

Multimodality Assessment of Congenital Coronary Anomalies

A Visual Clinical Atlas and Diagnostic Playbook

Distilled from the ASE Guidelines in
collaboration with SCAI, JSE, and SCMR.

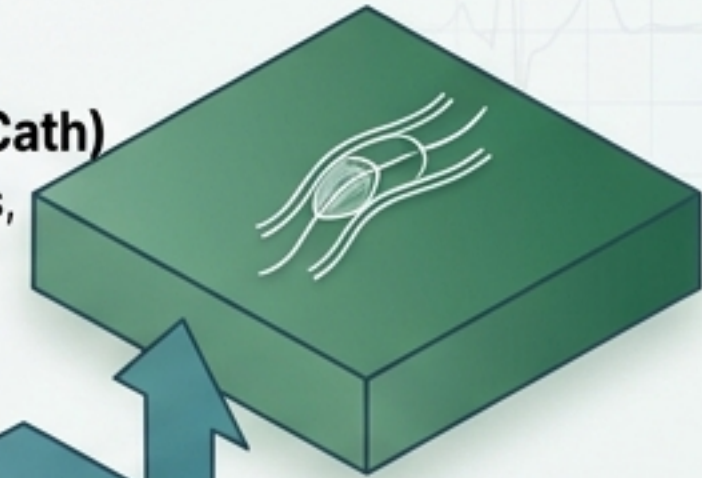
The Shift to Non-Invasive Multimodality Imaging



Prevalence of anomalous aortic origin associated with **sudden cardiac death** is estimated at 0.7%. **Prospective noninvasive screening** is now now the frontline defense.

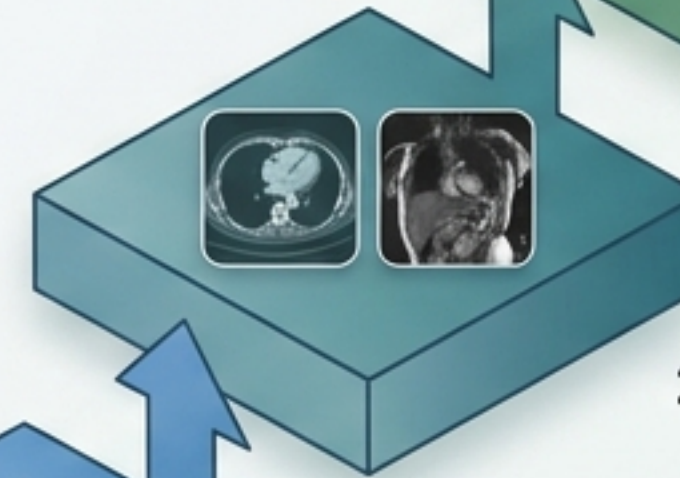
3. Interventional Planning: Cardiac Catheterization (Cath)

Precise pressure measurements, therapeutic planning only when necessary



2. Anatomical Confirmation: Cardiac CT (CCT) / Cardiac MR (CMR)

High-resolution 3D anatomy, dynamic function assessment























1. Universal Screening: Transthoracic Echocardiography (TTE)

Initial detection, non-invasive, widely available



The Modality Toolkit Matrix

	TTE	CMR	CCT	Cath
Spatial Resolution	 ++	 +++	 +++++	 +++++
Temporal Resolution	 +++	 ++	 ++	 +++
Evaluation of Ostia/Origin	 ++	 +++	 +++++	 +++
Distal Course/ Branches	 0	 ++	 +++	 +++++
Myocardial Ischemia	 ++	 +++	 0	 +++

Red Flag

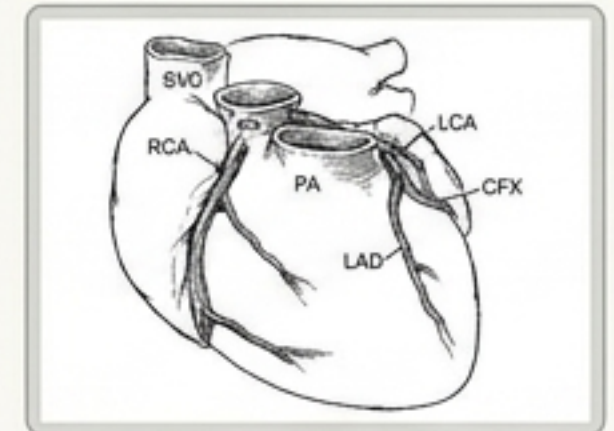
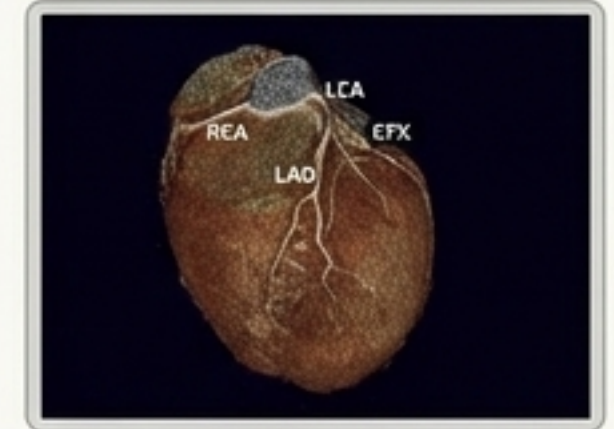
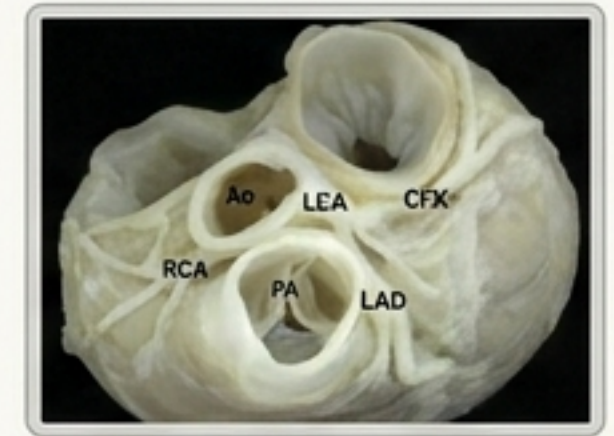
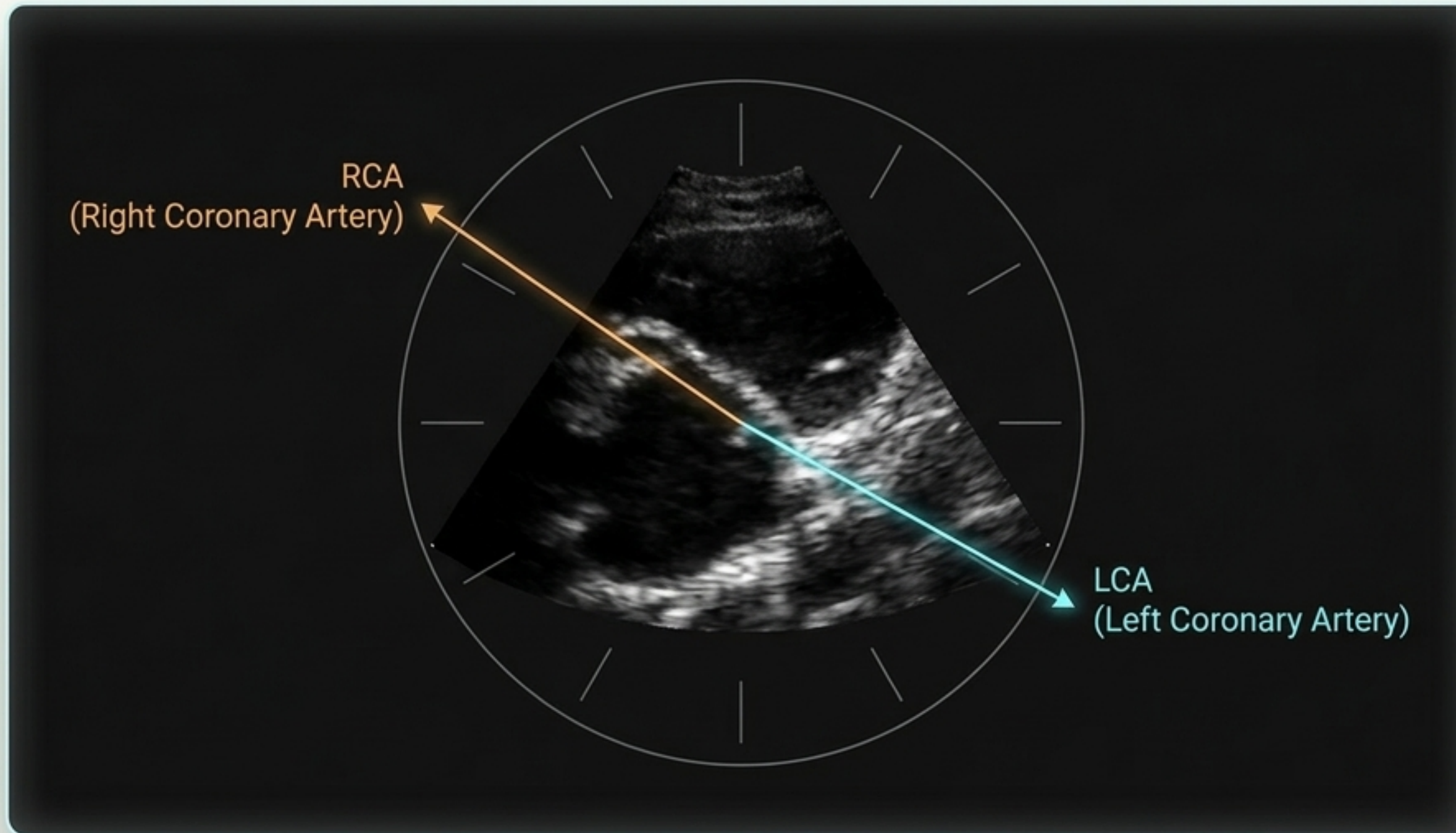
The Sedation/ Radiation Tradeoff

TTE: No radiation, moderate sedation need.

CCT: Radiation exposure, lower sedation need.

CMR: No radiation, high sedation need in pediatrics.

Defining the Baseline: The Normal Coronary Origin




Anatomical & CT References

Technical Spec Box: Optimal Imaging: Parasternal short-axis view, high-frequency transducer (4-12 MHz), lower Nyquist limit (20-40 cm/s) for low-velocity flow.

Clinical Presentation Matrix

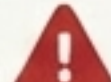
AAOCA

 **Primary Risk: Sudden Cardiac Death**

Diagnostic Hallmarks

✓ Oblique origin, intramural course.

ALCAPA

 **Primary Risk: Heart Failure / Dilated Cardiomyopathy**

Diagnostic Hallmarks

✓ Retrograde flow, RCA collaterals.

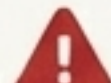
Coronary Fistulas (CAFs)

 **Primary Risk: Shunt volume overload / Steal ischemia**

Diagnostic Hallmarks

✓ Dilation, turbulent continuous flow.

SVAS-Associated

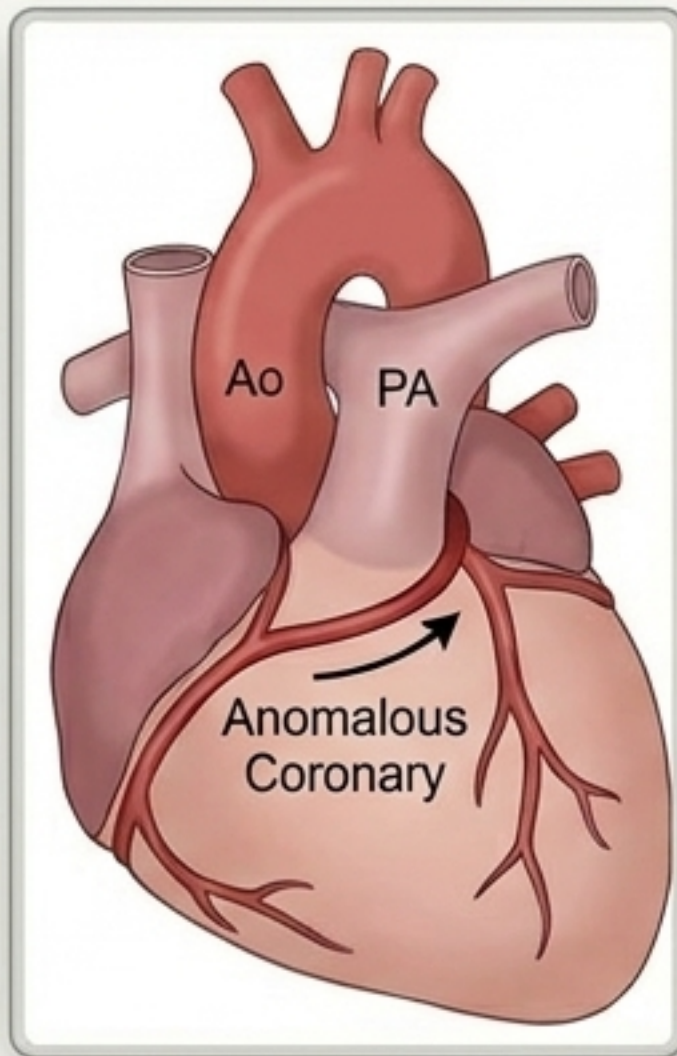
 **Primary Risk: Ostial stenosis / Anesthesia death**

Diagnostic Hallmarks

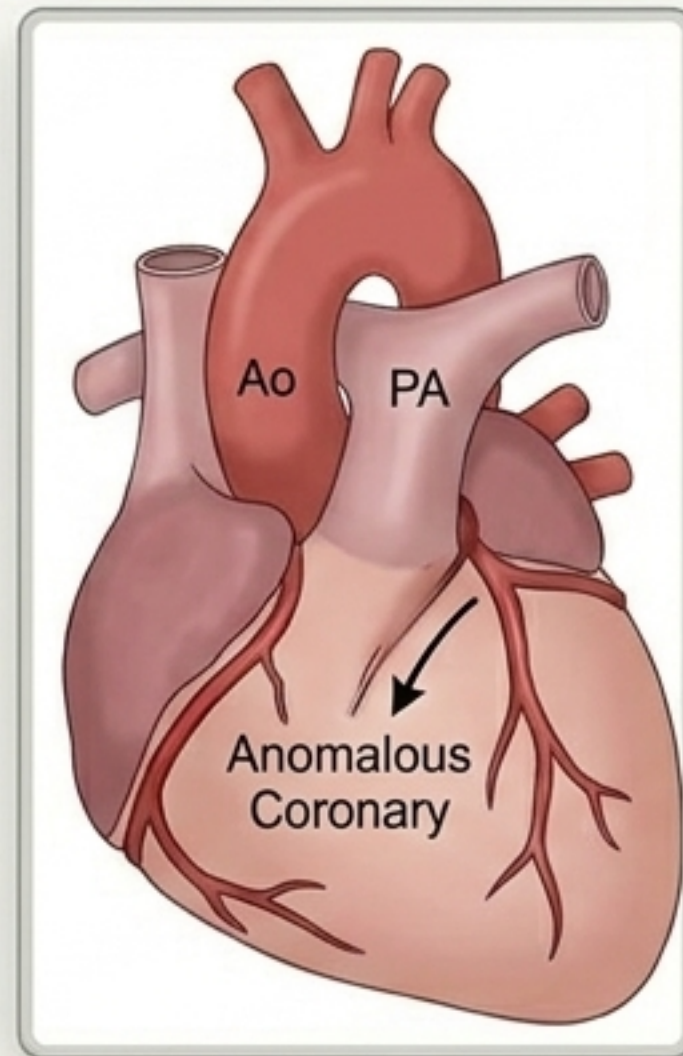
✓ Coronary hooding near sinotubular ridge.

AAOCA: Mapping Anomalous Aortic Origins

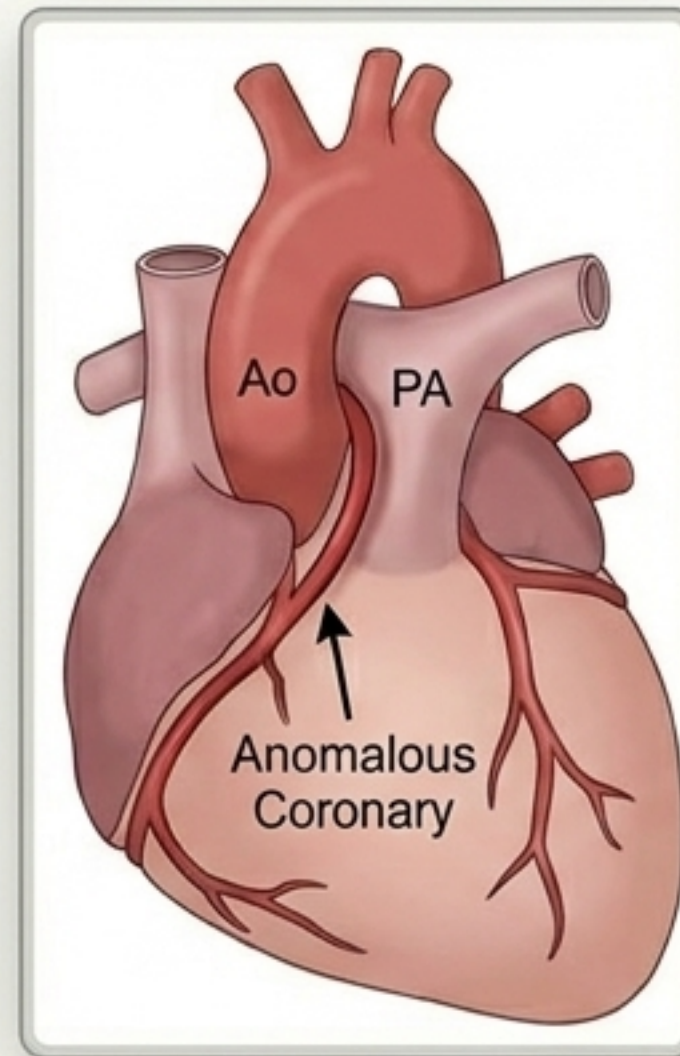
Anterior Course



Retro-aortic Course



Interarterial Course



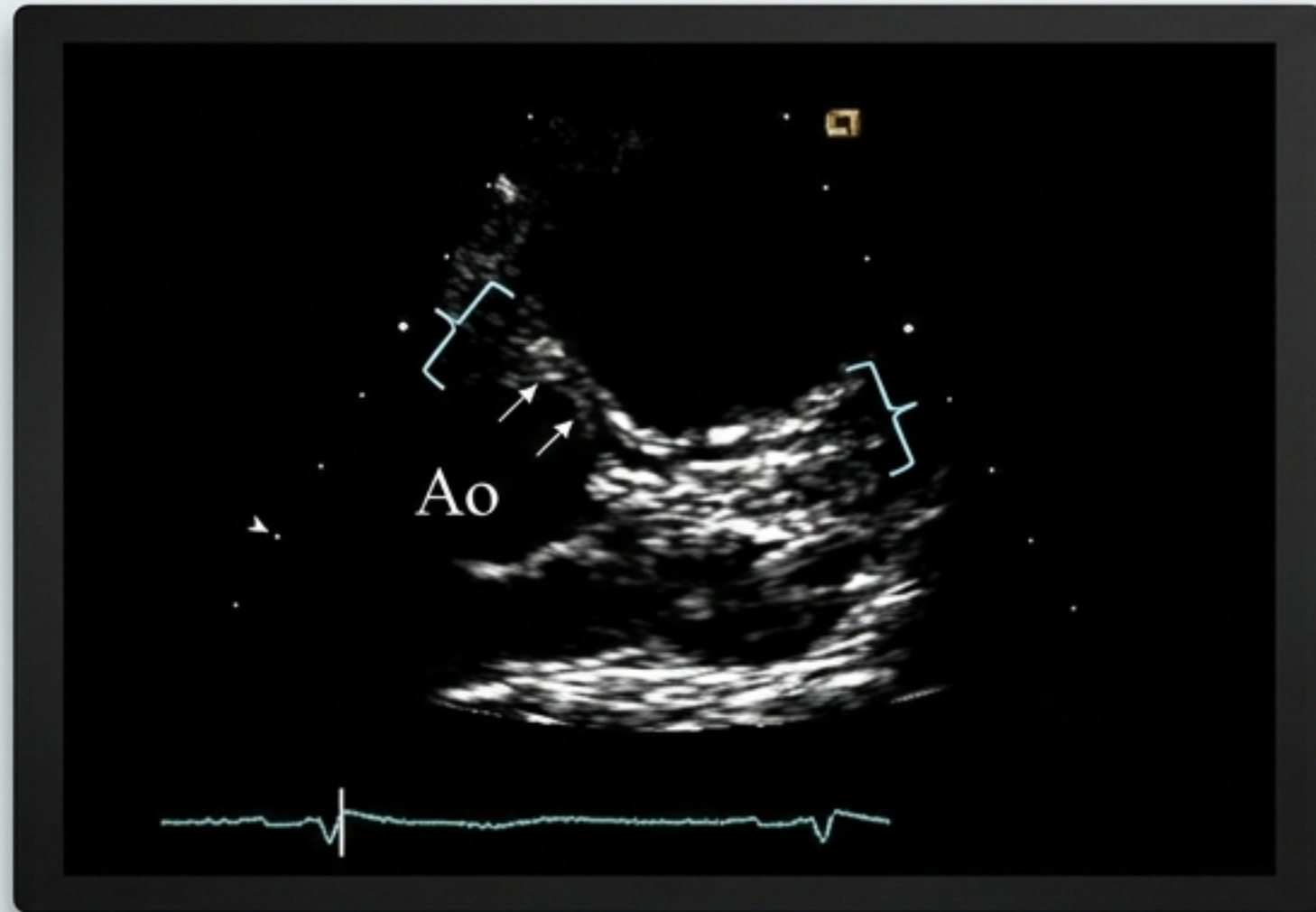
The Interarterial Danger Zone

Anomalies coursing between the aorta and pulmonary artery carry the highest risk for sudden death, especially during strenuous activity.

Etiology: ostial stenosis, acute angle takeoff, and lateral compression during increased cardiac output.

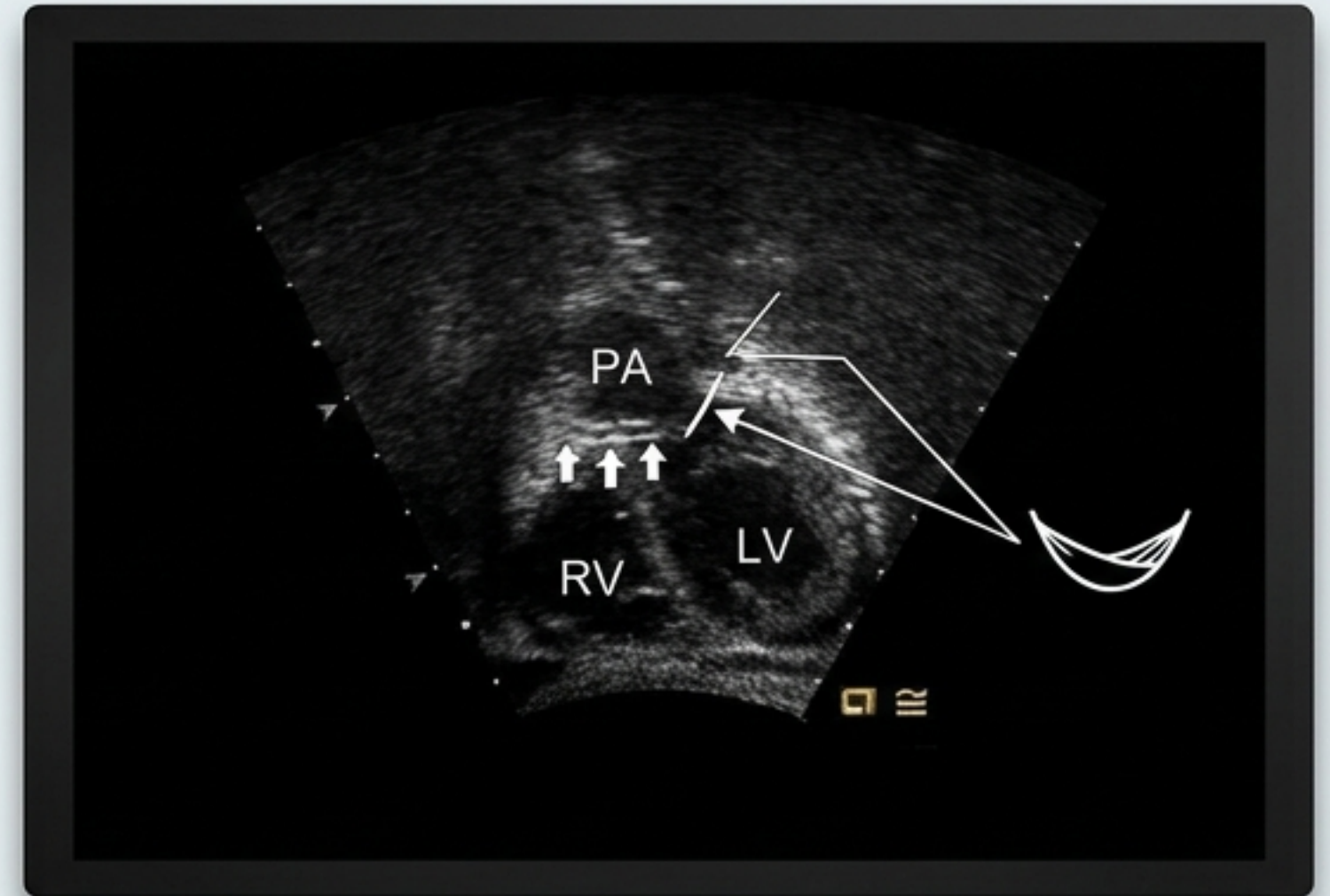
Interarterial Pathways: Intramural vs. Intraconal

Intramural Course



- Originates adjacent to commissure.
- Shares aortic tunica media.

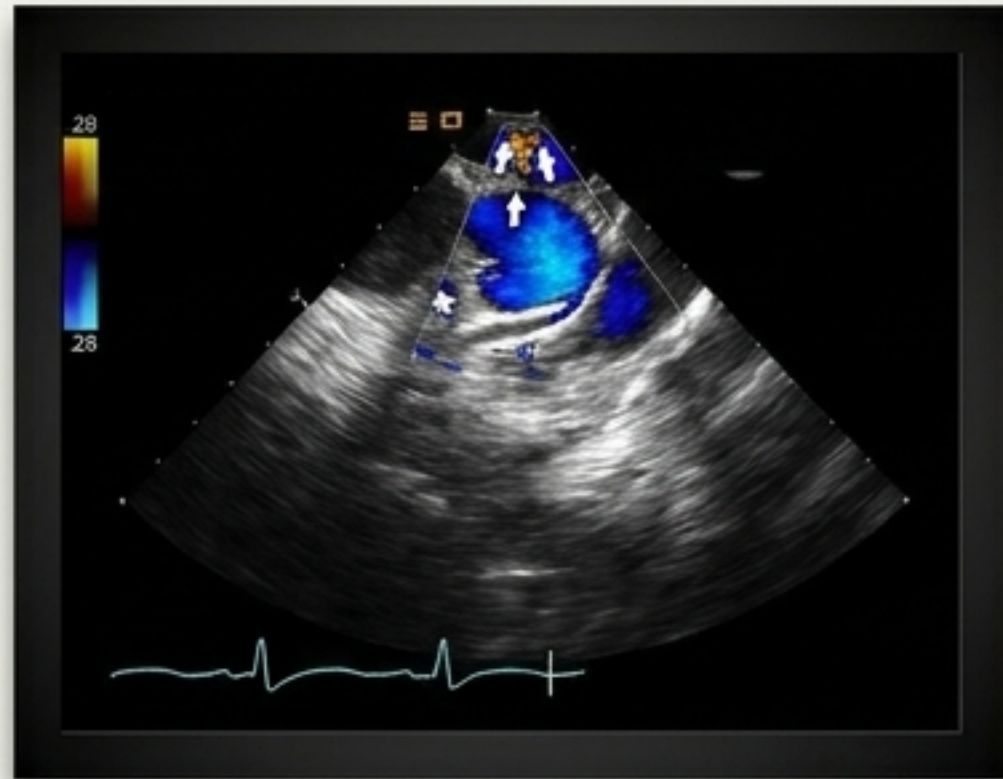
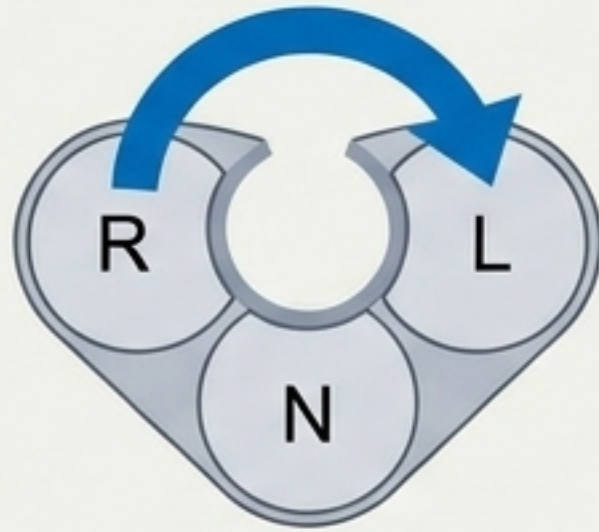
Intraconal Course



- Courses within the myocardial sulcus below the RVOT.
- Characterized by the "Hammock Sign".

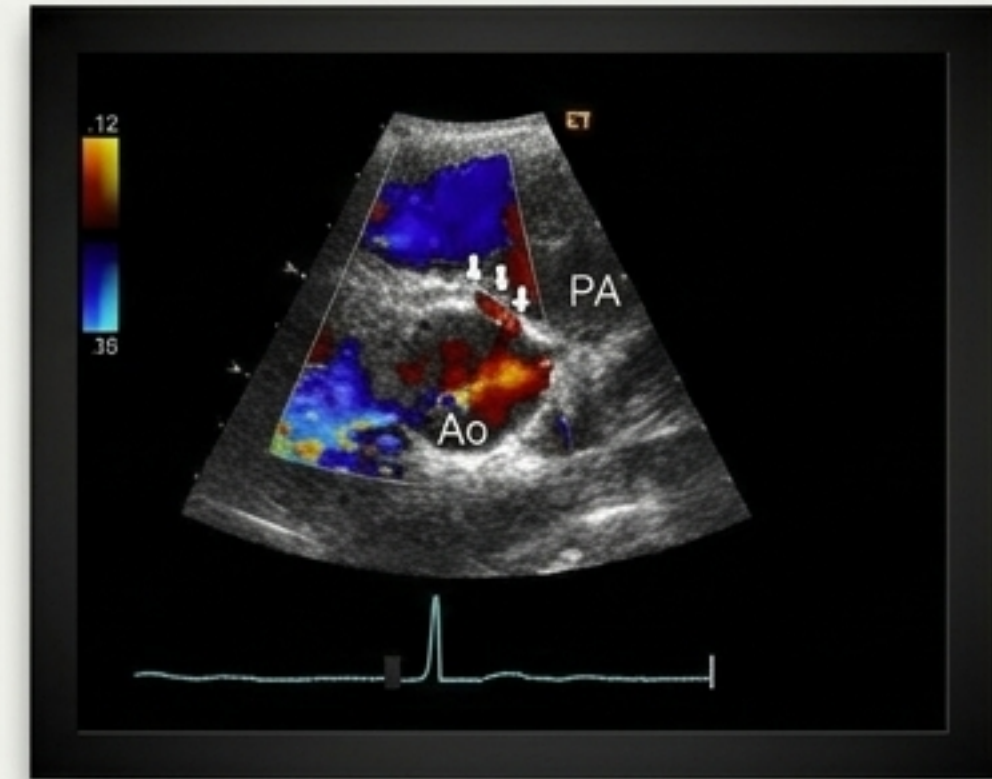
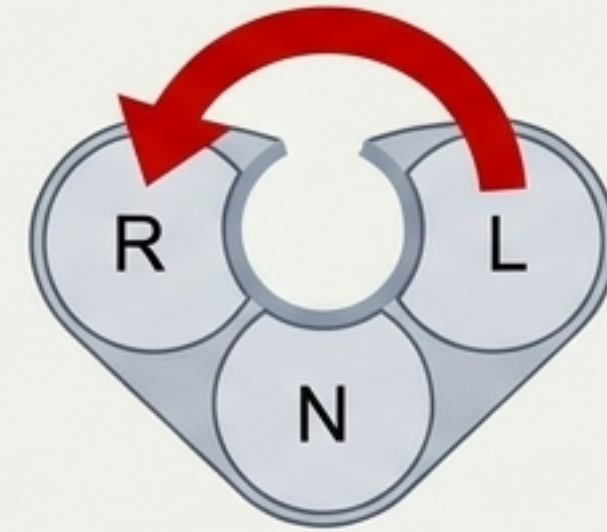
The Doppler Flow Reversal Schematic

AAOLCA (Left from Right Sinus)



Flow moving away from the right sinus toward the posterior left sinus. Color Doppler = BLUE.

AAORCA (Right from Left Sinus)



Flow moving toward the anterior right sinus from the left sinus. Color Doppler = RED.

AAOCA Imaging Strategy

Step 1: Universal Screen

Utilize TTE. Look for oblique origins and interarterial courses.



Step 2: Anatomical Confirmation

If interarterial AAOCA is identified, trigger CCT or CMR to delineate ostial origin, size, and precise intramural length.

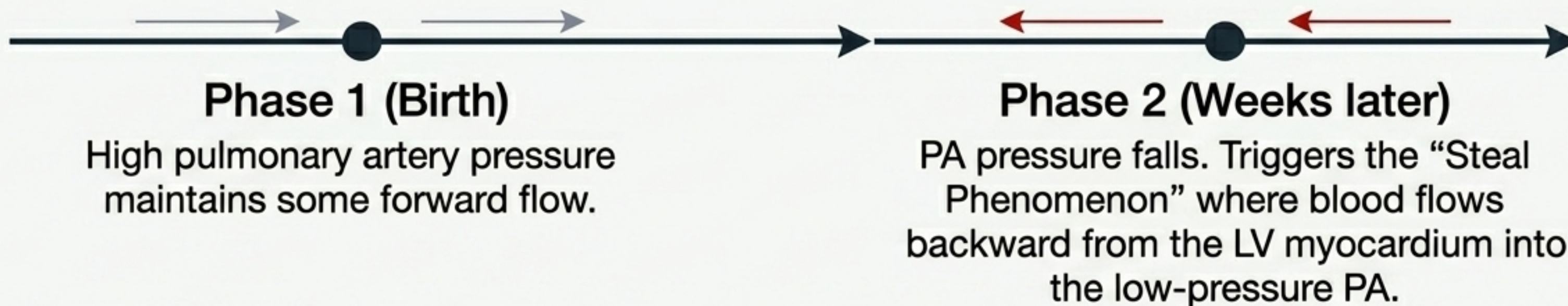


Step 3: Functional Assessment

Utilize resting and stress myocardial perfusion to identify ischemia or correlate symptoms.

ALCAPA: The Physiology of Ischemia

Transitional Circulation



The 90% Mortality Cliff

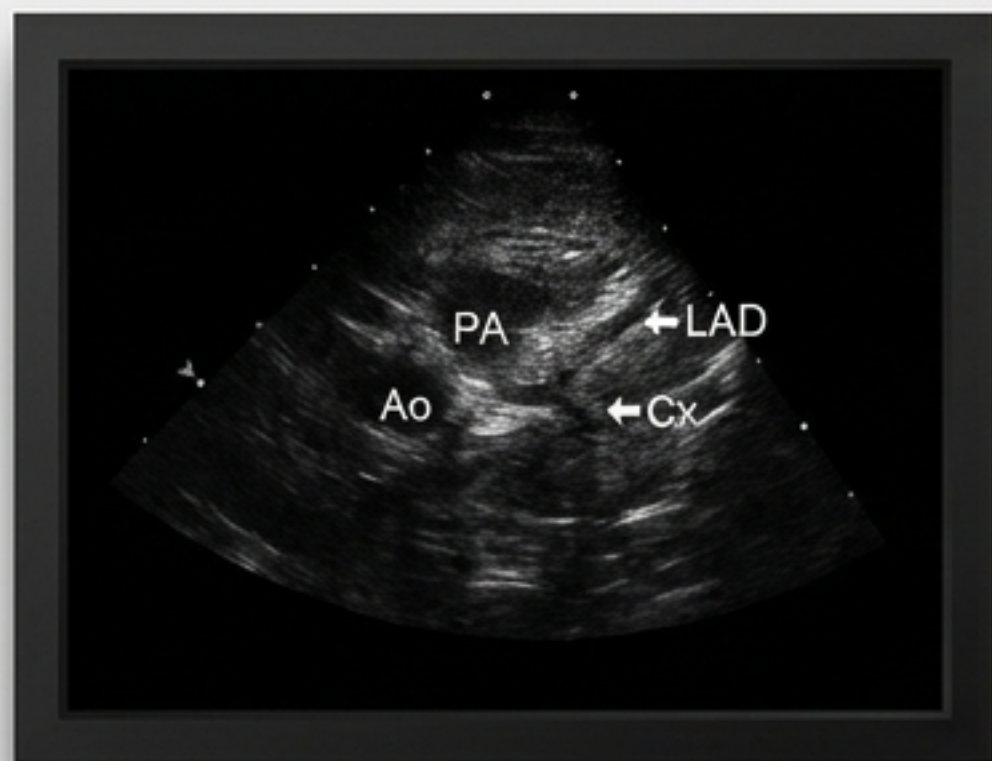
Infants with inadequate RCA collaterals develop profound ischemia, papillary muscle fibrosis, severe mitral regurgitation, and dilated cardiomyopathy. Without repair in year one, mortality is 90%.

Diagnostic Hallmarks of ALCAPA

1. The Origin (2D)



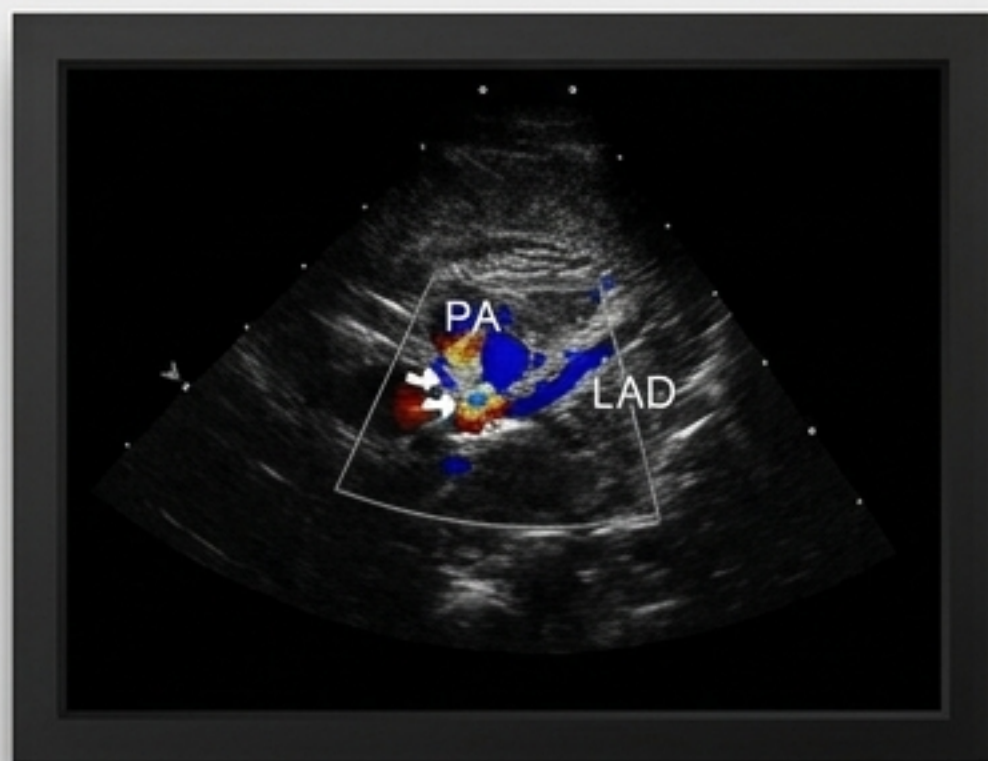
Direct visualization of LCA arising from posterior PA.



2. The Flow (Color Doppler)



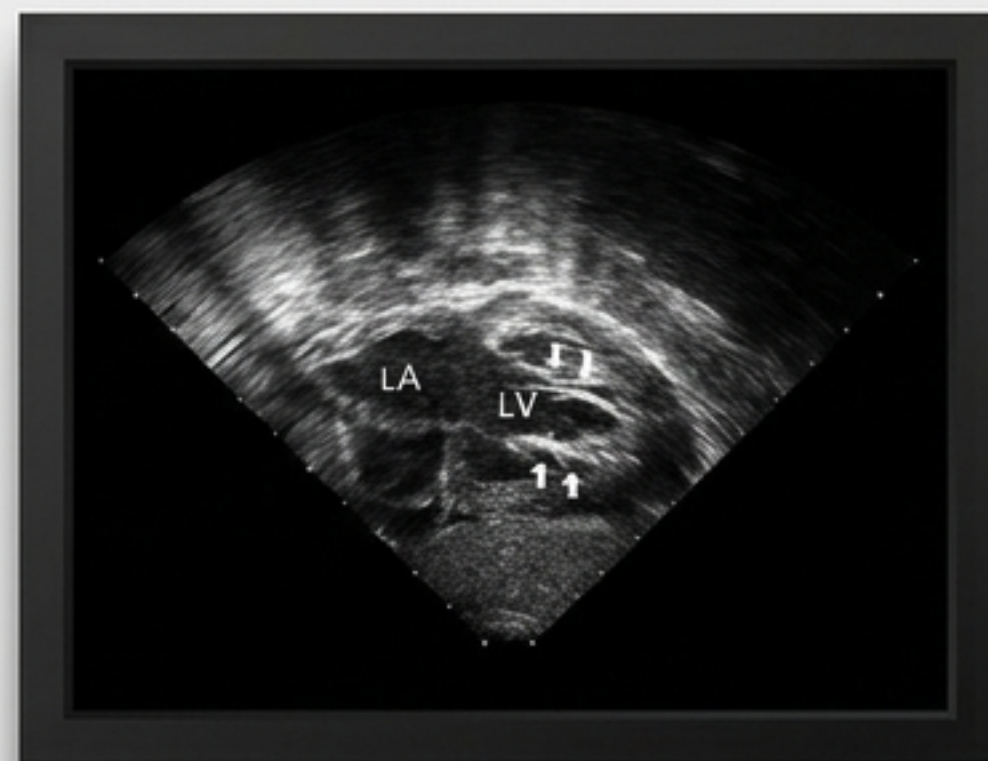
Retrograde flow into the PA. Emphasize abnormal BLUE signal moving away from aortic root.



3. The Compensation (Morphology)



Massive RCA dilation and prominent intramyocardial septal collaterals.



False dropouts can mimic normal LCA origin. Never rely on 2D still frames in isolation; always confirm flow direction.

ALCAPA Imaging Strategy

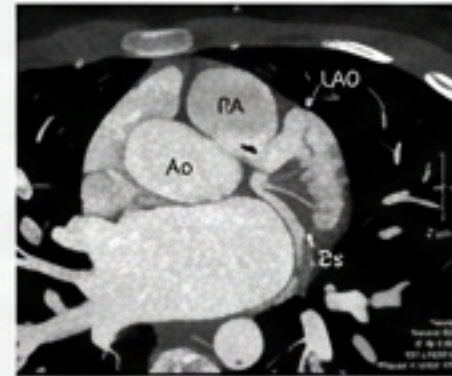
Step 1: Primary Diagnosis

TTE is the primary screening tool and is usually definitively diagnostic in infants.



Step 2: Complex Confirmation

Reserve CCT or CMR for cases with unclear anatomy or associated congenital heart disease.

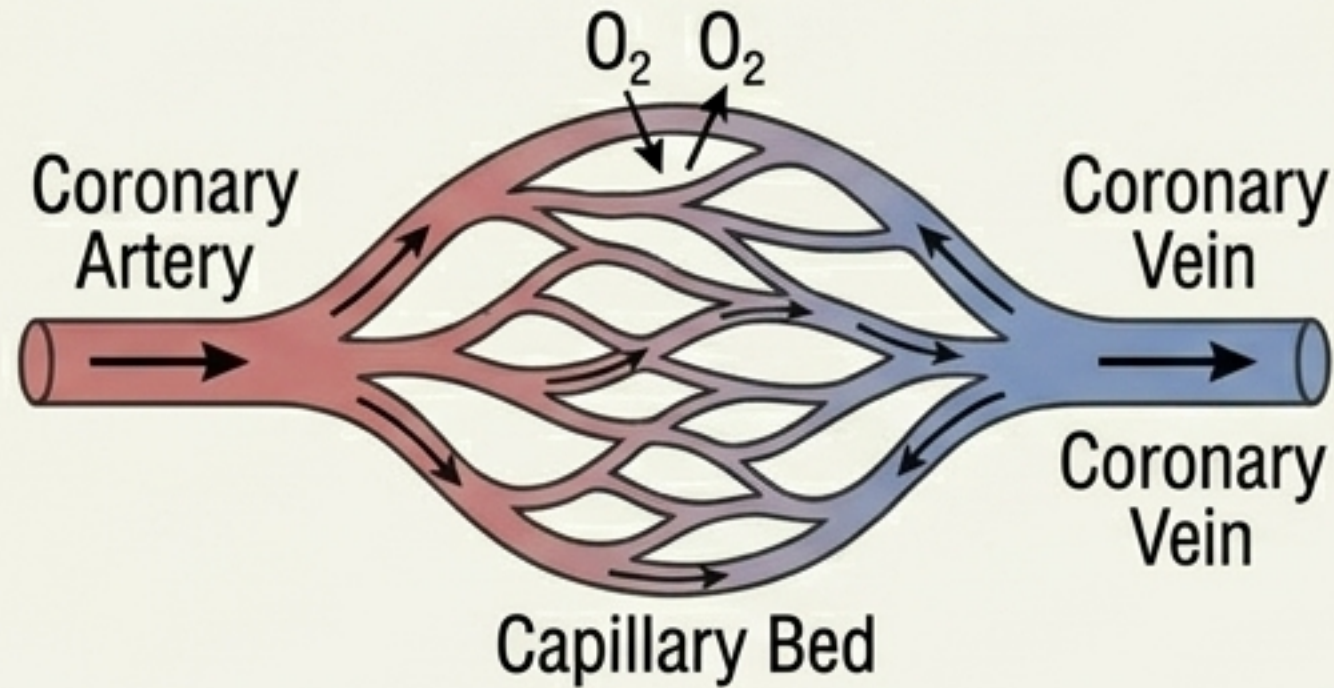


Step 3: Post-Op Monitoring

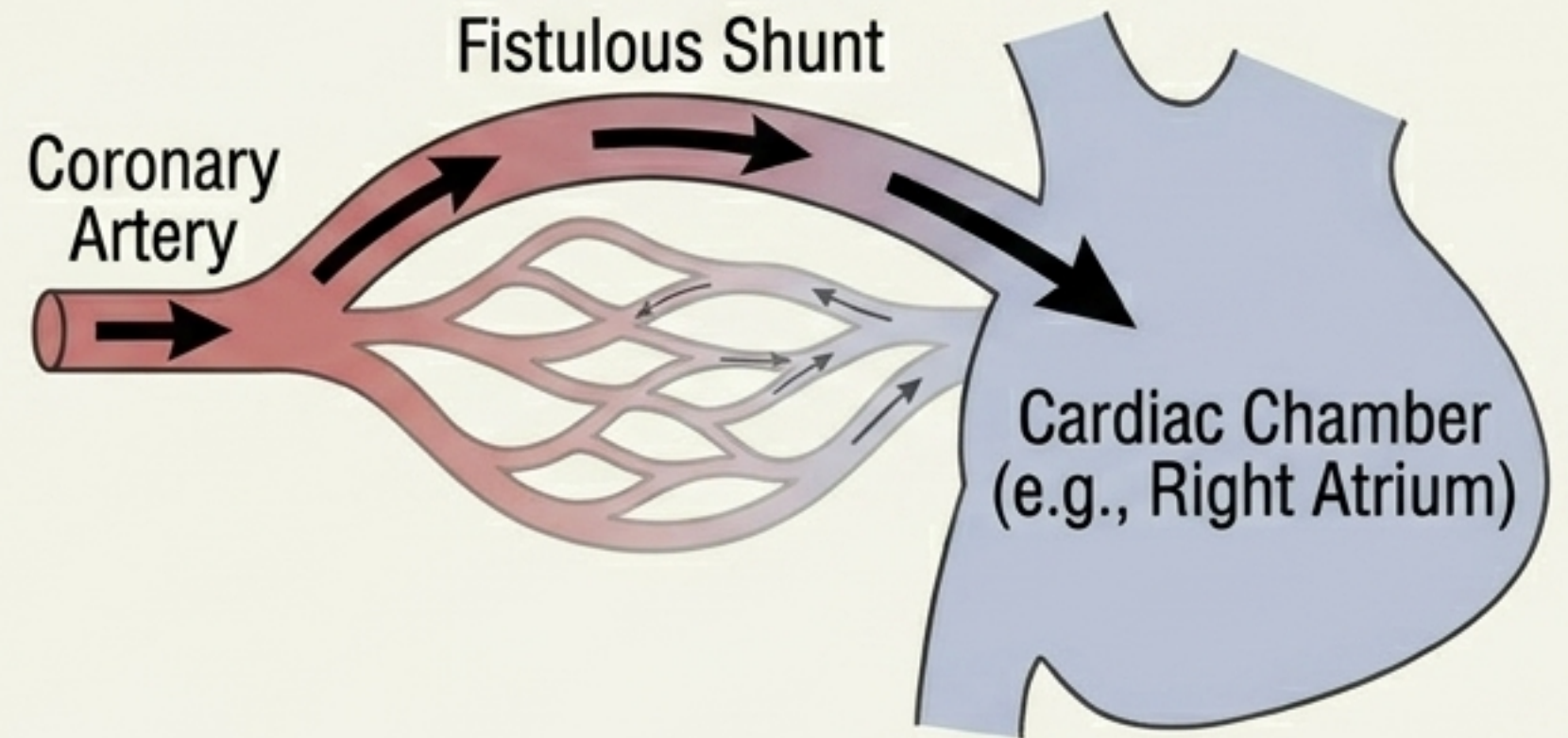
TTE for remodeling; CCT/CMR for residual stenosis, thrombosis, or perfusion defects.

The Mechanics of Coronary Artery Fistulas

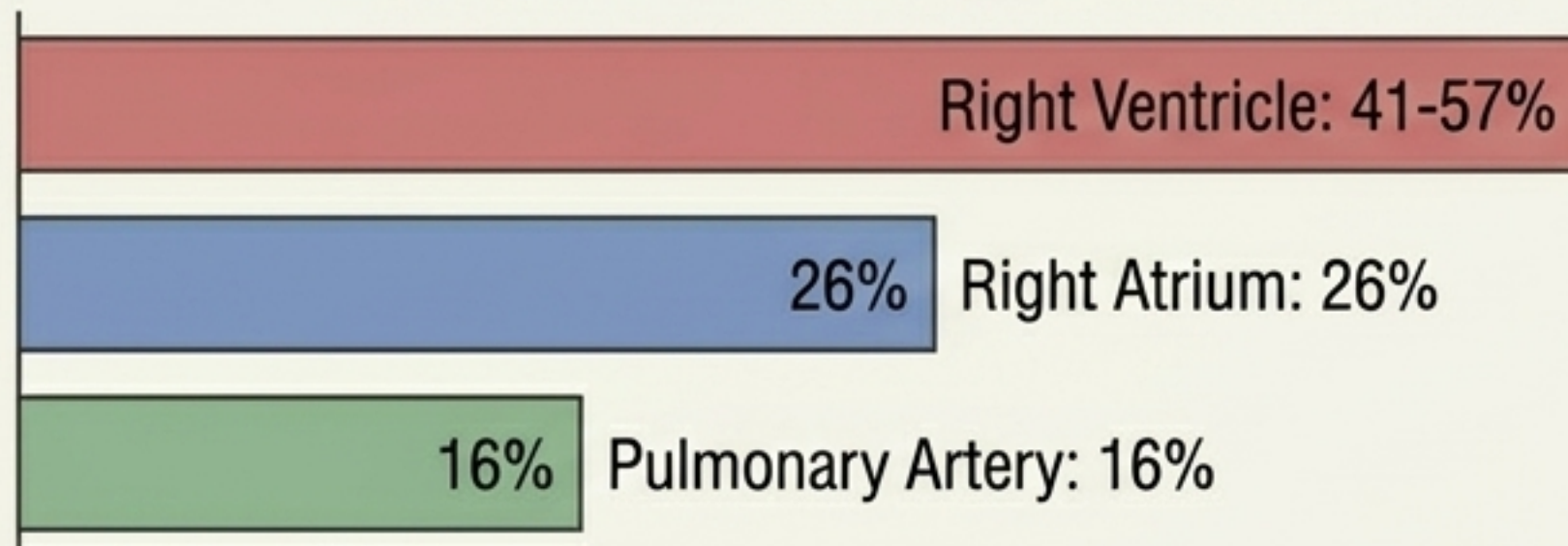
Normal Capillary Bed Flow



Fistulous "Bypass" Shunt



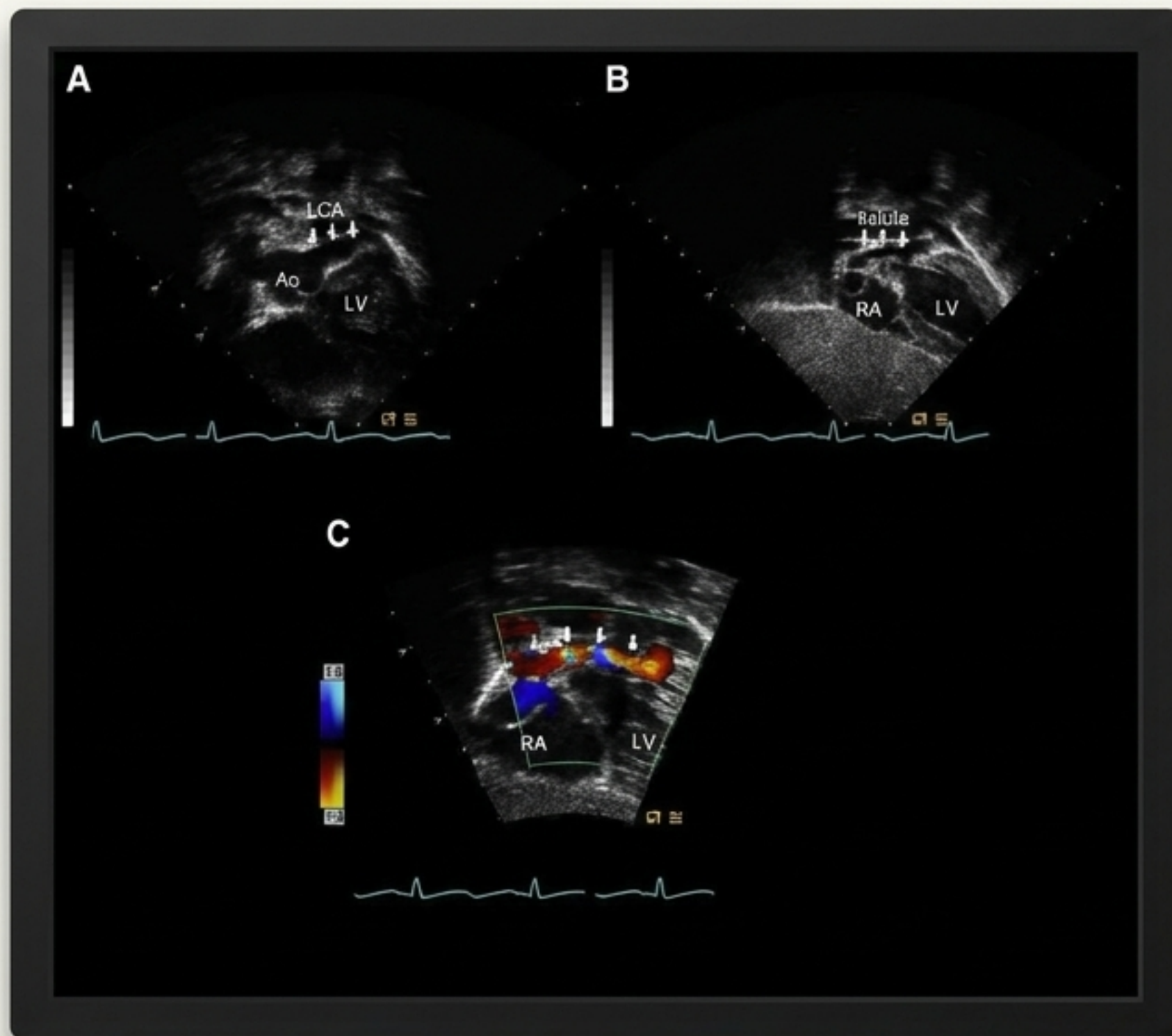
Most Common Termination Sites



Risk Profile

Structural Definition & Consequence:
Large shunts lead to volume overload, steal ischemia, and progressive aneurysmal dilation by bypassing the myocardial capillary bed.

Identifying and Monitoring CAFs



Diagnostic Rules of Thumb

Low-Pressure Exits (RA, RV, Coronary Sinus)

Produces a continuous, high-velocity turbulent flow signal.

High-Pressure Exits (LV)

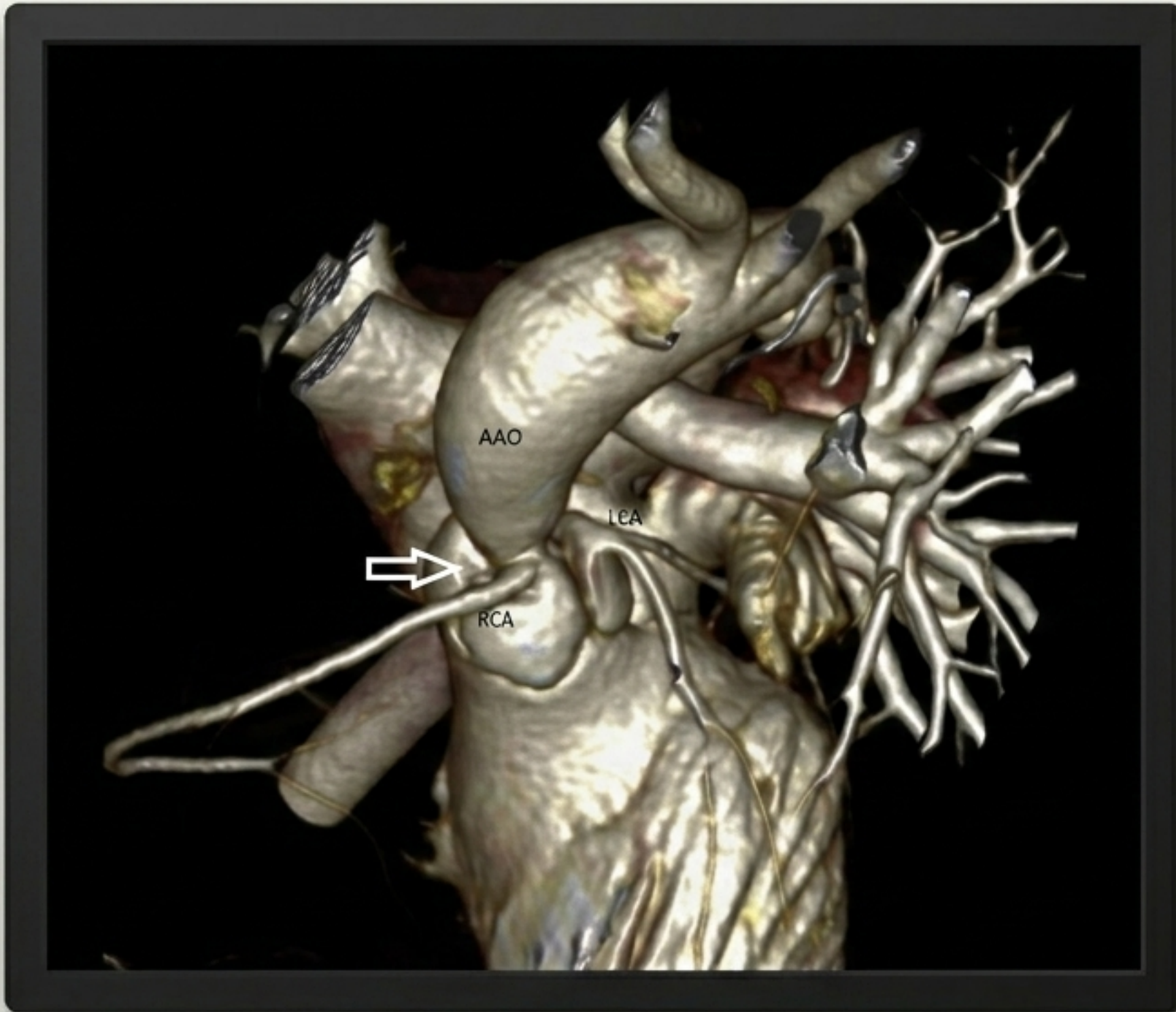
Shunt flow primarily restricted to diastole.

Post-Closure Alert

The Ectasia Risk

After surgical or device closure, residual dilation poses a high risk for stagnant blood flow and thrombosis. Continuous monitoring is required.

SVAS: Ostial Stenosis and Coronary Hooding



Pathology Breakdown

- Thickened sinus wall.
- Fusion of aortic leaflet edge to the sinotubular ridge (“Coronary Hooding”).
- Pre-stenotic high-pressure dilation.

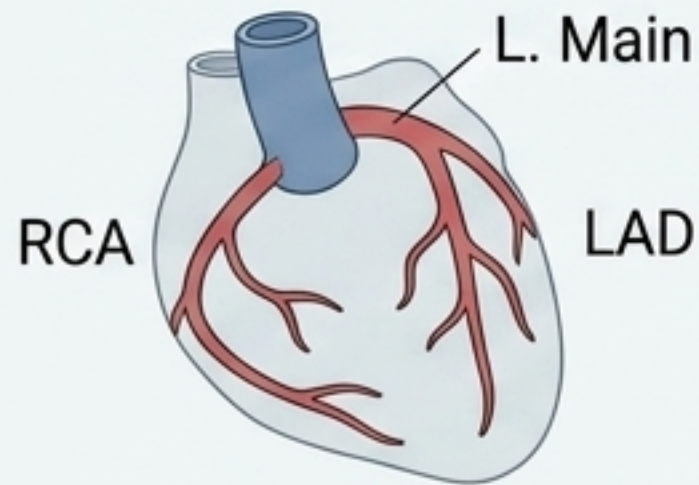
The Sedation Trap

Deep sedation or general anesthesia carries a massive risk of sudden cardiac death in SVAS patients (up to 79% of deaths linked to ischemia under anesthesia). This drastically limits the use of CMR and pushes TTE and fast-acquisition CCT to the forefront.

TGA: Anatomic Coronary Variations Map

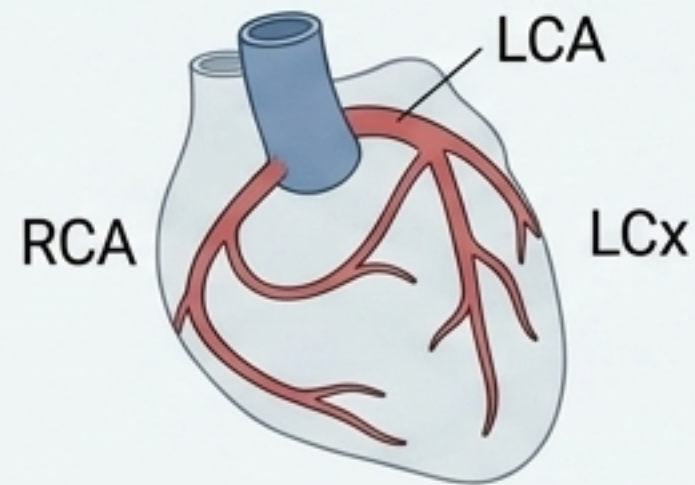
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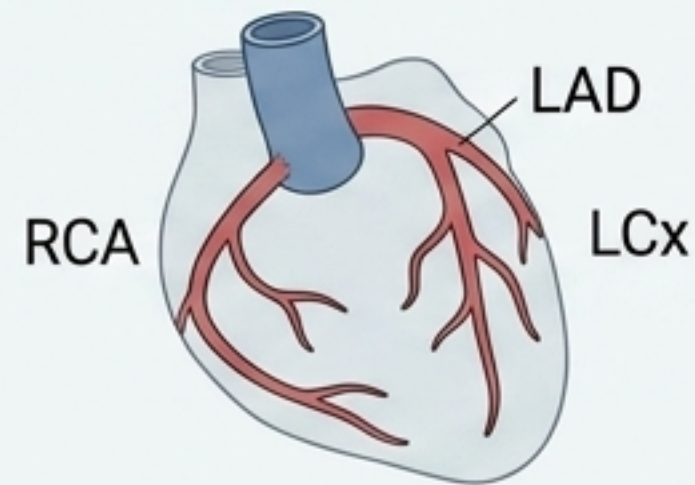
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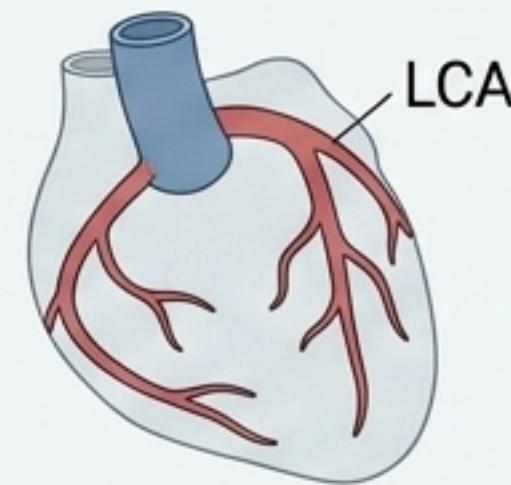
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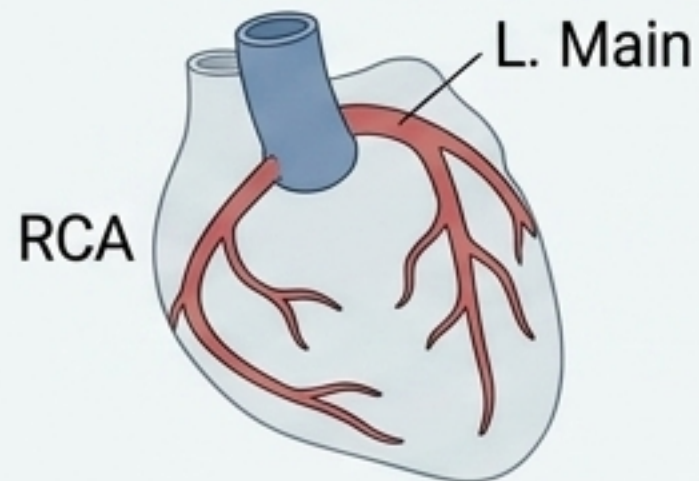
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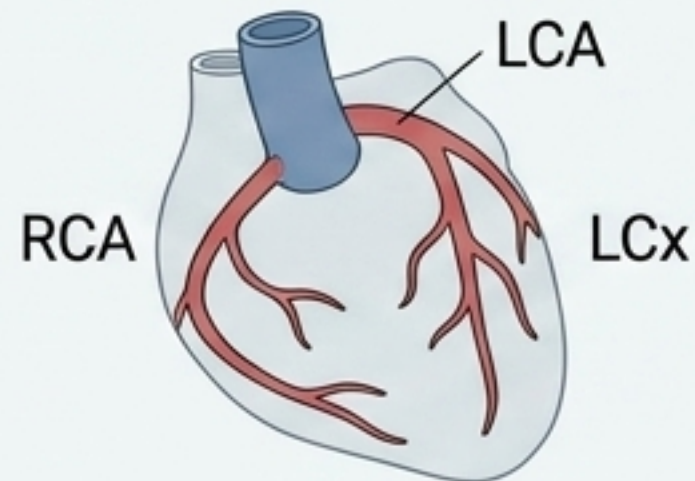
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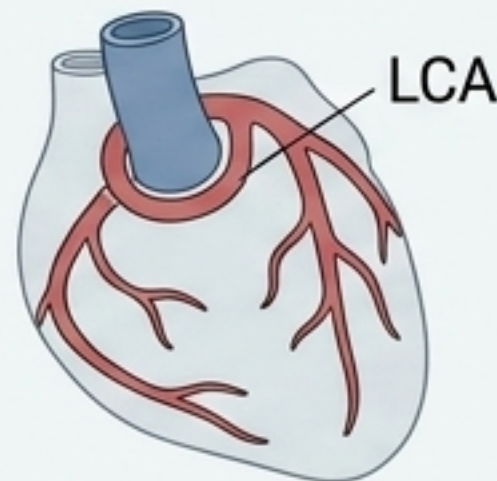
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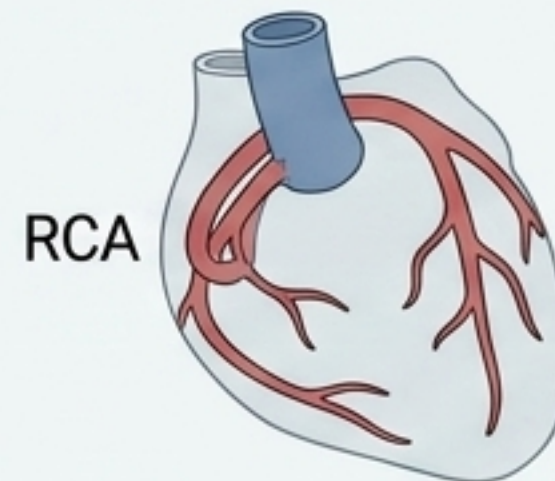
3%

G



0.75%

H



Context Note

Context: Accurate preoperative mapping of these variations is critical for the success of the Arterial Switch Operation, and CCT is highly effective for post-op surveillance of neo-aortic roots.

The Unified Imaging Decision Tree

