

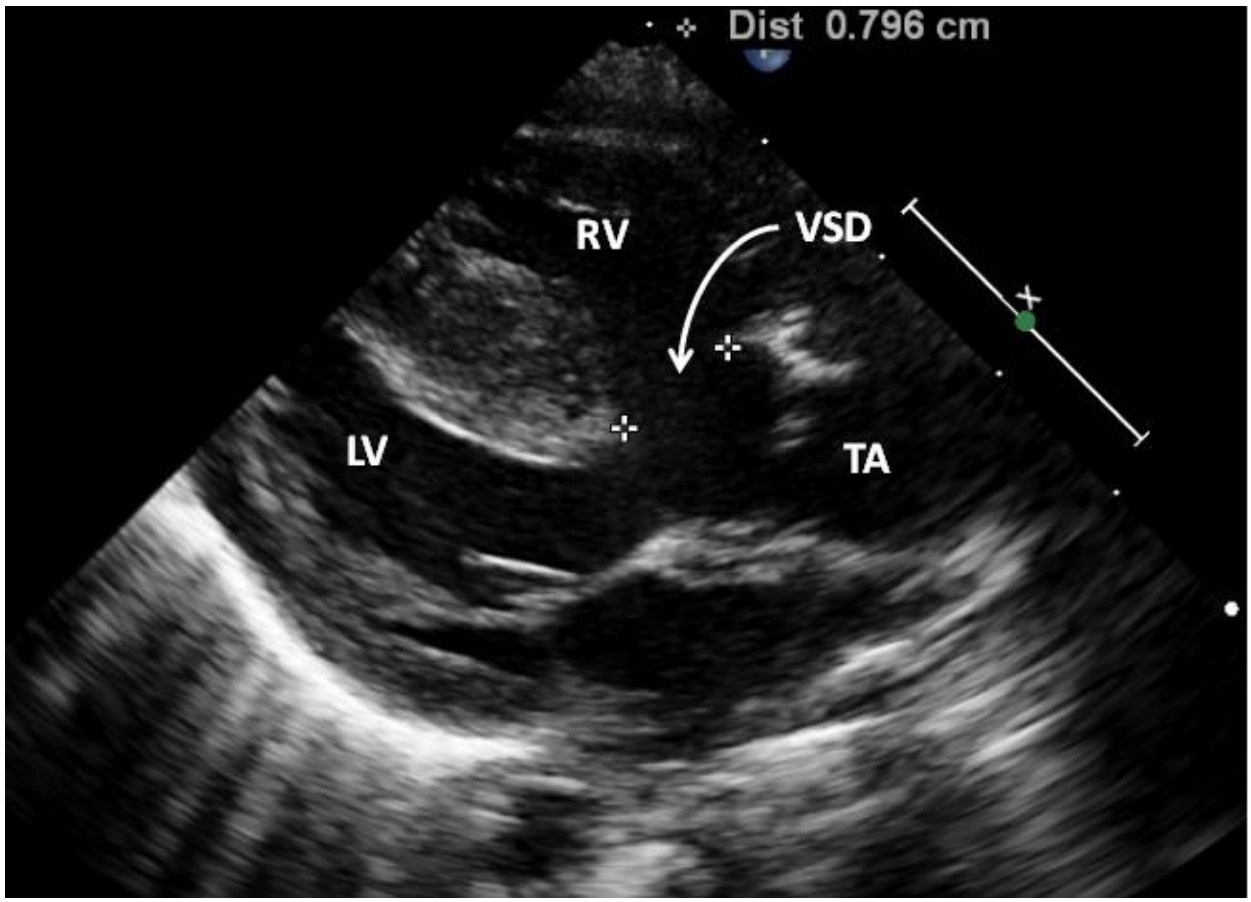
2. Echocardiographic Examination

STEP 1 — APICAL FOUR-CHAMBER

Evaluate: ventricular size, AV valves, large VSD, ventricular function.

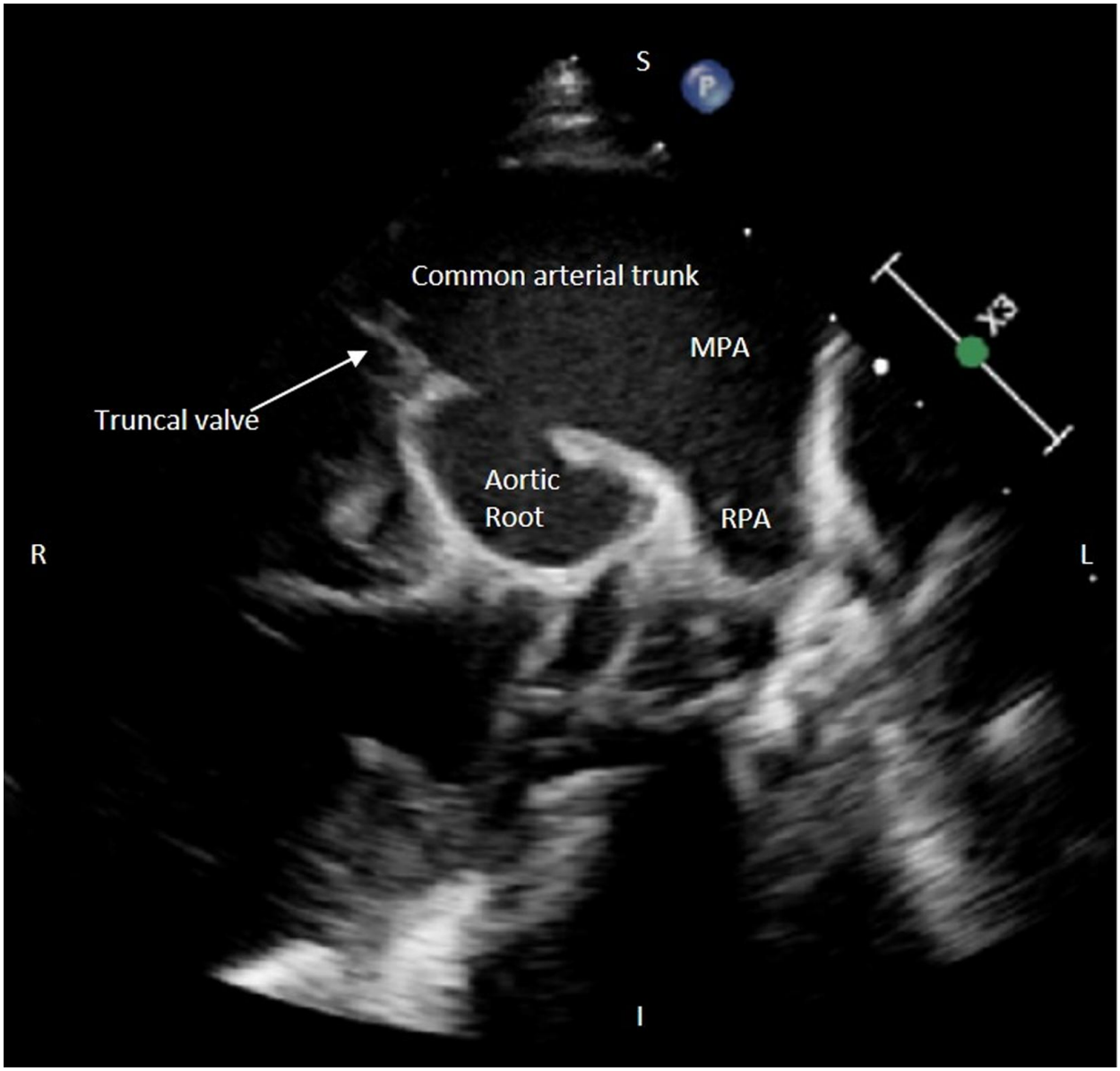
STEP 2 — PARASTERNAL LONG-AXIS (KEY VIEW)

Look for: single great artery, overriding truncal valve, large outlet VSD.



STEP 3 — PARASTERNAL SHORT-AXIS

Evaluate: truncal valve morphology, coronary artery origins, pulmonary artery origins.



STEP 4 — SUPRASTERNAL NOTCH

Assess: aortic arch, interrupted arch, coarctation, pulmonary artery branching.

HALLMARK ECHO FINDINGS — MAJOR DIAGNOSTIC FEATURES

- Single arterial trunk
- Large malalignment VSD
- Overriding truncal valve
- Single semilunar valve
- Pulmonary arteries arising from the truncus

These five findings establish the diagnosis.

TRUNCAL VALVE ASSESSMENT

The truncal valve may have three cusps, four cusps (most common abnormality), two cusps, or dysplastic leaflets.

Assess: valve morphology, regurgitation, stenosis.

TRUNCAL REGURGITATION

Evaluate: color Doppler jet, vena contracta, LV dilation, diastolic flow reversal.

Significant truncal regurgitation worsens prognosis.

VENTRICULAR SEPTAL DEFECT

The VSD is usually large, outlet type, nonrestrictive.

Measure: size, relationship to truncal valve, direction of shunt.

PULMONARY ARTERY ASSESSMENT

Identify: main pulmonary artery, right pulmonary artery, left pulmonary artery.

Document: site of origin, diameter, branch stenosis, discontinuity.

CORONARY ARTERIES

Always identify left and right coronary origin. Important before surgical repair.

ASSOCIATED LESIONS

- Interrupted aortic arch
- Right aortic arch
- ASD
- Persistent left SVC
- Coronary anomalies
- 22q11 deletion syndrome (DiGeorge syndrome)

DOPPLER FINDINGS

Color Doppler shows: large VSD flow, truncal valve regurgitation, pulmonary artery flow.

Continuous-wave Doppler used for: truncal stenosis, truncal regurgitation.

Differential Diagnosis

Condition	Echo Difference
Pulmonary atresia with VSD	Separate aorta present
Tetralogy of Fallot	Separate pulmonary valve
DORV	Two great arteries present
TGA	Two separate great arteries

SURGICAL PLANNING CHECKLIST

Before surgery document:

- Pulmonary artery anatomy
- Degree of truncal regurgitation

- Coronary origins
- Truncal valve morphology
- Interrupted arch
- Ventricular function

COMMON ECHO PITFALLS

- Missing separate pulmonary artery origins
- Confusing PA/VSD with truncus arteriosus
- Ignoring truncal valve regurgitation
- Not assessing coronary arteries
- Missing interrupted aortic arch

Single Ventricle Physiology

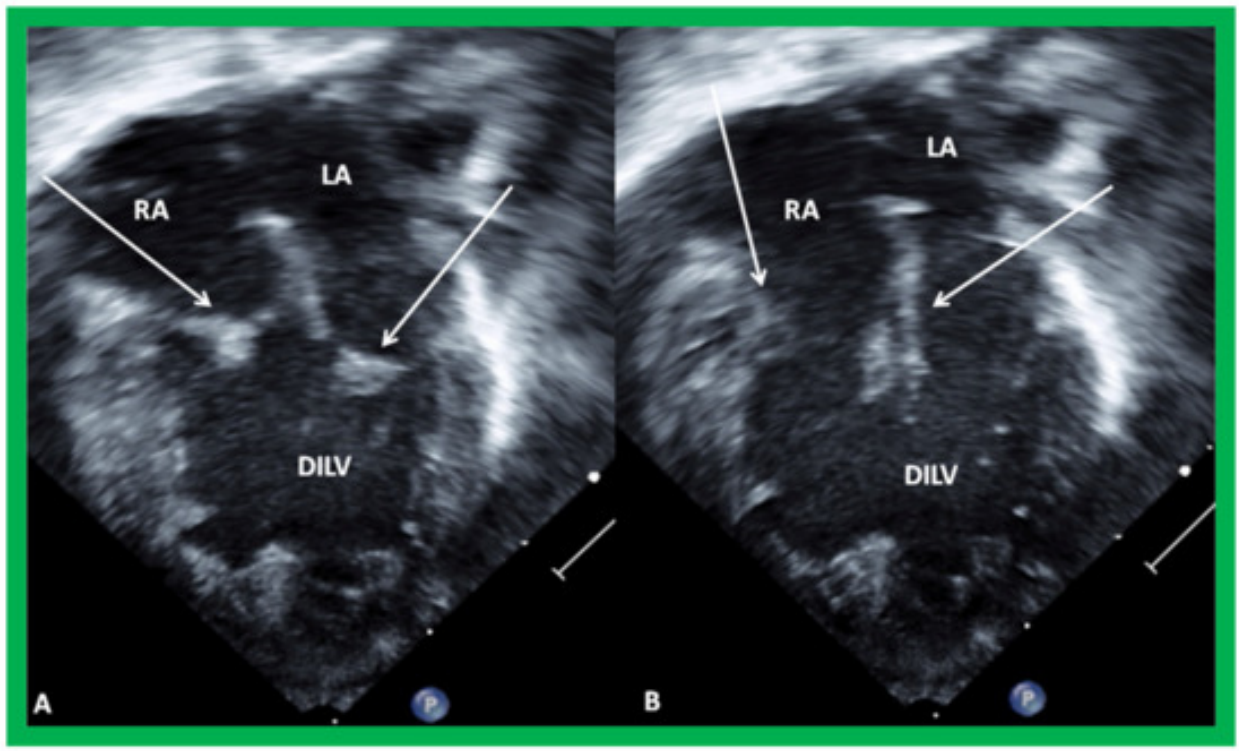
WHAT IS SINGLE VENTRICLE PHYSIOLOGY?

Only one functional pumping ventricle supports systemic circulation.

The second ventricle is: hypoplastic, non-functional, or absent.

Common examples include:

- Tricuspid atresia
- Hypoplastic left heart syndrome
- Double inlet left ventricle
- Unbalanced atrioventricular septal defect



HEMODYNAMIC CONCEPT

In single ventricle: one ventricle handles mixed blood, systemic and pulmonary circulations compete, flow balance determines symptoms.

NEONATAL PRESENTATION PATTERNS

Presentation	Physiology
Severe cyanosis	Low pulmonary flow
Shock after duct closure	Systemic obstruction
CHF at 3–6 weeks	High pulmonary flow
Relatively stable sats 75–85%	Balanced flow

ECHO GOALS IN ANY SINGLE VENTRICLE

1. Which ventricle is dominant?
2. Is pulmonary flow high, low, or balanced?
3. Is systemic outflow obstructed?
4. Is AV valve regurgitation present?
5. Is arch normal?
6. Is duct needed?

1. Step 1 — Identify Ventricular Morphology

Apical 4 Chamber — Assess: which ventricle is large? Is the other ventricle hypoplastic? AV valve number and morphology?

Morphologic LV: smooth septal surface, fine trabeculations.

Morphologic RV: coarse trabeculations, moderator band.

2. Step 2 — Determine Pulmonary Blood Flow

This determines neonatal stability.

LOW PULMONARY FLOW (CYANOTIC BABY)

Seen in: tricuspid atresia + PS, PA with intact septum, severe TOF variants.

Echo findings: pulmonary stenosis/atresia, duct-dependent pulmonary flow, small PA.

Management: PGE1.

HIGH PULMONARY FLOW (HEART FAILURE)

Seen in: unrestricted VSD, no pulmonary stenosis.

Echo findings: large VSD, dilated pulmonary arteries, volume overload.

Management: diuretics → PA band.

3. Step 3 — Evaluate Systemic Outflow

In single ventricle, systemic obstruction is catastrophic.

Check: aortic valve, LVOT, arch, coarctation. Especially critical in hypoplastic left heart syndrome.

4. Step 4 — AV Valve Assessment

AV valve regurgitation worsens prognosis.

Look for: moderate/severe regurgitation, annular dilation, leaflet dysplasia.

AV valve regurgitation = major surgical risk factor.

ECHO RED FLAGS

- Restrictive ASD
- Arch obstruction
- Severe AV valve regurgitation
- Poor ventricular function
- Duct-dependent systemic circulation

Glenn & Fontan — Echo Essentials

AFTER GLENN

- SVC flow to PA
- No SVC obstruction
- Low PA pressures

AFTER FONTAN

- No fenestration obstruction
- Good ventricular function
- No AV valve regurgitation
- No thrombus

MODULE 4

Acyanotic Congenital Heart Diseases

Topics
Complete Atrioventricular Septal Defect (CAVSD)
Atrial Septal Defect (ASD)
Primum ASD / PFO / Bubble Study / Sinus Venosus ASD / Atrial Septal Aneurysm
Ventricular Septal Defect (VSD)
Aortic Valve Abnormalities (BAV, AS, AR, Aortic Root)
Pulmonary Stenosis
Tricuspid Valve — Ebstein Anomaly
Mitral Valve — Prolapse & Scallop Localization

1. Complete Atrioventricular Septal Defect (CAVSD)

WHAT IS CAVSD?

Atrioventricular septal defect (Complete AVSD) is a defect characterized by:

- Common AV valve
- Primum ASD
- Inlet VSD
- Loss of normal AV septum

It is a **single atrioventricular junction defect**.

Strong association with: Down syndrome.

ANATOMY CONCEPT

Normally: mitral and tricuspid valves are separate; there is AV septal offset.

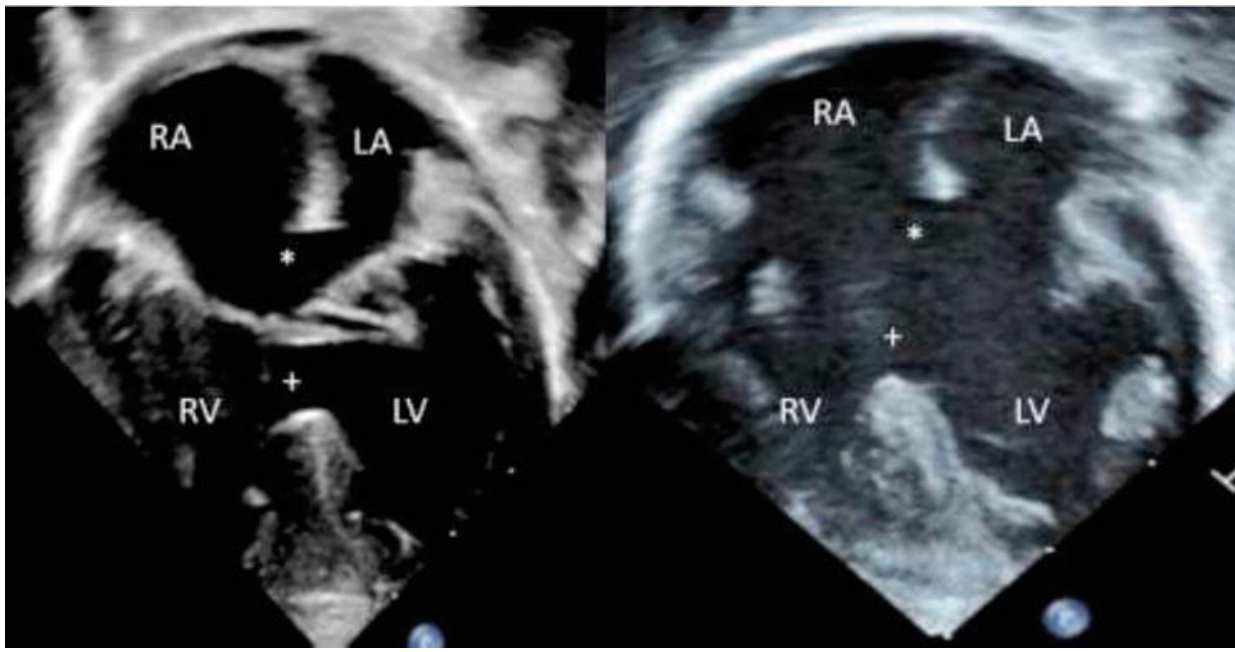
In CAVSD: no AV septal offset, single AV valve, large atrial + ventricular communication.

2. Echo Steps

APICAL 4 CHAMBER

Look for: common AV valve spanning both ventricles, no normal mitral–tricuspid offset, large primum ASD, inlet VSD, "single valve plane" appearance.

This view often makes the diagnosis immediately.



AV VALVE ANATOMY (CRITICAL FOR SURGERY)

The common AV valve has: superior bridging leaflet, inferior bridging leaflet, left mural leaflet, right mural leaflet.

Echo must assess: valve morphology, degree of regurgitation, balanced vs unbalanced ventricles.

Balanced CAVSD — both ventricles adequate size, biventricular repair possible.

Unbalanced CAVSD — one ventricle small, may require single ventricle pathway.

HEMODYNAMICS

High pulmonary blood flow: large L → R shunt, pulmonary overcirculation, CHF at 4–8 weeks.

Rare neonatal cyanosis unless: severe pulmonary stenosis, unbalanced physiology.

DOPPLER EVALUATION

Color Doppler: large atrial shunt, large ventricular shunt, AV valve regurgitation jets.

CW Doppler: evaluate AV valve regurgitation severity.

KEY ECHO CHECKLIST

When diagnosing CAVSD, document:

1. Type: complete vs partial
2. Balanced or unbalanced
3. AV valve regurgitation severity
4. Ventricular size adequacy
5. LVOT obstruction (important!)
6. Pulmonary hypertension signs
7. Associated lesions, situs and interrupted IVC

LVOT OBSTRUCTION — IMPORTANT TRAP

In some cases: elongated LVOT, "Goose-neck" deformity. May cause subaortic obstruction.

Always check LVOT gradient.

DIFFERENTIATING COMPLETE VS PARTIAL AVSD

Feature	Complete	Partial
AV valve	Single	Two valves
VSD	Present	Absent
ASD	Primum	Primum
Neonatal CHF	Common	Less common

SURGICAL PERSPECTIVE (ECHO GUIDES STRATEGY)

Balanced CAVSD: early repair (3–6 months), patch ASD + VSD, AV valve cleft repair.

Unbalanced CAVSD: may require single ventricle palliation.

Echo determines operability.

RED FLAGS ON ECHO

- Severe AV valve regurgitation
- Ventricular imbalance
- LVOT obstruction
- Elevated PA pressures
- Poor ventricular function

Atrial Septal Defect (ASD)

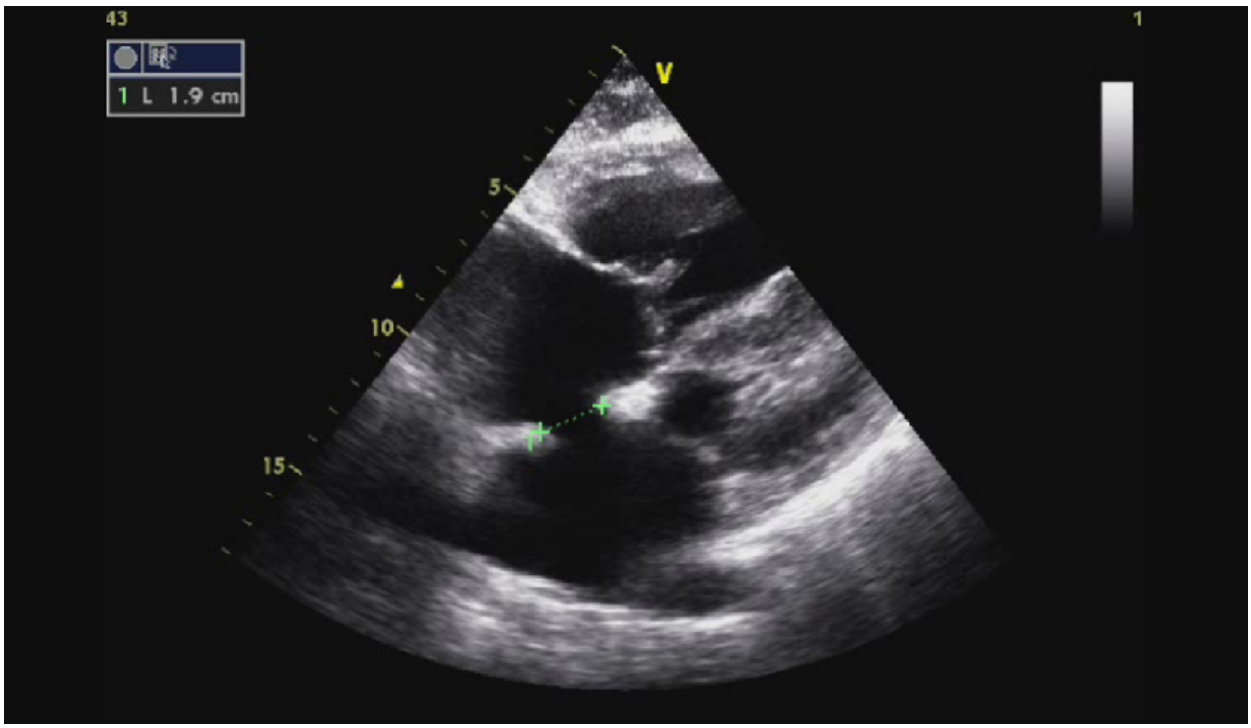
Atrial Septal Defect (ASD) is a communication between the right and left atria causing **left-to-right shunt** at atrial level.

It results in: right atrial enlargement, right ventricular volume overload, increased pulmonary blood flow.

TYPES OF ASD

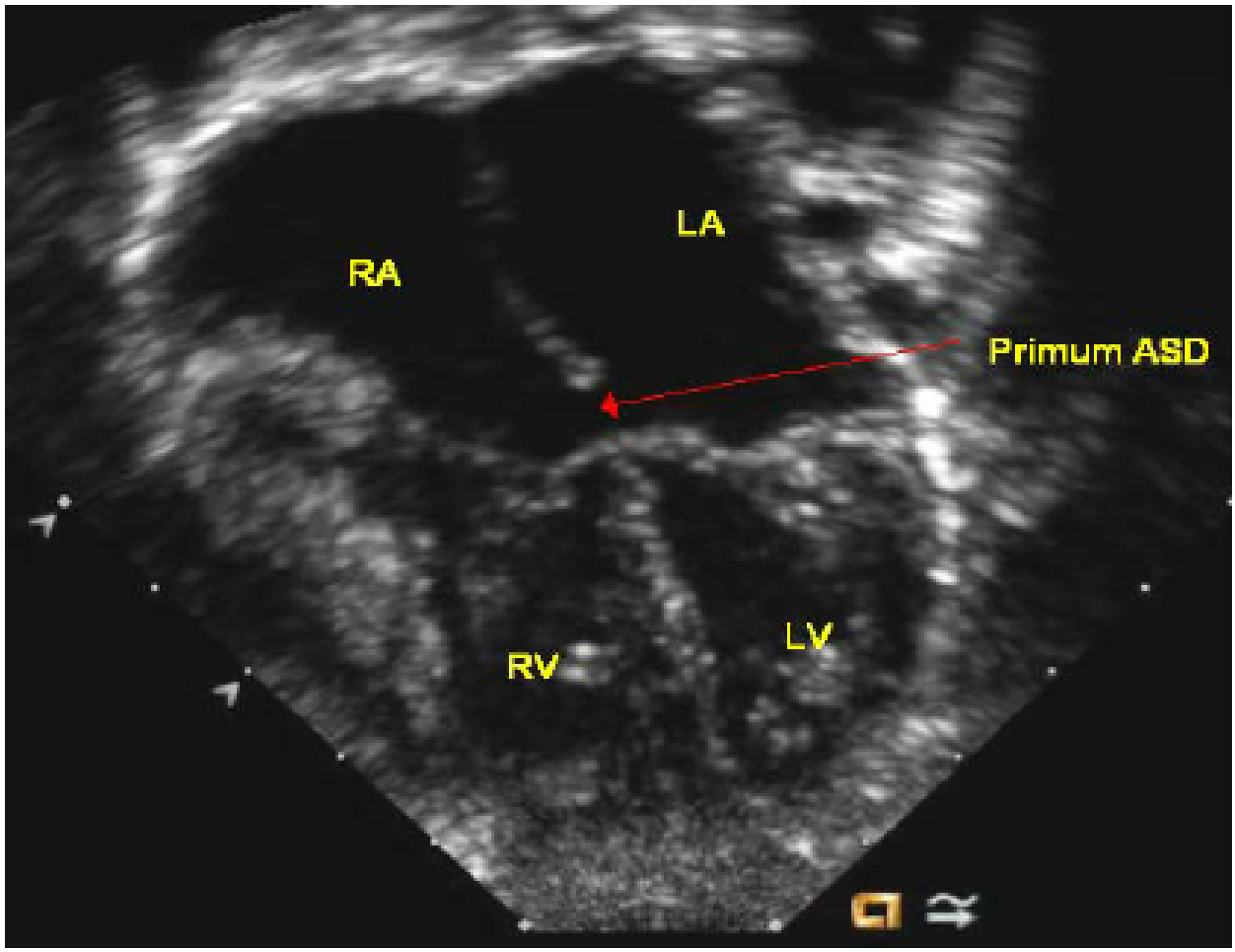
Secundum ASD (Most Common — 70–80%)

Located at **fossa ovalis**, central septum. Suitable for **device closure**.



1. Primum ASD (AV Septal Type)

Low septum. Part of **AVSD spectrum**. Associated with: mitral cleft.



2. Echo Diagnosis — Stepwise Approach

STEP 1: SUBCOSTAL VIEW (BEST IN CHILDREN)

Look for defect in septum, assess size, use color Doppler.

STEP 2: APICAL 4-CHAMBER

Check RV enlargement, evaluate AV valves.

STEP 3: MEASURE

- Defect diameter
- Septal rims (for device suitability)
- RV size
- PA pressure (TR jet)

ECHO CLUES OF SIGNIFICANT ASD

- RV dilatation
- Septal flattening (volume overload)
- Increased PA flow
- Qp:Qs > 1.5:1

WHEN TO CLOSE?

Indications: RV enlargement, significant shunt, symptoms.

Closure options: device (Secundum only), surgery (Primum / Sinus venosus).

Best age: 2–5 years (if significant).

3. Diagnostic Pitfalls

Patent Foramen Ovale (PFO) mistaken as ASD → small, flap-like, no RV dilation.

Sinus venosus missed → always check pulmonary veins.

Underestimating size → use multiple views.

ONE-LINE CLINICAL MEMORY

ASD = RV volume overload disease. If RV is normal size, think again.

Think of dropout / thin IAS / PFO.

PFO Assessment

STEP 1: CONFIRM IT IS TRULY A PFO (NOT ASD)

Essential echo criteria: flap-like tunnel, no true septal tissue deficiency, no RV enlargement, shunt only with Valsalva.

If RV is dilated → reconsider ASD.

STEP 2: PERFORM PROPER BUBBLE STUDY

- Inject agitated saline
- Look for bubbles crossing into LA within 3 beats
- Observe RA opacification
- Repeat with Valsalva

Positive = bubbles in LA within 3 cardiac cycles.

STEP 3: ASSESS HIGH-RISK ANATOMY

High-risk PFO features:

- Large shunt (many bubbles)
- Atrial septal aneurysm
- Long tunnel (>8–10 mm)
- Prominent Eustachian valve

These increase stroke risk.

Bubble Study — Step-by-Step Guide

1. Practical Echo Protocol

PREPARATION

- Use 10 ml saline
- Add 1 ml air
- Agitate between two syringes
- Inject rapidly via peripheral vein

IMAGING VIEW

Best views: apical 4-chamber, subcostal 4-chamber, TEE (if detailed assessment needed).

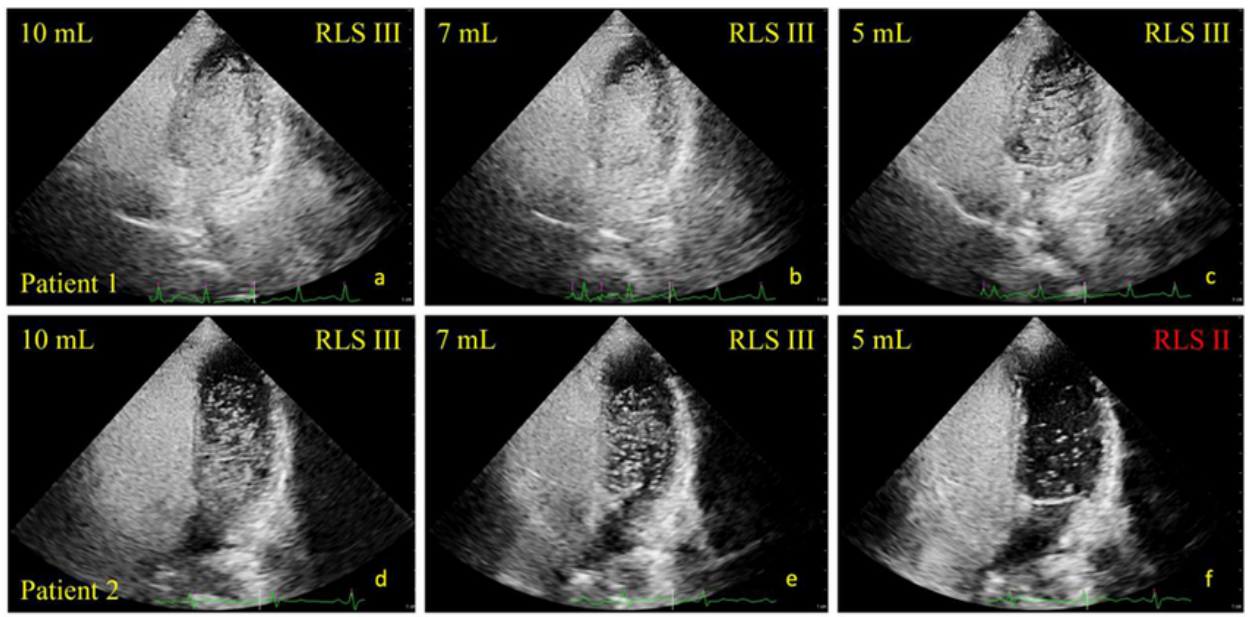
INTERPRETATION TIMELINE

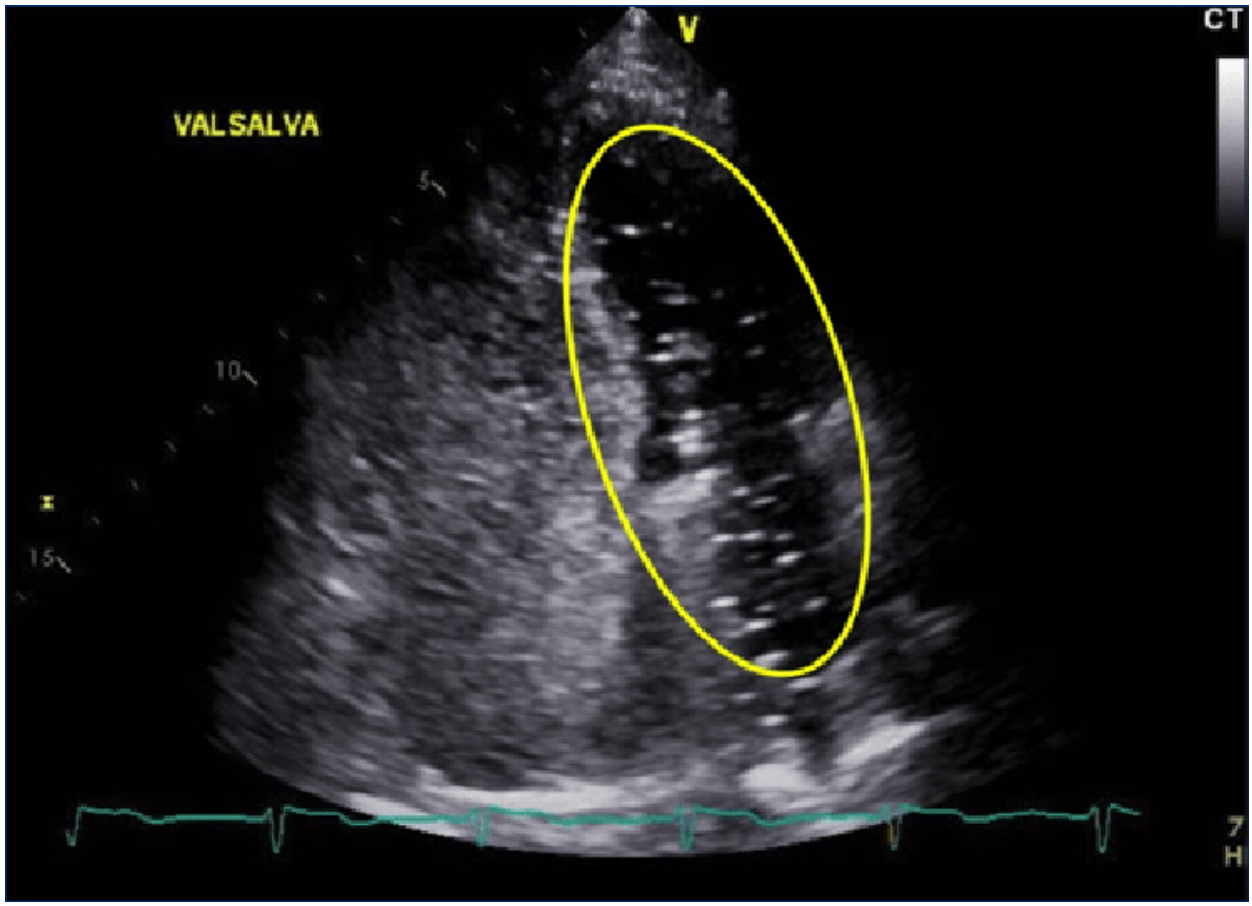
Timing of Bubbles in LA	Meaning
Within 3 beats	Intracardiac shunt (PFO/ASD)
After 3–5 beats	Pulmonary AV malformation

VALSALVA MANEUVER

Essential for PFO detection. Mechanism: increases RA pressure, opens flap, forces bubbles across.

If no Valsalva → false negative study.




V
CT
VALSALVA
10
15
7
H

Sinus Venosus ASD

1. The Commonly Missed ASD

ANATOMY

Located near: SVC–RA junction (most common), IVC–RA junction (less common).

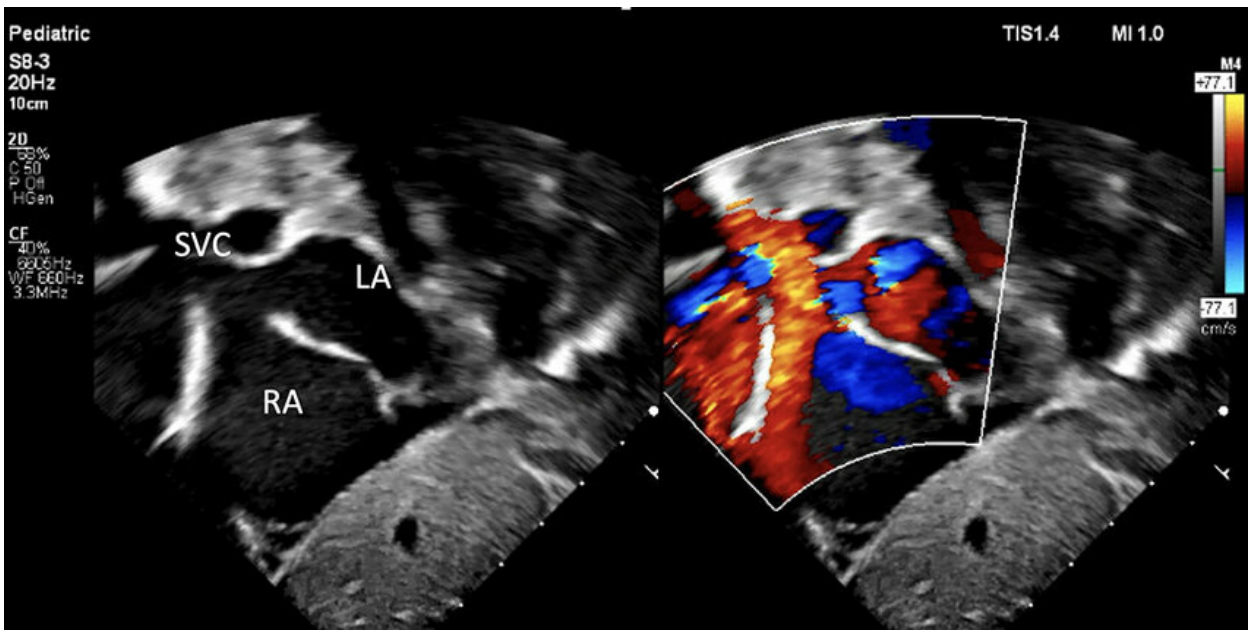
Almost always associated with: partial anomalous pulmonary venous return (PAPVR).

KEY FINDINGS

- High septal defect near SVC
- Difficult to see on routine apical views
- RV dilation
- Dilated SVC
- Missing right upper pulmonary vein entering LA

BEST VIEWS

Subcostal bicaval view, high parasternal view. TEE for confirmation.



DIAGNOSTIC CLUE

If RV dilated BUT no obvious secundum defect → search for sinus venosus defect.

ASSOCIATED LESION

Most common: right upper pulmonary vein drains into SVC.

If pulmonary veins not visualized → incomplete study.

WHY IMPORTANT?

Device closure NOT possible. Requires surgical correction.

ECHO CHECKLIST FOR SINUS VENOSUS ASD

- RV size assessed
- SVC region carefully examined
- Pulmonary veins traced
- PAPVR excluded

Atrial Septal Aneurysm (ASA)

Atrial Septal Aneurysm (ASA) is excessive mobility and bulging of the interatrial septum (usually septum primum) into either atrium.

It is: a structural abnormality, often associated with PFO, a risk modifier for embolic stroke.

DIAGNOSTIC CRITERIA (ECHO DEFINITION)

ASA is diagnosed when:

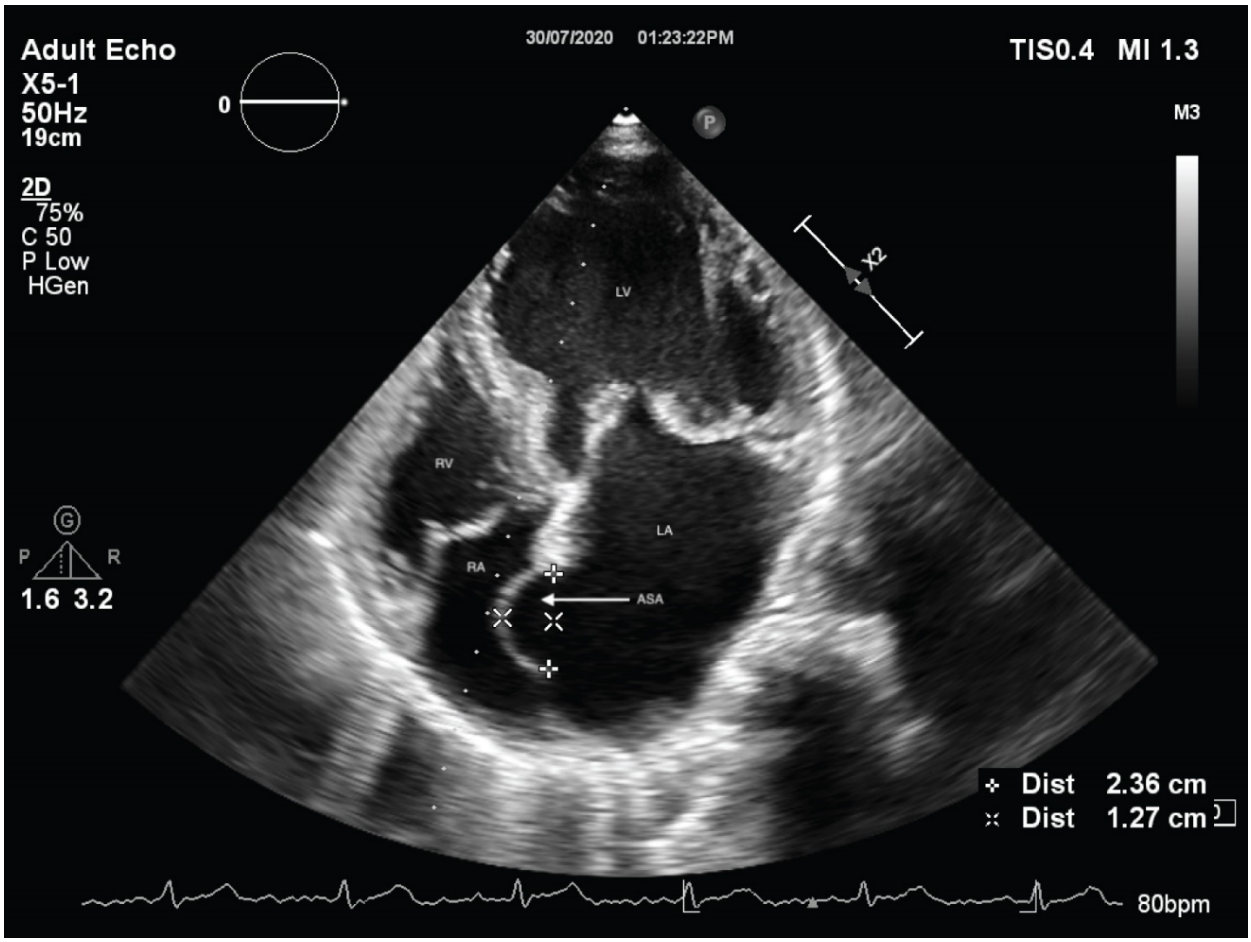
- Septal excursion ≥ 10–15 mm
- Excessive phasic movement during cardiac cycle
- Base width ≥ 15 mm

Bulging may occur: into RA, into LA, or bidirectionally.

TYPICAL ECHO FINDINGS

- Redundant floppy septum
- Oscillation with respiration
- May appear "balloon-like"
- Often seen better on TEE

Best view: subcostal (children), apical 4-chamber, TEE (gold standard).



PATHOPHYSIOLOGY

ASA itself usually does NOT cause: volume overload, pulmonary hypertension, RV enlargement.

It becomes clinically important when associated with: PFO, small ASD, thrombus formation.

1. Clinical Significance

STROKE RISK

ASA + PFO = significantly increased risk of: paradoxical embolism, cryptogenic stroke. Especially in young adults.

ARRHYTHMIAS

May be associated with: atrial ectopy, atrial fibrillation. Due to abnormal septal tissue.

BUBBLE STUDY IN ASA

Often reveals: right-to-left shunt, intermittent shunting, shunt increases with Valsalva.

WHEN TO INTERVENE?

ASA alone: no treatment required.

ASA + PFO + Stroke: consider PFO closure.

ASA + thrombus: anticoagulation.

2. Common Diagnostic Pitfalls

Mistaking ASA for ASD → look for continuous shunt and RV enlargement.

Overcalling mild septal mobility → must meet excursion criteria.

Missing associated PFO → always perform bubble study.

Ventricular Septal Defect (VSD)

Ventricular Septal Defect (VSD) is a communication between the right and left ventricles.

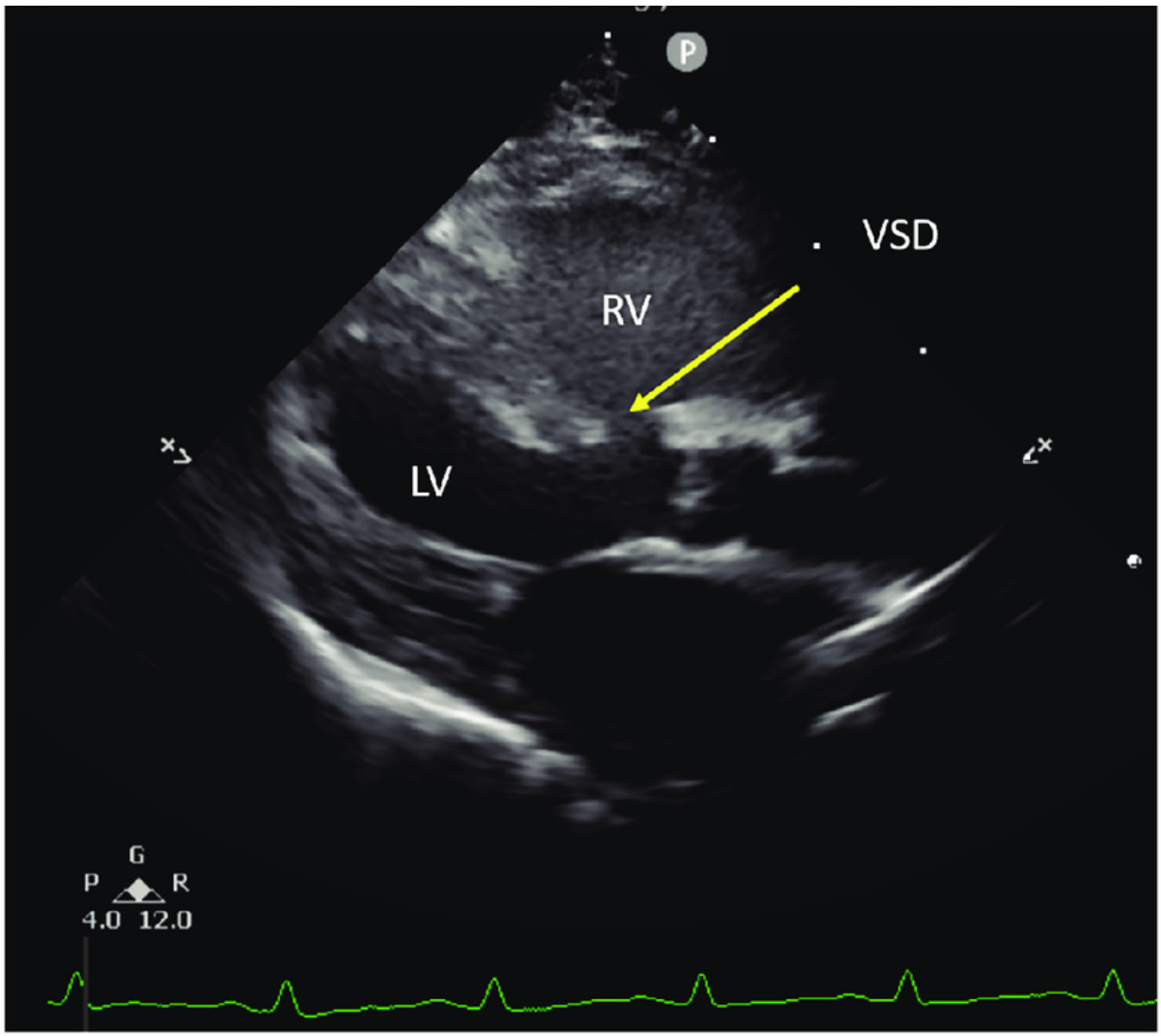
Most common congenital heart defect worldwide. Causes **left-to-right shunt** (initially).

1. Types of VSD

1. PERIMEMBRANOUS VSD (MOST COMMON ~70%)

Located beneath aortic valve, adjacent to tricuspid septal leaflet.

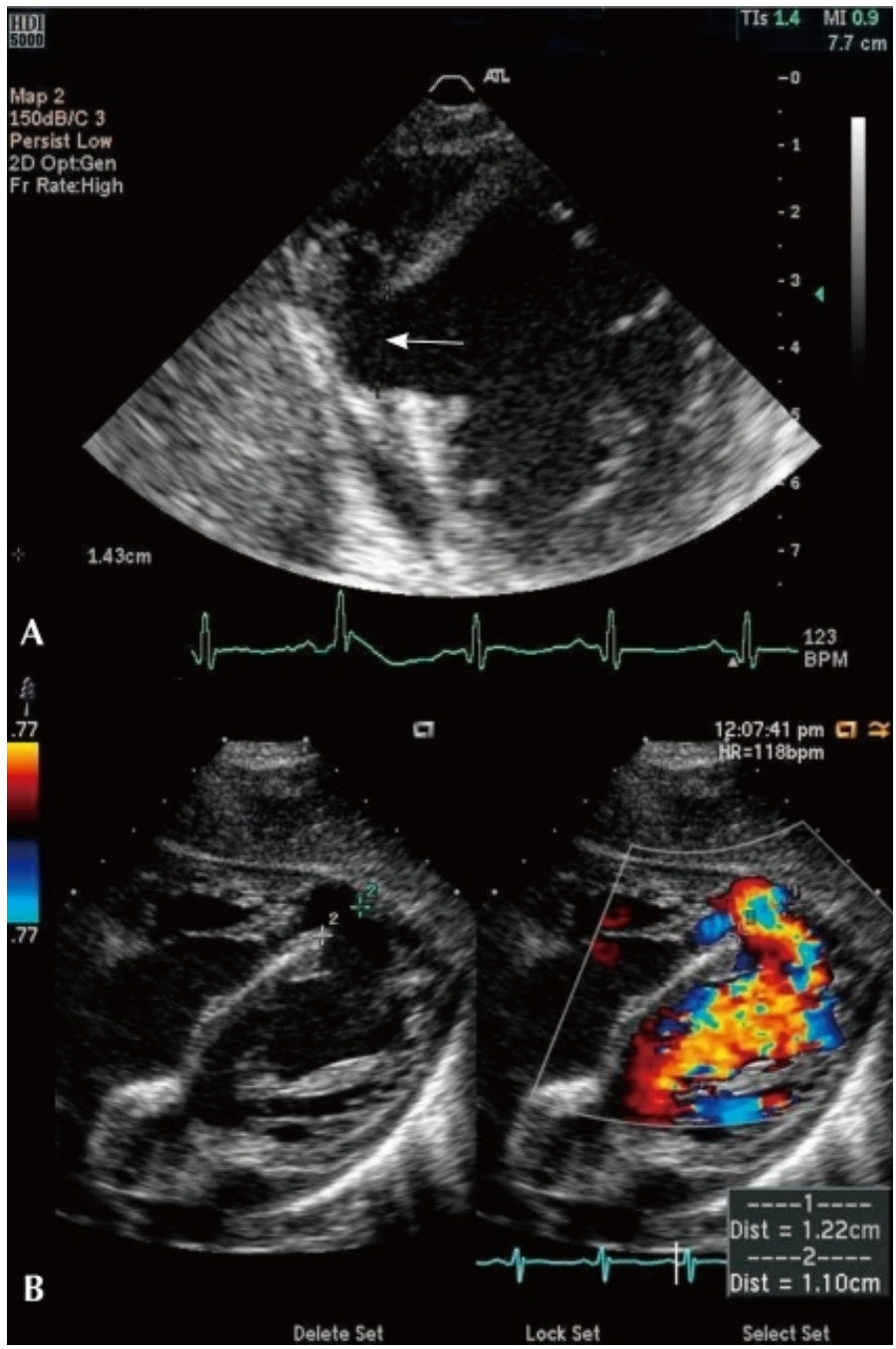
Echo clues: jet near aortic valve, risk of aortic cusp prolapse, may partially close by tricuspid tissue/aneurysm.



MUSCULAR VSD

Anywhere in muscular septum, apical or mid-septal.

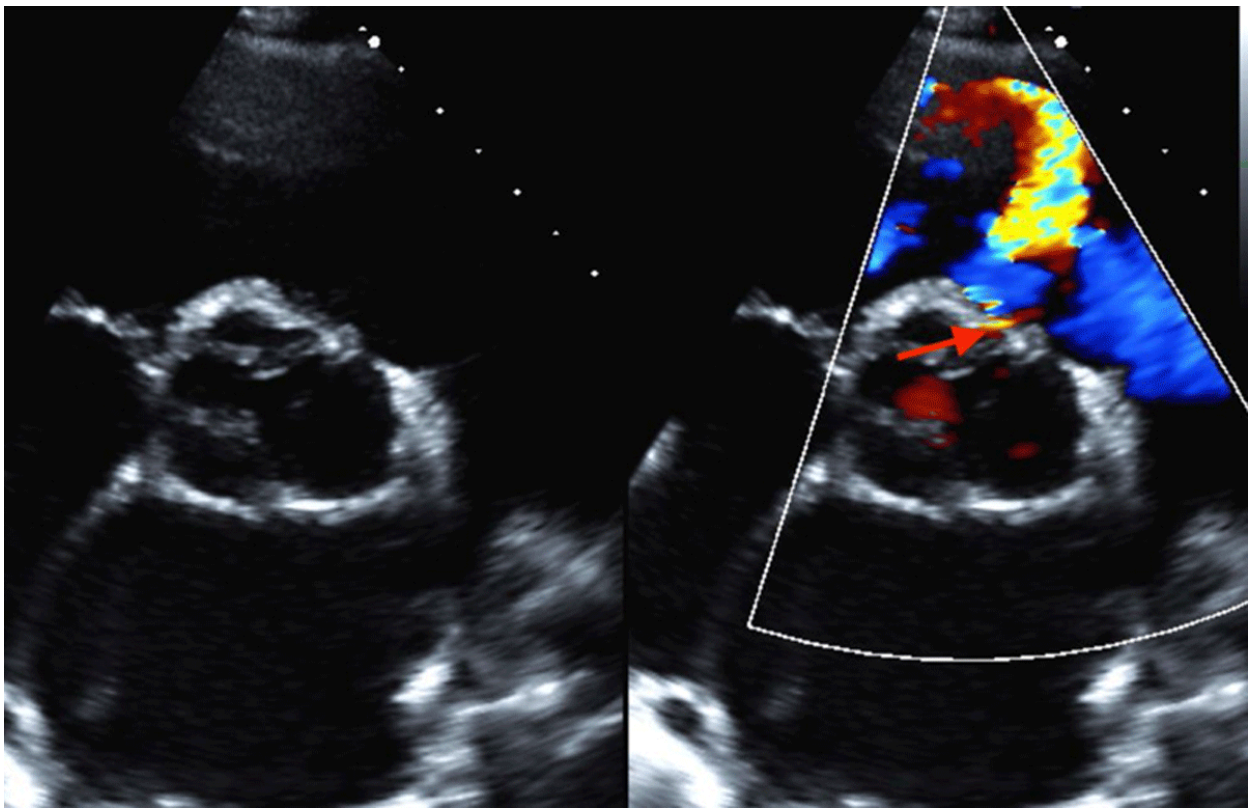
Echo clues: often small, may be multiple ("Swiss cheese"), high spontaneous closure rate.



3. OUTLET (SUPRACRISTAL / DOUBLY COMMITTED) VSD

Beneath pulmonary valve. Common in Asian population.

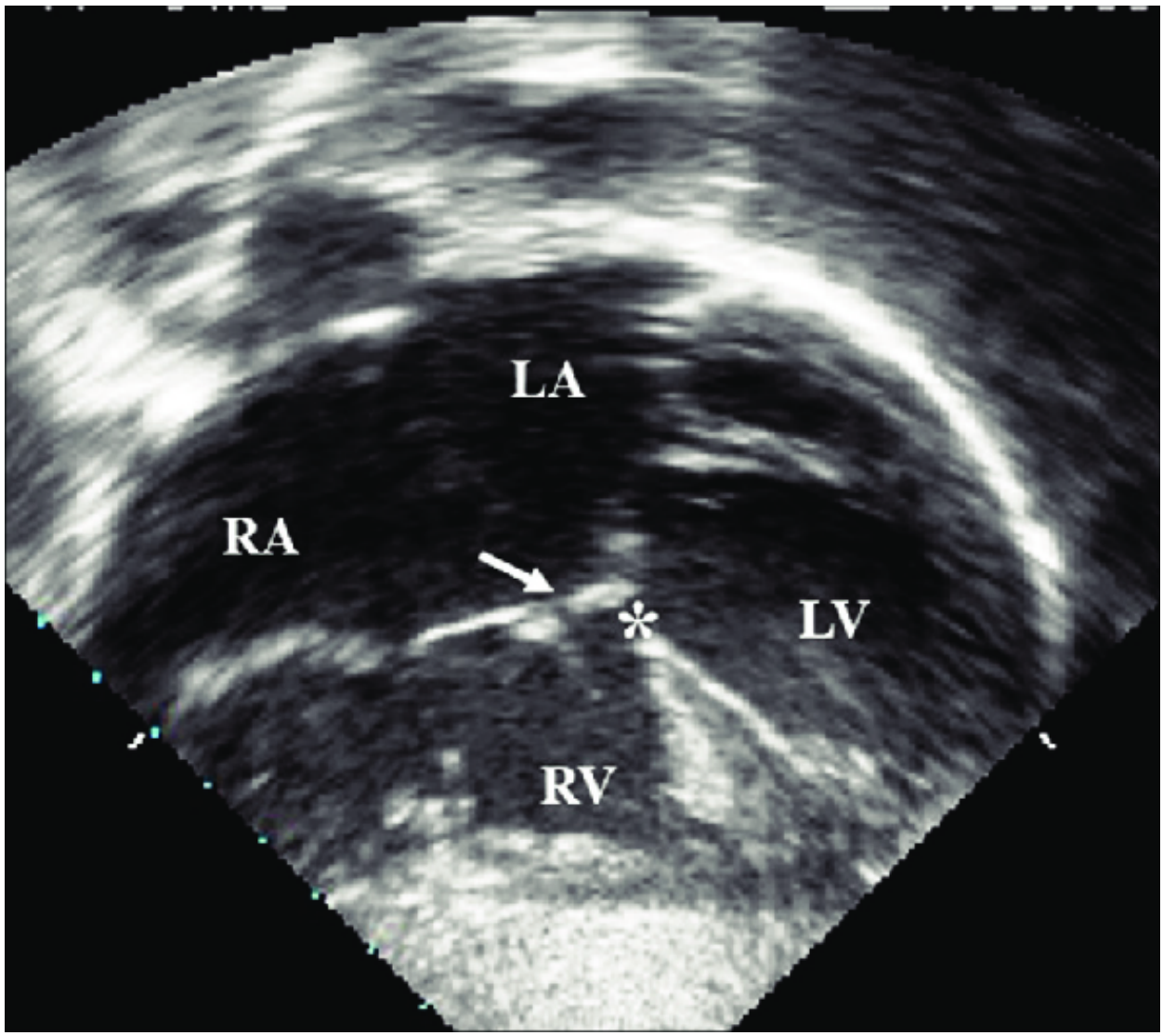
Echo clues: aortic cusp prolapse, aortic regurgitation risk.



4. INLET VSD

Posterior septum beneath TV leaflet. Part of AVSD spectrum.

Associated with: Down syndrome.



HEMODYNAMICS

Initially: LV pressure > RV pressure → left-to-right shunt.

Consequences: increased pulmonary blood flow, LA enlargement, LV volume overload, pulmonary hypertension (if large).

Late stage: right-to-left shunt → Eisenmenger.

VSD SIZE DETERMINES PHYSIOLOGY

Size	Hemodynamics	Clinical
Small (Restrictive)	High gradient	Loud murmur
Moderate	Increased flow	CHF signs
Large (Non-restrictive)	Equal LV & RV pressure	Early PH

Important Rule: smaller VSD = louder murmur; larger VSD = softer murmur.

2. Echo Assessment — Stepwise

STEP 1: IDENTIFY LOCATION

Best views: parasternal long axis, parasternal short axis, apical 4-chamber, subcostal view (best in children).

STEP 2: MEASURE SIZE

Compare VSD diameter to aortic annulus.

Small: < 1/3 aortic annulus. Moderate: 1/3–2/3. Large: > 2/3.

STEP 3: DOPPLER GRADIENT

Measure peak velocity across VSD.

Gradient = $4V^2$

High velocity (>4 m/s) → restrictive. Low velocity (<2 m/s) → large VSD.

STEP 4: ASSESS CHAMBER ENLARGEMENT

- LA size
- LV size
- RV pressure
- PA size

STEP 5: ASSESS AORTIC VALVE AND ANEURYSMAL TISSUE

- Aortic cusp prolapse
- Aortic regurgitation
- Aneurysmal tissue

COMPLICATIONS

- Pulmonary hypertension
- Eisenmenger syndrome
- Aortic regurgitation
- RVOT obstruction/DCRV
- Endocarditis
- Arrhythmias

WHEN TO CLOSE?

Indications: large VSD with CHF, failure to thrive, significant LV dilation, aortic cusp prolapse, pulmonary hypertension (before irreversible).

Best timing: 3–6 months (if large).

ECHO RED FLAGS

RV pressure systemic • Bidirectional shunt • D-shaped LV • Reduced RV function • Aortic valve prolapse

3. Diagnostic Pitfalls

- Missing small muscular VSD
- Underestimating PH
- Confusing TR jet with VSD jet (Gerbode defect)
- Not checking for multiple defects

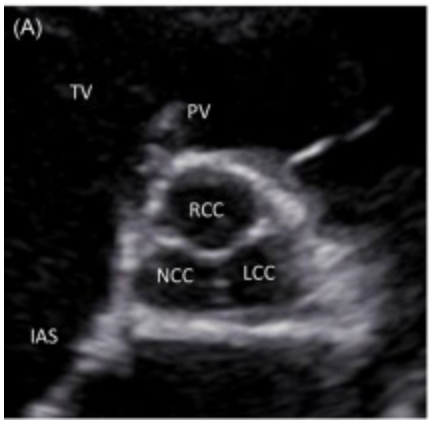
Valve Abnormalities in Children — Aortic Valve

NORMAL AORTIC VALVE ANATOMY

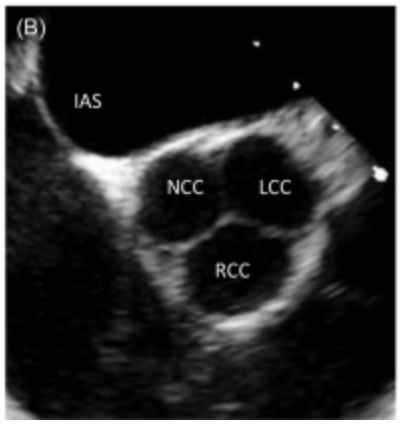
The normal aortic valve has three cusps: right coronary cusp (RCC), left coronary cusp (LCC), non-coronary cusp (NCC).

During systole, the valve opens in a triangular / Mercedes-Benz pattern.

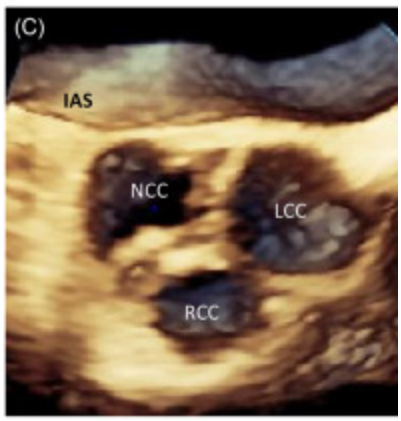
2D TTE. Parasternal short axis view of the aortic valve



2D TEE. Mid-esophageal 50° short axis view of the aortic valve



3D TEE. Short axis view of the aortic valve from ventricular aspect



Common Aortic Valve Abnormalities in Children

Lesion	Frequency
Bicuspid aortic valve	Most common
Congenital valvar aortic stenosis	
Unicuspid aortic valve	Rare
Quadricuspid aortic valve	Rare
Aortic regurgitation	Associated lesion
Subaortic membrane	Important differential

1. Bicuspid Aortic Valve (BAV)

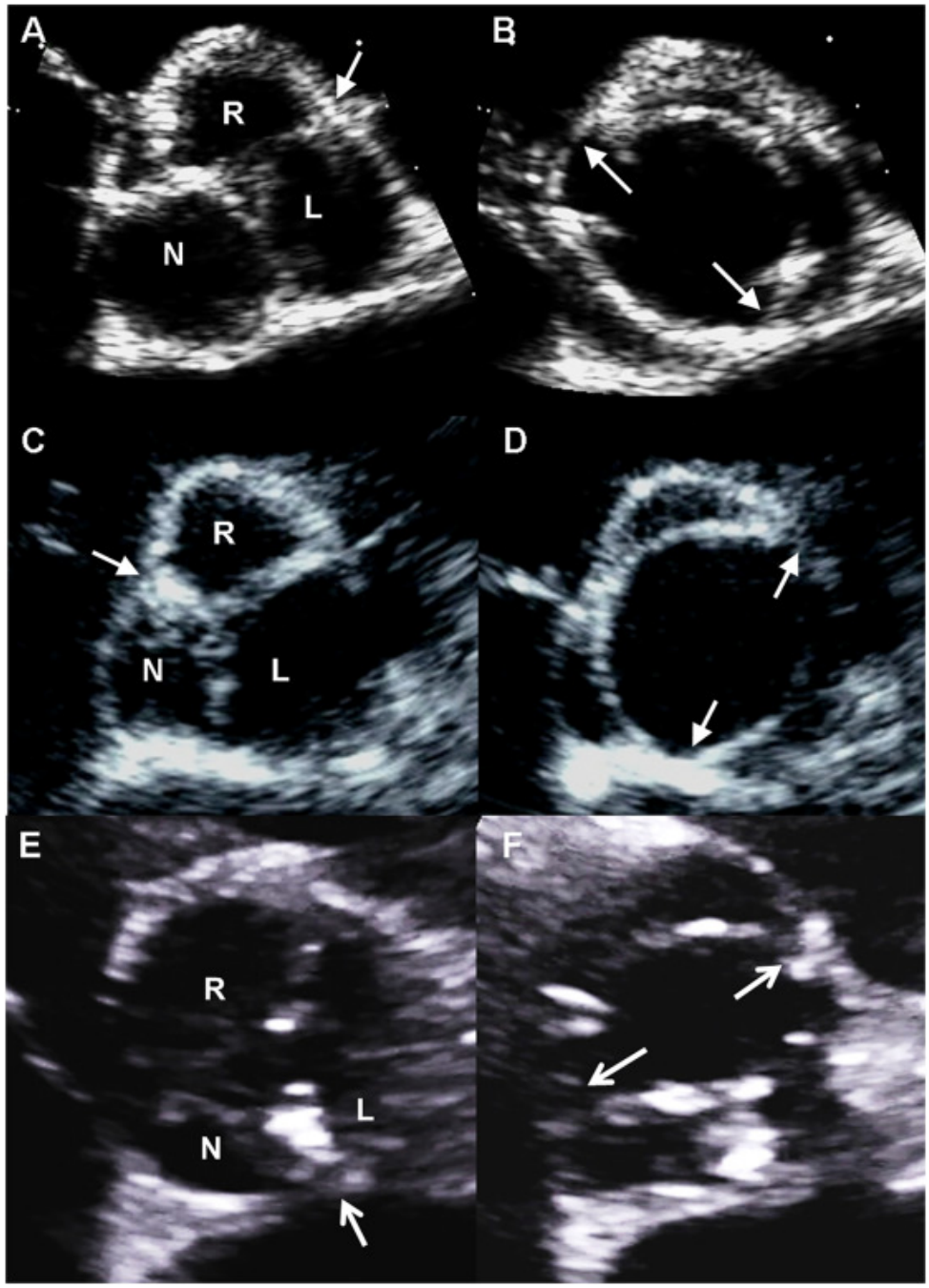
KEY ECHO FINDINGS

- Two cusps instead of three
- Fish-mouth opening in systole
- Raphe may be present
- Eccentric closure line in diastole

Best View — Parasternal short-axis at the aortic valve level.

Common Fusion Pattern

Fusion	Frequency
RCC + LCC	Most common
RCC + NCC	Less common



ASSOCIATED FINDINGS IN BAV

Always evaluate: coarctation of the aorta, ascending aortic dilatation, aortic root dilatation, aortic stenosis, aortic regurgitation.

IMPORTANT PEARL

Every child with BAV should have the entire aortic arch assessed.

2. Congenital Aortic Stenosis

MORPHOLOGY

- Thickened cusps
- Doming systolic opening
- Reduced excursion

DOPPLER ASSESSMENT

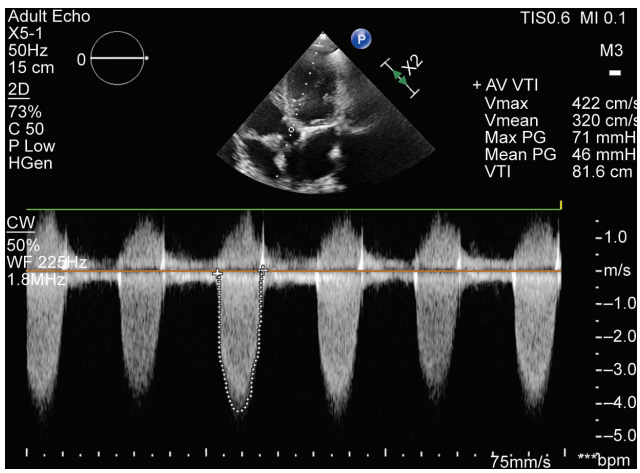
Modified Bernoulli equation. Use CW Doppler.

Gradient = $4V^2$

where V = peak aortic velocity

SEVERITY (PEDIATRIC APPROXIMATION)

Peak Gradient	Severity
< 25 mmHg (mean 20)	Mild
25–49 mmHg (mean 40)	Moderate
50–69 mmHg (mean 40)	Moderate-Severe
≥ 70 mmHg (mean 40)	Severe



3. Neonatal Critical Aortic Stenosis

Echo findings:

- Poor LV function
- Small antegrade aortic flow
- Thick dysplastic valve
- Mitral regurgitation
- Duct-dependent systemic circulation

CRITICAL AS — EMERGENCY

A low Doppler gradient does not exclude critical AS if the LV is failing.

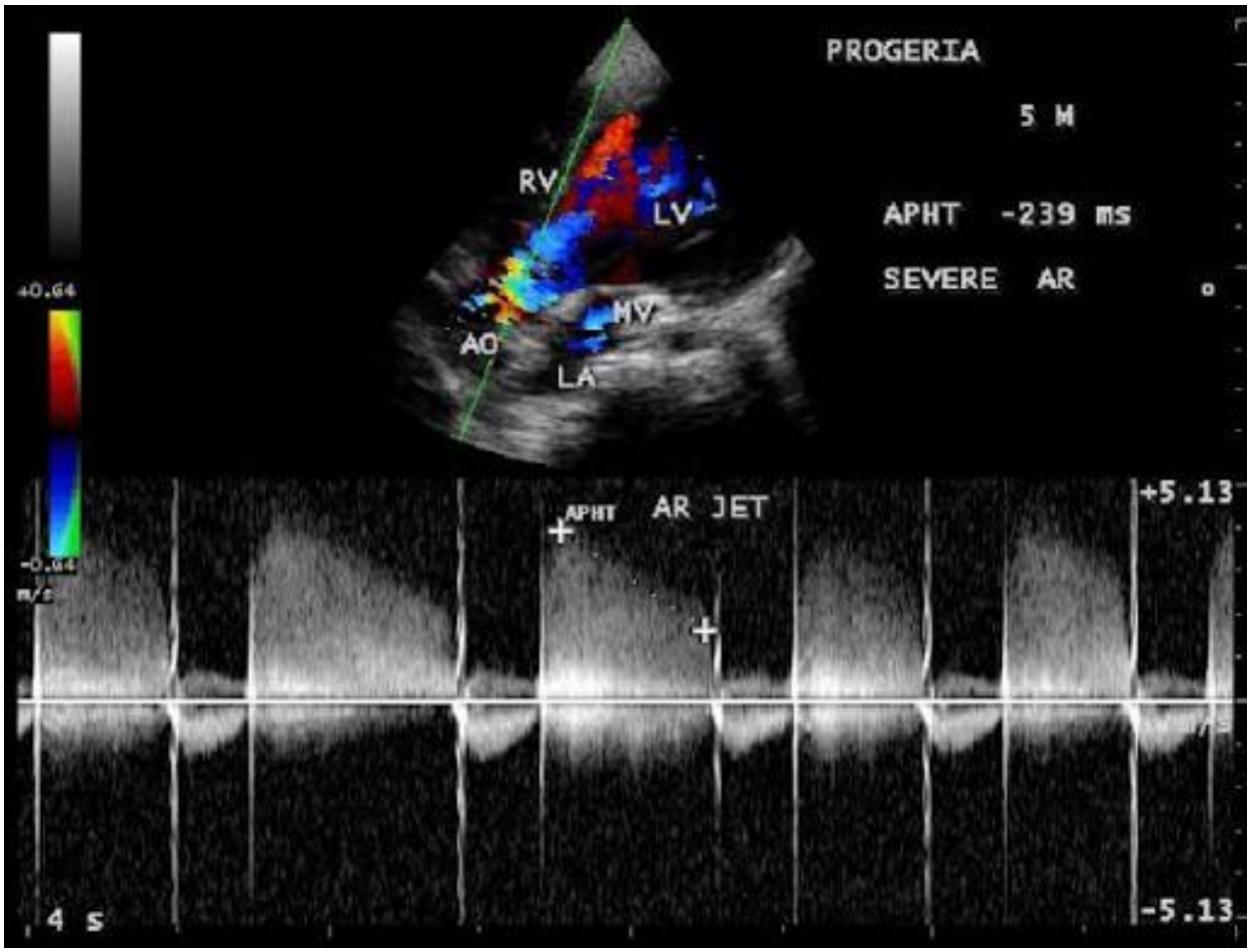
4. Aortic Regurgitation (AR)

COLOR DOPPLER

Assess: jet width, jet length, vena contracta, diastolic flow reversal.

ECHO SIGNS OF SIGNIFICANT AR

- LV dilatation
- Holodiastolic flow reversal in descending aorta
- Dense CW Doppler signal
- Wide vena contracta



UNICUSPID AORTIC VALVE

Usually presents with: severe AS, neonatal critical obstruction, marked valve thickening, single eccentric opening.

QUADRICUSPID AORTIC VALVE

Rare lesion. Echo clue: four cusps visible in short-axis view. Most commonly causes aortic regurgitation rather than stenosis.

5. Aortic Root and Ascending Aorta

Measure: aortic annulus, sinus of Valsalva, sinotubular junction, ascending aorta. Use Z-scores according to body surface area.

Important in: bicuspid aortic valve, connective tissue disorders, progressive aortopathy.

Condition	Key Echo Difference
Valvar AS	Jet begins at valve
Subaortic membrane	Membrane below valve
Supravalvar aortic stenosis	Narrowing above valve
Hypertrophic cardiomyopathy	Dynamic LVOT obstruction

COMMON ECHO PITFALLS

- Misdiagnosing BAV in diastole only
- Underestimating AS due to poor alignment
- Missing associated coarctation
- Ignoring ascending aortic measurements
- Grading AR by color jet area alone

6. Must Know

- Bicuspid aortic valve is the most common congenital valve abnormality.
- Parasternal short-axis is the best view for valve morphology.
- RCC-LCC fusion is the commonest BAV pattern.
- Always assess the aortic arch in BAV.
- Low gradient does not exclude critical neonatal AS.
- Holodiastolic flow reversal suggests significant AR.
- Subaortic membrane is an important mimic of valvar AS.

GOLDEN RULE

Every child with bicuspid aortic valve deserves a complete assessment of the aortic root, ascending aorta, and aortic arch.

Pulmonary Stenosis (PS) — Echocardiographic Findings

Pulmonary stenosis is obstruction to blood flow from the right ventricle to the pulmonary artery.

It may occur at three levels:

Level	Example
Subvalvar	Infundibular stenosis
Valvar	Most common
Supravalvar	MPA / branch PA stenosis

Valvar PS accounts for the vast majority of pediatric cases.

NORMAL PULMONARY VALVE ANATOMY

Normal valve has three thin cusps with wide systolic opening.

Best views: parasternal short-axis, parasternal RVOT view, subcostal sagittal view.

1. Valvar Pulmonary Stenosis — Morphology

KEY ECHO FINDINGS

- Thickened pulmonary valve cusps
- Doming systolic opening
- Reduced excursion
- Post-stenotic dilatation of the main PA

BEST DOPPLER VIEW

Use the view with the highest velocity: parasternal short-axis, RV inflow-outflow view, subcostal view, high parasternal view.

Modified Bernoulli equation, CW Doppler.

Gradient = $4V^2$

where V = peak pulmonary valve velocity

SEVERITY OF PULMONARY STENOSIS

Peak Gradient	Severity
< 36 mmHg	Mild
36–64 mmHg	Moderate
64 mmHg	Severe
80–100 mmHg	Critical / Very Severe

RIGHT VENTRICULAR CHANGES

- RV hypertrophy
- RV dilation (late)
- Septal flattening
- Right atrial enlargement
- Post-stenotic MPA dilatation

2. Critical Neonatal Pulmonary Stenosis

Echo findings:

- Dysplastic thick pulmonary valve
- Very small valve opening
- Hypoplastic RV
- Severe RV hypertrophy
- Right-to-left shunt across ASD/PFO
- Duct-dependent pulmonary circulation

IMPORTANT

Critical Neonatal PS: a low Doppler gradient may occur if RV output is severely reduced.

DYSPLASTIC PULMONARY VALVE

Common in Noonan syndrome.

Features: markedly thick cusps, minimal doming, poor mobility, often severe, balloon valvotomy may be less effective.

SUBVALVAR (INFUNDIBULAR) PULMONARY STENOSIS

Echo clues: narrowing below the valve, turbulence in RVOT, dynamic obstruction possible, common in TOF.

SUPRAVALVAR PULMONARY STENOSIS

Sites: main pulmonary artery, right pulmonary artery, left pulmonary artery.

Associated conditions: Williams syndrome, Alagille syndrome.

ASSOCIATED LESIONS

Always search for: ASD / PFO, Tetralogy of Fallot, Noonan syndrome, pulmonary valve dysplasia, branch PA stenosis.

Lesion	Echo Clue
Valvar PS	Doming valve
Infundibular PS	Narrowing below valve
Supravalvar PS	Narrowing above valve
Pulmonary atresia	No antegrade flow
Tetralogy of Fallot	VSD + overriding aorta

INDICATIONS FOR BALLOON PULMONARY VALVOTOMY

Generally indicated for: peak gradient ≥ 50–60 mmHg, symptoms, RV hypertrophy progression, cyanosis in critical PS.

Balloon valvotomy is the treatment of choice for typical valvar PS.

POST-VALVOTOMY ECHO

Assess: residual gradient, pulmonary regurgitation, RV size, RV function, branch PA flow.

3. Must Know

- Valvar PS is the commonest form of pulmonary stenosis.
- Doming pulmonary valve is the classic echo sign.
- Use CW Doppler and the highest velocity obtained.
- Peak gradient >64 mmHg = severe PS.
- Post-stenotic MPA dilatation supports valvar PS.
- Low gradient does not exclude critical neonatal PS.
- Noonan syndrome is strongly associated with dysplastic pulmonary valve.

GOLDEN RULE

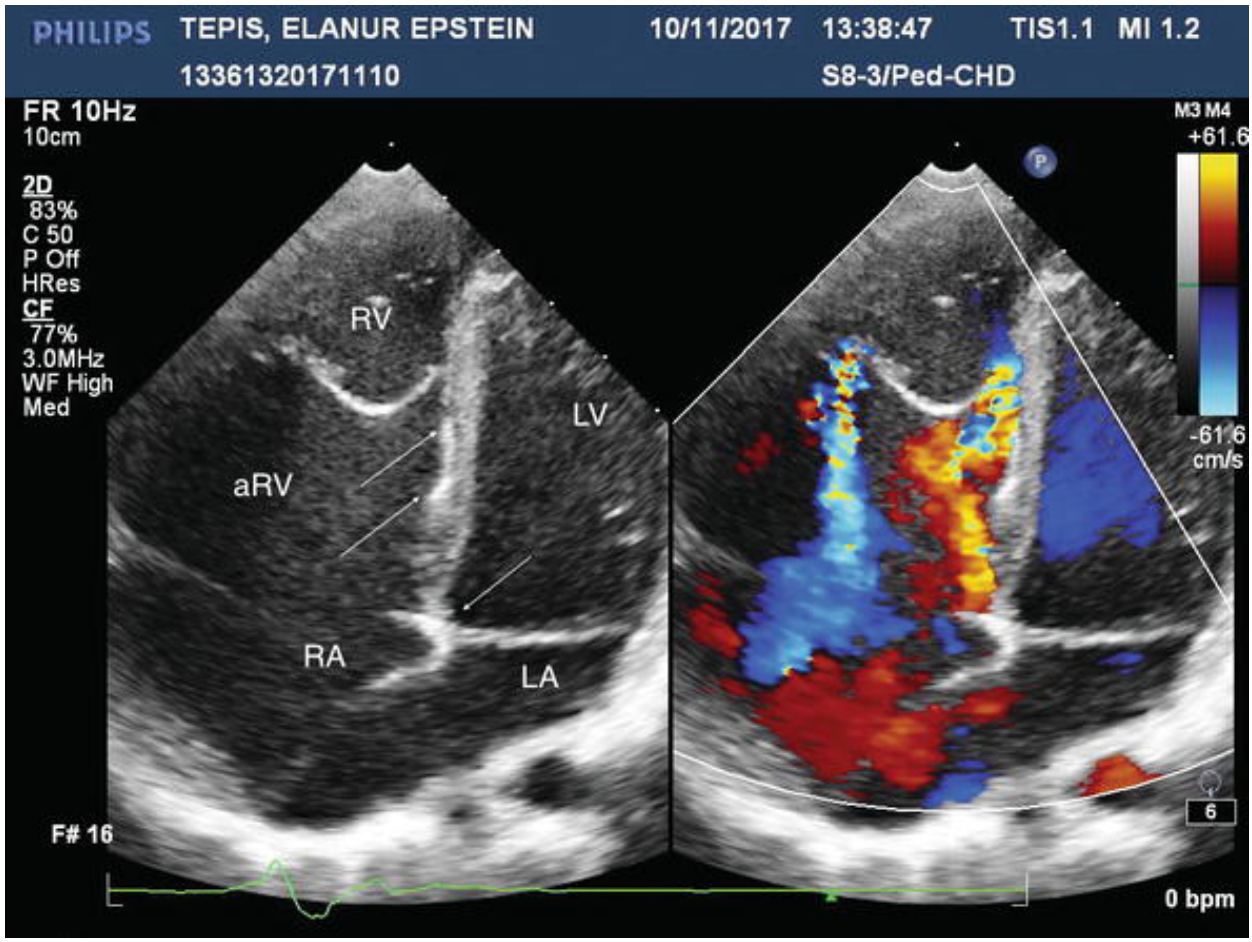
A doming pulmonary valve with post-stenotic pulmonary artery dilatation is valvar pulmonary stenosis until proven otherwise.

Tricuspid Valve Abnormalities — Ebstein Anomaly

Ebstein anomaly is a congenital malformation characterized by apical displacement of the septal and posterior tricuspid valve leaflets into the right ventricle.

This creates:

- A massively enlarged right atrium
- An "atrialized" portion of the right ventricle
- A small functional RV
- Significant tricuspid regurgitation



CLINICAL PRESENTATION

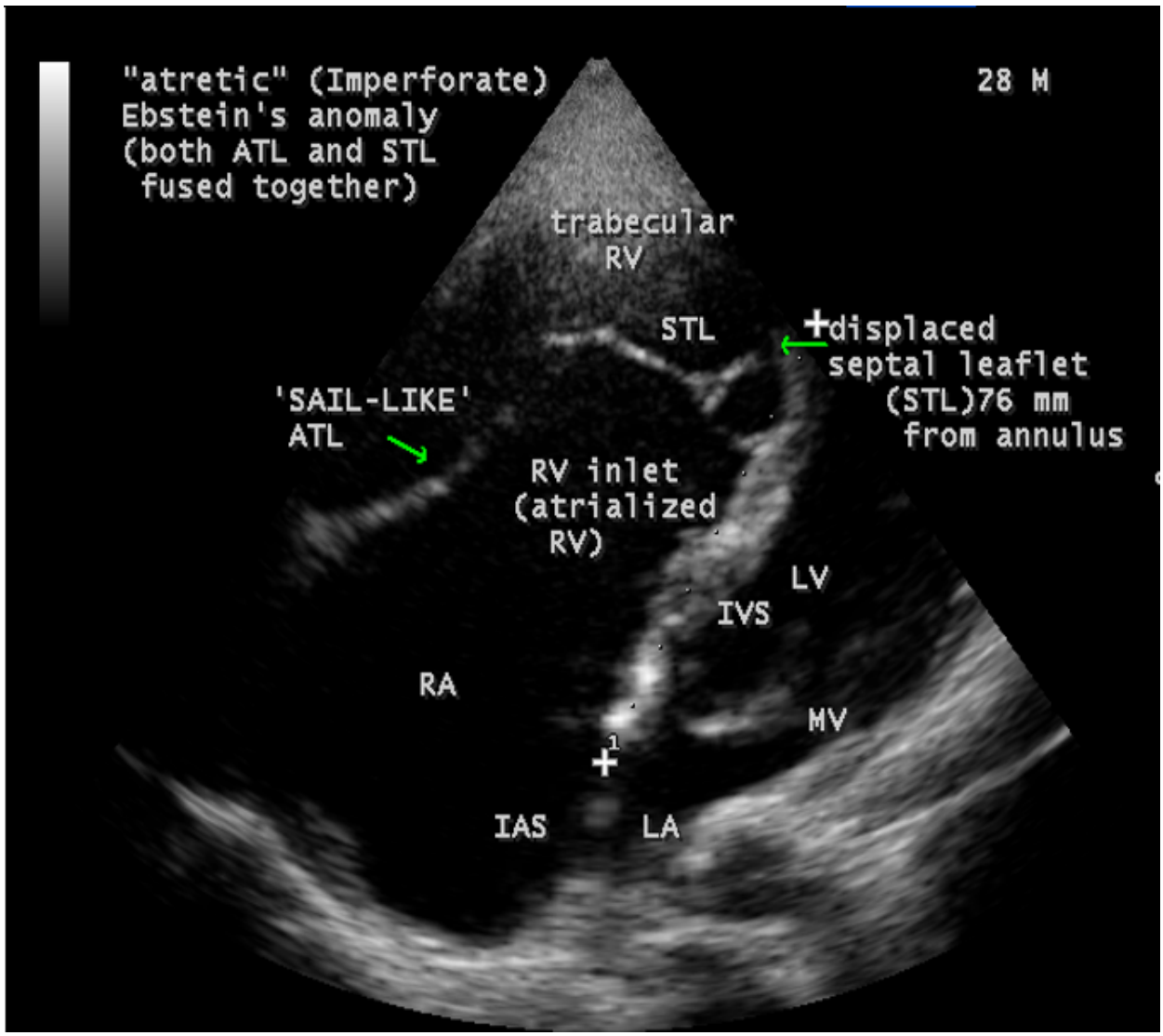
Age	Presentation
Neonate	Severe cyanosis, heart failure
Infant	Tachypnea, poor feeding
Child	Murmur, exercise intolerance
Adolescent	Palpitations, arrhythmias

Severity varies widely.

1. Key Echocardiographic Views

APICAL 4-CHAMBER — MOST IMPORTANT

Look for: apical displacement of septal leaflet, large right atrium, atrialized RV, small functional RV, TR jet.



DIAGNOSTIC MEASUREMENT — APICAL DISPLACEMENT INDEX

Measure the distance between: mitral valve septal insertion, tricuspid valve septal insertion.

Diagnostic criterion: ≥ 8 mm/m² body surface area.

This is the classic echocardiographic criterion.

HALLMARK ECHO FINDINGS

Finding	Importance
Apical septal leaflet displacement	Diagnostic
Massive RA enlargement	Very common
Atrialized RV	Characteristic
Small functional RV	Severity marker
Severe TR	Hemodynamic impact
ASD/PFO	Causes cyanosis

TRICUSPID REGURGITATION ASSESSMENT

Evaluate: jet area, vena contracta, hepatic vein flow reversal, CW Doppler density, RA enlargement.

ASSOCIATED LESIONS

Frequently associated with: Atrial Septal Defect, Patent Foramen Ovale, Wolff–Parkinson–White syndrome, pulmonary stenosis, right ventricular outflow tract obstruction.

IMPORTANT PEARL

Up to 20–30% may have accessory pathways.

2. Neonatal Ebstein Anomaly

Neonates may present with: severe cyanosis, functional pulmonary atresia, duct-dependent pulmonary circulation, massive cardiomegaly.

Echo clues: huge RA, tiny functional RV, severe TR, minimal antegrade pulmonary flow, right-to-left atrial shunt.

CELERMAJER INDEX (SEVERITY ASSESSMENT)

Ratio of: (RA + atrialized RV area) ÷ (functional RV + LV + LA area)

Index	Severity
< 0.5	Mild
0.5–0.99	Moderate
1.0–1.49	Severe
≥ 1.5	Very severe

Higher index = worse prognosis.

DIFFERENTIAL DIAGNOSIS

Condition	Key Difference
Ebstein anomaly	Apical displacement present
Tricuspid valve dysplasia	No significant displacement
Uhl anomaly	Thin RV wall, no displaced valve
Severe tricuspid regurgitation	Normal valve insertion
Pulmonary atresia with intact septum	Hypoplastic RV without displaced TV

COMMON ECHO PITFALLS

- Measuring displacement in a non-apical view
- Confusing severe TR with Ebstein anomaly
- Ignoring body surface area indexing
- Missing atrial-level shunt
- Underestimating functional RV size

MANAGEMENT IMPLICATIONS

Medical — diuretics, antiarrhythmics, prostaglandin (if duct-dependent).

Surgical — cone reconstruction, tricuspid valve repair, valve replacement, one-and-a-half ventricle repair, Fontan pathway (severe cases).

3. Pearls

- The apical 4-chamber view is the key diagnostic view.
- Diagnostic criterion: ≥8 mm/m² apical displacement.
- Massive RA enlargement is the most striking feature.
- Severe TR is common.
- ASD/PFO frequently causes cyanosis.
- WPW syndrome is an important association.
- Neonatal severe Ebstein anomaly may behave like pulmonary atresia.

ECHO CHECKLIST BEFORE ENDING STUDY

- Measure tricuspid displacement
- Index to BSA
- Assess RA size
- Identify atrialized RV
- Measure functional RV
- Grade TR
- Search for ASD/PFO
- Assess pulmonary flow
- Evaluate RV function

Mitral Valve Abnormalities — Mitral Valve Prolapse

Mitral Valve Prolapse (MVP) is the **systolic displacement of one or both mitral valve leaflets into the left atrium** during ventricular systole.

It is the **most common valvular abnormality** in children and adults.

MVP may occur as: isolated abnormality, familial disorder, connective tissue disease, associated congenital heart disease.

The mitral valve consists of: anterior leaflet, posterior leaflet, mitral annulus, chordae tendineae, papillary muscles.

Normal function requires all five components to work together.

ETIOLOGY

Primary MVP — degenerative valve disease, myxomatous degeneration, familial MVP.

Secondary MVP — Marfan syndrome, Loeys-Dietz syndrome, Ehlers-Danlos syndrome, hypertrophic cardiomyopathy, congenital mitral valve abnormalities, rheumatic heart disease (rare cause of prolapse).

ECHO DEFINITION

The diagnosis should **only be made in the Parasternal Long-Axis (PLAX) view**.

Diagnostic criterion: mitral leaflet displacement ≥2 mm beyond the mitral annular plane during systole.

CLINICAL PEARL

Never diagnose MVP using only the apical four-chamber view because the saddle-shaped mitral annulus can create a false impression of prolapse.

1. Echo Views

PARASTERNAL LONG AXIS (MOST IMPORTANT)

Confirms prolapse, measures leaflet displacement, detects leaflet thickening.

APICAL FOUR CHAMBER

Useful for: mitral regurgitation, leaflet motion, LV size. Do not diagnose MVP from this view alone.

PARASTERNAL SHORT AXIS

Assesses: commissures, leaflet morphology, chordal rupture.

APICAL LONG AXIS (3 CHAMBER)

Excellent for: posterior leaflet prolapse, MR jet direction.

CLASSIC VS NON-CLASSIC MVP

Feature	Classic MVP	Non-Classic MVP
Leaflet thickness	≥5 mm	<5 mm
Leaflet prolapse	≥2 mm	≥2 mm
Myxomatous degeneration	Present	Minimal
MR risk	Higher	Lower
Surgical risk	Higher	Lower

Classic MVP carries a greater risk of progressive mitral regurgitation.

ECHO FEATURES

- Leaflet displacement into LA
- Thickened leaflets
- Redundant leaflet tissue
- Elongated chordae
- Mitral annular dilatation
- Mitral regurgitation

COLOR DOPPLER ASSESSMENT

Evaluate: origin of MR, direction of jet, severity, pulmonary vein flow.

Jet direction helps identify the prolapsing leaflet.

Prolapsing Leaflet	MR Jet Direction
Posterior leaflet	Anterior jet
Anterior leaflet	Posterior jet
Bileaflet	Central or eccentric

QUANTIFYING MITRAL REGURGITATION

Assess: vena contracta, PISA (if feasible), pulmonary vein flow, left atrial size, LV size, continuous-wave Doppler profile.

Never grade MR based on color jet area alone.

ASSOCIATED FINDINGS

MVP may be associated with: mitral annular disjunction, chordal elongation, chordal rupture, flail leaflet, left atrial enlargement, left ventricular enlargement.

COMPLICATIONS

- Progressive mitral regurgitation
- Infective endocarditis (uncommon)
- Chordal rupture
- Flail mitral leaflet
- Atrial fibrillation
- Heart failure
- Ventricular arrhythmias (rare)
- Sudden cardiac death (very rare, high-risk subgroup)

PEDIATRIC CONSIDERATIONS

Most children: mild prolapse, minimal MR, excellent prognosis.

Surgery is rarely required during childhood unless severe MR develops.

DIFFERENTIAL DIAGNOSIS

Differentiate MVP from: mitral valve billowing, flail leaflet, rheumatic mitral disease, mitral cleft, redundant chordae, hypertrophic cardiomyopathy.

SURGICAL INDICATIONS

Consider surgery if: severe mitral regurgitation, symptoms, progressive LV dilatation, declining LV systolic function, new atrial fibrillation, pulmonary hypertension.

Mitral valve repair is preferred over replacement whenever feasible.

2. Clinical Pearls

REMEMBER THE "2–5 RULE"

- ≥2 mm systolic displacement = Mitral valve prolapse
- ≥5 mm leaflet thickness = Classic MVP

JET RULE

- Posterior leaflet prolapse → Anterior MR jet
- Anterior leaflet prolapse → Posterior MR jet

GOLDEN RULE

Never diagnose mitral valve prolapse from the apical four-chamber view alone. The parasternal long-axis view is the diagnostic standard.

MITRAL VALVE PROLAPSE (MVP)

Definition: Displacement of one or both mitral leaflets into the left atrium during systole **≥ 2 mm** above the mitral annular plane.

1. EPIDEMIOLOGY

- Most common valvular abnormality in young adults.
- Prevalence: 1-3% in general population.
- More common in females.

2. PATHOPHYSIOLOGY

- Results from myxomatous degeneration of the valve.
- Leaflet thickening, redundancy, and elongation of chordae.
- Leads to leaflet billowing/prolapse.
- May cause mitral regurgitation (MR) of variable severity.

3. ETIOLOGY

- Myxomatous (degenerative) – most common.
- Familial / connective tissue disorders (e.g., Marfan, Ehlers-Danlos).
- Secondary to ischemia, infection, or other conditions (rare).

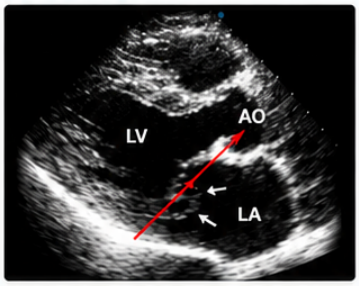
4. ECHOCARDIOGRAPHIC DIAGNOSIS

Diagnostic Criteria:

- Systolic displacement of mitral leaflets(≥ 2 mm above the annular plane
- Seen in parasternal long-axis, apical 2-chamber, or apical 4-chamber views

Best Views:

- Parasternal long-axis (PLAX)
- Apical 2-chamber
- Apical 4-chamber
- Apical 3-chamber



Displacement of posterior mitral leaflet ≥ 2 mm above the annular plane (fred line) into the LA during systole.

5. CLASSIFICATION (Based on Leaflet Thickness)

Non-classic MVP	Leaflet thickness ≤ 5 mm
Classic MVP	Leaflet thickness ≥ 5 mm

(Assess in parasternal long-axis view in mid-systole)

6. ASSOCIATED FINDINGS

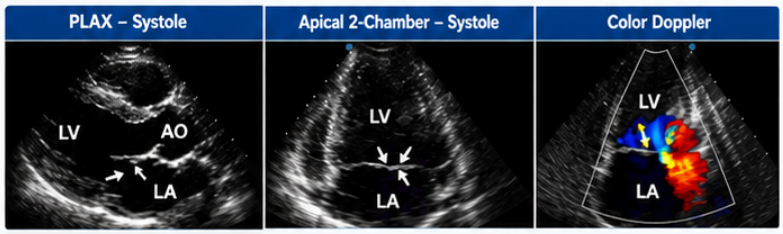
- Mitral regurgitation (mild to severe).
- Left atrial enlargement (if significant MR).
- Chordae tendineae elongation and thickening.
- Aortic root dilation (occasionally).
- Arrhythmias (e.g., palpitations).

7. CLINICAL FEATURES

- Often asymptomatic.
- May present with palpitations, atypical chest pain, fatigue.
- Murmur: late systolic click + late systolic murmur.
- In classic MVP: risk of arrhythmias and rare complications (e.g., infective endocarditis, stroke).

8. ECHOCARDIOGRAPHIC FEATURES

- Leaflet thickening and redundancy.
- Leaflet prolapse (≥ 2 mm above annular plane).
- Mild systolic "bowing" into LA.
- Mitral regurgitation (jet direction posteriorly or centrally).
- May be "bileaflet" or single-leaflet prolapse.



9. ASSESSMENT OF MITRAL REGURGITATION

Severity	• Eccentric, posteriorly-directed or central. • Assess using standard MR criteria: – Jet area, vena contracta, PISA, EROA, regurgitant volume – Pulmonary vein flow (systolic flow blunting in significant MR)
Complications	Left atrial enlargement, LV dilation, arrhythmias.

10. DIFFERENTIAL DIAGNOSIS

- Rheumatic mitral leaflets without prolapse.
- Rheumatic mitral valve disease.
- Endocarditis.
- Chordal rupture (flail leaflet).
- Normal variant (mild leaflet redundancy).

11. MANAGEMENT

- Asymptomatic, no significant MR: Reassurance, periodic follow-up.
- Symptomatic or significant MR: Medications, monitoring, or surgery (if severe MR or complications).
- Endocarditis prophylaxis: Not routinely recommended (except high-risk patients).

12. KEY POINTS

- ✓ MVP is defined as systolic displacement ≥ 2 mm above annular plane.
- ✓ Assess leaflet thickness: ≤ 5 mm = non-classic, ≥ 5 mm = classic MVP.
- ✓ Commonly associated with mitral regurgitation.
- ✓ Most patients have a benign course.
- ✓ Regular follow-up with echocardiography is important.

RED FLAGS | Severe MR | LV dysfunction | Atrial fibrillation | Recurrent embolic events | Symptoms (dyspnea, palpitations)

Echo Pearls for Identifying Mitral Valve Scallops (A1–A3 & P1–P3)

Localization of mitral valve scallops is essential for **mitral valve repair, transcatheter edge-to-edge repair (MitraClip), and surgical planning**. The key is to recognize which echo view displays which portion of the valve.

ALWAYS IMAGINE THE MITRAL VALVE FROM THE LEFT ATRIUM

The mitral valve is divided into **six scallops**.

ANTERIOR LEAFLET: A1 A2 A3 | POSTERIOR LEAFLET: P1 P2 P3

- A1/P1 = Anterolateral commissure
- A2/P2 = Middle (central)
- A3/P3 = Posteromedial commissure

Memory Rule: numbers increase from lateral → medial.

PARASTERNAL LONG-AXIS (PLAX) ALWAYS SHOWS THE MIDDLE

PLAX is the most important view for MVP.

Usually visualizes: A2, P2.

MVP seen in PLAX usually involves A2 or P2. PLAX = Middle scallops (A2–P2).

APICAL FOUR-CHAMBER SWEEPS FROM LATERAL TO MEDIAL

As the probe is swept: left side of screen → A1/P1; middle → A2/P2; right side → A3/P3.

During an apical sweep, the scallop number increases from 1 to 3.

APICAL LONG-AXIS (THREE-CHAMBER)

This view again primarily shows A2, P2.

It is excellent for: posterior leaflet prolapse, MR jet direction, chordal rupture.

POSTERIOR LEAFLET PROLAPSE PRODUCES AN ANTERIOR JET

This is one of the most reliable localization clues.

Prolapsing Leaflet	MR Jet Direction
P1/P2/P3	Anterior
A1/A2/A3	Posterior
Bileaflet	Central or multiple jets

Remember: the MR jet points away from the prolapsing leaflet.

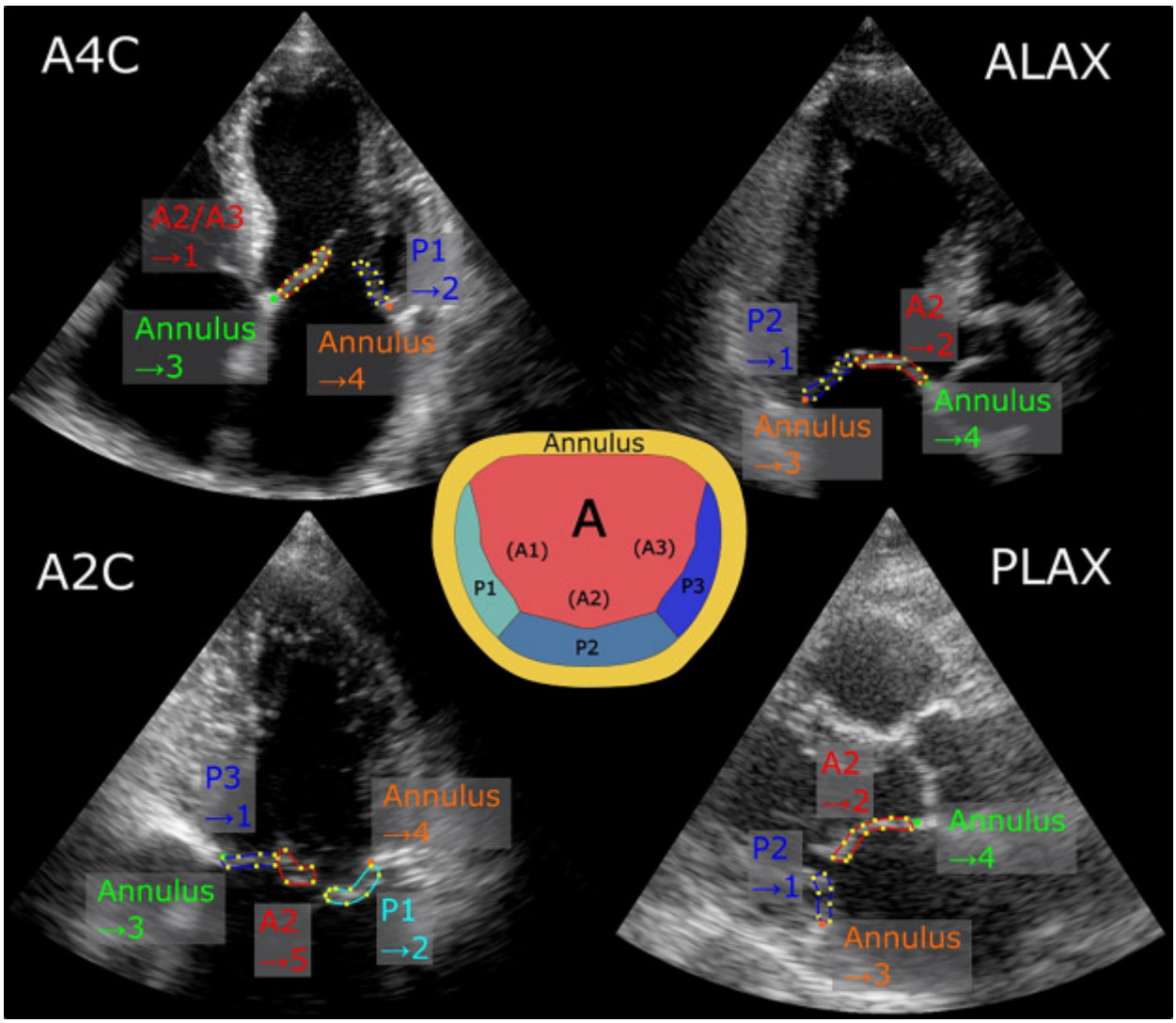
THE MIDDLE SCALLOPS ARE THE MOST COMMON SITE

Approximately 70–80% of degenerative MVP involves: P2.

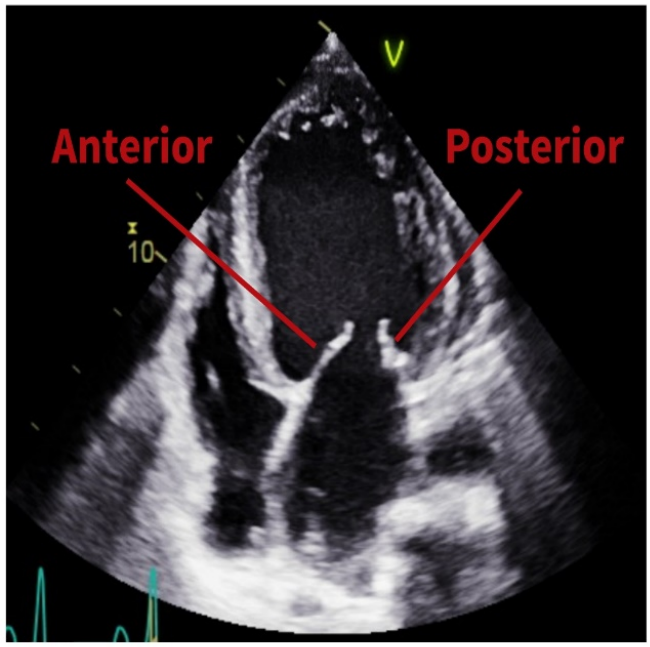
If you see isolated prolapse, think P2 first.

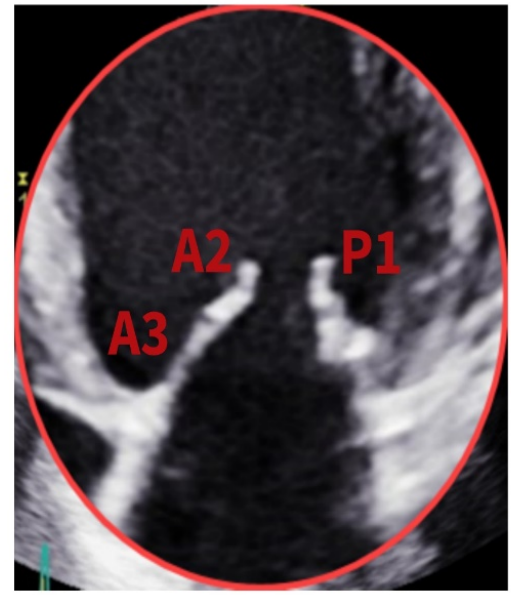
WHICH VIEW BEST SHOWS EACH SCALLOP?

Scallop	Best Echo View
A1	Apical 4-chamber (lateral sweep), TEE commissural view
A2	PLAX, Apical Long-Axis (3-chamber), TEE Long-Axis
A3	Apical 4-chamber (medial sweep), TEE commissural view
P1	Apical 4-chamber (lateral), TEE commissural view
P2	PLAX, Apical Long-Axis, TEE Long-Axis
P3	Apical 4-chamber (medial), TEE commissural view



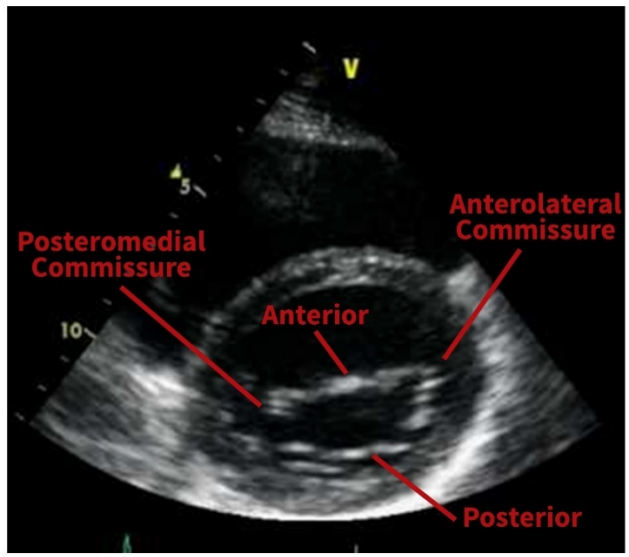
Apical 4 Chamber

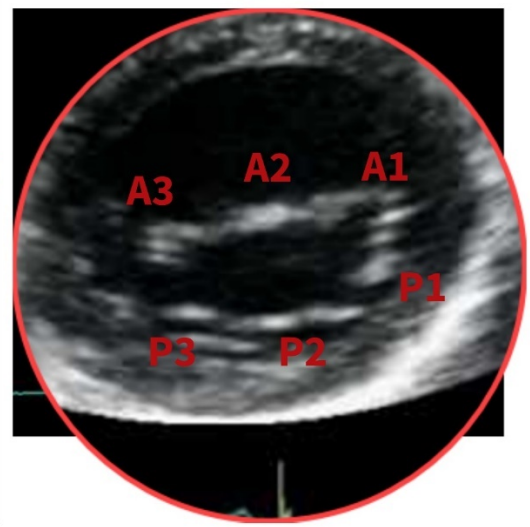




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Parasternal Long Axis





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SURGICAL MEMORY TRICK

Imagine the mitral valve as a clock viewed from the left atrium.

Lateral → Medial: A1 → A2 → A3, P1 → P2 → P3.

Moving toward the interatrial septum means moving toward A3/P3.

REMEMBER THE COMMISSURES

Anterolateral commissure — between A1 and P1.

Posteromedial commissure — between A3 and P3.

If prolapse is near a commissure, specify: anterolateral commissural prolapse, posteromedial commissural prolapse.

- PLAX = A2–P2 (the central scallops).
- P2 is the most common site of degenerative mitral valve prolapse.
- Posterior leaflet prolapse → anterior MR jet.
- Anterior leaflet prolapse → posterior MR jet.
- TEE is the gold standard for precise scallop localization before mitral valve repair.
- Always sweep the valve from lateral to medial to avoid missing isolated A1/P1 or A3/P3 lesions.

GOLDEN RULE

Think "Middle in PLAX, Lateral-to-Medial in Apical Sweep, All Six in TEE Commissural View."

MODULE 5

Miscellaneous

Topics
Pulmonary Hypertension in CHD
Pericardial Effusion
Myocarditis
Myocardial Stunning
Takotsubo Cardiomyopathy
IVC Collapsibility and Distensibility
Coronary Artery Abnormalities
Coronary Artery Fistula (CAF)
ALCAPA
Kawasaki Disease
Cardiomyopathies in Children
Cardiac Tumors in Children

1. Pulmonary Hypertension in Congenital Heart Disease

DEFINITION

Pulmonary Hypertension (PH) in CHD is defined as:

Mean pulmonary artery pressure ≥ 20 mmHg with pulmonary vascular resistance (PVR) ≥ 3 Wood units

In congenital lesions, PH usually results from: chronic left-to-right shunts, pulmonary overcirculation, progressive pulmonary vascular remodeling.

CLASSIFICATION OF PH IN CHD

Hyperkinetic PH — high pulmonary flow (reversible stage). Examples: large VSD, large ASD, PDA.

Obstructive Pulmonary Vascular Disease — elevated PVR (partially reversible).

Eisenmenger Syndrome — irreversible pulmonary vascular disease, right-to-left shunt.

PH After Defect Repair — persistent elevated PVR despite correction.

Common CHD Causing PH — large VSD, large ASD, PDA, AVSD, truncus arteriosus, TAPVR (obstructed).

2. Echo Assessment of PH in CHD

STEP 1: ESTIMATE RV SYSTOLIC PRESSURE

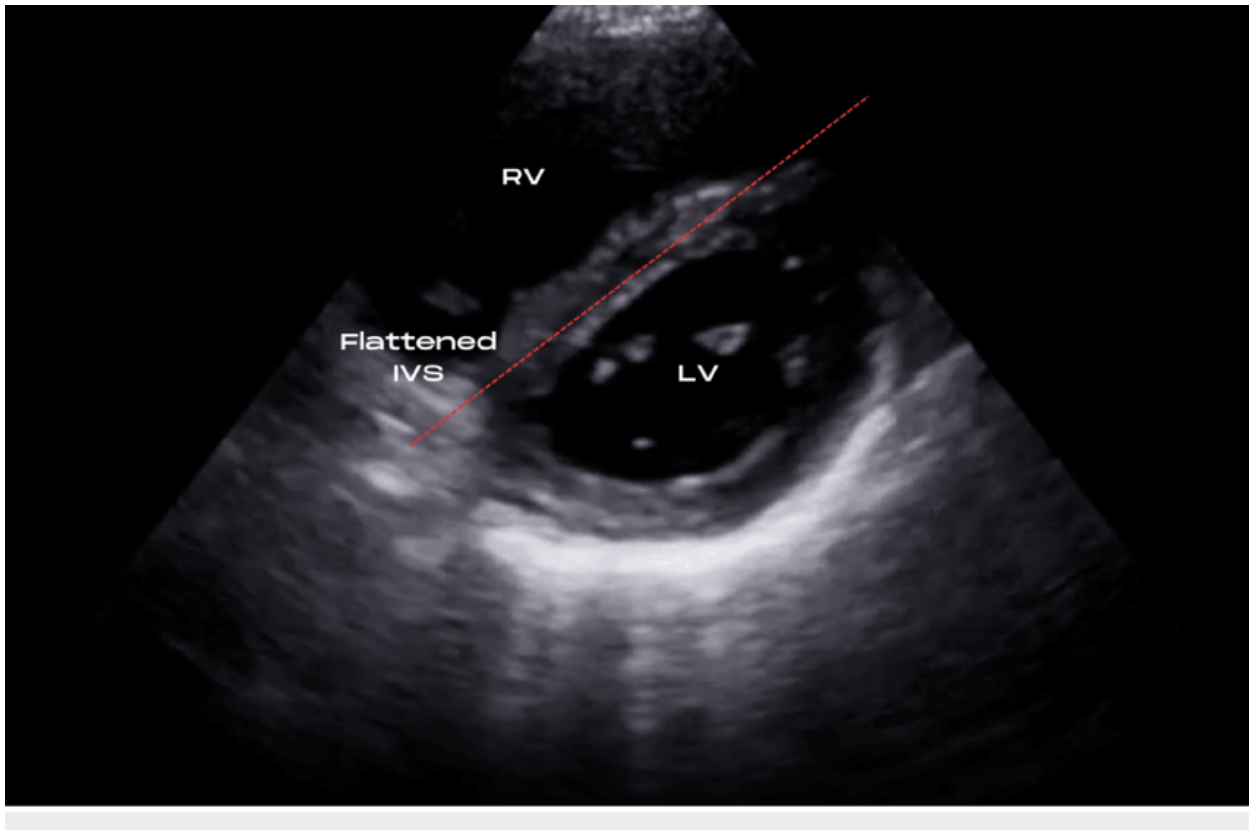
Measure TR jet velocity.

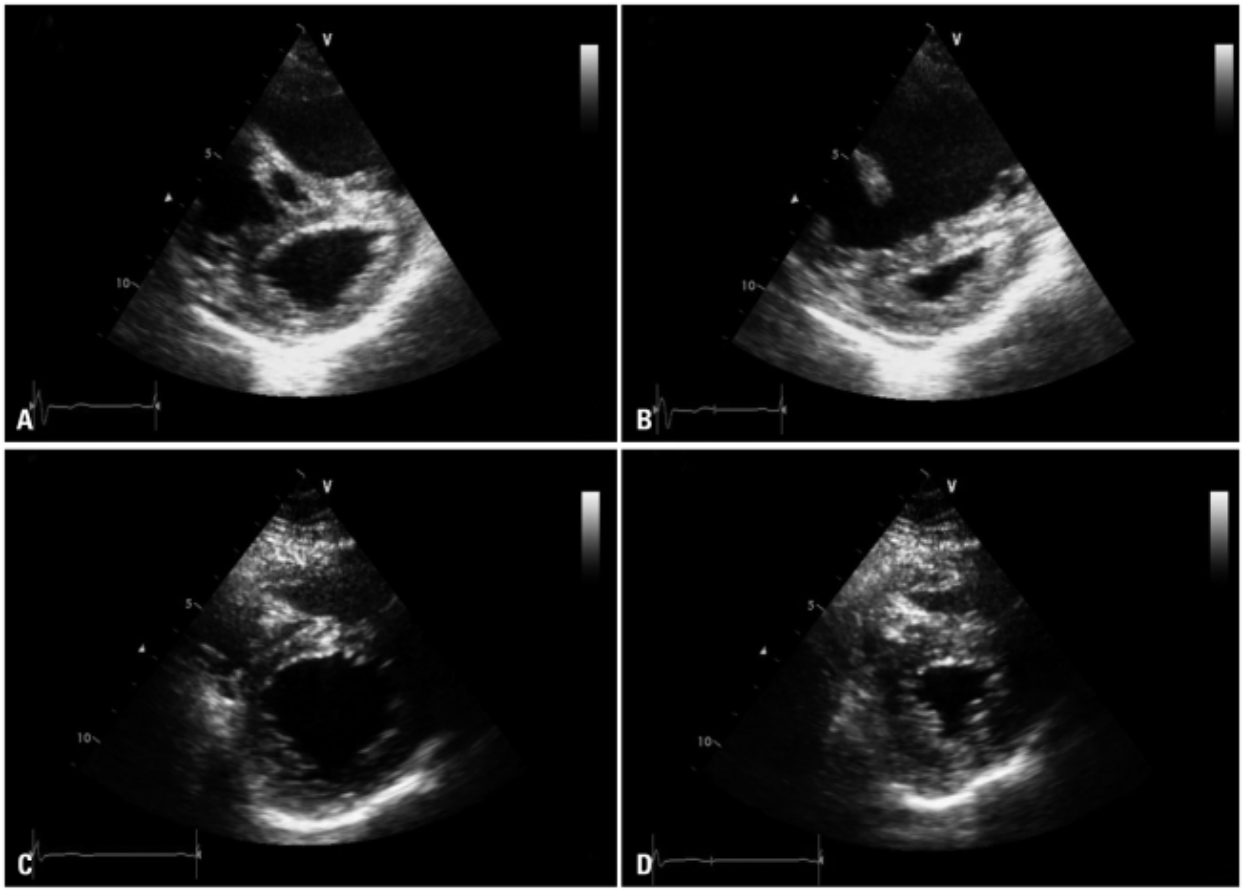
RVSP = 4(V^2) + RA pressure

If RVSP ≈ systemic pressure → severe PH.

STEP 2: LOOK AT RV MORPHOLOGY & IVS — SEVERE PH SIGNS

- RV hypertrophy
- RV dilation
- Interventricular septal flattening
- D-shaped LV in short axis
- Reduced RV function





STEP 3: PULMONARY ARTERY ASSESSMENT

- Dilated main PA
- Dilated branch PAs
- Reduced PA acceleration time

Short PA acceleration time (<100 ms) suggests high pressure.

STEP 4: SHUNT DIRECTION ASSESSMENT

Shunt Direction	Meaning
Left → Right	Flow stage (reversible)
Bidirectional	Advanced disease
Right → Left	Eisenmenger

3. PH in Specific Lesions

VSD with PH — Early: left → right. Late: bidirectional → right → left. If RV pressure = LV pressure → severe PH.

ASD with PH — late complication; RV dilated early; pulmonary vascular disease develops slowly.

PDA with PH — right → left shunt through PDA causes differential cyanosis (cyanosis lower limbs, upper limbs pink).

4. Eisenmenger Syndrome

Advanced irreversible PH with shunt reversal.

Echo findings: severe RV hypertrophy, systemic-level RV pressure, right → left shunt, dilated PA.

Clinical: cyanosis, clubbing, polycythemia. Closure contraindicated.

ECHO MARKERS OF IRREVERSIBILITY

- Severe RV hypertrophy
- Reduced RV function
- Right-to-left shunt
- Small LV cavity (due to low preload)
- No significant left-to-right flow

Cardiac catheterization required for final decision.

Echo signs of reversibility: LV dominant, left-to-right shunt.

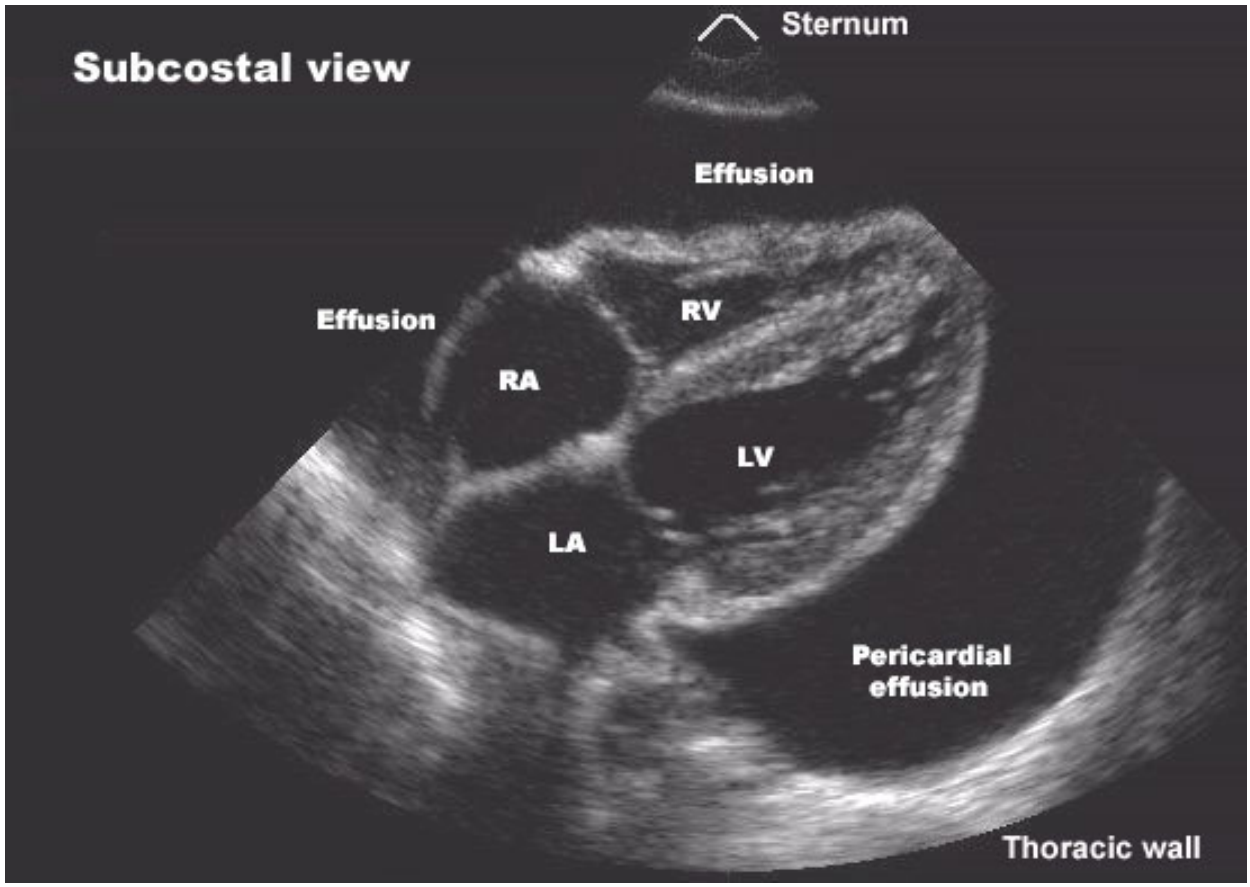
Pericardial Effusion

Pericardial effusion is the accumulation of fluid in the pericardial space surrounding the heart.

In children, causes include: viral pericarditis, post-cardiac surgery, tuberculosis (regional relevance), malignancy, renal failure, autoimmune disease, hypothyroidism, neonatal hydrops.

ECHO APPEARANCE — SUBCOSTAL VIEW (BEST INITIAL VIEW IN CHILDREN)

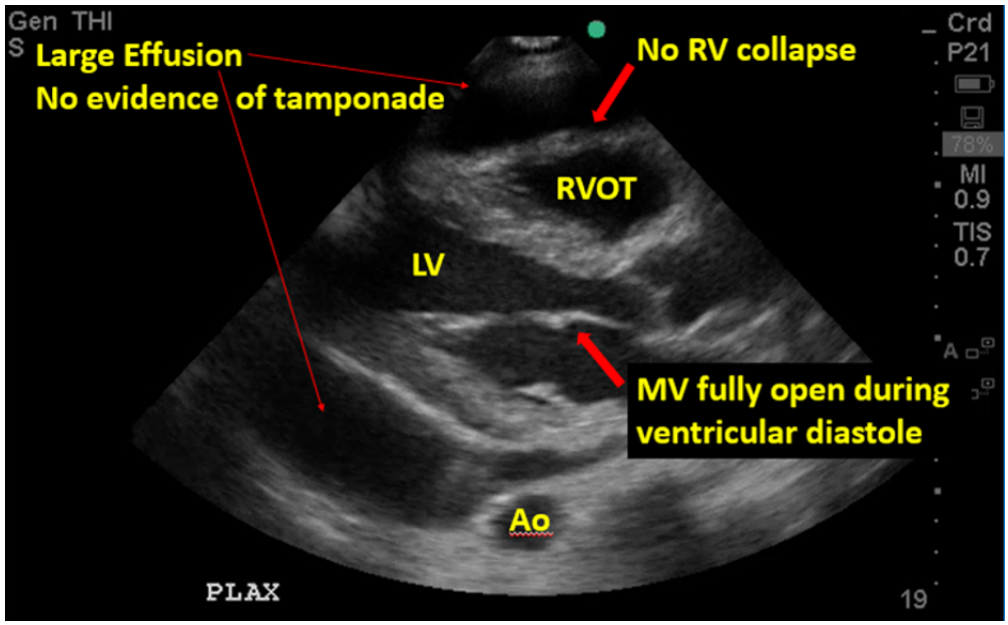
- Anechoic (black) space around heart
- Fluid between epicardium and pericardium
- May be circumferential or localized



HOW TO DIFFERENTIATE FROM PLEURAL EFFUSION

Key trick: pericardial effusion lies **anterior to descending aorta**; pleural effusion lies **posterior to descending aorta**.

Parasternal long axis view helps.



GRADING THE EFFUSION (ECHO-BASED)

Measure diastolic separation:

Size	Separation
Small	<10 mm
Moderate	10–20 mm
Large	>20 mm

In pediatrics, always interpret relative to body size.

1. The Critical Question: Is There Tamponade?

Tamponade is a clinical diagnosis supported by echo findings.

ECHO SIGNS OF TAMPONADE

1. RA Collapse (Early Sign) — RA wall collapses during systole.

2. RV Diastolic Collapse (More Specific) — RV free wall collapses in early diastole.

3. IVC Plethora — dilated IVC, <50% inspiratory collapse.

4. Exaggerated Respiratory Variation — mitral inflow variation >25%, tricuspid inflow variation >40%.

PEDIATRIC-SPECIFIC CONSIDERATIONS

Children may compensate better early. Be alert if: tachycardia, hepatomegaly, hypotension (late sign), muffled heart sounds, pulsus paradoxus.

In neonates: tamponade may present with sudden deterioration.

POST-OPERATIVE EFFUSION

After cardiac surgery: effusion may be loculated, posterior collections common, clots may appear echogenic.

Always sweep multiple views.

EFFUSION VS EPICARDIAL FAT

Epicardial fat — more echogenic, moves with heart, located anteriorly.

Effusion — anechoic, circumferential, clear fluid space.

WHEN TO DRAIN?

Urgent pericardiocentesis if: tamponade physiology, rapidly increasing effusion, hemodynamic instability.

Stable large effusion without tamponade → monitor.

RED FLAGS

Hypotension • RV diastolic collapse • Dilated IVC • Rapid clinical deterioration

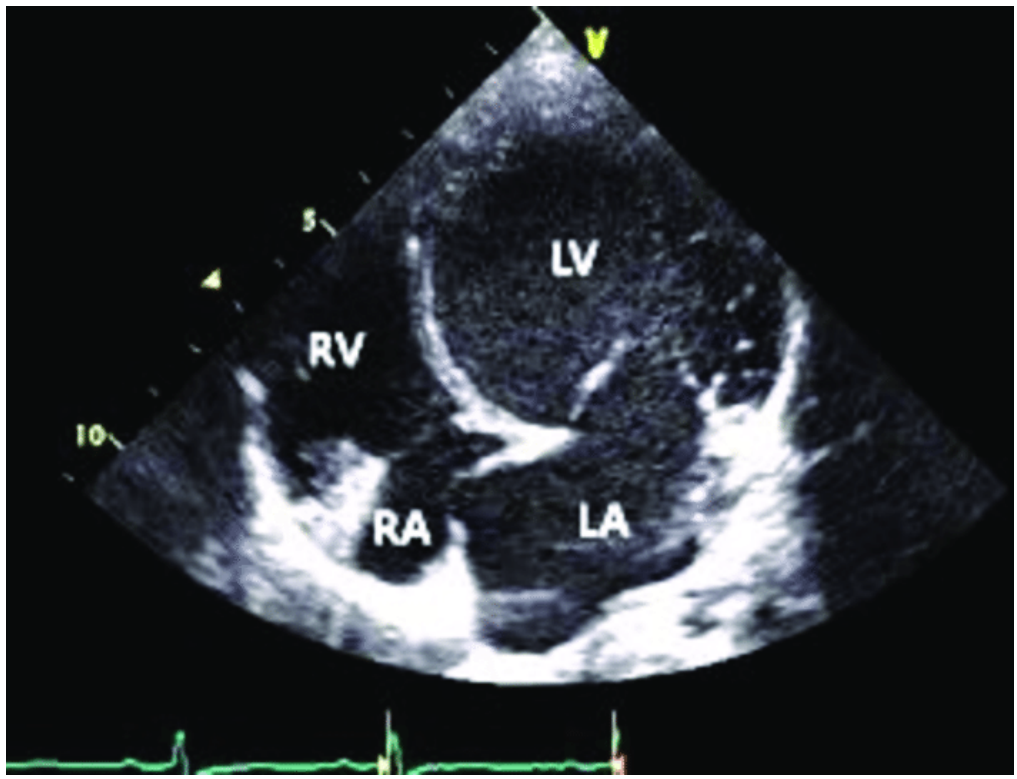
Myocarditis

Myocarditis is inflammation of the myocardium, most commonly viral in children.

Common causes: viral (Coxsackie, Adenovirus, Parvovirus, etc.), post-infectious immune response, autoimmune disease, drug-related, MIS-C (post-COVID inflammatory syndrome).

It may present as: acute heart failure, cardiogenic shock, dilated cardiomyopathy phenotype, arrhythmias.

ECHO FINDINGS — APICAL 4-CHAMBER VIEW



Look for: dilated LV, global hypokinesia, reduced ejection fraction, mild pericardial effusion (common).

KEY ECHO PATTERNS — GLOBAL LV DYSFUNCTION (MOST COMMON)

Diffuse reduced contractility, EF often <40%. LV dilated, no regional wall motion pattern like coronary disease.

Biventricular failure = worse prognosis.

RV INVOLVEMENT

RV may show: dilatation, reduced TAPSE, TR, septal flattening.

FULMINANT MYOCARDITIS

Severe LV dysfunction, thickened ventricular walls (edema), small LV cavity, rapid hemodynamic collapse.

Important: walls appear thick due to inflammation, not hypertrophy.

SERIAL ECHO IS CRITICAL

In myocarditis, function may worsen rapidly or improve dramatically within days. Repeat echo every 24–48 hrs in unstable patients.

DILATED CARDIOMYOPATHY PHENOTYPE

If subacute: dilated LV, thinned walls, severe systolic dysfunction. May resemble chronic cardiomyopathy.

LVNC-like: varying degree of LV dysfunction, dilated LV, recesses formation.

DOPPLER FINDINGS

Reduced LVOT VTI, functional mitral regurgitation, elevated filling pressures, TR from RV dysfunction.

Pericardial Effusion — mild effusion common, large effusion uncommon but possible. EF not accurate in effusion.

DIFFERENTIATING MYOCARDITIS FROM OTHER CAUSES

Condition	Key Feature
ALCAPA	Regional dysfunction
Kawasaki	Coronary dilation present

Always evaluate coronaries, AS and CoA in children.

RED FLAGS ON ECHO

EF <30% • Severe RV dysfunction • LV thrombus • Rising pulmonary pressures • Progressive dilation

Myocardial Stunning

Myocardial stunning is a temporary, post-ischemic mechanical dysfunction of the left ventricle (LV) causing reversible regional wall motion abnormalities.

PATHOPHYSIOLOGY AND CAUSES

Transient Ischemia — short-term reduction or total occlusion of coronary blood flow followed by successful reperfusion.

Cellular Mechanisms — free radical production, intracellular calcium overload, and myofilament desensitization.

Triggers — exercise-induced stress, unstable angina, or acute myocardial infarction treated early.

CLINICAL RECOGNITION AND MANAGEMENT

Diagnosis — identified using dobutamine echocardiography or scintigraphy to demonstrate viable, reversible tissue.

Prognosis — contractile function recovers entirely over hours, days, or weeks without permanent necrosis.

Treatment — standard guideline-directed therapy for underlying coronary artery disease, including beta-blockers.

Takotsubo Cardiomyopathy

Takotsubo cardiomyopathy, also known as broken-heart syndrome or stress-induced cardiomyopathy, is a temporary heart condition. It features sudden chest pain, shortness of breath, and temporary weakening of the heart's main pumping chamber.

KEY SIGNS AND TRIGGERS

Common triggers — severe emotional stress (like losing a loved one) or physical stress (like a sudden illness or accident).

Main symptoms — chest pain and trouble breathing that feel just like a heart attack.

DIAGNOSIS AND RECOVERY

Blocked arteries — unlike a real heart attack, tests show no major blockages in the heart blood vessels.

Heart shape — the left ventricle changes shape and looks like a round Japanese octopus trap (*takotsubo*).

Healing time — most people get better completely within one to two months with standard supportive care and heart medications.

Inferior Vena Cava (IVC) Collapsibility and Distensibility

IVC collapsibility and distensibility measure how the main vein's diameter changes during breathing to estimate right atrial pressure and predict fluid responsiveness. Use collapsibility for spontaneous breathing and distensibility for mechanical ventilation.

SPONTANEOUS BREATHING (COLLAPSIBILITY INDEX)

Concept — negative pressure during inspiration draws blood into the chest, causing the IVC to collapse.

Collapsibility Index = [IVC max (expiration) − IVC min (inspiration)] / IVC max (expiration) × 100

Clinical Meaning — a collapsibility index greater than 50% with an IVC diameter under 2.1 cm suggests low right atrial pressure (0–5 mm Hg) and possible fluid depletion. A flat or minimally collapsing IVC indicates high pressure or volume overload.

MECHANICAL VENTILATION (DISTENSIBILITY INDEX)

Concept — positive pressure ventilation pushes air in, increasing chest pressure during inspiration and expanding the IVC.

Distensibility Index = [IVC max (inspiration) − IVC min (expiration)] / IVC min (expiration) × 100

Clinical Meaning — a distensibility index greater than 18% strongly suggests that a mechanically ventilated patient will increase their cardiac output if given intravenous fluids (fluid responsiveness).

Coronary Artery Abnormalities in Children

Common Coronary Abnormalities in Children

Lesion	Echo Importance
Coronary artery fistula	Commonest symptomatic anomaly
ALCAPA	Surgical emergency
ARCAPA	Rare
Anomalous coronary origin from opposite sinus	Sudden death risk
Single coronary artery	Variable
Kawasaki aneurysm	Important acquired lesion

Although coronary abnormalities are uncommon, they may cause: sudden cardiac death, myocardial ischemia, heart failure, ventricular dysfunction, arrhythmias, syncope, continuous murmurs.

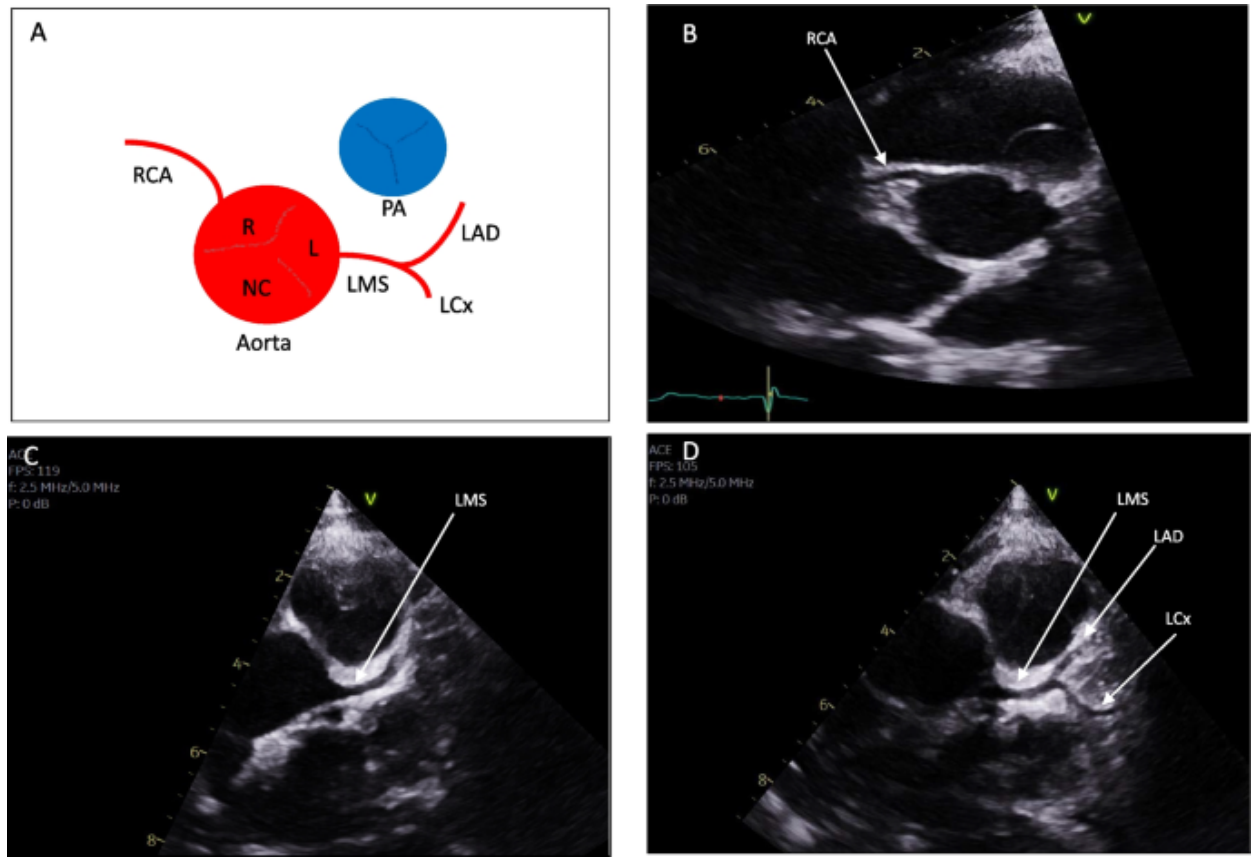
GOLDEN RULE

Every complete pediatric echocardiogram should document the origin of both coronary arteries.

NORMAL CORONARY ANATOMY

Left Main Coronary Artery (LMCA) — Origin: left coronary sinus. Divides into: Left Anterior Descending (LAD), Left Circumflex (LCx).

Right Coronary Artery (RCA) — Origin: right coronary sinus. Courses in right atrioventricular groove.



1. Echo Views for Coronary Arteries

PARASTERNAL SHORT-AXIS AT AORTIC VALVE (BEST VIEW)

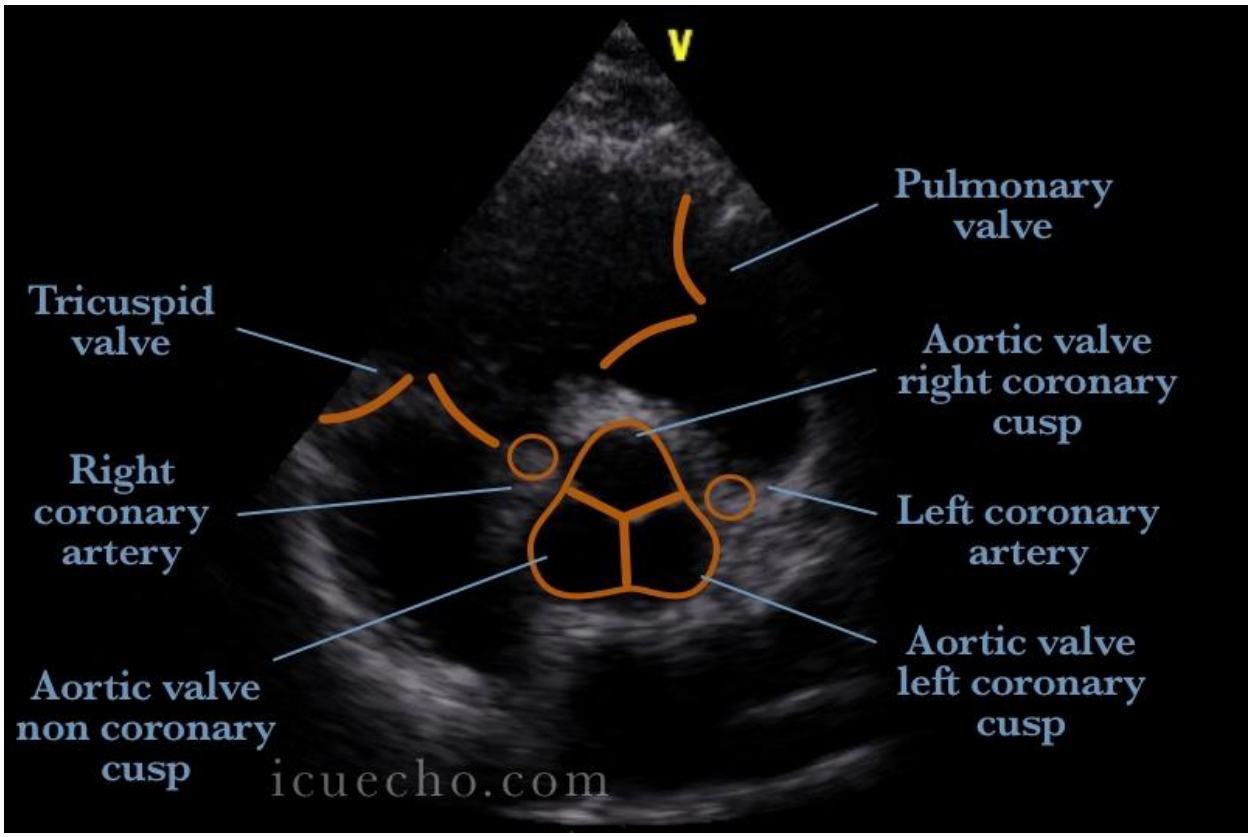
This is the **single most important view**.

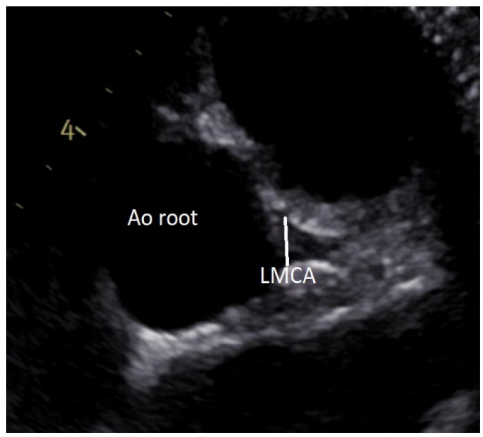
Visualize: left coronary ostium, right coronary ostium, direction of flow.

ADDITIONAL VIEWS

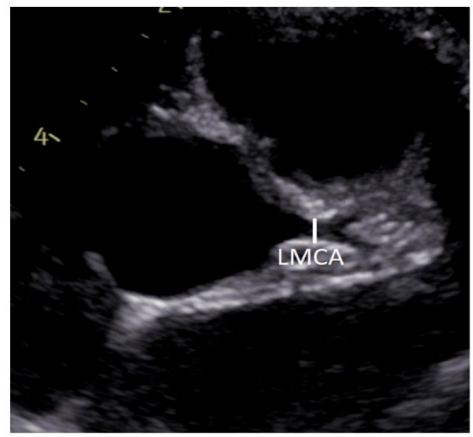
- Parasternal Long Axis
- High Parasternal
- Apical Long Axis
- Modified Short Axis

Color Doppler improves visualization.

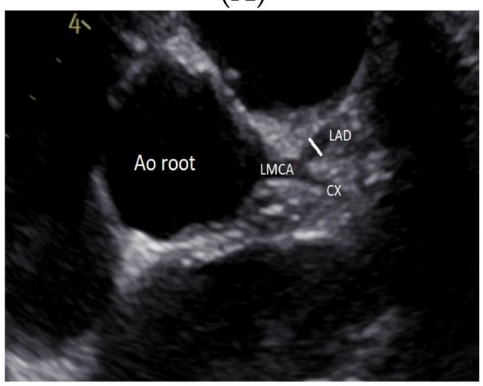



Ao root
LMCA

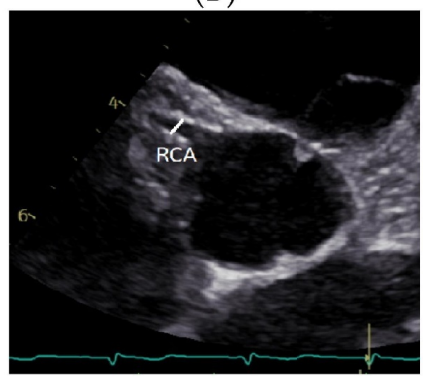
(A)


LMCA

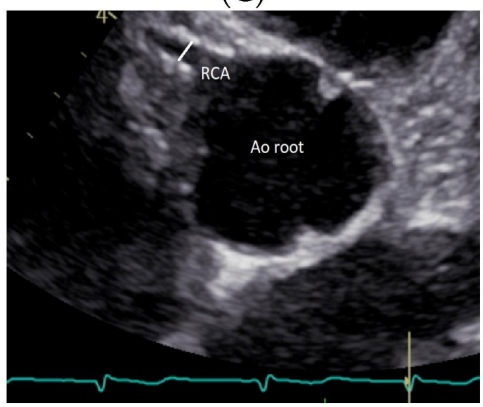
(B)


Ao root
LAD
LMCA
CX

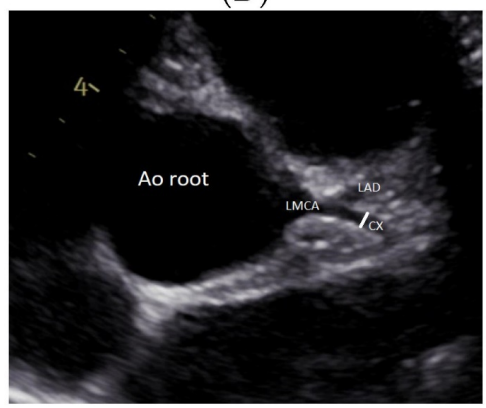
(C)


RCA

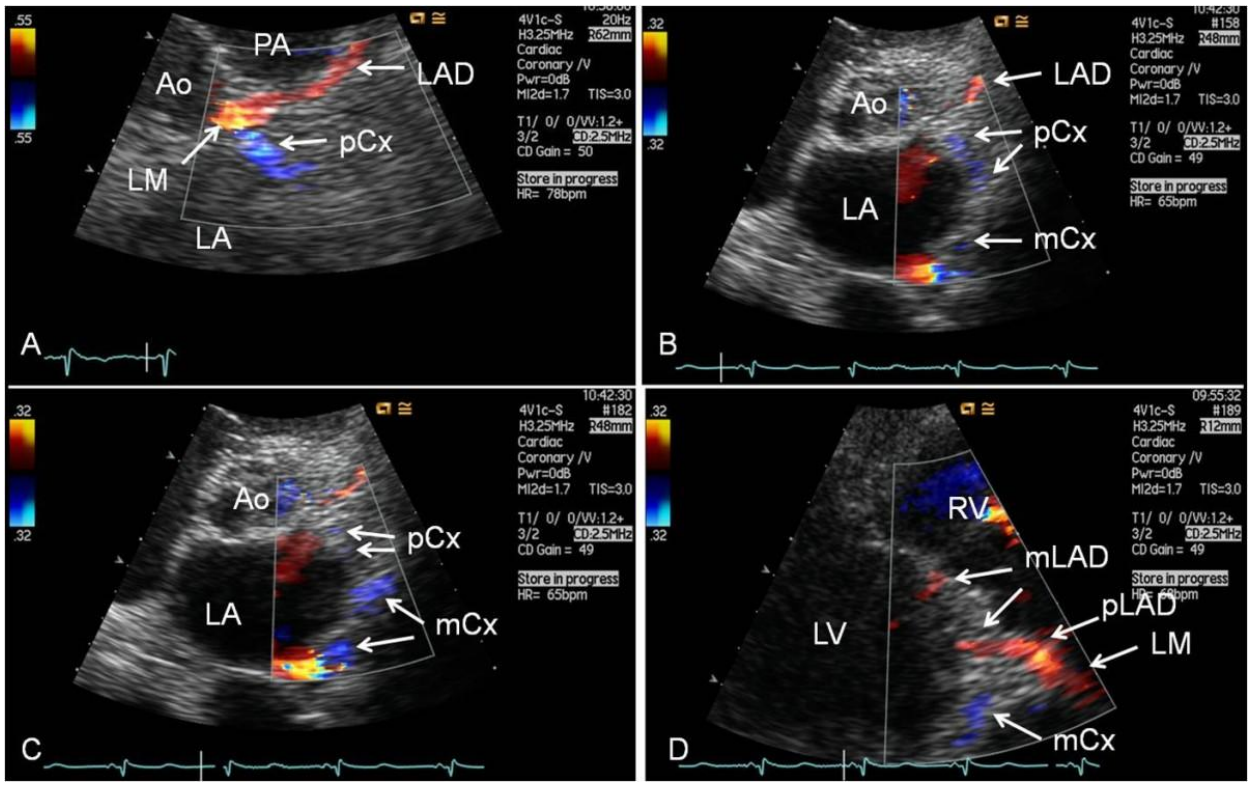
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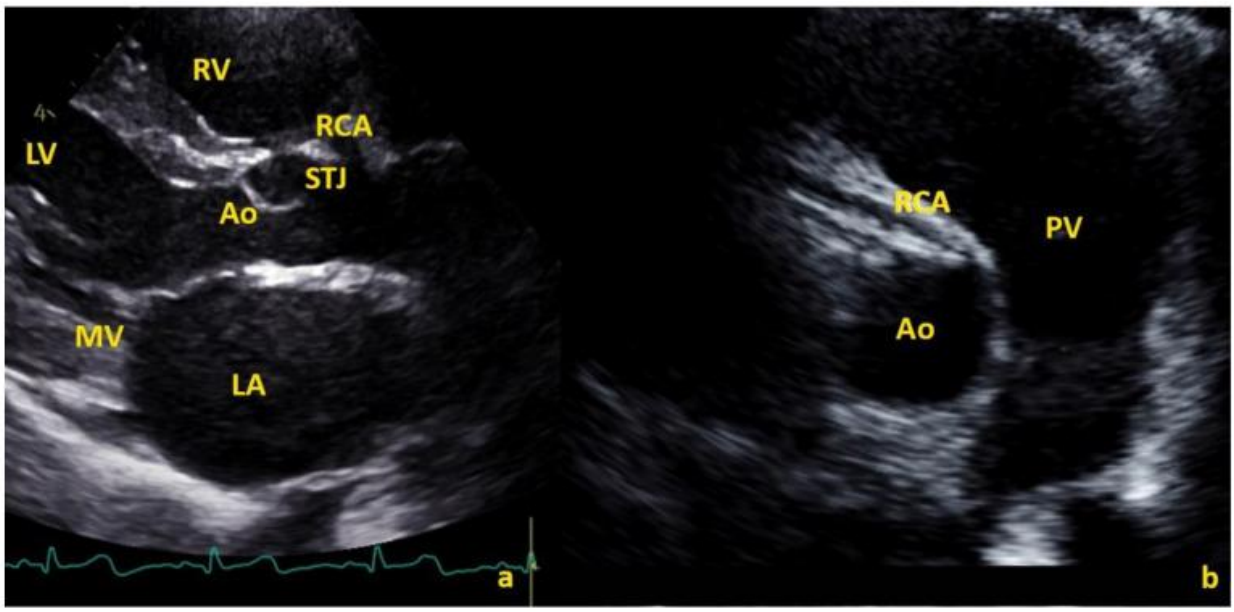

RCA
Ao root

(E)


Ao root
LAD
LMCA
CX

(F)





STEPWISE CORONARY EXAMINATION

Step 1 — Identify: right coronary sinus, left coronary sinus, non-coronary sinus.

Step 2 — Confirm: LMCA arises from left sinus; RCA arises from right sinus.

Step 3 — Follow proximal course. Assess: diameter, branching, color Doppler flow.

Step 4 — Look for: dilatation, abnormal origin, abnormal flow, fistulous communication.

Coronary Artery Fistula (CAF)

DEFINITION

A coronary artery fistula is an **abnormal communication between a coronary artery and a cardiac chamber or great vessel**, bypassing the myocardial capillary bed.

COMMON SITES OF DRAINAGE

Most drain into: right ventricle, right atrium, pulmonary artery.

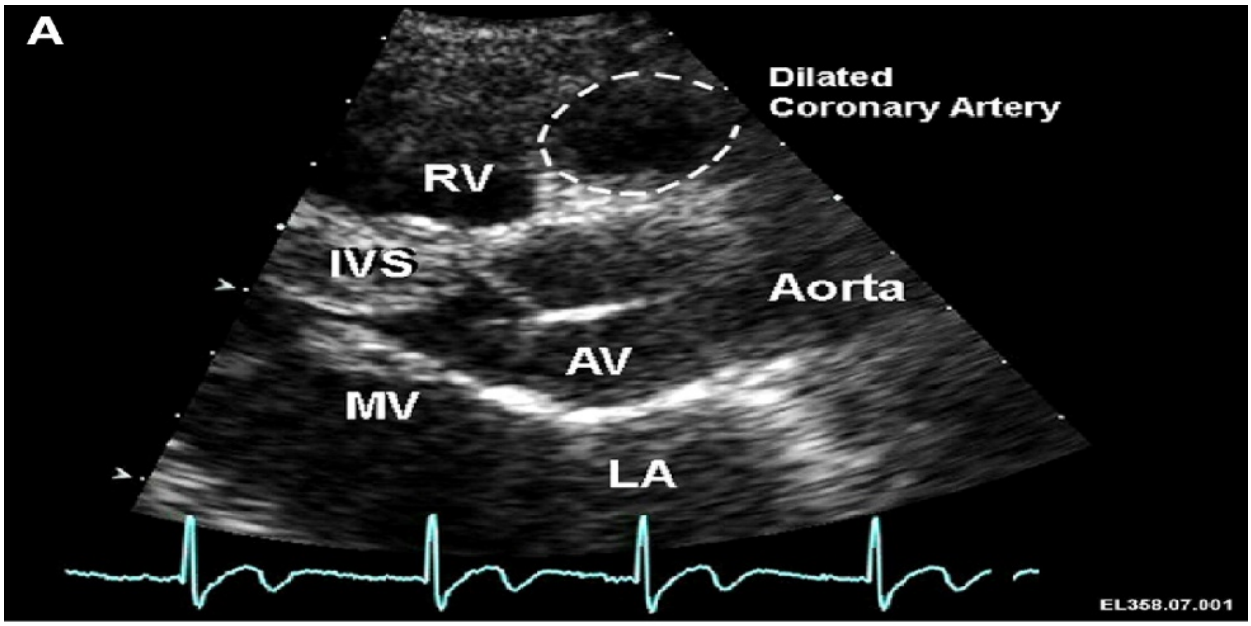
Less commonly: coronary sinus, left atrium, left ventricle.

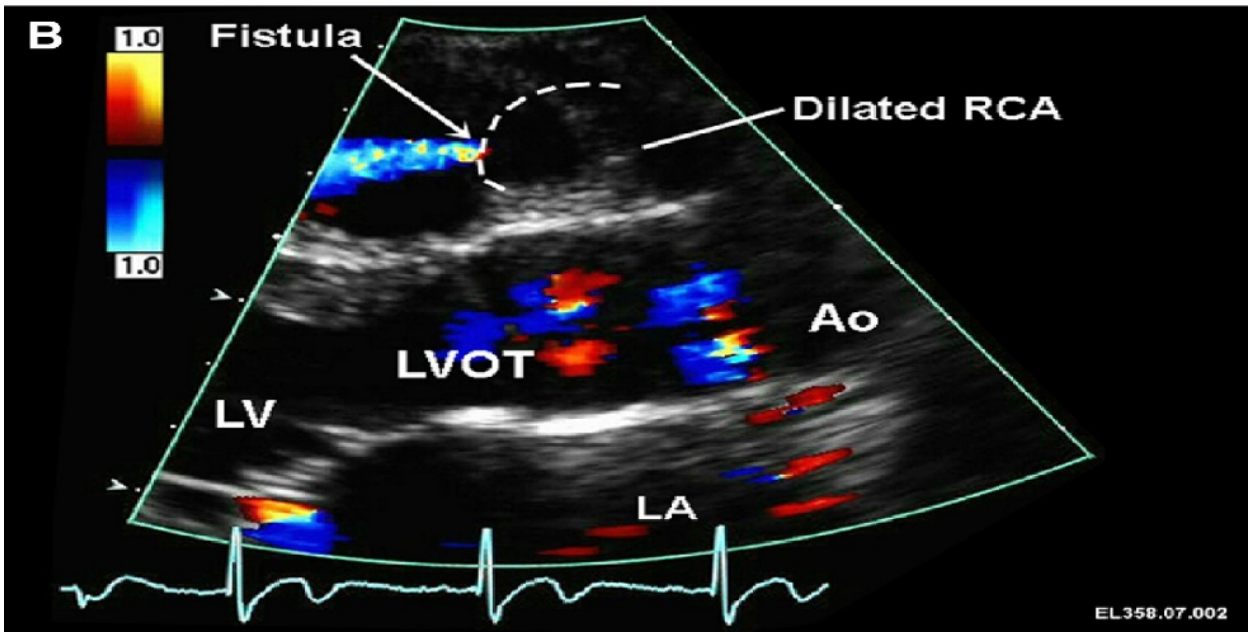
TWO MAJOR FINDINGS

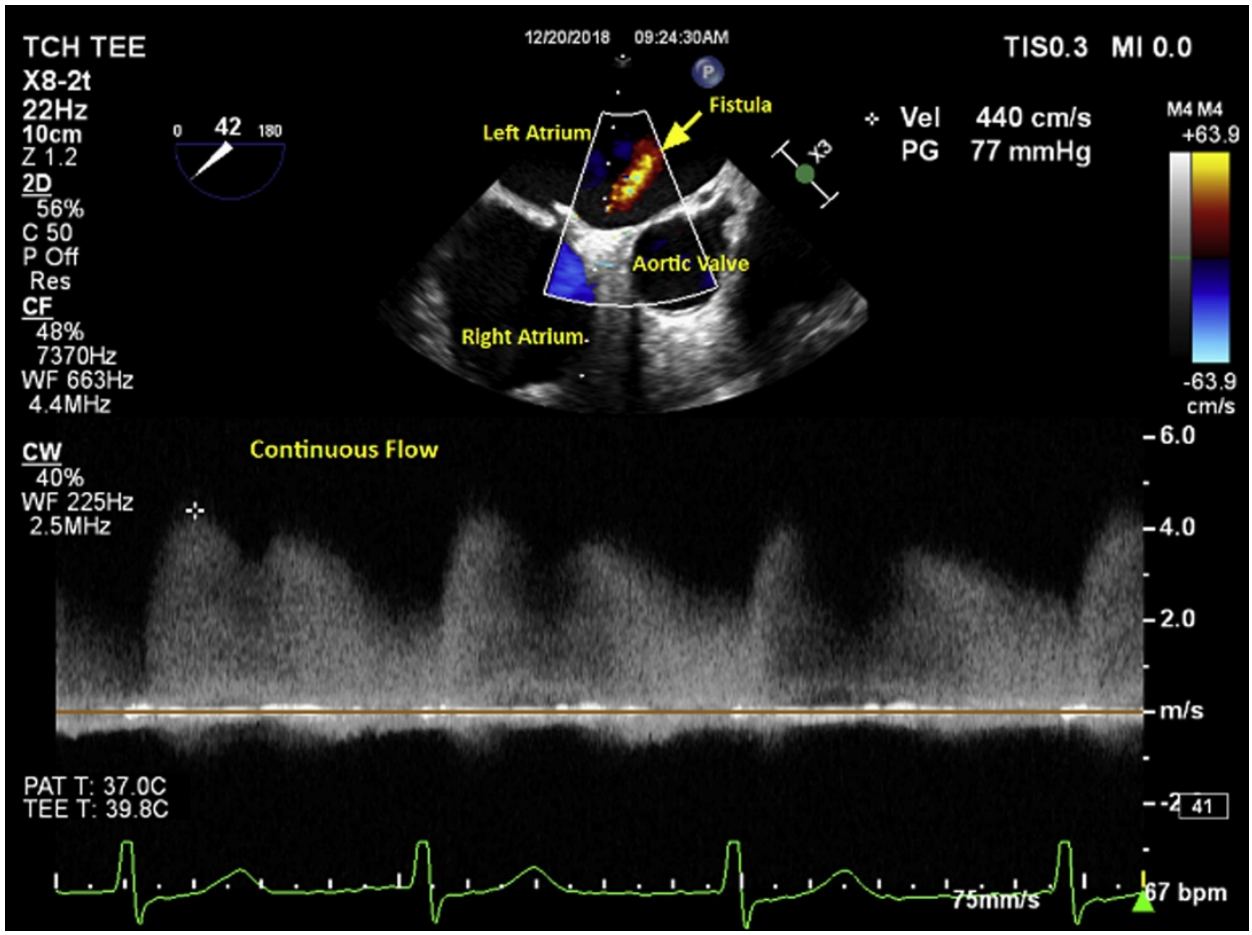
1. Dilated Feeding Coronary — large tortuous vessel, much larger than expected.

2. Continuous Color Flow — continuous turbulent flow entering: RV, RA, PA.

Color Doppler is diagnostic. Unlike PDA, flow follows the fistulous tract.


A
Dilated
Coronary Artery
RV
IVS
Aorta
AV
MV
LA
EL358.07.001


B
1.0
1.0
Fistula
Dilated RCA
Ao
LVOT
LV
LA
EL358.07.002



HEMODYNAMICS

Large fistula causes:

Coronary steal phenomenon → Reduced myocardial perfusion → LV dysfunction → Heart failure

CLINICAL PRESENTATION

Small fistula — usually asymptomatic.

Large fistula — continuous murmur, failure to thrive, heart failure, myocardial ischemia.

INDICATIONS FOR CLOSURE

- Large fistula
- Symptoms
- LV volume overload
- Myocardial ischemia
- Progressive coronary dilatation

Closure may be: catheter device, surgery.

ALCAPA (Anomalous Left Coronary Artery from Pulmonary Artery)

WHY IMPORTANT?

One of the most important diagnoses in pediatric echocardiography.

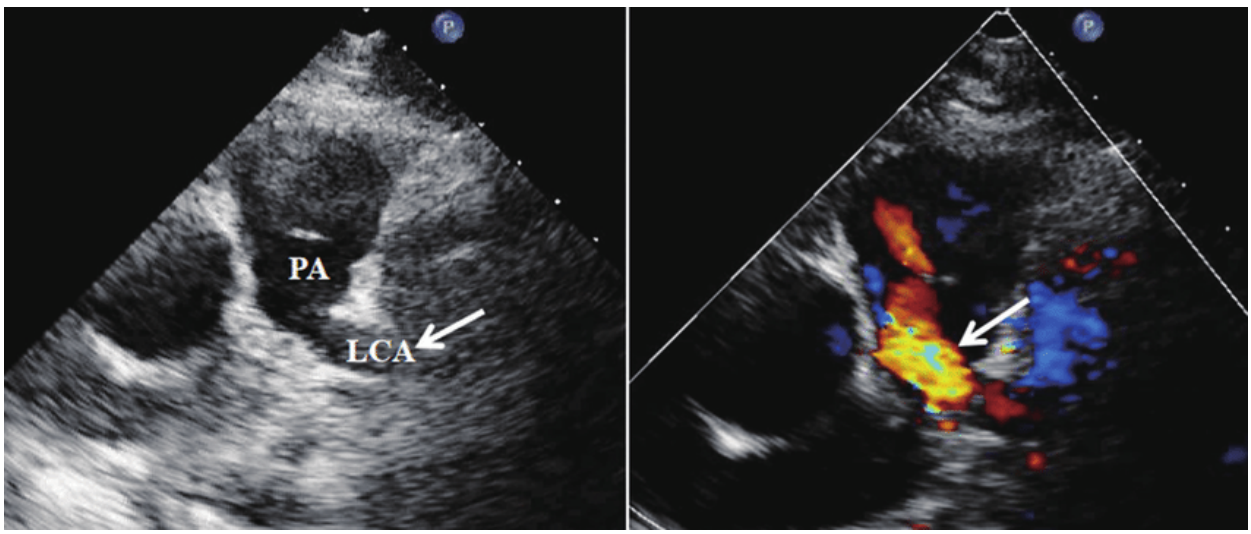
Without surgery: mortality approaches **90% during infancy**.

ECHO CLUES — PARASTERNAL SHORT AXIS

LMCA absent from left coronary sinus.

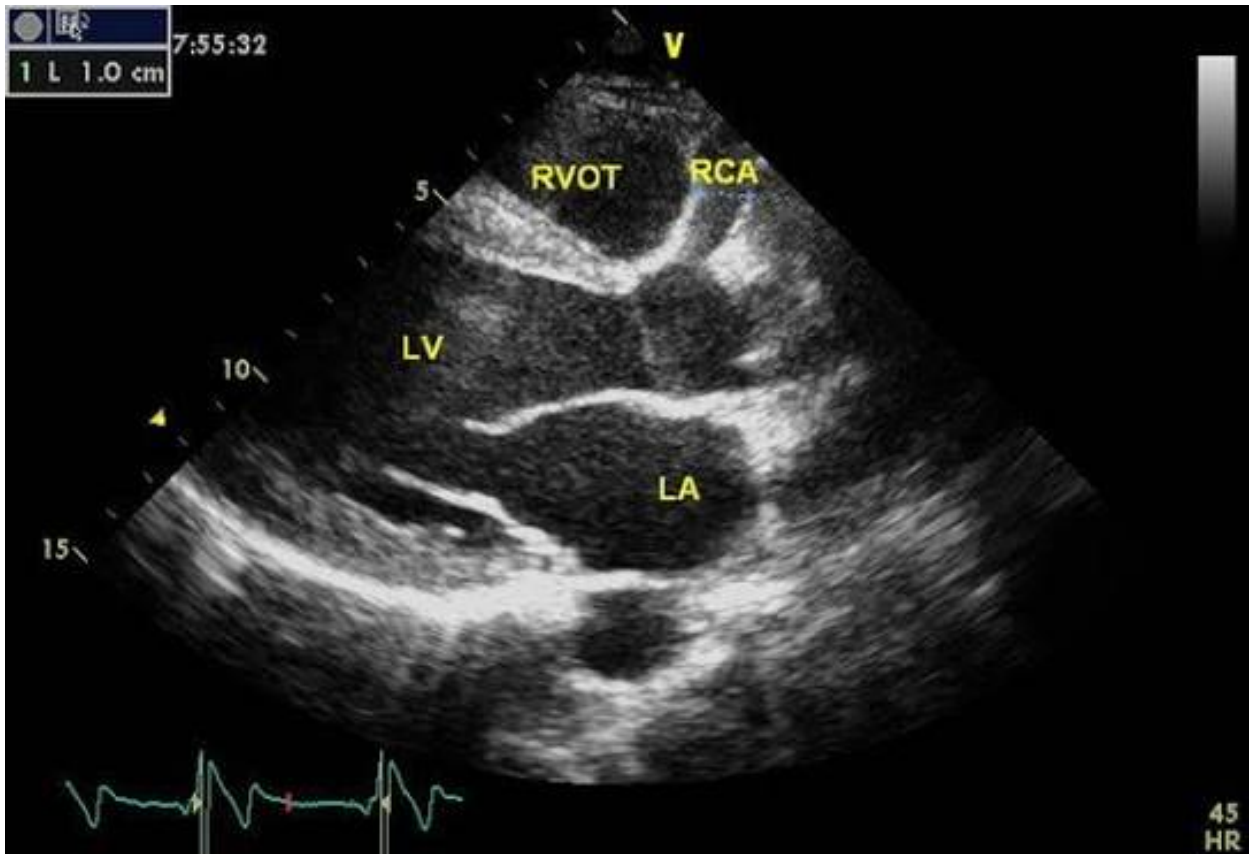
COLOR DOPPLER

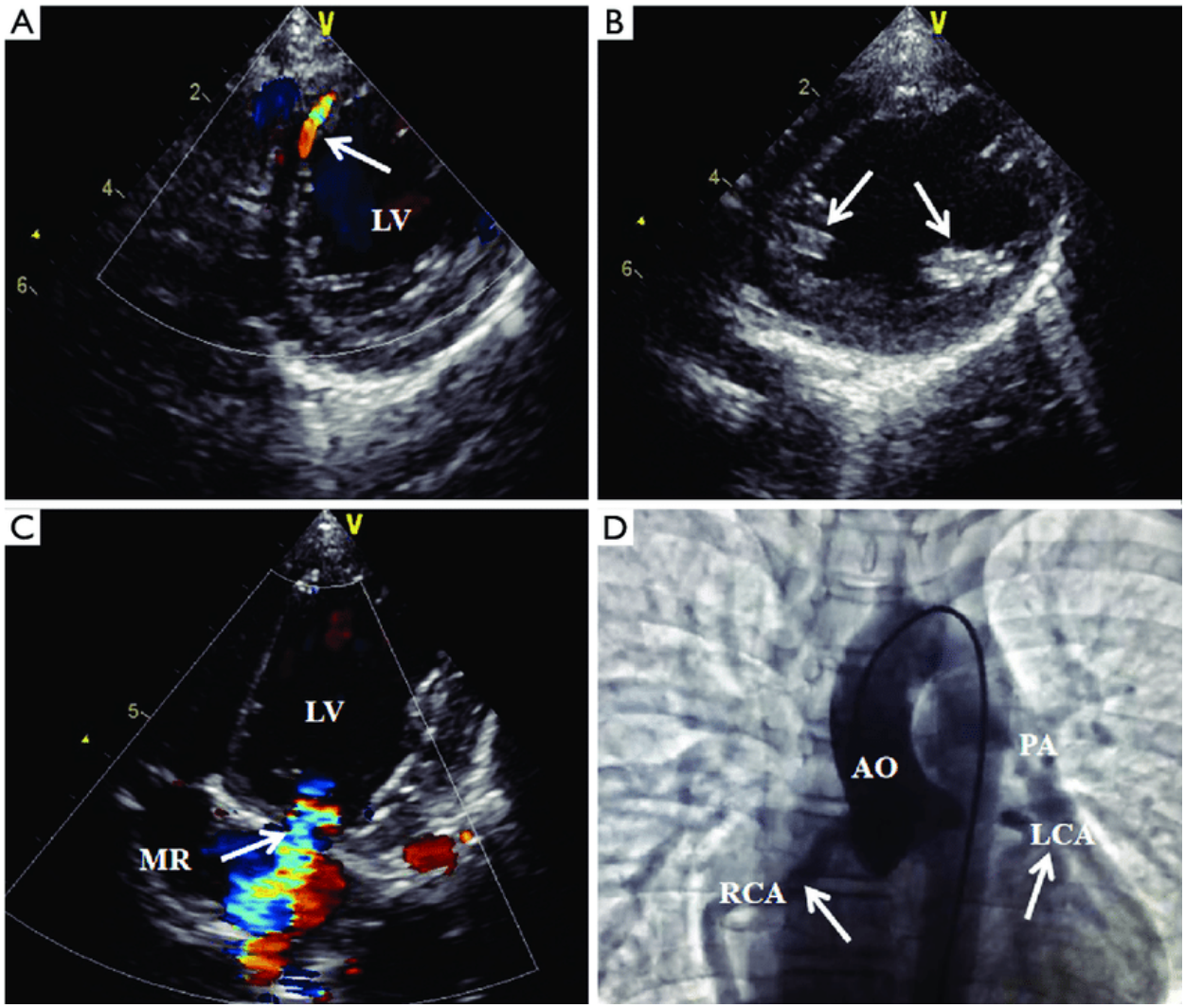
Reverse flow from LMCA → into pulmonary artery
This is the hallmark.



ADDITIONAL FINDINGS

- Dilated RCA
- Extensive collateral vessels
- LV dysfunction
- Dilated LV
- Papillary muscle echogenicity
- Mitral regurgitation





CLASSIC CLINICAL PICTURE

Infant, 2–4 months — poor feeding, excessive cry/colic, sweating, heart failure.

Echo — dilated RCA, absent LMCA origin, MR, poor LV function.

THINK

ALCAPA until proven otherwise.

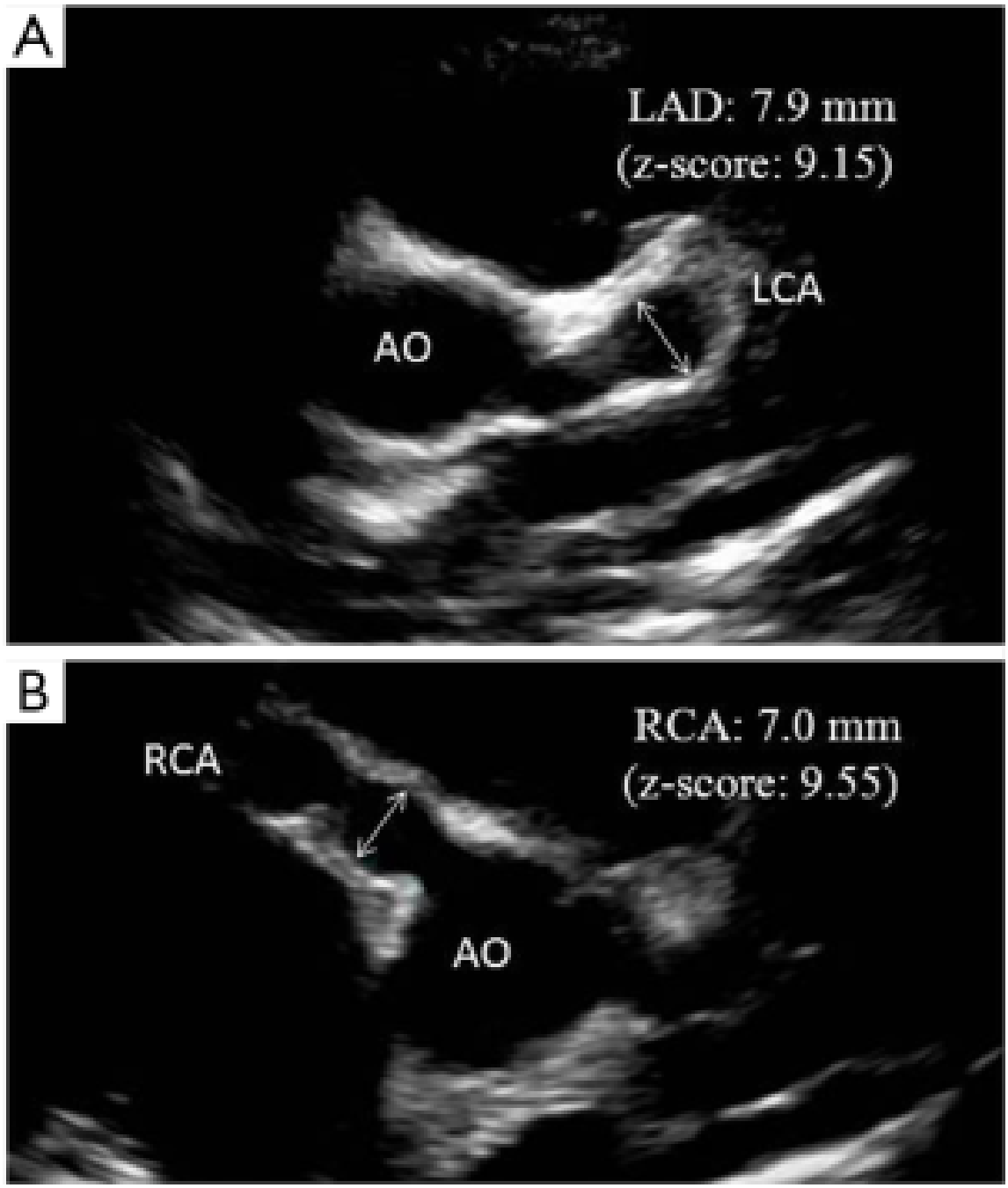
Kawasaki Disease

Most common acquired coronary abnormality.

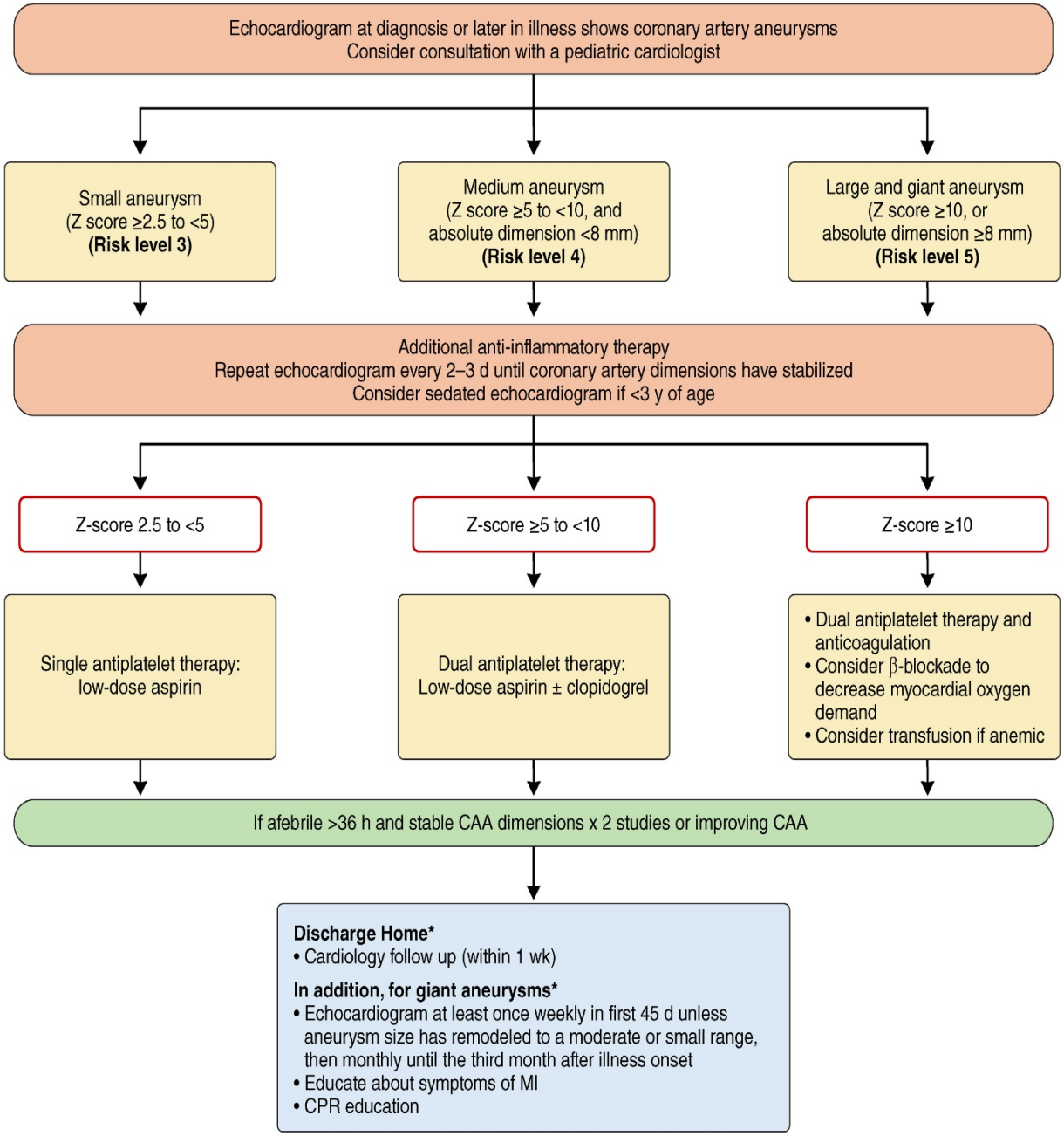
Echo findings: coronary dilatation, coronary aneurysm, thrombus, stenosis.

Measure: LMCA, LAD, RCA.

Use **Z-scores** according to body surface area.



MANAGEMENT OF CORONARY ARTERY DILATATION IN KAWASAKI DISEASE



Cardiomyopathies in Children

1. Echocardiographic Evaluation — A Practical, Stepwise Approach

Cardiomyopathy refers to a primary disease of the myocardium resulting in abnormal ventricular structure or function that cannot be explained solely by congenital heart disease, hypertension, or coronary artery disease.

CLASSIFICATION OF PEDIATRIC CARDIOMYOPATHY

Type	Main Echo Finding
Dilated Cardiomyopathy (DCM)	Dilated ventricle + poor systolic function
Hypertrophic Cardiomyopathy (HCM)	Thick ventricular walls with preserved or hyperdynamic function
Restrictive Cardiomyopathy (RCM)	Severe atrial enlargement with normal ventricular size
Left Ventricular Non-compaction (LVNC)	Prominent trabeculations with deep recesses
Arrhythmogenic Cardiomyopathy	RV dilation and dysfunction

STEPWISE ECHO APPROACH — EVERY STUDY SHOULD ANSWER SIX QUESTIONS

1. Is the ventricle dilated?

↓

2. Is myocardial thickness normal?

↓

3. Is systolic function preserved?

↓

4. Is diastolic function abnormal?

↓

5. Are atria enlarged?

↓

6. Is there associated thrombus, valve disease or pericardial effusion?

ESSENTIAL ECHO VIEWS

Always obtain: parasternal long axis, parasternal short axis, apical four chamber, apical two chamber, apical five chamber, subcostal four chamber, M-mode, tissue Doppler, color Doppler, spectral Doppler.

MEASUREMENTS TO OBTAIN

Left ventricle — LVEDD (Z-score), LVESD, septal thickness, posterior wall thickness.

Function — fractional shortening, ejection fraction (Simpson biplane), MAPSE, tissue Doppler S', global longitudinal strain (if available).

Right ventricle — TAPSE, FAC, RV dimensions.

Atria — LA volume index, RA area.

2. Dilated Cardiomyopathy (DCM)

Dilated cardiomyopathy is characterized by: dilated left ventricle, reduced systolic function, absence of significant congenital or valvular disease.

COMMON CAUSES

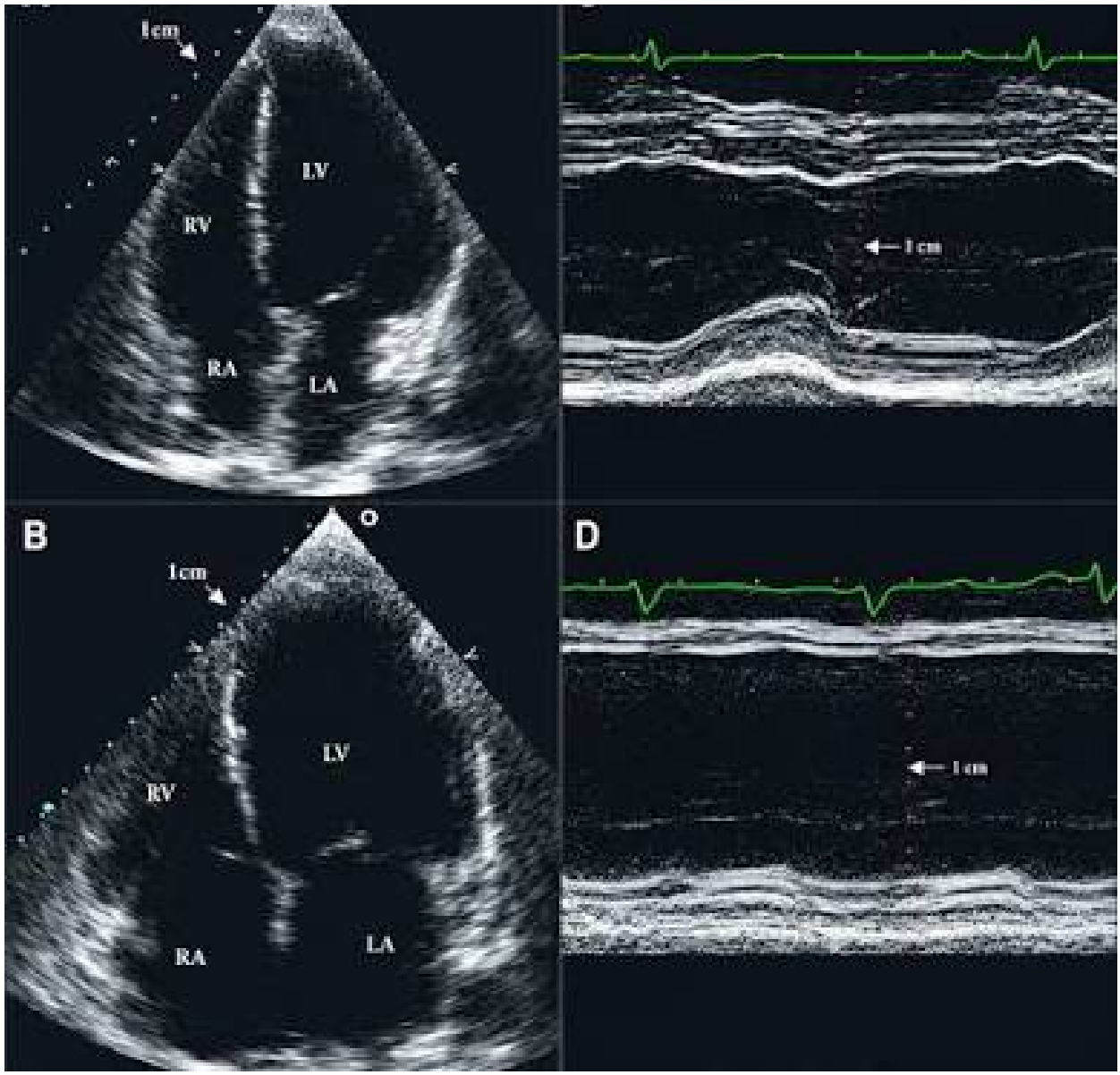
- Viral myocarditis
- Genetic disease
- Neuromuscular disorders
- Metabolic disease
- Anthracycline toxicity
- Idiopathic

ECHO FINDINGS — PARASTERNAL LONG AXIS

Observe: enlarged LV cavity, thin ventricular walls, poor inward motion.

M-mode shows reduced shortening, low fractional shortening.

Apical Four Chamber — LV enlargement, functional mitral regurgitation, left atrial enlargement.



Doppler Findings — reduced LVOT VTI, low cardiac output, mitral regurgitation.

QUANTIFYING LV FUNCTION

Category	EF
Normal	55%
Mild dysfunction	45–54%
Moderate	30–44%
Severe	<30%

ECHO CLUES

- Globular heart
- Poor contractility
- Spontaneous echo contrast
- LV thrombus
- Functional MR

CLINICAL PEARL

Always look carefully for LV thrombus before discharge.

3. Hypertrophic Cardiomyopathy (HCM)

CAUSES

- Sarcomeric mutations
- Pompe disease
- Friedreich ataxia
- Infant of diabetic mother
- Metabolic disorders

TYPES

- Asymmetric septal hypertrophy
- Concentric hypertrophy
- Apical hypertrophy (rare)

ECHOCARDIOGRAPHIC CRITERIA

Asymmetric Septal Hypertrophy (ASH)

- Maximal wall thickness ≥ 15 mm in one or more LV segments, typically the interventricular septum
- Septal-to-posterior wall ratio > 1.3 in normotensive patients (or > 1.5 with history of hypertension)
- Disproportionate thickening of the septum compared to the posterior free wall, often with small LV cavity and dynamic outflow tract obstruction (SAM of the mitral valve)

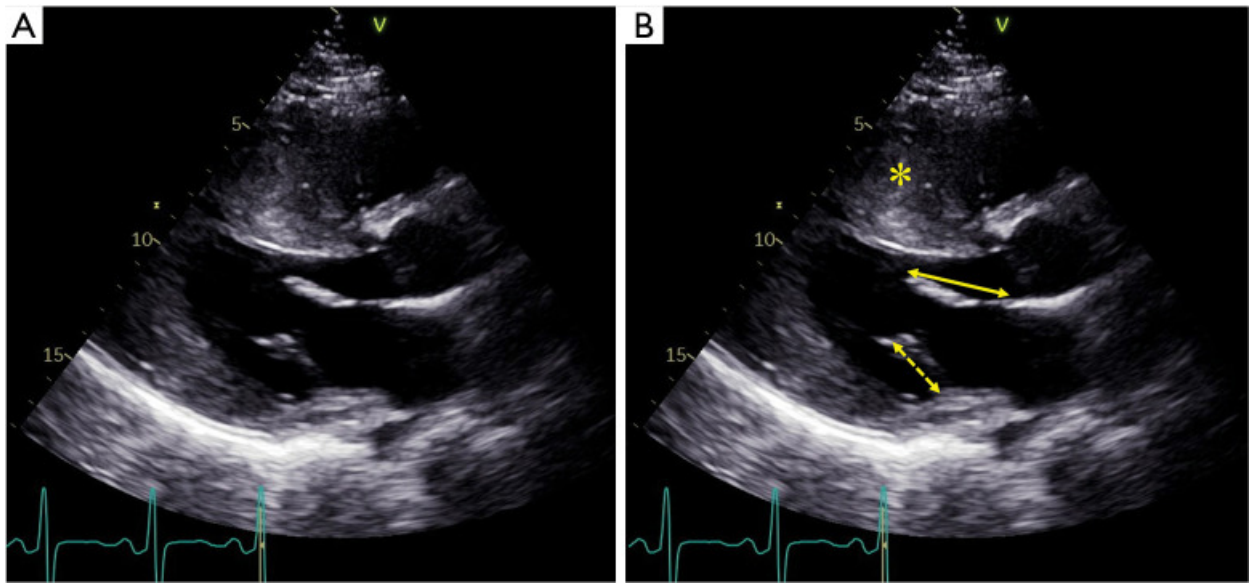
Concentric Hypertrophy

- Diffuse, uniform thickening of all LV walls with increased relative wall thickness (RWT > 0.42) and reduced chamber size
- Septal-to-posterior wall thickness ratio close to 1.0 (symmetrical thickening)
- Usually secondary to chronic pressure overload (systemic hypertension, aortic stenosis), though infiltrative processes like cardiac amyloidosis can also manifest symmetrically

HOCM: DYNAMIC LVOT OBSTRUCTION

Look for: systolic anterior motion (SAM), mitral-septal contact, late-peaking Doppler velocity.

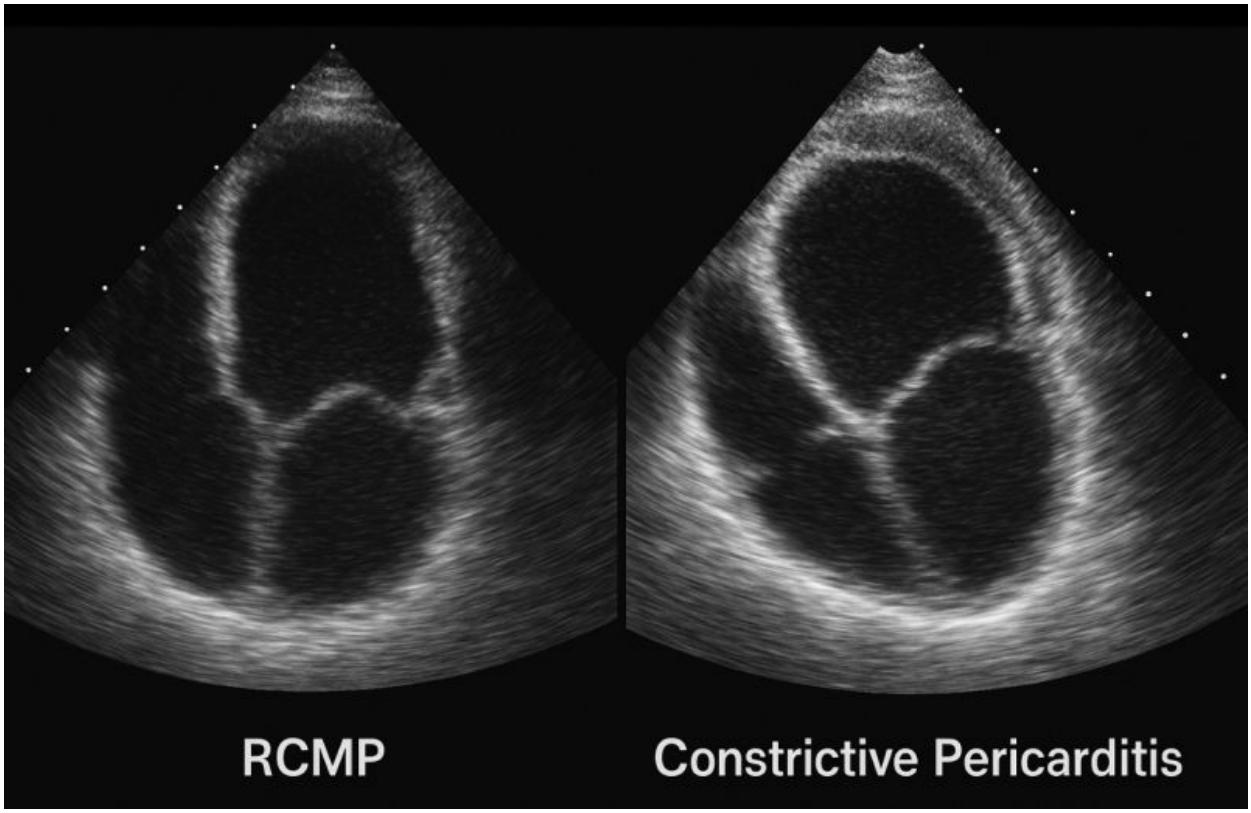
Doppler — late-peaking dagger-shaped LVOT jet, mitral regurgitation.



4. Restrictive Cardiomyopathy

The hallmark is: massive biatrial enlargement.

Ventricles: normal size, normal wall thickness, normal EF.



Doppler Findings — restrictive filling pattern, high E velocity, low A velocity, short deceleration time, elevated E/e'.

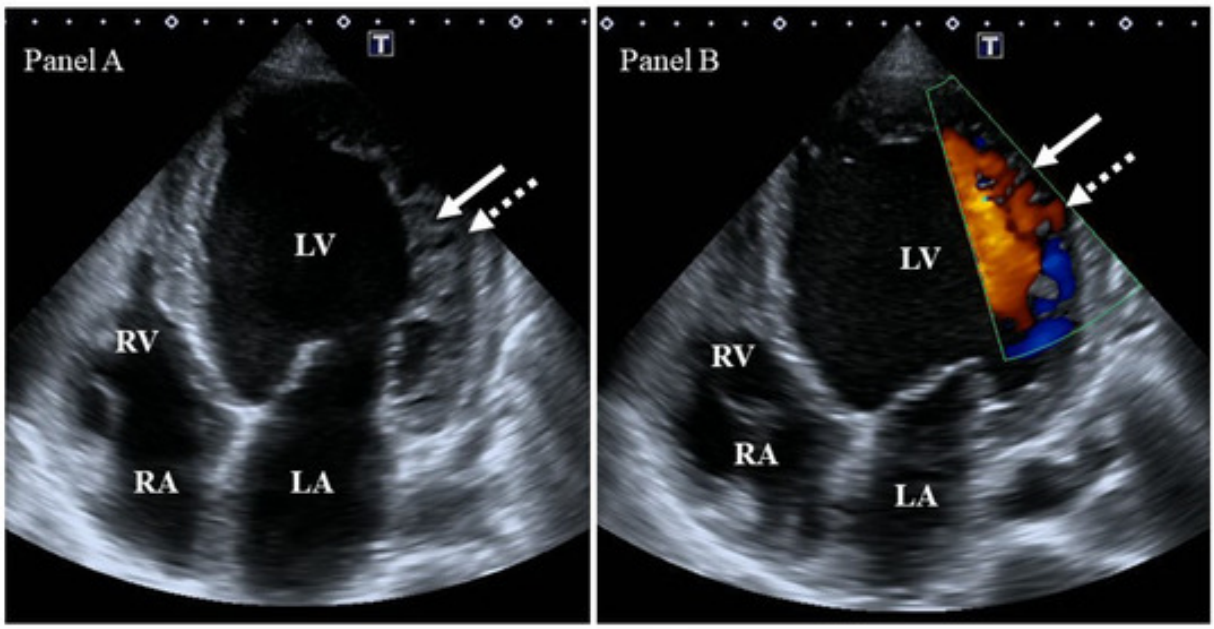
Tissue Doppler — reduced e'.

CLINICAL PEARL

Large atria with normal ventricles should always suggest restrictive cardiomyopathy.

5. Left Ventricular Non-Compaction (LVNC)

Prominent trabeculations, deep recesses, color Doppler entering recesses.



DIAGNOSTIC CRITERIA

Non-compacted : compacted myocardium ratio > 2 at end systole (Jenni criteria).

COMMON SITES

- Apex
- Inferior wall
- Lateral wall

ASSOCIATED FINDINGS

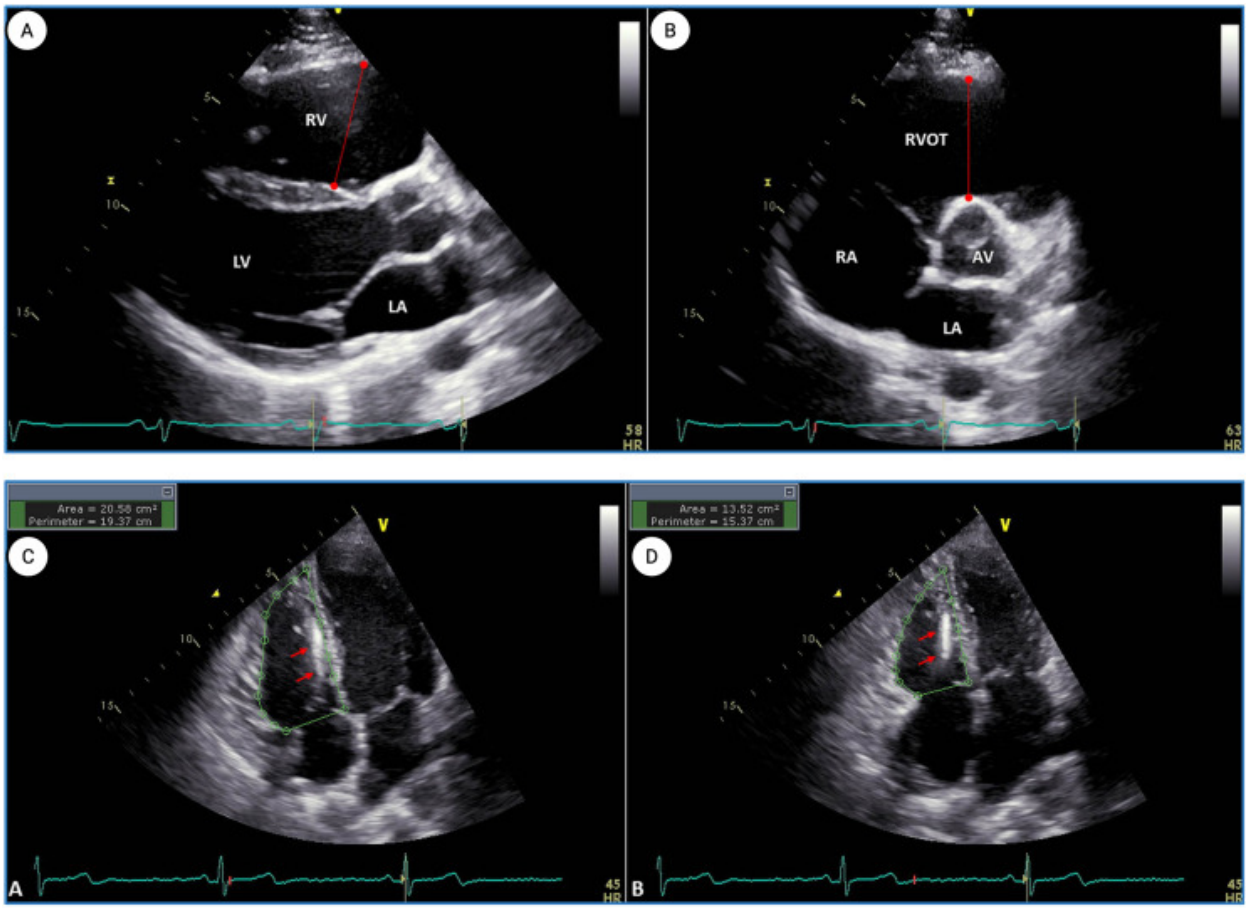
- LV dysfunction
- MR
- Thrombus
- Arrhythmias

6. Arrhythmogenic Cardiomyopathy

Usually affects: right ventricle.

ECHO FINDINGS

- RV dilation
- Regional akinesia
- RV aneurysm
- Reduced TAPSE
- RV dysfunction



ASSESSMENT OF DIASTOLIC FUNCTION

Always evaluate: mitral inflow, pulmonary vein flow, tissue Doppler, LA volume, E/e'.

ASSESSMENT OF RIGHT VENTRICULAR FUNCTION

Measure: TAPSE, FAC, S', RV strain.

MYOCARDIAL STRAIN IMAGING

Global longitudinal strain detects dysfunction before EF falls.

Normal pediatric GLS: approximately −20%.

Reduced strain indicates early myocardial disease.

THROMBUS ASSESSMENT

Always inspect: LV apex, LA appendage (if visible), RV apex.

DIFFERENTIAL DIAGNOSIS

Disease	Echo Finding
Myocarditis	Dilated LV + edema + variable EF
Anomalous coronary artery	Regional dysfunction
Severe Aortic Stenosis	Pressure overload hypertrophy
Endocardial fibroelastosis	Bright endocardium
Athlete's heart	Symmetric hypertrophy with normal diastolic function

7. Practical Reporting Template

Indication: Evaluation of ventricular dysfunction.

Findings

- Situs normal.
- AV and VA concordance.
- LV markedly dilated (LVEDD Z-score +4.2).
- Global hypokinesia.
- EF 28%.
- Mild functional mitral regurgitation.
- No LV thrombus.
- RV systolic function preserved (TAPSE normal).
- No pericardial effusion.

Impression: Severe dilated cardiomyopathy with markedly reduced systolic function. No intracardiac thrombus. Findings correlate with advanced heart failure.

DIAGNOSTIC PITFALLS

- Calling myocarditis "idiopathic DCM" without clinical correlation.
- Measuring EF from one view only.
- Missing LV thrombus.
- Ignoring RV function.
- Forgetting tissue Doppler.
- Not using Z-scores in children.
- Confusing physiological hypertrophy with HCM.

8. High-Yield Echo Pearls

- Dilated LV + poor EF = Think DCM.
- Thick septum + SAM = Think HCM.
- Huge atria + normal ventricles = Think RCM.
- Apical trabeculations + deep recesses = Think LVNC.
- RV dilation + regional dysfunction = Think arrhythmogenic cardiomyopathy.
- Always assess both systolic and diastolic function.
- Search carefully for intracardiac thrombus in severe ventricular dysfunction.

Cardiac Tumors in Children

INTRODUCTION

Primary cardiac tumors in children are rare, with an estimated incidence of **0.001–0.03%** in autopsy series. Unlike adults, where metastatic tumors predominate, **most pediatric cardiac tumors are benign**.

Many are detected prenatally or during infancy because of: heart murmur, arrhythmia, heart failure, cyanosis, incidental echocardiographic findings.

Echocardiography is the **first-line imaging modality** for diagnosis, defining tumor location, size, hemodynamic impact, and guiding treatment.

CLASSIFICATION OF PEDIATRIC CARDIAC TUMORS

Tumor	Frequency	Typical Age	Common Location
Rhabdomyoma	Most common	Fetal/Infancy	Ventricles
Fibroma	Second most common	Infancy/Childhood	LV free wall or septum
Myxoma	Rare	Older children	Left atrium
Teratoma	Neonates	Pericardium	Base of heart
Hemangioma	Rare	Any age	Anywhere
Papillary Fibroelastoma	Rare	Older children	Valves
Malignant Tumors (Sarcoma, Lymphoma)	Very rare	Variable	Any chamber

STEPWISE ECHOCARDIOGRAPHIC APPROACH

Step 1 — Where is the mass located?

↓

Step 2 — Single or multiple?

↓

Step 3 — Attached to: valve? septum? pericardium?

↓

Step 4 — Is it mobile or fixed?

↓

Step 5 — Does it obstruct blood flow?

↓

Step 6 — Does it cause valve dysfunction?

↓

Step 7 — Is there pericardial effusion?

↓

Step 8 — Could this be thrombus rather than tumor?

1. Rhabdomyoma

Rhabdomyoma is the **most common pediatric cardiac tumor**, accounting for approximately 50–60% of cases.

It is a benign hamartomatous tumor arising from cardiac muscle.

CLINICAL ASSOCIATIONS

The most important association: **Tuberous sclerosis complex**.

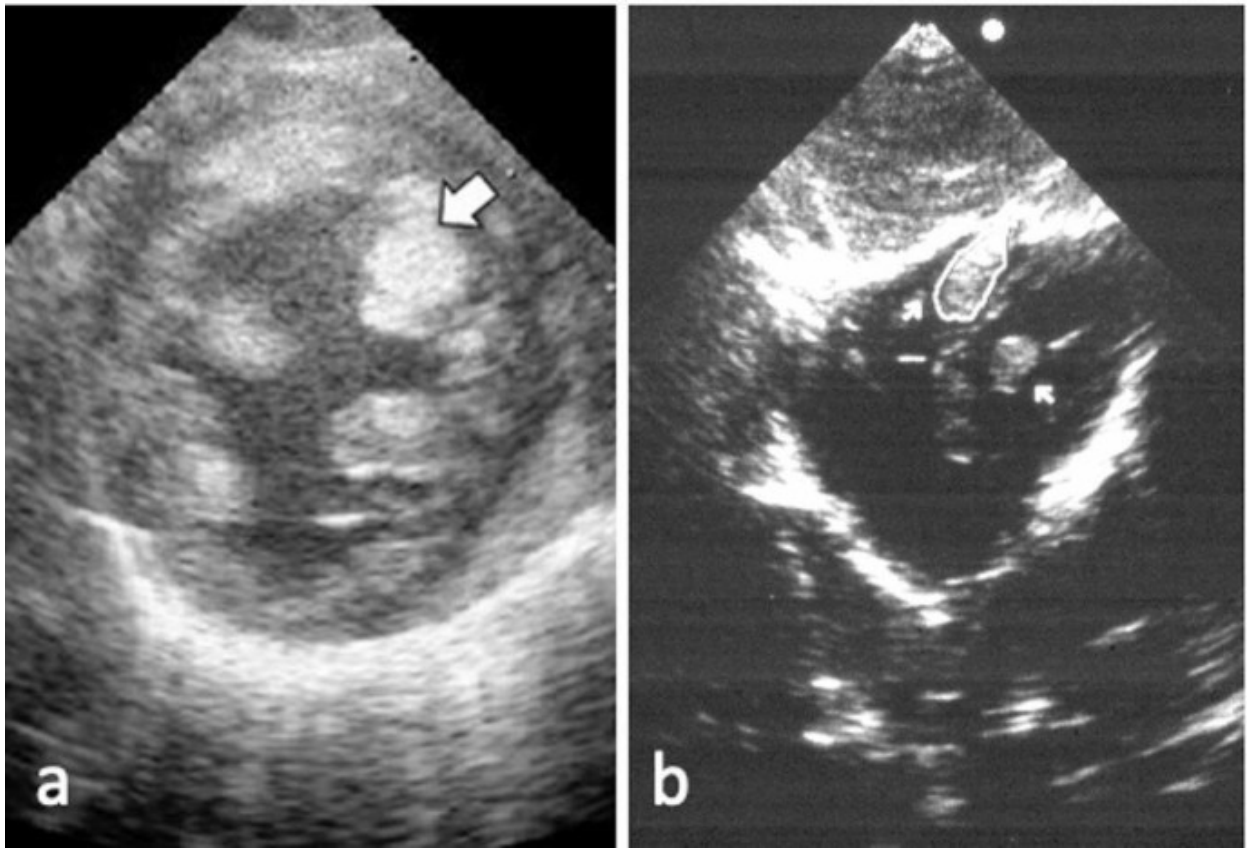
Approximately 70–90% of children with multiple rhabdomyomas have tuberous sclerosis.

COMMON LOCATIONS

- Ventricular septum
- LV free wall
- RV free wall
- Multiple ventricular masses

ECHO FINDINGS — APPEARANCE

- Multiple
- Well circumscribed
- Homogeneous
- Highly echogenic
- Intramyocardial



Common Pattern — numerous "white balls" scattered throughout both ventricles.

Doppler Assessment — evaluate for LVOT obstruction, RVOT obstruction, valve interference, reduced ventricular filling.

CLINICAL PEARLS

Most rhabdomyomas regress spontaneously during infancy. Surgery is reserved for severe obstruction, refractory arrhythmias, heart failure.

2. Fibroma

Fibroma is the second most common pediatric cardiac tumor.

Unlike rhabdomyoma, fibromas do **not regress**.

Location — usually solitary. Most common sites: LV free wall, interventricular septum.

Echo Findings — large, homogeneous, bright echogenic mass, well-defined margins. Often causes ventricular distortion.

Clinical Features — children may present with: ventricular tachycardia, heart failure, sudden cardiac death.

Echo Clues — solitary, large, intramural, calcification may be present.

3. Myxoma

Myxoma is the most common primary cardiac tumor in adults but is uncommon in children.

Typical Location — left atrium. Attached to: interatrial septum, near the fossa ovalis.

Echo Findings — pedunculated, mobile, gelatinous appearance, moves through mitral valve during diastole.

Doppler Findings — may produce mitral inflow obstruction, transmitral gradient.

Clinical Features — children may present with: syncope, stroke, embolism, constitutional symptoms.

CLINICAL PEARL

A mobile atrial mass attached to the interatrial septum is a myxoma until proven otherwise.

4. Teratoma

A congenital germ-cell tumor usually arising from the pericardium.

Echo Findings — large extracardiac mass, mixed echogenicity, cystic areas, calcification, large pericardial effusion.

5. Malignant Cardiac Tumors

Very uncommon in children.

Examples: sarcoma, lymphoma, metastatic neuroblastoma.

Echo Findings — large, irregular, poorly defined, rapid growth, myocardial invasion, pericardial effusion, reduced ventricular function.

6. Differential Diagnosis — Not Every Intracardiac Mass Is a Tumor

Condition	Echo Clues
Thrombus	Avascular, often in areas of poor ventricular function or around central lines
Vegetation	Valve attachment, irregular, associated with infective endocarditis
Chiari network	Thin, highly mobile structure in RA
Eustachian valve	Normal anatomical variant
Moderator band	Normal RV structure
Lipomatous hypertrophy	Fatty septal thickening

ASSESSMENT OF HEMODYNAMIC EFFECTS

Always determine whether the tumor causes: LVOT obstruction, RVOT obstruction, AV valve obstruction, semilunar valve obstruction, inflow obstruction, pericardial tamponade, ventricular dysfunction.

ARRHYTHMIA ASSESSMENT

Tumors involving the septum may produce: complete heart block, ventricular tachycardia, supraventricular tachycardia, premature ventricular contractions.

Correlate with ECG and Holter monitoring.

ROLE OF ADVANCED IMAGING

When echocardiography is inconclusive:

Cardiac MRI — best for tissue characterization, myocardial infiltration, tumor extent.

Cardiac CT — useful for calcification, coronary artery relationships, surgical planning.

7. Practical Reporting Template

Indication: Evaluation of intracardiac mass.

Findings

- Situs solitus with normal AV and VA connections.
- A well-circumscribed, homogeneous echogenic mass measuring 24 × 18 mm is present within the interventricular septum.
- No significant LVOT or RVOT obstruction.
- Normal biventricular systolic function.
- No valvular dysfunction.
- No pericardial effusion.

- Findings are most consistent with cardiac rhabdomyoma.

Impression: Multiple intramyocardial echogenic masses consistent with rhabdomyomas. Recommend evaluation for tuberous sclerosis and clinical follow-up with serial echocardiography.

DIAGNOSTIC PITFALLS

- Mistaking thrombus for tumor.
- Missing small valve tumors.
- Ignoring Doppler evidence of obstruction.
- Failing to assess pericardial effusion.
- Not examining all cardiac chambers.
- Assuming every ventricular mass is a rhabdomyoma.
- Forgetting the association between multiple rhabdomyomas and tuberous sclerosis.

8. High-Yield Echo Pearls

- Multiple ventricular masses in an infant → Think rhabdomyoma.
- Solitary intramural LV mass → Think fibroma.
- Mobile left atrial pedunculated mass → Think myxoma.
- Pericardial mass with large effusion in a neonate → Think teratoma.
- Valve-attached, highly mobile lesion → Think papillary fibroelastoma.
- Always use Color Doppler to assess inflow and outflow obstruction.
- Evaluate ventricular function and rhythm disturbances in every patient with a cardiac tumor.

9. Summary

Primary cardiac tumors in children are rare but clinically significant. Most are benign, with **rhabdomyoma** being the most common and strongly associated with **tuberous sclerosis complex**. Echocardiography is the cornerstone of diagnosis, allowing precise assessment of tumor location, morphology, mobility, hemodynamic effects, ventricular function, and associated complications. A structured, stepwise echocardiographic approach helps distinguish different tumor types, guides further imaging when needed, and supports timely medical or surgical management.

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