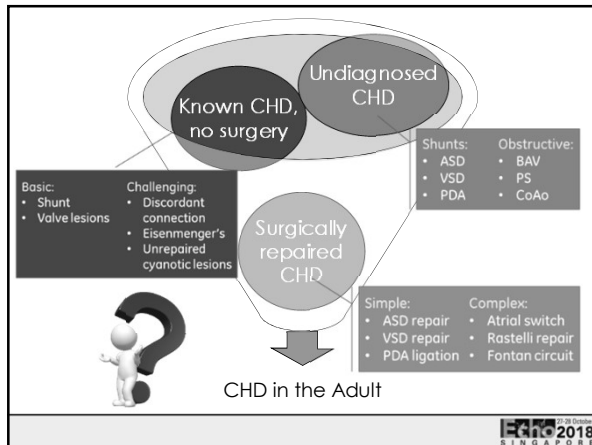


Tips and Tricks for Assessing Adult Congenital Heart Disease

Bonita Anderson

DMU (Cardiac), MAppISc (Med Ultrasound), ACS, AMS, FASE





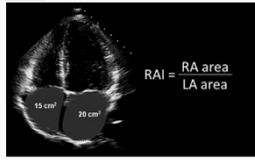
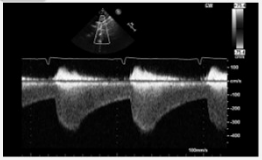
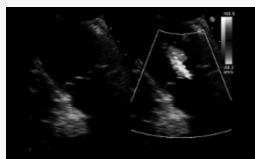

Echo may be a challenge.....

Adult with CHD

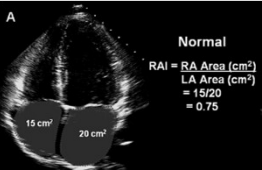
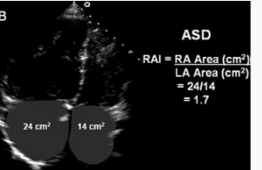


Echocardiographer



<p>1 Relative Atrial Index [RAI]</p> 	<p>2 CWD Descending Aorta</p> 
<p>3 PR Post TOF Repair</p> 	<p>4 Segmental Approach</p> 

1 Relative Atrial Index [RAI]

<p>A Normal</p> <p>RAI = RA Area (cm²) / LA Area (cm²) = 15/20 = 0.75</p> 	<p>B ASD</p> <p>RAI = RA Area (cm²) / LA Area (cm²) = 24/14 = 1.7</p> 
-------------------------------------------------------------------------------------------------------------------------------------------------------------------------------------------	---------------------------------------------------------------------------------------------------------------------------------------------------------------------------------------

Cutoff value > **0.92** predicted patients with ASDs vs. matched controls with 99.1% sensitivity & 90.5% specificity

Kelly NF, Scalia GM. J Am Soc Echocardiogr. 2010 Mar;23(3):275-81

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Types of ASDs

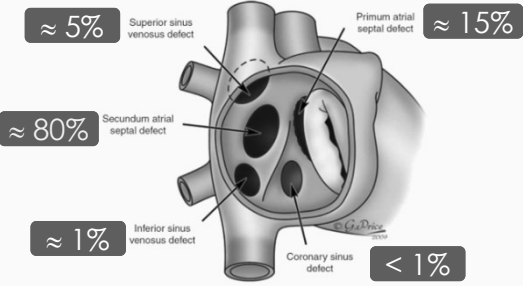
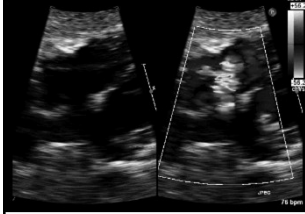


Image from Sridharan S., Price G., Tann O, et al. Cardiovascular MRI in Congenital Heart Disease: An Imaging Atlas. 2010

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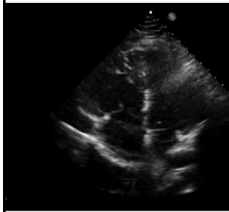
L-R Shunting across an ASD



Right heart volume overload
↓
RA & RV dilatation
↓
Pulmonary hypertension
↓
RV failure

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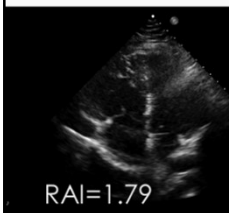
Limitations for Detecting an ASD



- "Drop-out" of IAS (Ap4-ch)
- Suboptimal image quality
- Use of limited standard imaging planes
- Location of the defect

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Limitations for Detecting an ASD



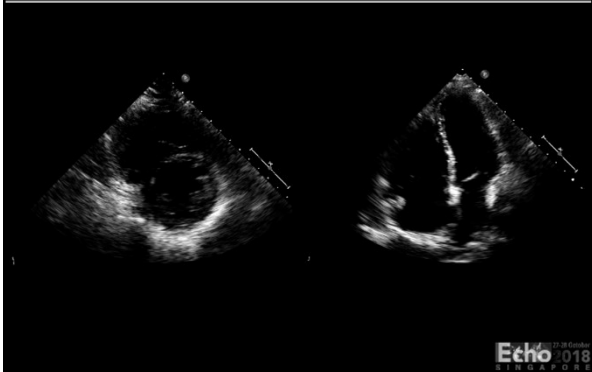
f IAS (Ap4-
y

WARNING!
RAI elevated
Is there an ASD?

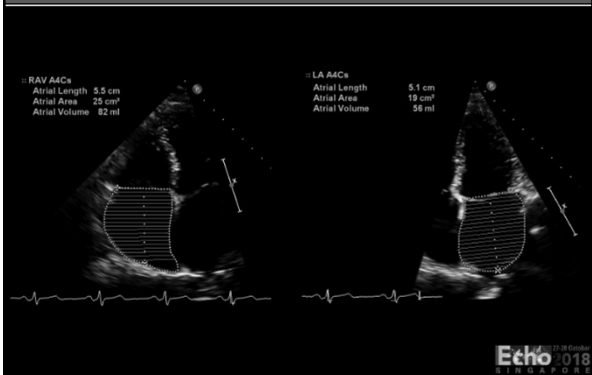
location of the defect

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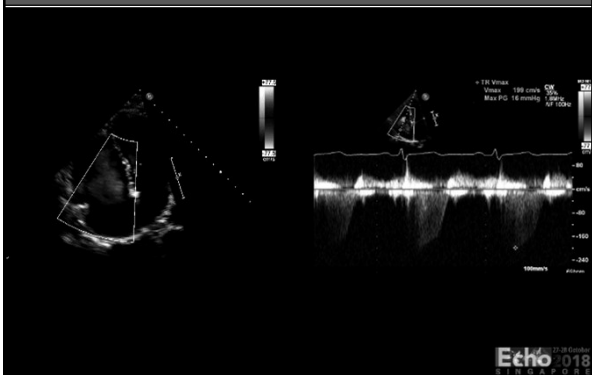
54 y.o. female, dilated right heart on outside echo



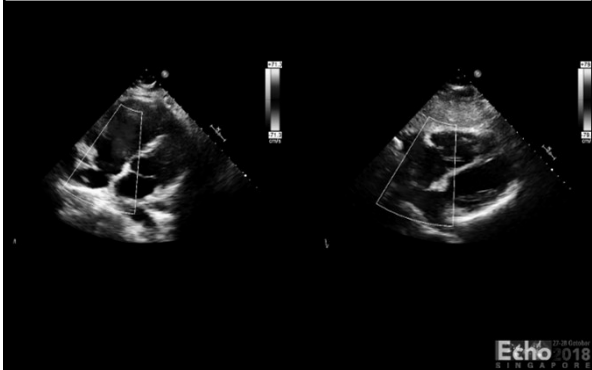
54 y.o. female, dilated right heart on outside echo



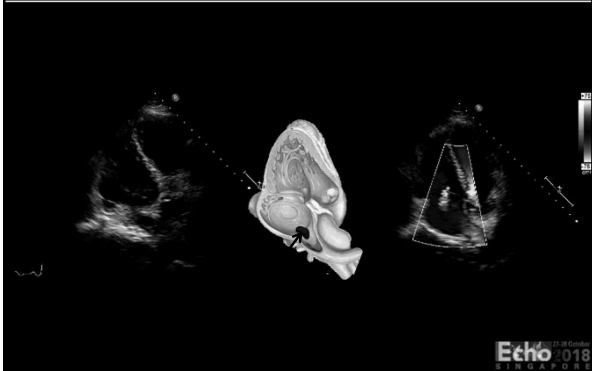
54 y.o. female, dilated right heart on outside echo



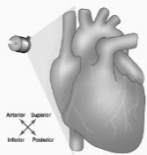
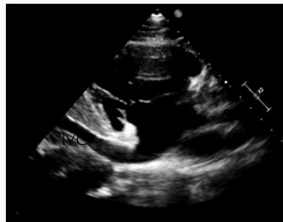
54 y.o. female, dilated right heart on outside echo



54 y.o. female, dilated right heart on outside echo



 Imaging tip - Look from the Right Sternal Edge



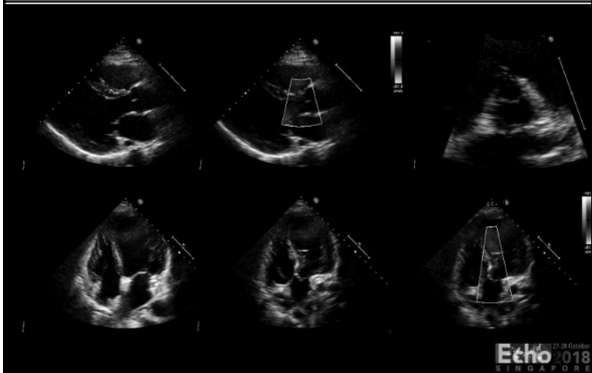
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2 CW Doppler Descending Aorta



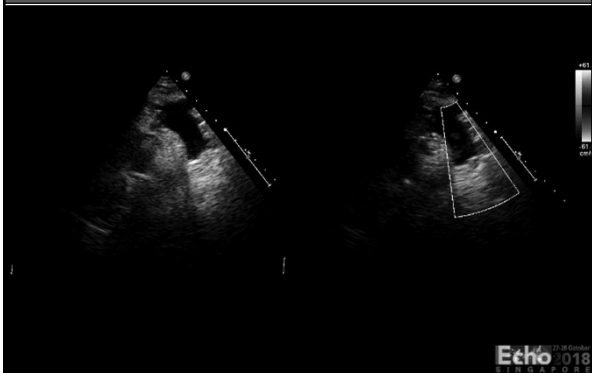
Echo 2018 SINGAPORE

23 y.o. female, routine echo prior to RFA



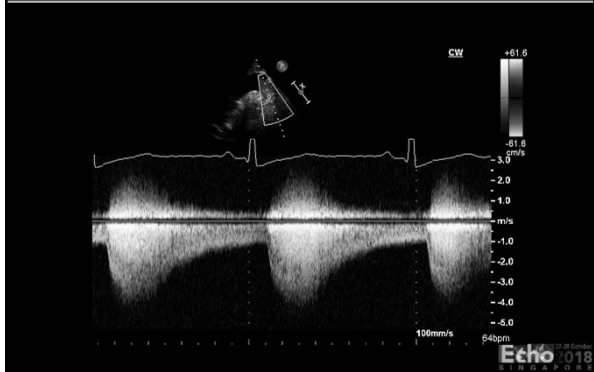
Echo 2018 SINGAPORE

23 y.o. female, routine echo prior to RFA

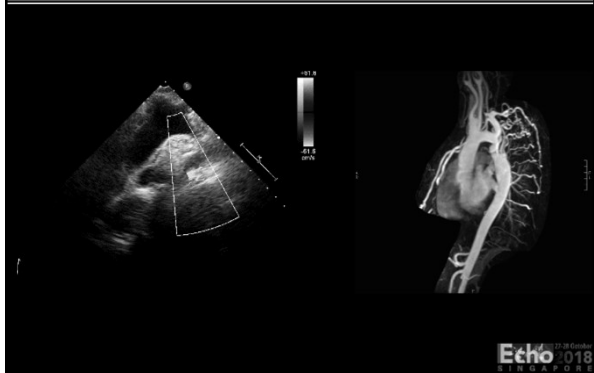


Echo 2018 SINGAPORE

23 y.o. female, routine echo prior to RFA



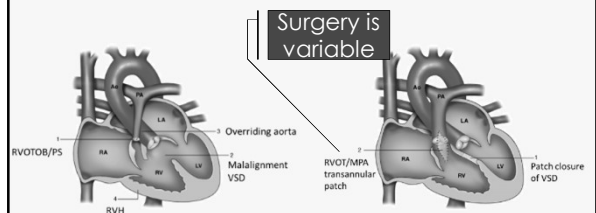
23 y.o. female, routine echo prior to RFA



 Don't forget the Abdominal Aorta



3 PR Severity post ToF Repair



Images from Mullins CE, Mayer DC: Congenital Heart Disease: A Diagrammatic Atlas. New York, Wiley-Liss, 1988



3 PR Severity post ToF Repair

Table 1 Structural and functional abnormalities encountered in repaired ToF

Structural Abnormalities	Functional Abnormalities
Obstruction to LV output	RV volume overload
Partial or complete removal of pulmonary valve tissue	RV pressure overload
Infundibular stenosis	RV outflow obstruction
Resection of fibromuscular muscle bundles	RV outflow tract stenosis
Right coronary artery	RV outflow tract obstruction
VSD patch	RV outflow tract stenosis
Residual or recurrent lesions	RV outflow tract obstruction
RV outflow tract obstruction	RV outflow tract stenosis
Main or branch pulmonary artery stenosis	RV outflow tract stenosis
Ventricular septal defect	RV outflow tract stenosis
Atrial septal defect	RV outflow tract stenosis
Acquired lesions	RV outflow tract stenosis
Tricuspid valve abnormalities	RV outflow tract stenosis
RV outflow tract aneurysm	RV outflow tract stenosis
RV failure	RV outflow tract stenosis
Associated anomalies	RV outflow tract stenosis
Dilated aorta	RV outflow tract stenosis
Associated congenital cardiovascular anomalies	RV outflow tract stenosis
Associated genetic and non-cardiac anomalies	RV outflow tract stenosis

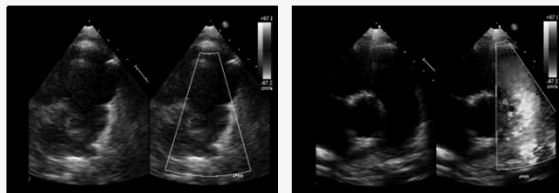
- Significant PR frequently missed on Echo
- Chronic severe PR leads to RV dilatation & dysfunction, exercise intolerance, ↑ risk arrhythmias & SCD
- PVR too late: RV dysfunction irreversible, pts. vulnerable to arrhythmias & SCD

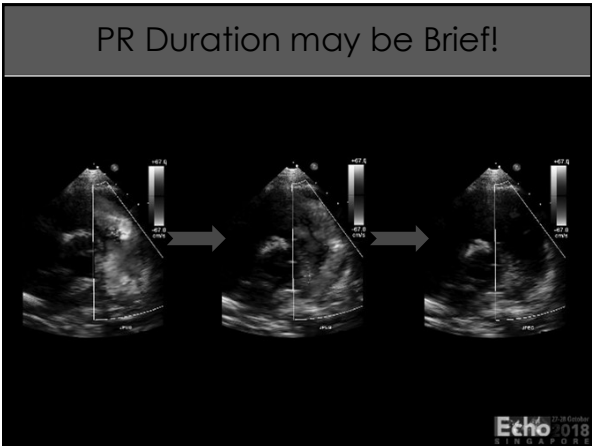
Geva T. J Cardiovasc Magn Reson. 2011 Jan 20;13:9.

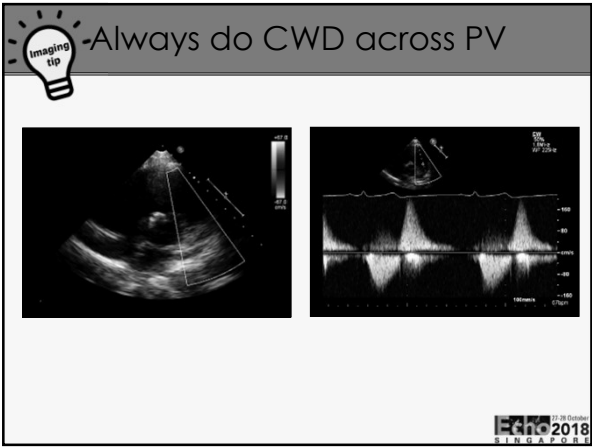


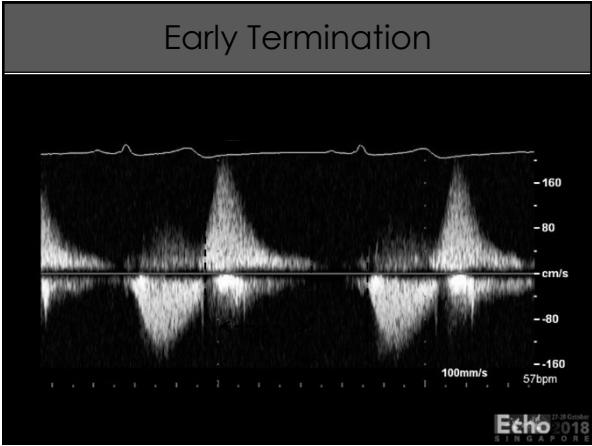
“Free”/“Wide Open” PR

Low velocity, laminar flow of short duration



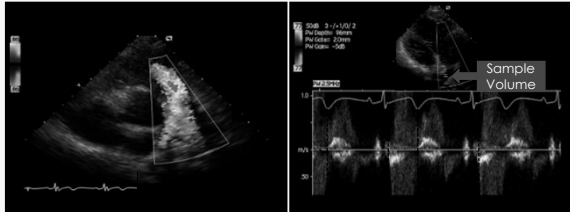






Imaging tip

Look at PA Branches for Diastolic Flow Reversal (DFR)



DFR: 3.1-10/2
PR Depth: 10mm
PR Gain: 20mm
PR Gain: -5dB

Sample Volume

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DFR in PA Branches Best Predictor of Severe PR

CONGENITAL HEART DISEASE

Two-Dimensional and Doppler Echocardiography Reliably Predict Severe Pulmonary Regurgitation as Quantified by Cardiac Magnetic Resonance

Renella P, J Am Soc Echocardiogr 2010;23:880-6.

Background: The gradient pulmonary regurgitation (PGR) severity by two-dimensional (2D) and Doppler echocardiography in the presence of Congenital Heart Disease (CHD) may vary significantly from that of the PGR in the normal heart. The objective of this study was to determine the best 2D and Doppler echocardiographic indicators of severe PGR.

Methods: Thirty-two patients with history of PGR in pulmonary valve disease with prior pulmonary valve replacement or transcatheter or surgical aortic valve replacement (AVR) and Doppler echocardiography and cardiac magnetic resonance (CMR) measurements. Left ventricular and right ventricular dimensions were used to estimate PGR severity. Left ventricular and right ventricular dimensions were used to estimate PGR severity. PGR severity was defined as severe if the ratio of PGR to the main pulmonary artery (MPA) jet width was $\geq 50\%$ of the pulmonary artery. PGR severity was defined as severe if the ratio of PGR to the MPA jet width was $\geq 50\%$ of the pulmonary artery. PGR severity was defined as severe if the ratio of PGR to the MPA jet width was $\geq 50\%$ of the pulmonary artery.

Results: With the exception of PGR index, all indices were significant independent predictors of severe PGR. The best echocardiographic predictor of severe PGR was Doppler echocardiography.

Conclusions: Two-dimensional and Doppler echocardiography reliably identified severe PGR in this cohort. (J Am Soc Echocardiogr 2010;23:880-6.)

Keywords: Pulmonary regurgitation; Echocardiography; Cardiac magnetic resonance

Parameter	Sensitivity	Specificity	PPV	NPV
MPA diastolic flow reversal	100%	39%	59%	100%
BPA diastolic flow reversal	87%	87%	87%	87%
PR jet/pulmonary annular diameter ratio $\geq 50\%$	94%	74%	76%	93%
PdT < 100 ms	90%	64%	78%	82%
PRi < 0.77	73%	47%	58%	64%

BPA, Branch pulmonary arteries; MPA, main pulmonary artery.

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Imaging tip

**PRESUME SEVERE PR
UNTIL PROVEN OTHERWISE**

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4

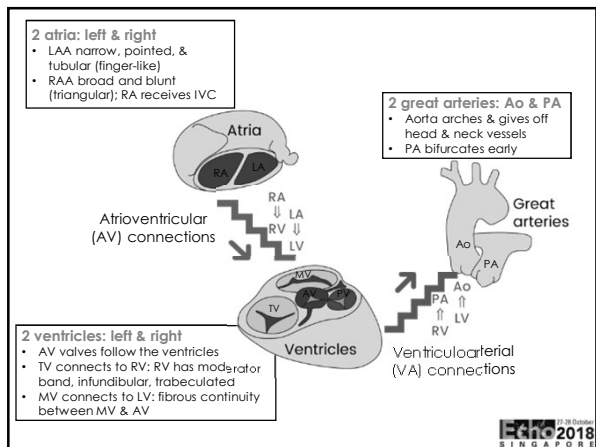
Segmental Approach

3 storied house

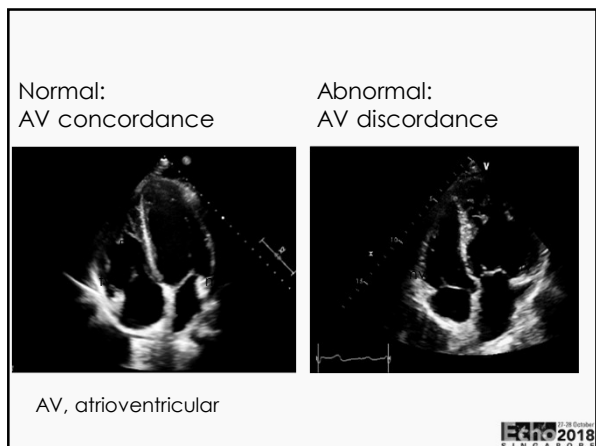


- 3 floors:
 - Atria
 - Ventricles
 - Great arteries
- 2 Staircases:
 - Atrioventricular connections
 - Ventriculoarterial connections
- 2 Entrances:
 - Systemic veins
 - Pulmonary veins

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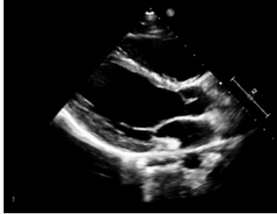


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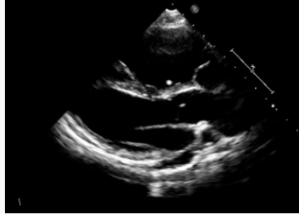


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Normally related GA:
PLAX => Only one seen



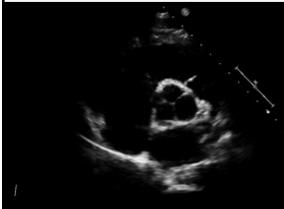
Abnormally related GA:
PLAX => Side-by-side



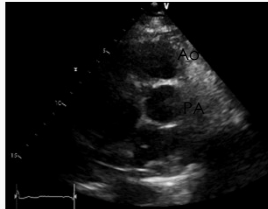
GA, great arteries

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Normally related GA:
"sausage-circle"



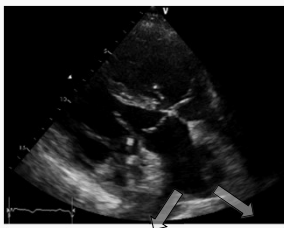
Abnormally related GA:
both valves seen in SAX



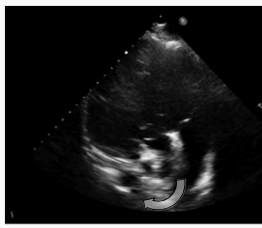
GA, great arteries

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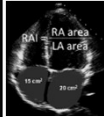
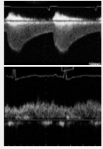
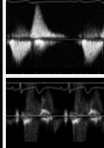

PA Bifurcates Early



Aorta Arches




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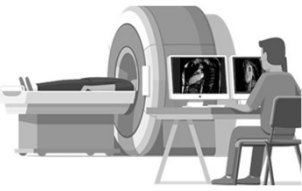
<p>1 Relative Atrial Index [RAI]</p>  <ul style="list-style-type: none"> • RAI > 0.92: ? ASD • Off-axis imaging & panning with colour Doppler crucial 	<p>2 CWD Descending Aorta</p>  <ul style="list-style-type: none"> • Sawtooth profile (diastolic tail) = severe coarctation • Check abdominal aorta for damped or continuous flow
<p>3 PR Post TOF Repair</p>  <ul style="list-style-type: none"> • Presume severe PR until proven otherwise • CWD of PR jet & PWD in PA branches essential 	<p>4 Segmental Approach</p>  <ul style="list-style-type: none"> • Identify chambers & arteries • Identify AV & VA connections • Start at the apical window

Know the limitations & refer Onwards.....

Specialist Centre



Alternative Imaging



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References & Further Reading

General

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References & Further Reading

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Tetralogy of Fallot:

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- Li W, et al. **Doppler-echocardiographic assessment of pulmonary regurgitation in adults with repaired tetralogy of Fallot: comparison with cardiovascular magnetic resonance imaging.** *Am Heart J.* 2004 Jan;147(1):165-72.
- Renella P, et al. **Two-dimensional and Doppler echocardiography reliably predict severe pulmonary regurgitation as quantified by cardiac magnetic resonance.** *J Am Soc Echocardiogr.* 2010 Aug;23(8):890-6.
- Valente AM, et al. **Multimodality imaging guidelines for patients with repaired tetralogy of Fallot: a report from the American Society of Echocardiography: developed in collaboration with the Society for Cardiovascular Magnetic Resonance and the Society for Pediatric Radiology.** *J Am Soc Echocardiogr.* 2014 Feb;27(2):111-41.