Echocardiography in pulmonary atresia with intact ventricular septum

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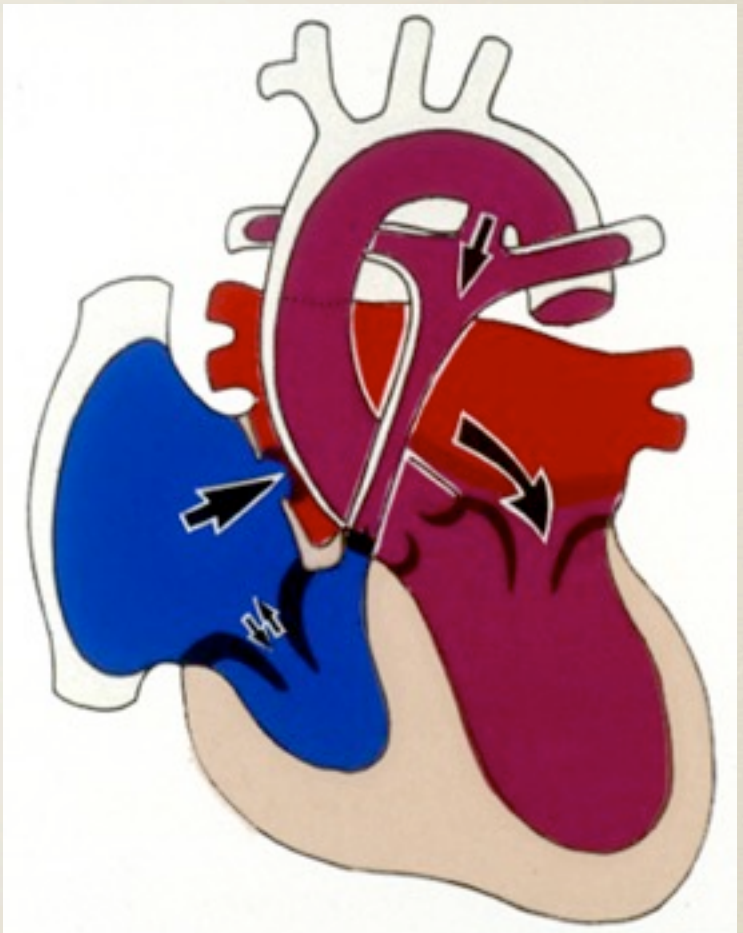
PAIVS- Anatomy and physiology

* Congenital disease of TV, RV and pulmonary valve
* Atresia of the pulmonary valve
* Blood passes from:
  * RA to LA to LV via ASD/ PFO
  * Blood enters PAs retrogradely via duct
  * RA to RV and exits via TR & coronary fistulae



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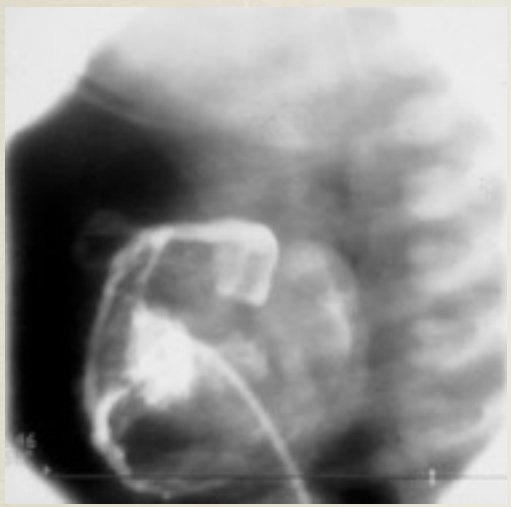
PAIVS: Presentation

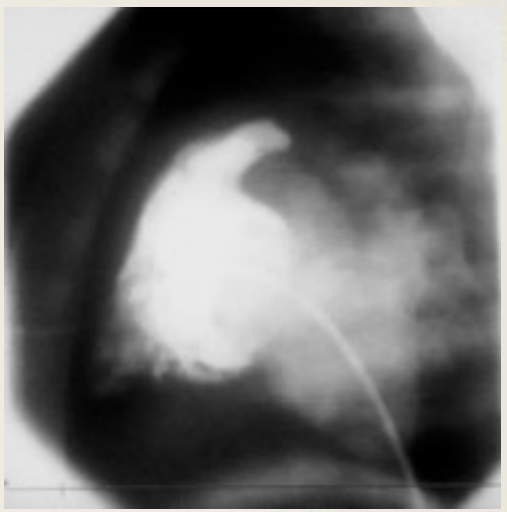
- Prenatally
  - Abnormal 4 chamber view
- Postnatally when duct shuts
  - Cyanosis with saturations in upper limb = lower limb
  - Good pulses
  - Tricuspid regurgitant murmur
  - Often quite well initially, then eventual collapse/ acidosis

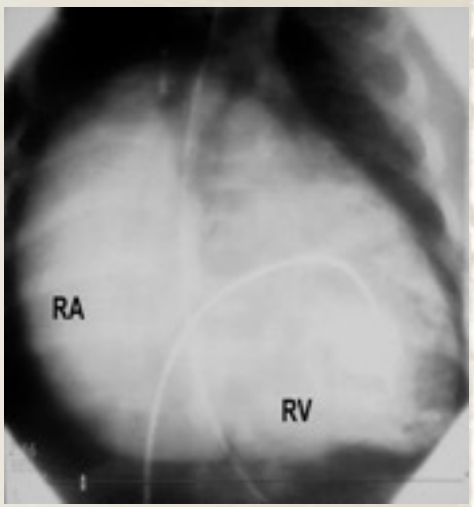
PAIVS: Initial management

* Hyperoxic test
* Prostaglandin
* Ventilate only if needed
* Correct acidosis
* Arrange transport
* Establish exact diagnosis

PAIVS: Wide variation in morphology







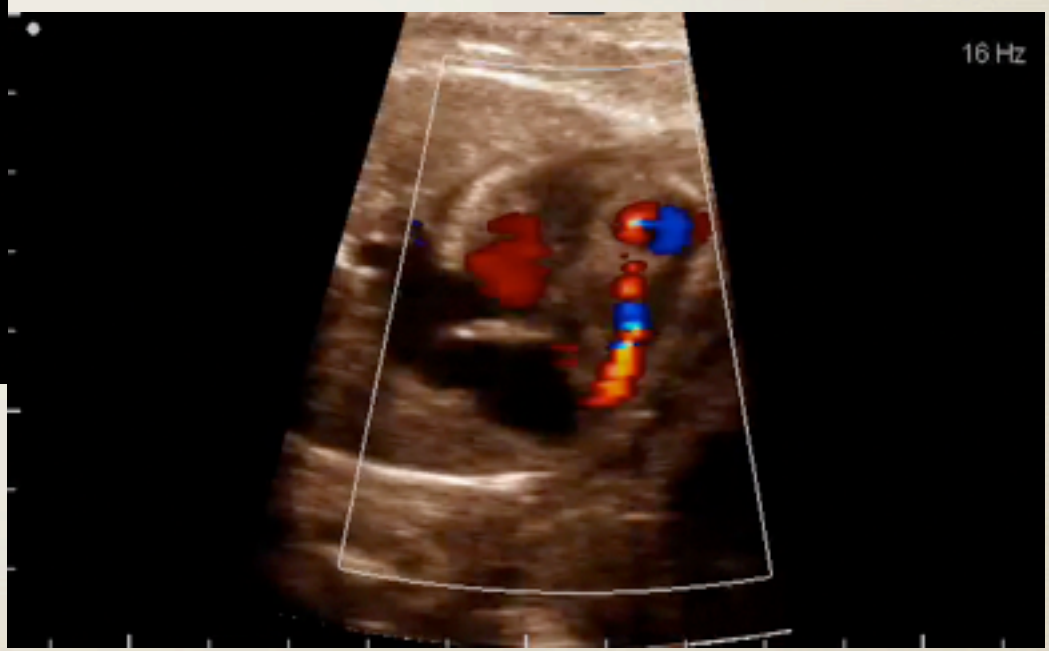
PAIVS: Aims of echocardiography

* Confirm diagnosis
* Document all morphologic features systematically
* Decide on initial palliation bearing in mind longterm goals of separating the systemic and pulmonary circulations
  * Biventricular repair
  * 1.5 ventricle repair (RVOT reconstruction and cavo-pulmonary anastomosis)
  * Univentricular repair (TCPC/ Fontan)

PAIVS: Ultimate goals of management

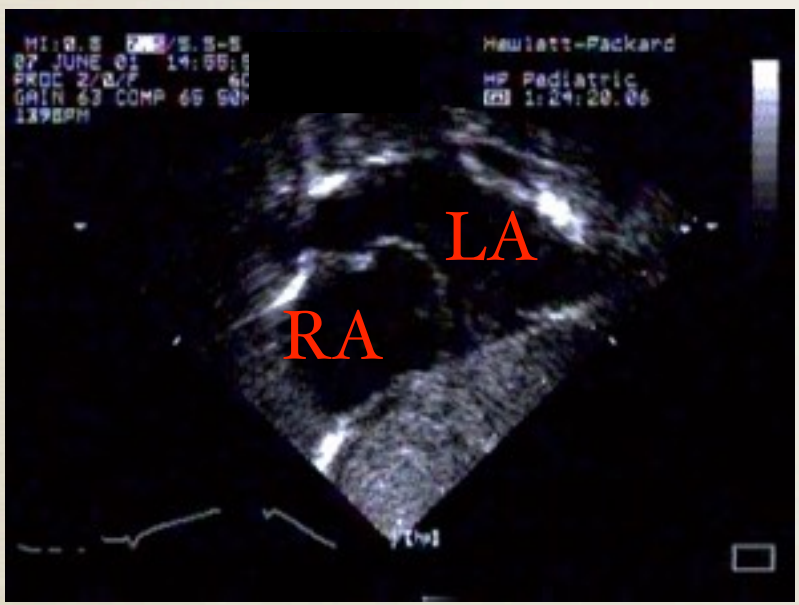
	Biventricular Route	Borderline	Univentricular Route
TV z-score	>-2.5	-2.5 to -5	<-5
RV Morphology	Tripartite	Bipartite	Unipartite
RVDI	>0.35	<0.35	<0.35 and muscular atresia
Presence of RV infundibulum	Yes	Small and narrow	No
RV fistulae	Nil/Minor	Minor/Major	RVDCC
Treatment at Presentation	RF perforation or closed surgical valvotomy	RF perforation or surgical valvotomy/RVOT patch and consider PDA stent/BTS	Palliative procedure- BTS +/- BAS

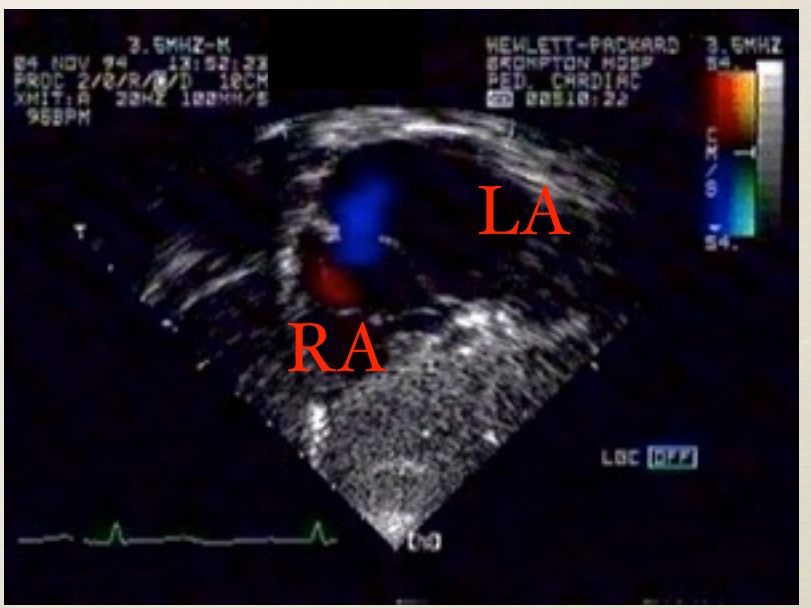
Fetal diagnosis



Range of morphology: atrial septum

- Restrictive/ non-restrictive
- Biventricular/ univentricular repair
- Balloon septostomy/ septectomy if univentricular





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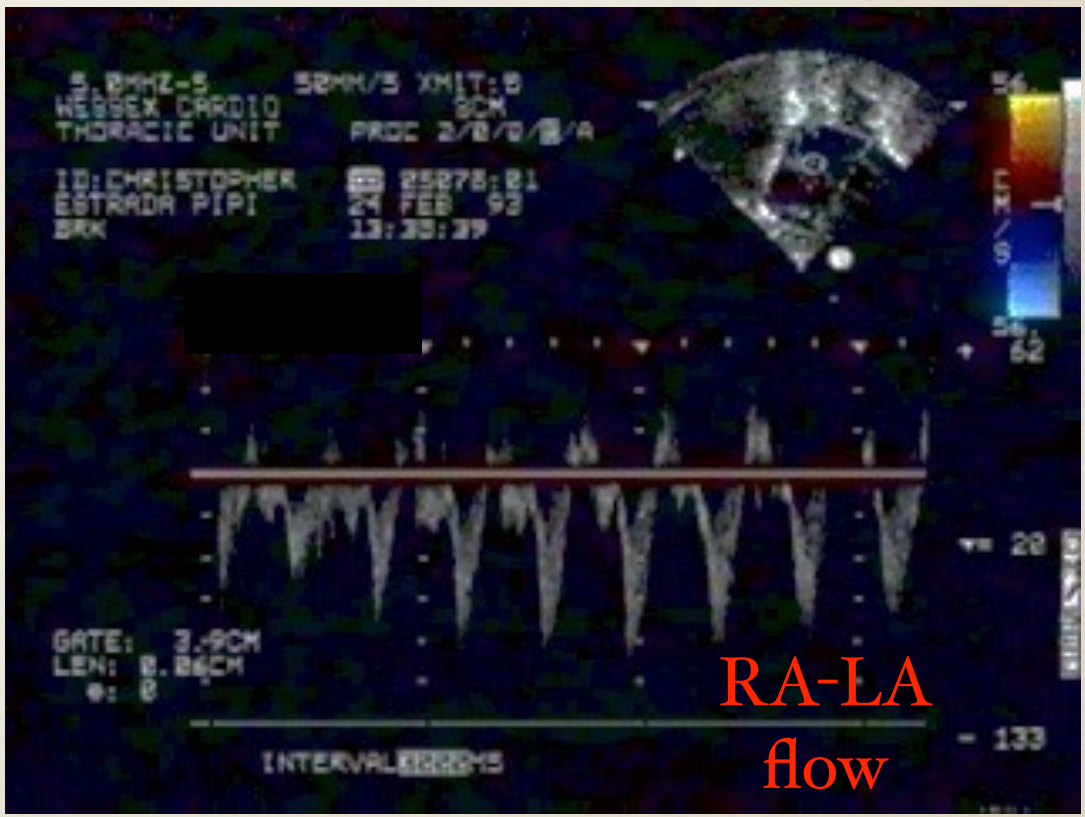


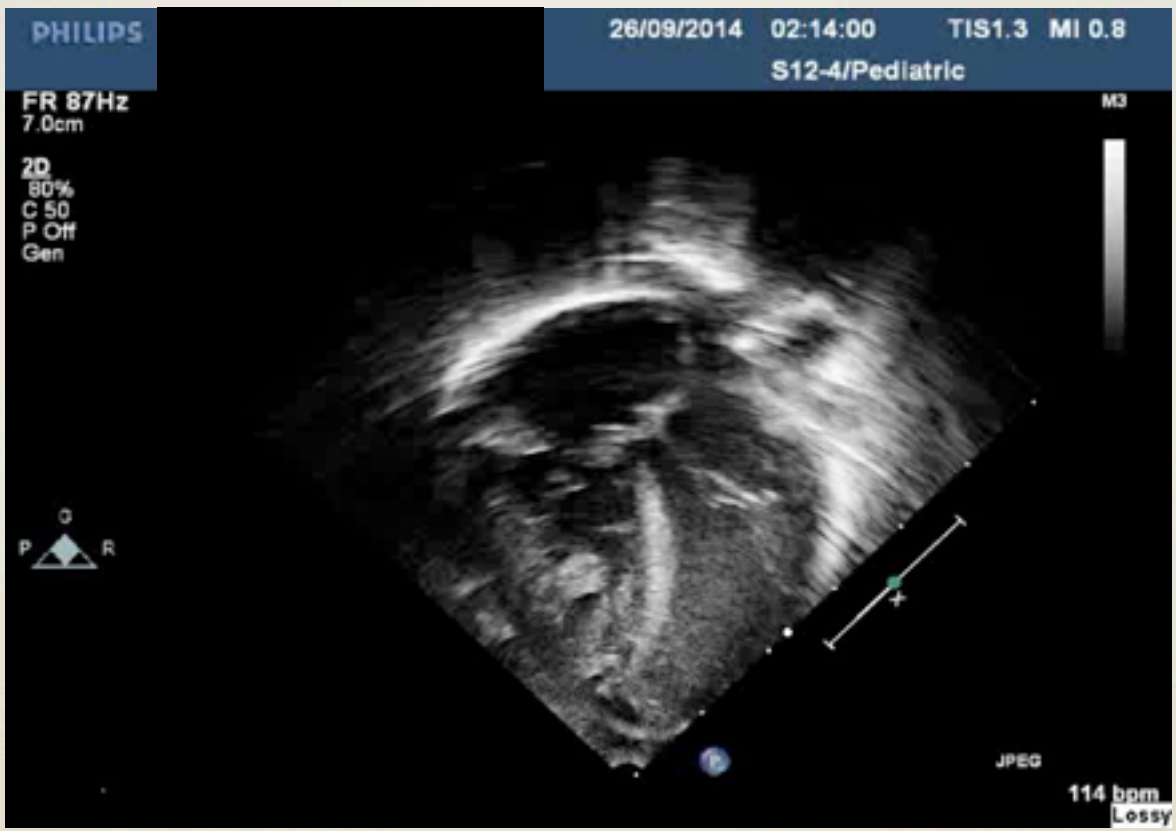
Range of morphology: tricuspid valve

- TV dysplasia common
- Ebstein malformation
- Tricuspid regurgitation
- TV size: surrogate for RV volume?

Tricuspid valve dysplasia

[redacted]

Tricuspid valve dysplasia



Tricuspid valve dysplasia

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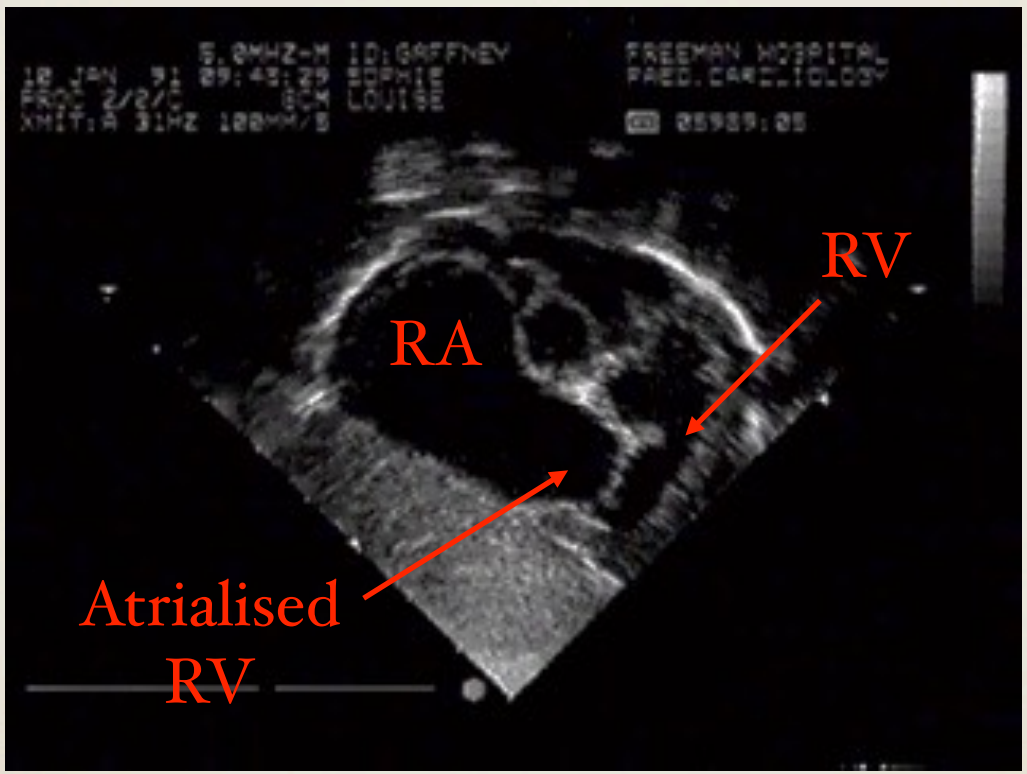
Ebstein malformation



10% all PAIVS cases

Daubeney 2002

Tricuspid regurgitation

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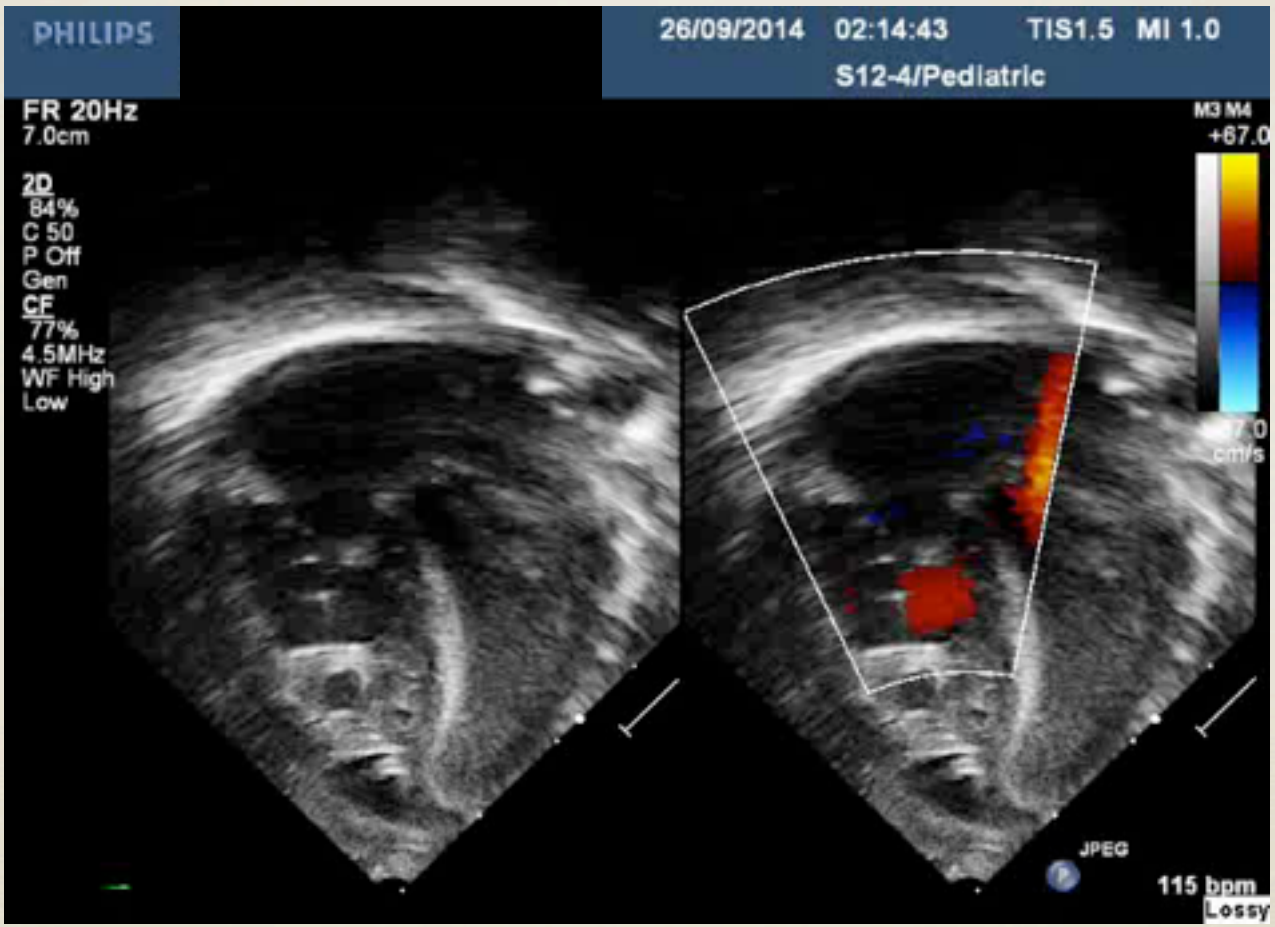
Tricuspid regurgitation



Tricuspid regurgitation

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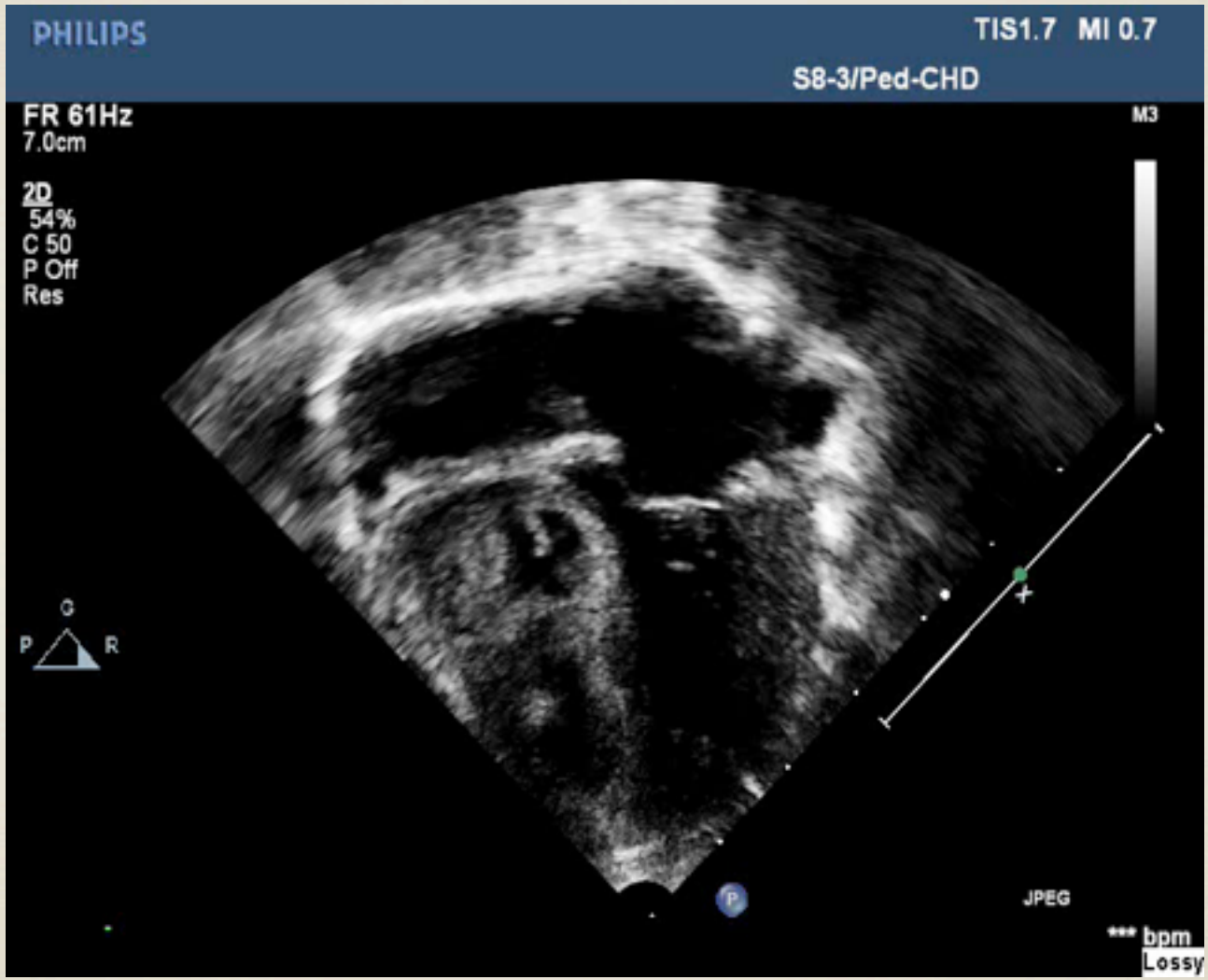
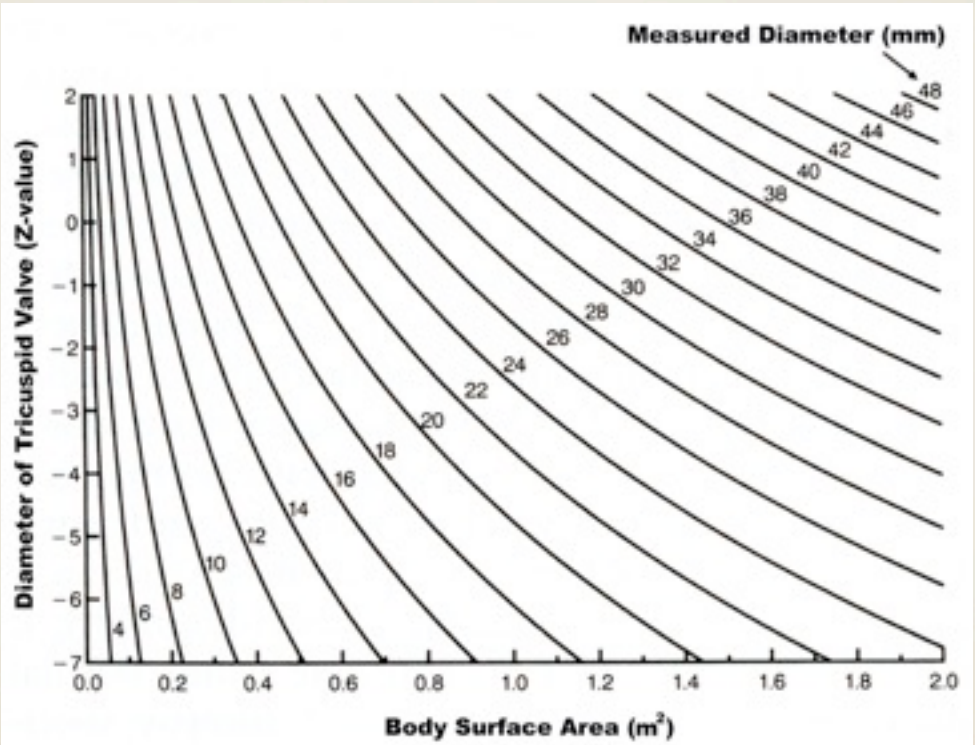
Hypoplastic tricuspid valve

Hypoplastic tricuspid valve



Hypoplastic tricuspid valve

Tricuspid valve Z score



Cardiology in the Young

Daubeney 1999

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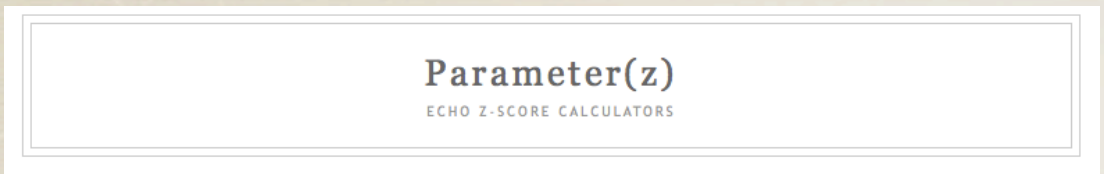
Z score calculators: Parameter(z)



Pediatric and Fetal Echo Z-Score Calculators

WWW!

Announcing a new parameterz.com site with new references! All development has moved over to this new platform and I will no longer be developing for this site (Blogger). That's a good thing.

App

The ParameterZ web app. A lot of information in a little package. Log in with your Google account.

Aortic Root Z-Scores

Calculate BSA-adjusted z-scores of the aortic annulus and sinuses of Valsalva using data published by Boston Children's Hospital. (Updated 11/2008: added percentiles).

Also see: consolidated aortic root z-score calculator

Ascending Aorta Z-Scores

Z-Scores of the ascending aorta (AAO) and aortic root. Data from the Children's Heart Center, Halifax, Nova Scotia, published in 2006 and from Hôpital Bichat, Paris, France published in 2010

Cardiac Valve Z-Scores

Calculate BSA-adjusted z-scores of the mitral valve, tricuspid valve, aortic valve, and pulmonary valve using data published by Cincinnati Children's Hospital. This data is also used to create dynamic z-score tables.

Coronary Artery Z-Scores

Calculate BSA-adjusted z-scores of the proximal coronary arteries (for patients with Kawasaki disease) using data published by Boston Children's Hospital and Children's National Medical Center, Washington, DC and Montreal, Canada (also calculates aortic valve standardized coronary z-scores). Also, have a look at my comparison and charting tool.

Fetal Echo Z-Scores

Calculate gestational age-adjusted z-scores for fetal echo (left ventricle, mitral valve, aortic valve, aortic arch, etc.) using data published by The Royal Brompton Hospital and Boston Children's Hospital.

LVEDV Z-Scores

BSA-adjusted z-scores for LVEDV in patients < 3 y.o. using the 5/6 Area Length (Bullet) formula. (sorry about the offsite link- I'm trying something different)

LV Mass Z-Scores

Calculate height-adjusted z-scores of left ventricular mass (m-mode derived calculation) using data published by Montreal Children's Hospital.

Z-Scores of Cardiac Structures | Detroit Data

Calculate the z-scores of 21 common 2D and M-Mode echo measurements, related to body surface area. Measurement sites include the mitral valve, left ventricle, aortic valve, aortic arch, pulmonary valve, and pulmonary arteries. Data is from 782 patients evaluated at the Children's Hospital of Michigan.

Height (cm): 55
Weight (kg): 3
BSA formula: Haycock 0.21 M^2

Site	Measured (cm)	Mean	Range	Z-Score
RVD:		1.02	(0.69 - 1.52)	
IVSd:		0.37	(0.26 - 0.52)	
IVSs:		0.49	(0.36 - 0.66)	
LVIDd:		1.87	(1.59 - 2.21)	
LVIDs:		1.16	(0.94 - 1.43)	
LVPWd:		0.29	(0.21 - 0.40)	
LVPWs:		0.55	(0.43 - 0.71)	
Aortic Annulus:		0.69	(0.58 - 0.81)	
Sinuses:		0.97	(0.81 - 1.16)	
ST Junction:		0.76	(0.61 - 0.95)	
Transverse Arch:		0.78	(0.61 - 1.00)	
Isthmus:		0.55	(0.42 - 0.72)	
Distal Arch:		0.59	(0.45 - 0.77)	
Ao at Diaphragm:		0.59	(0.47 - 0.73)	
Pulmonary Annulus:		0.78	(0.61 - 1.01)	
MPA:		0.82	(0.64 - 1.06)	
RPA:		0.48	(0.36 - 0.63)	
LPA:		0.44	(0.34 - 0.58)	
Mitral Annulus:		1.20	(0.94 - 1.53)	
Tricuspid Annulus:	0.8	1.31	(0.96 - 1.79)	-2.59

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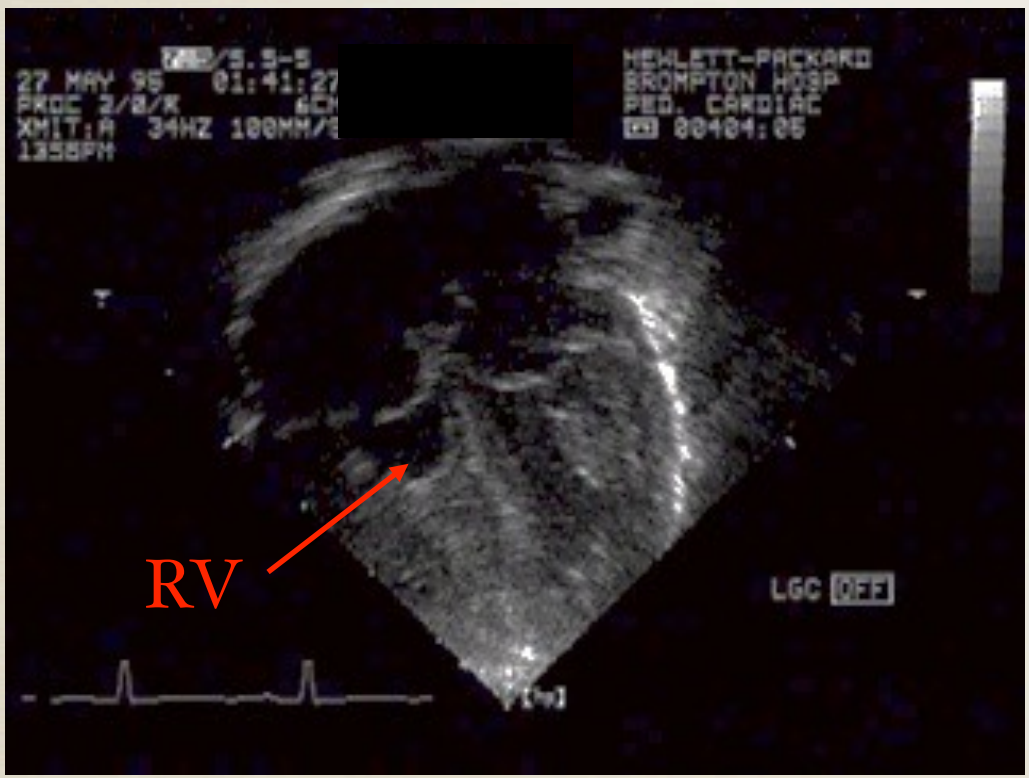
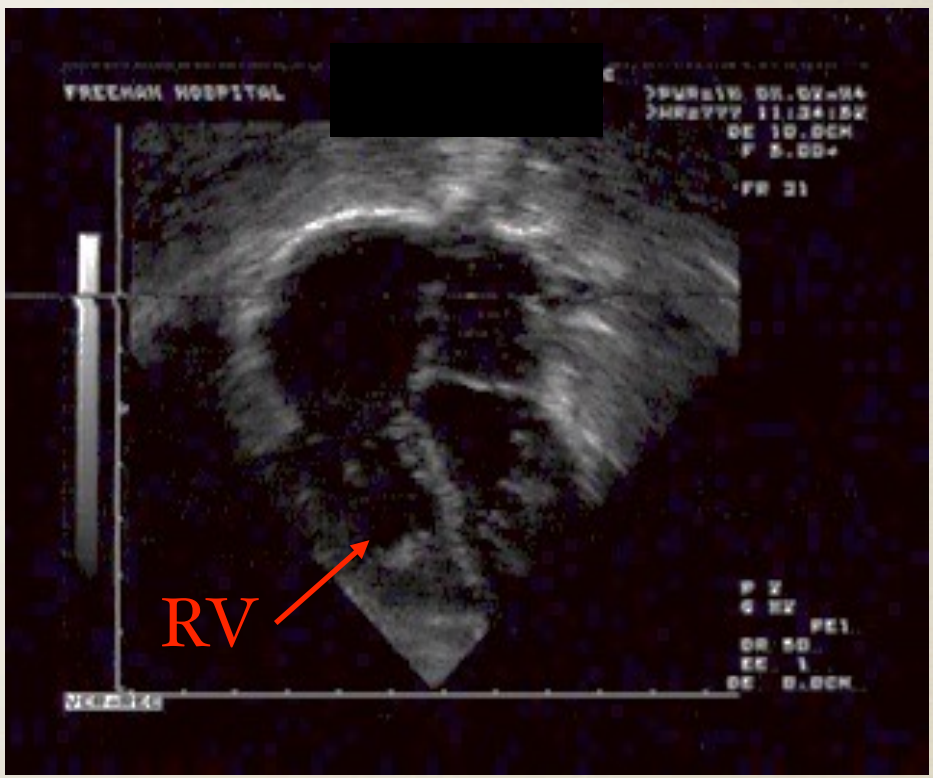
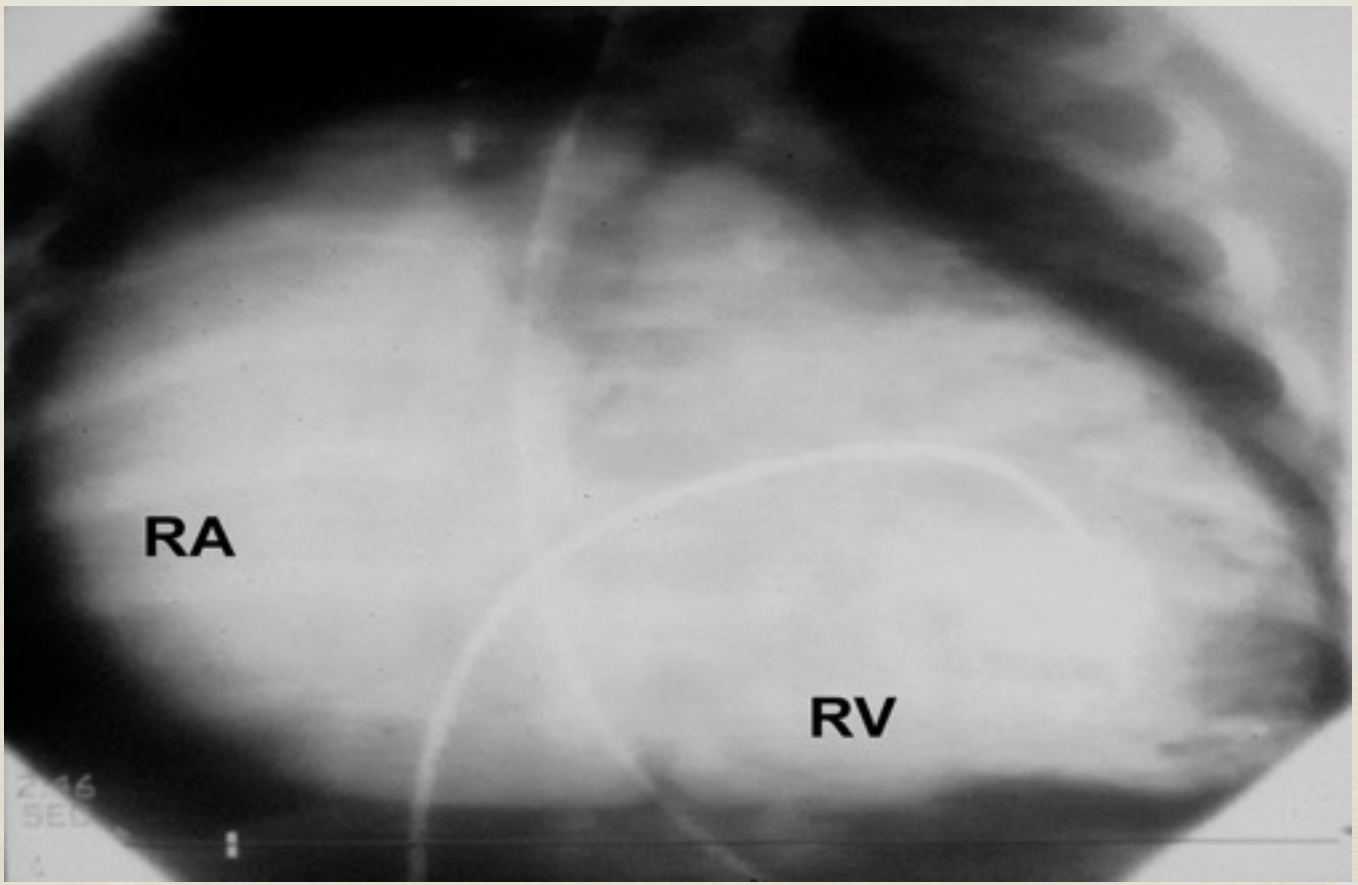
Right ventricle size and morphology





Right Ventricle: Size and morphology



Right Ventricle: Size and morphology

- RV inlet: Surrogate for RV volume
- Partite classification
  - Tripartite 59%
  - Bipartite 33.5%
  - Unipartite 7.5%

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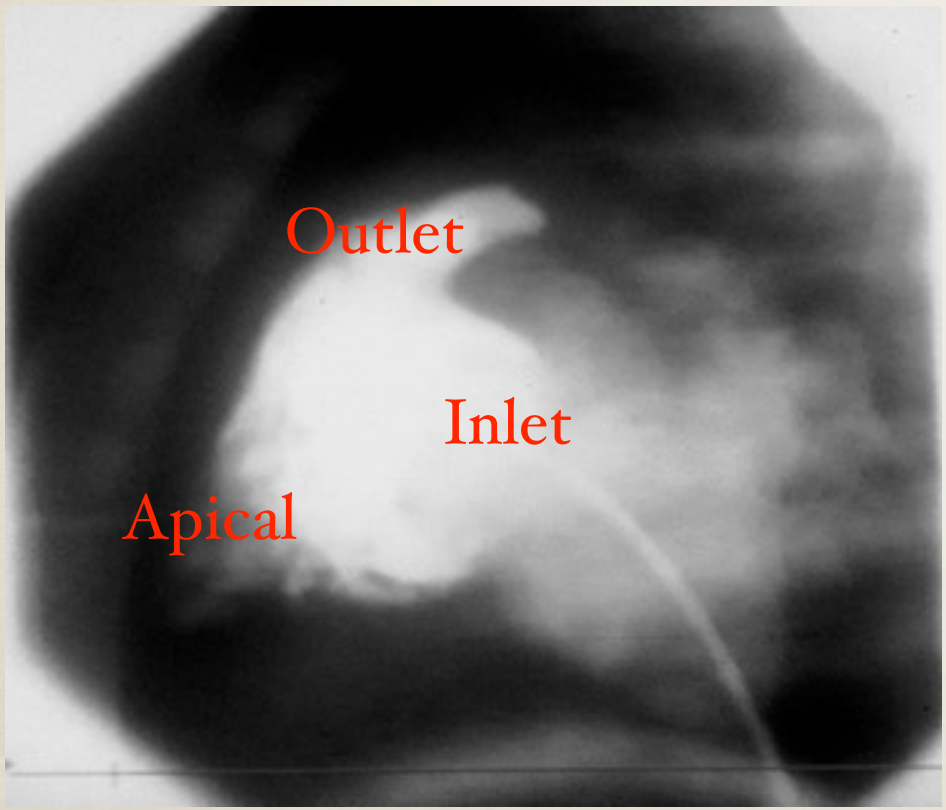
Daubeney 2002

Tripartite right ventricle



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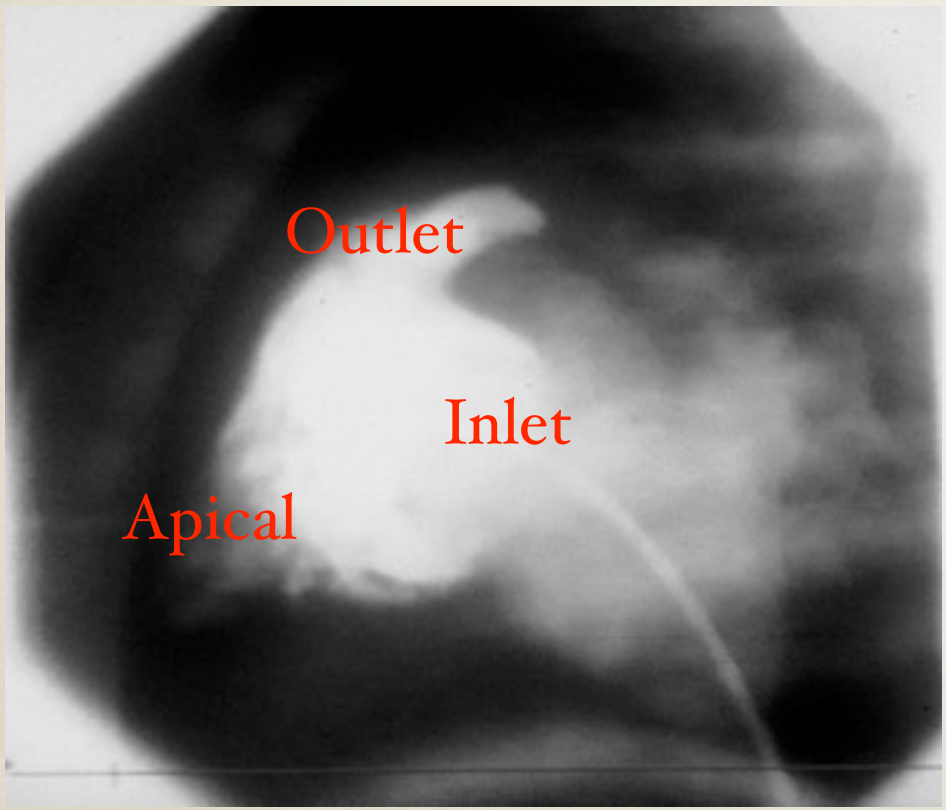
Tripartite right ventricle



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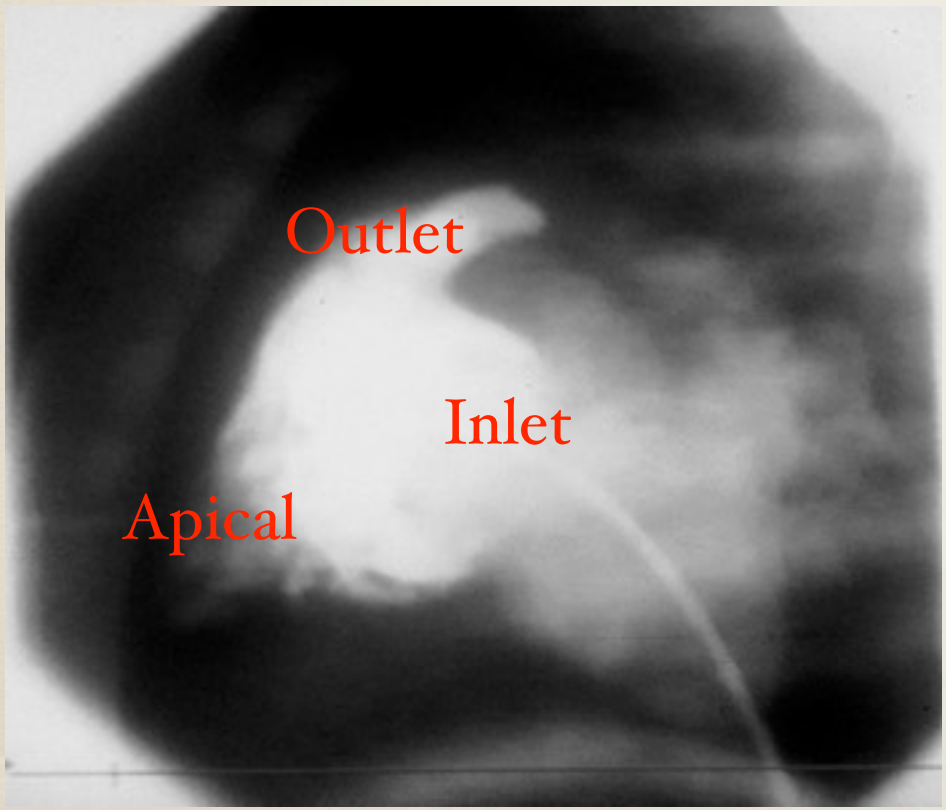

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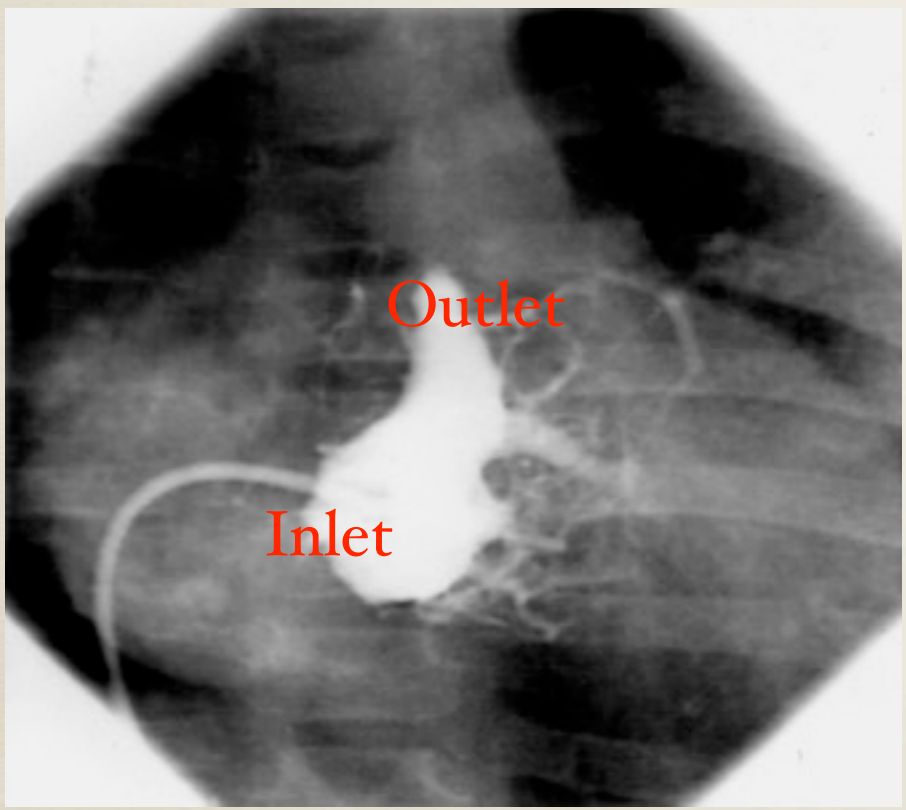
Bipartite right ventricle



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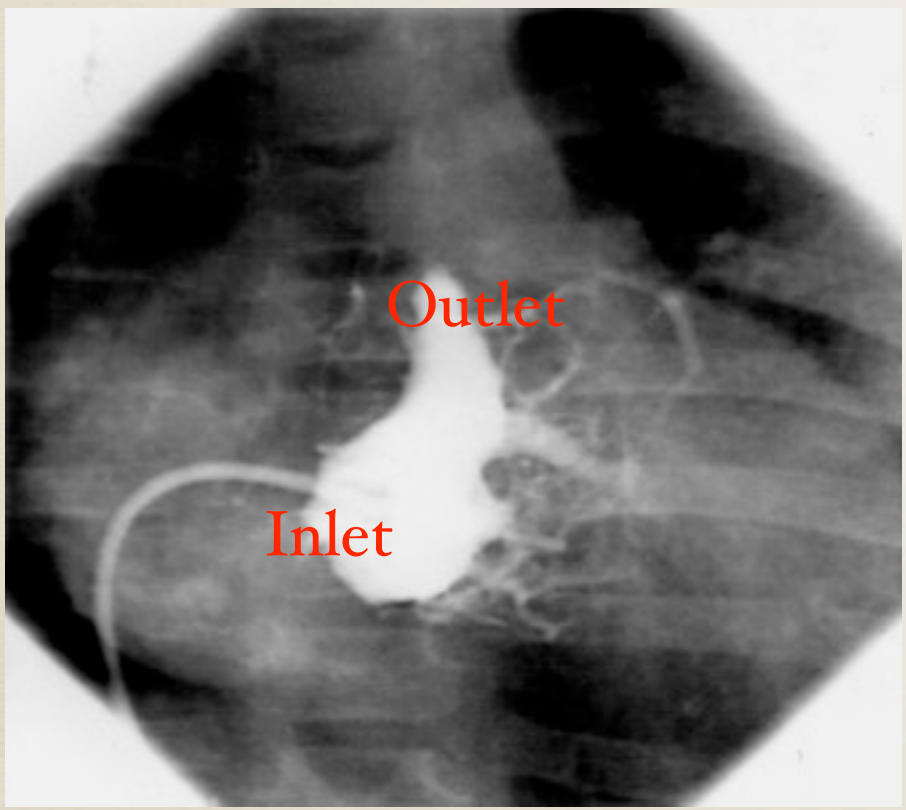
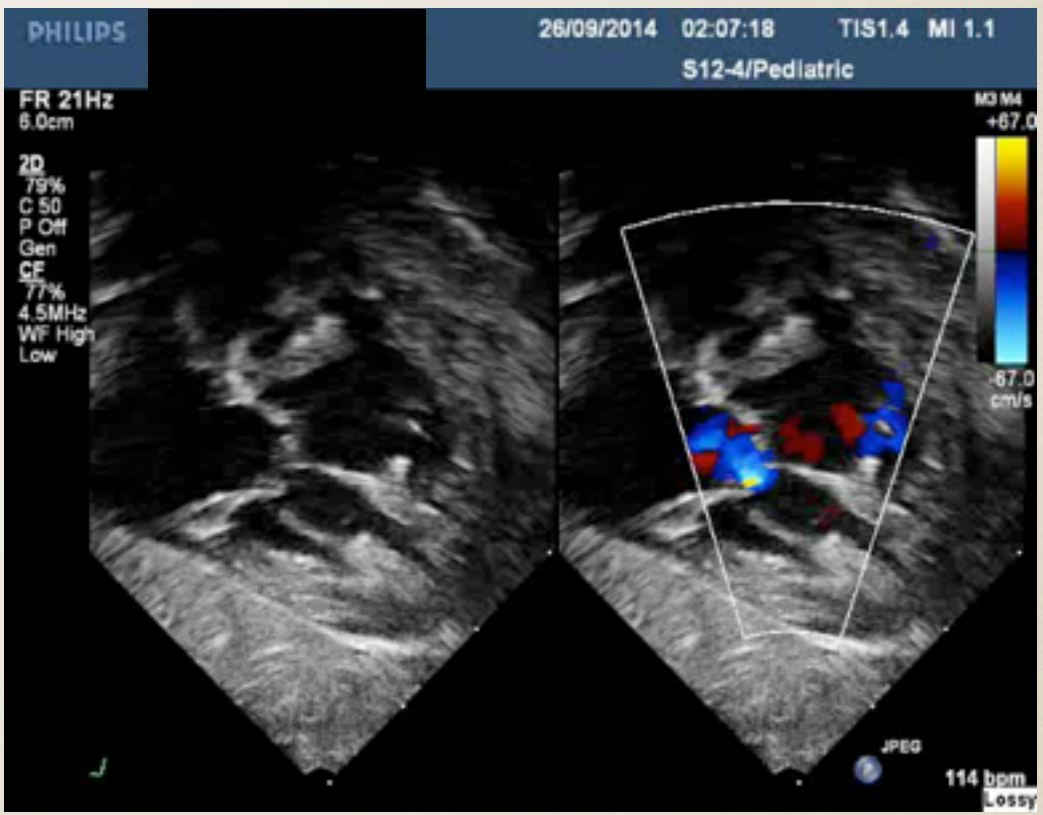
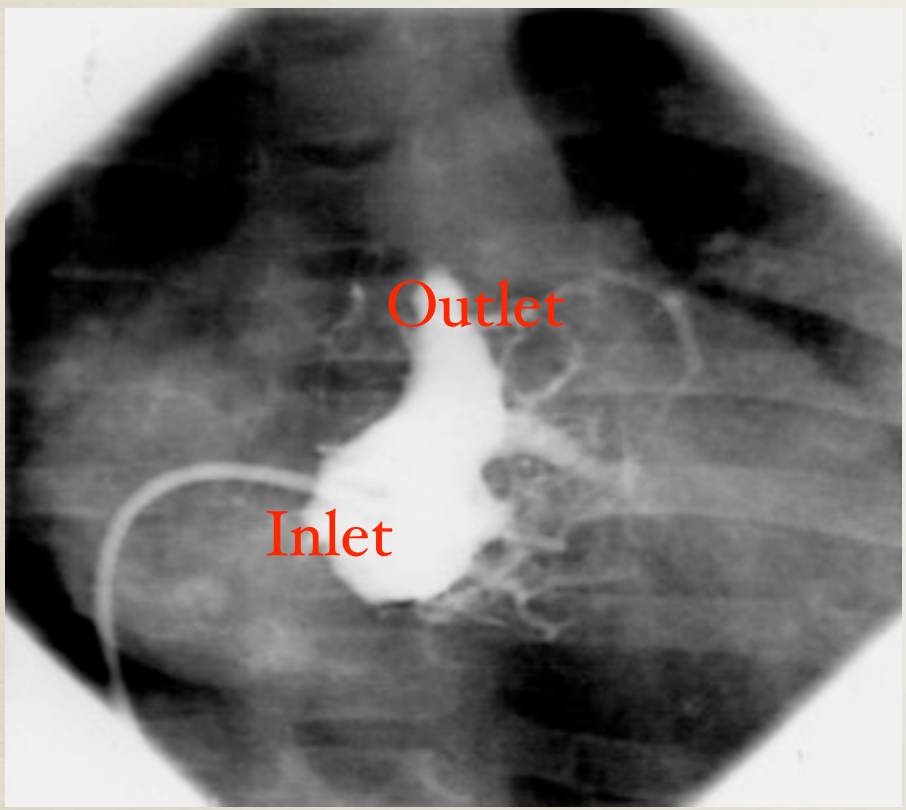
Bipartite right ventricle





Bipartite right ventricle



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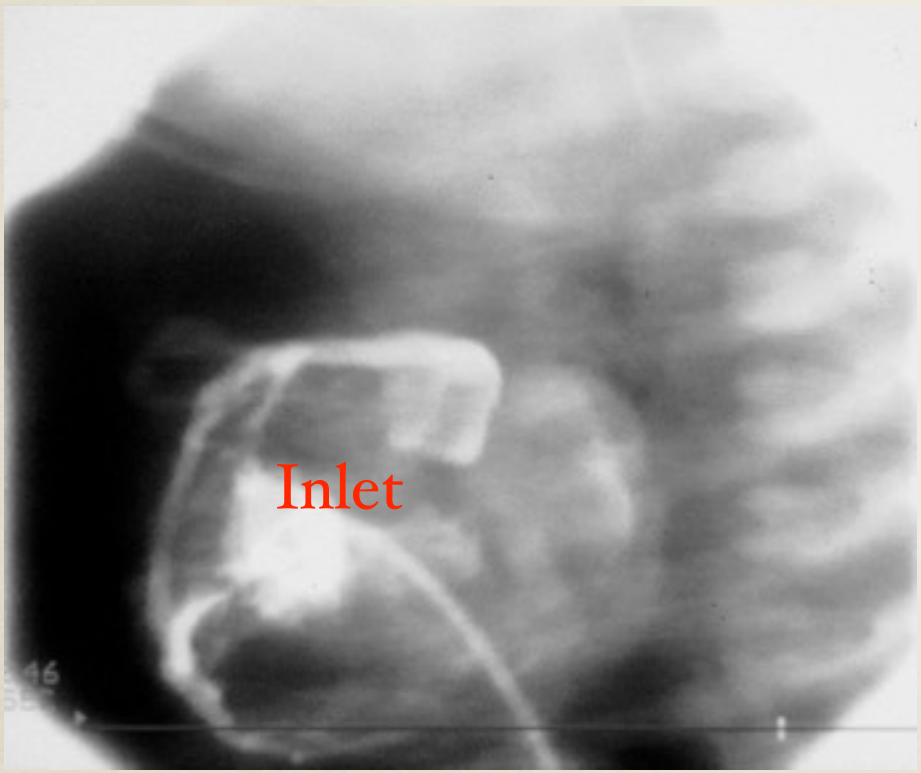
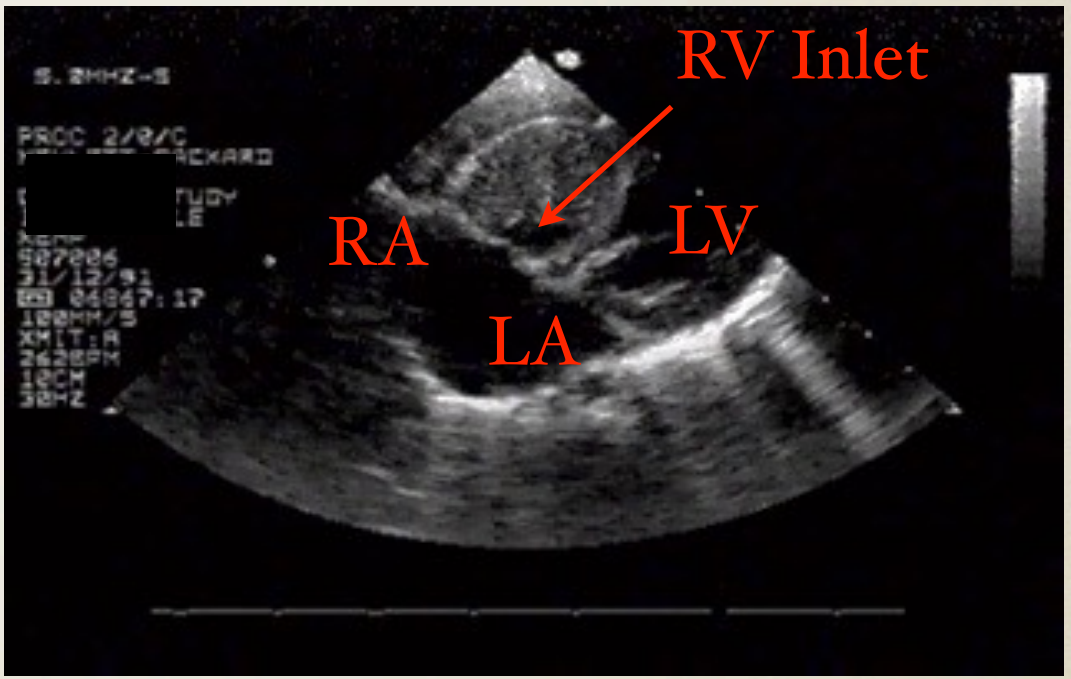
Unipartite right ventricle





Range of morphology: Pulmonary valve

Muscular 25%
Membranous 75%

[redacted]

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Range of morphology: Pulmonary valve

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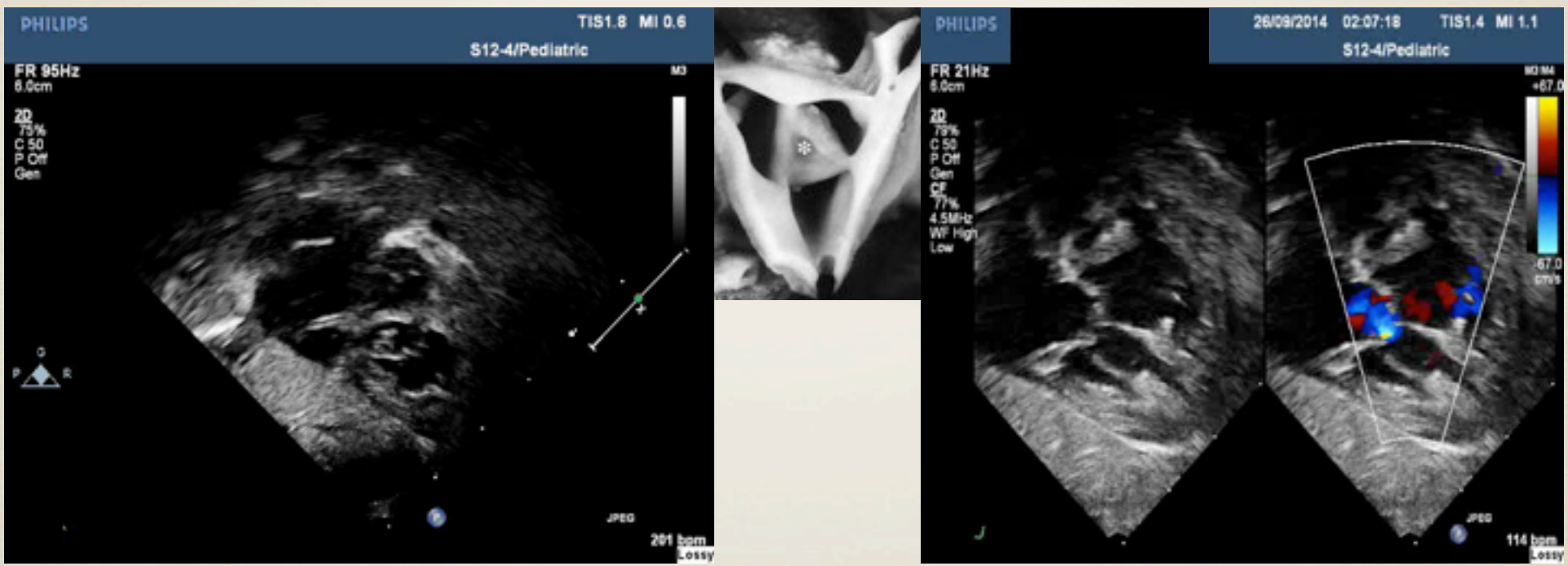
Membranous 75%



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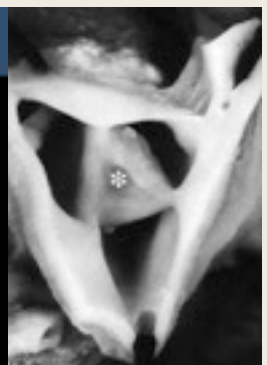

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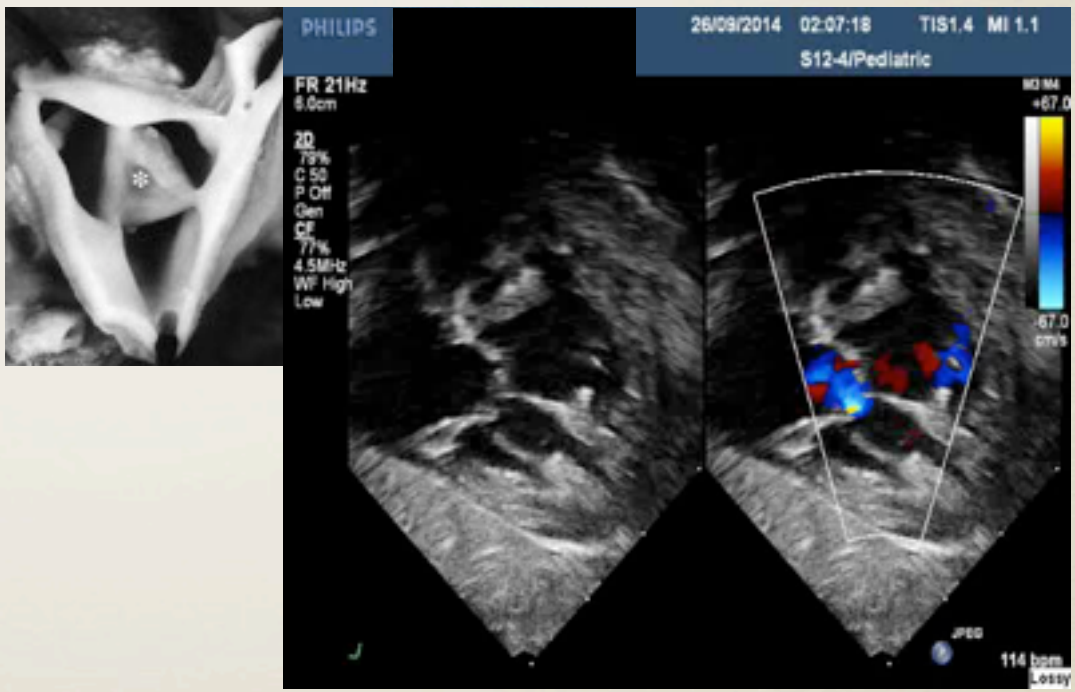

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RV coronary connections: Fistulae

Fistulae 46%

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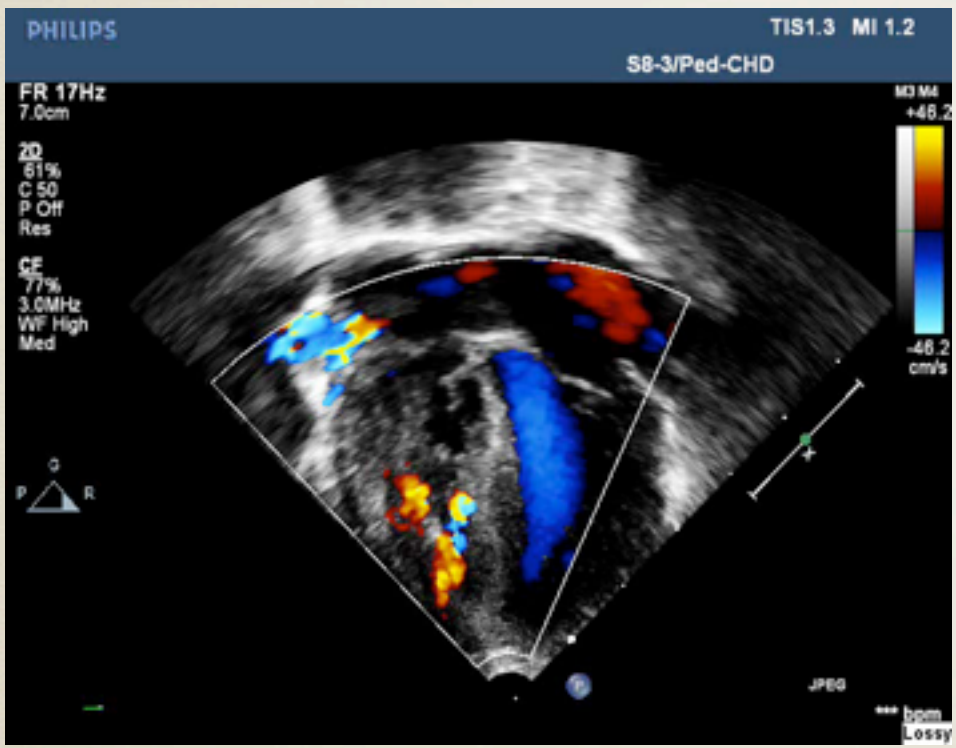
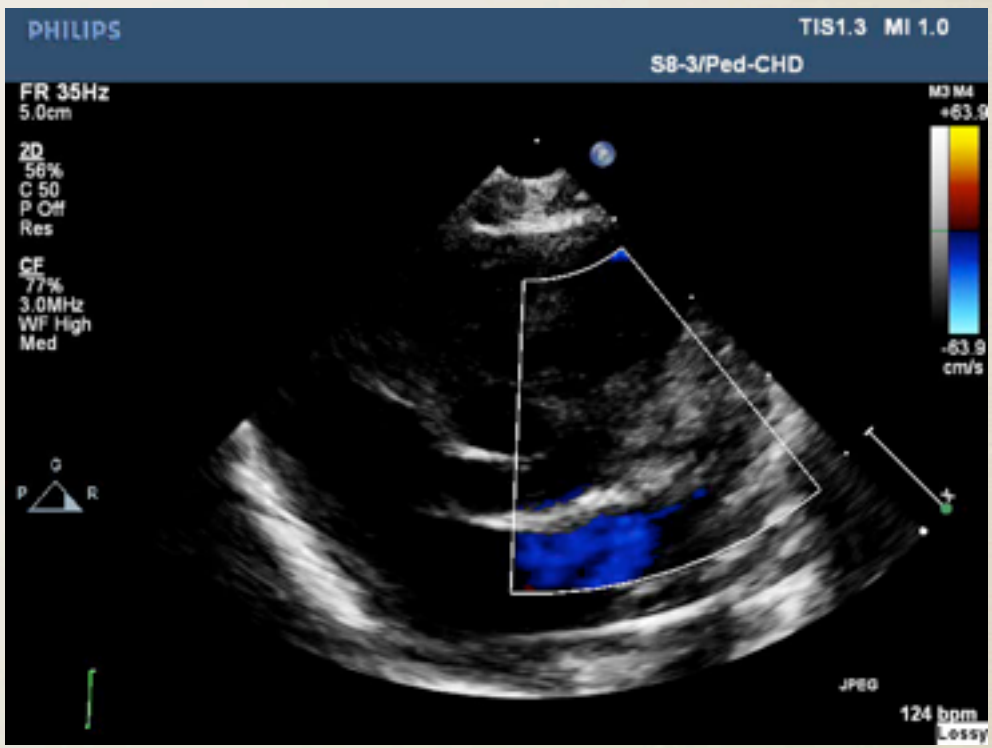
RV coronary connections: Fistulae





Fistulae 46%

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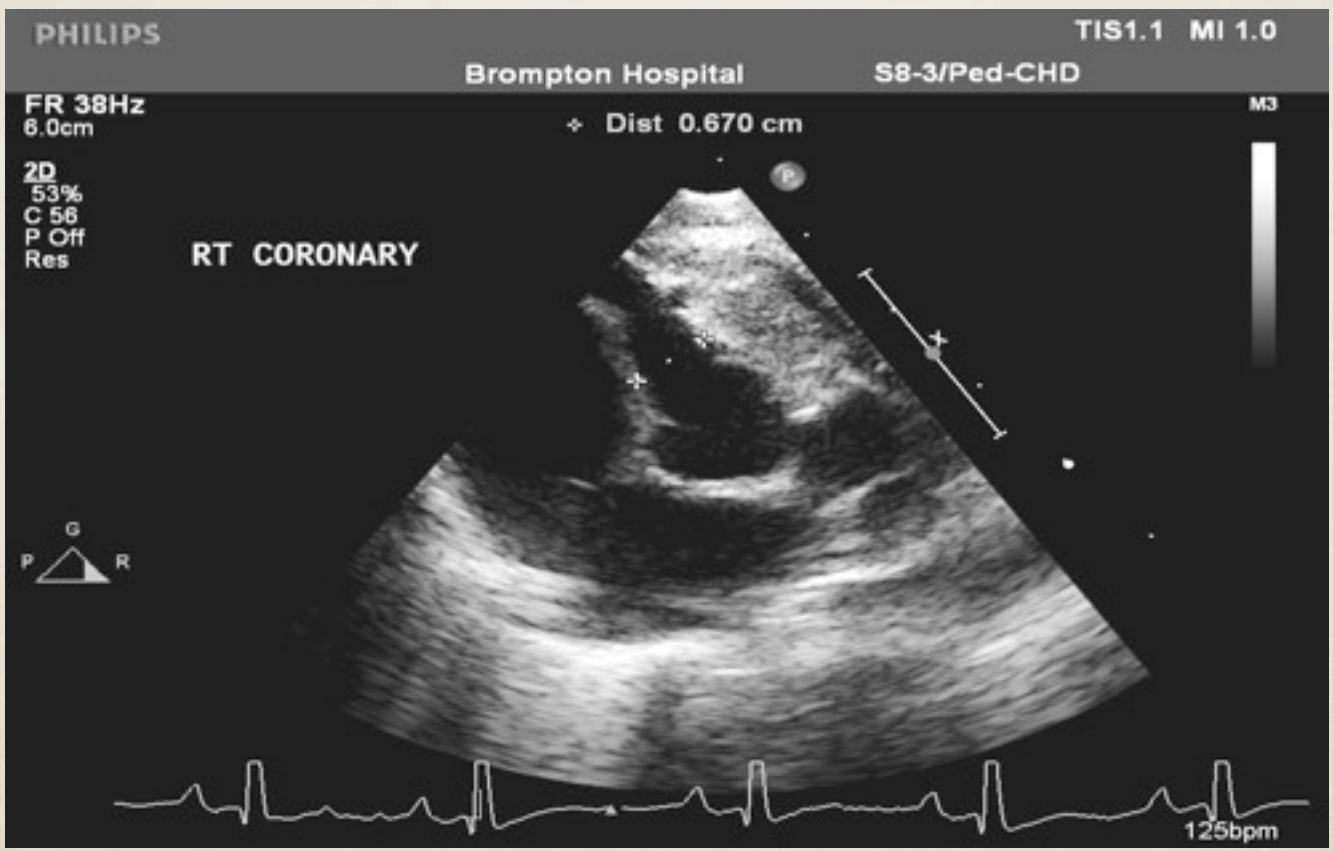
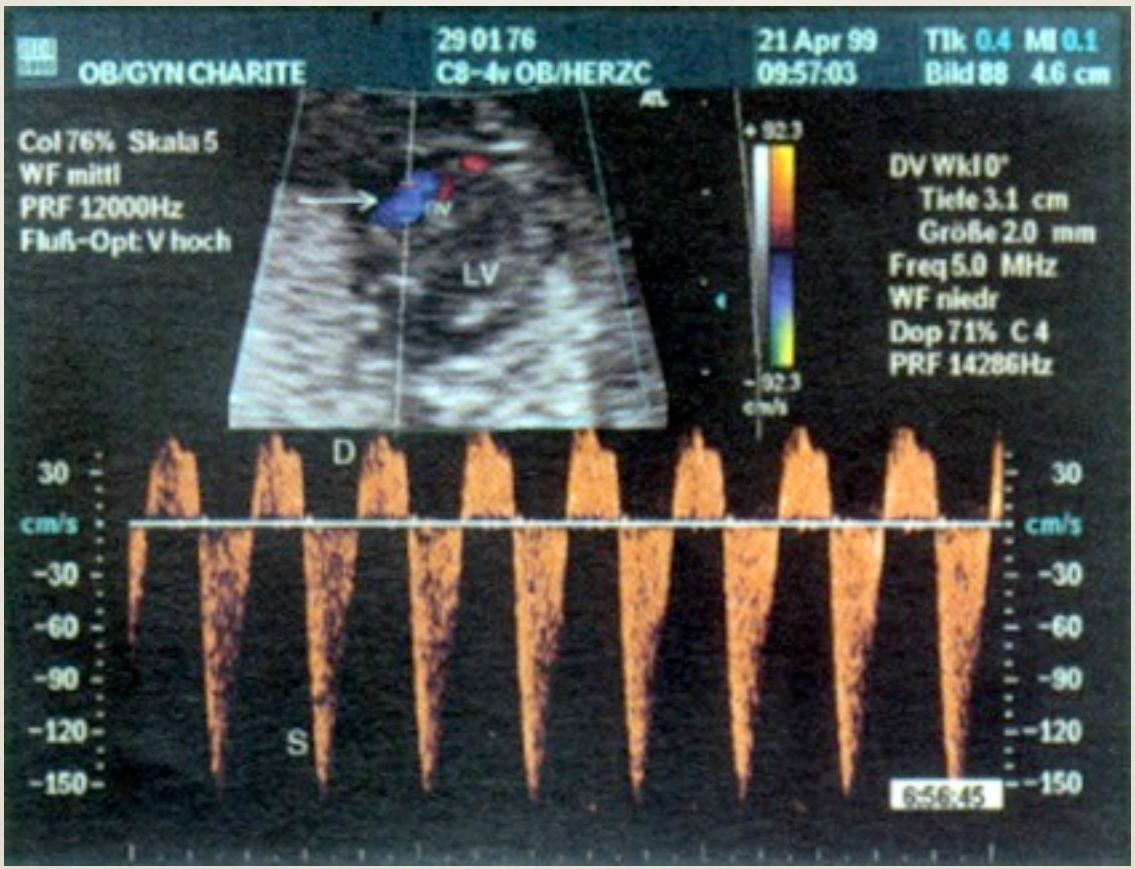
Findings suspicious of fistulae



Fistulae in prenatal life

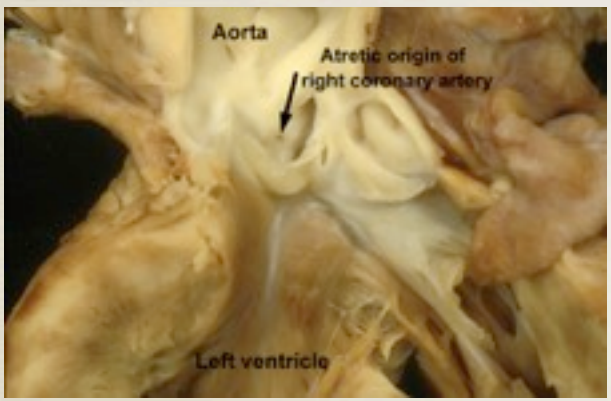


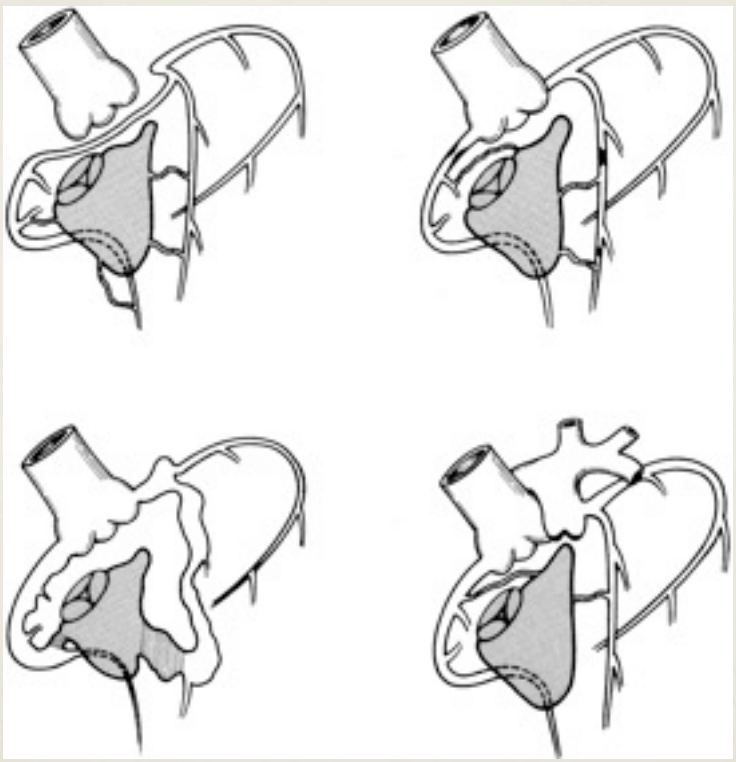
RV fistulae

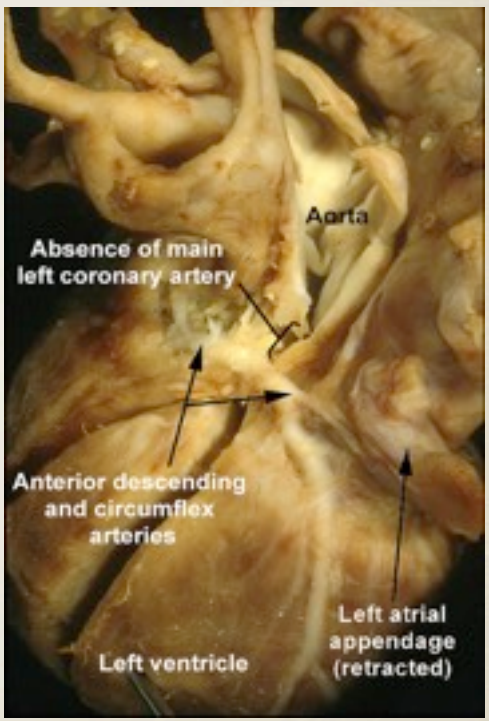
	RV fistulae	RV dep	Frequency
Grade 0	Nil	No	**46%** (Range 25-55%)
Grade 1	Small and insignificant	No	**18%** (8-21%)
Grade 2	Significant, filling the aortic root from RV injection	Few	**20%** (18-23%)
Grade 3	Associated with one interruption in a major coronary artery	Some	**10%** (4-20%)
Grade 4	Associated with interruptions in two major coronary arteries	All	**5%** (2-15%)

Chubb and Daubeney 2012

RV coronary connections: RV dependence







RV dependence 7.5%

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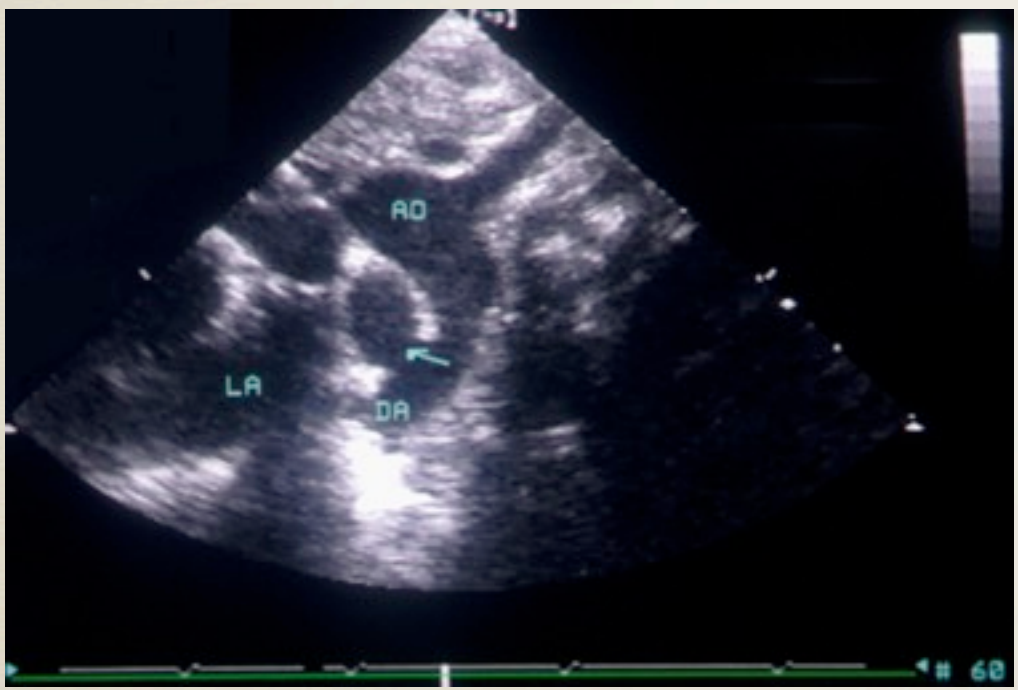
Pulmonary arteries and arterial

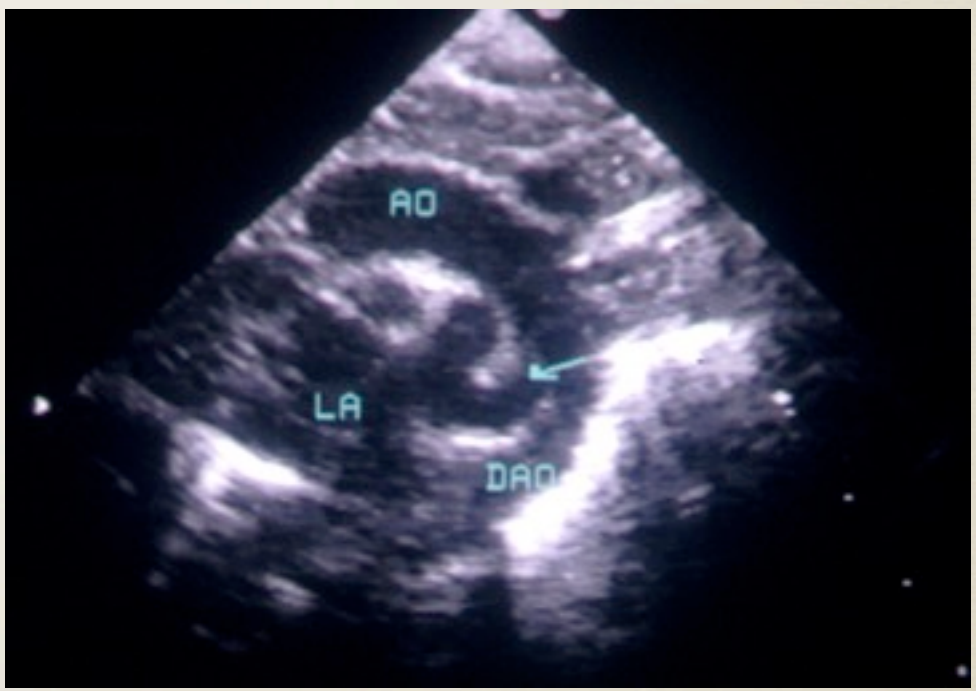
- Pulmonary arteries
  - Normal
  - Hypoplastic
- Arterial duct
  - Normal angled
  - Acute angled (earlier lesion in pregnancy)

Arterial ducts in PAIVS

Normal



Acute Angled



Left heart abnormalities

- Mitral valve
- LV outflow especially septal bulge
- Aortic valve
- LV function global and regional wall abnormalities

Spectrum of morphology: Co-variance

	Dilated RV	Mild RV Hypoplasia	Moderate RV Hypoplasia	Severe RV Hypoplasia
RV morphology	Huge and thin walled	Tripartite	'Bipartite'	'Unipartite'
Timing of closure of PV in utero	Any	Late → Early		
Tricuspid valve	Large (z>0)	Normal/ small (z=0 to -2)	Very small (z=-2 to -5)	Tiny (z<-5)
Angle of arterial duct	Obtuse/ Acute	Obtuse	Obtuse/ Acute	Acute
RV fistulae	Not present	Not present/ mild	Mild/ major	Major/ RVDCC

Chubb and Daubeney 2012

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PAIVS decision-making: Treatment algorithms

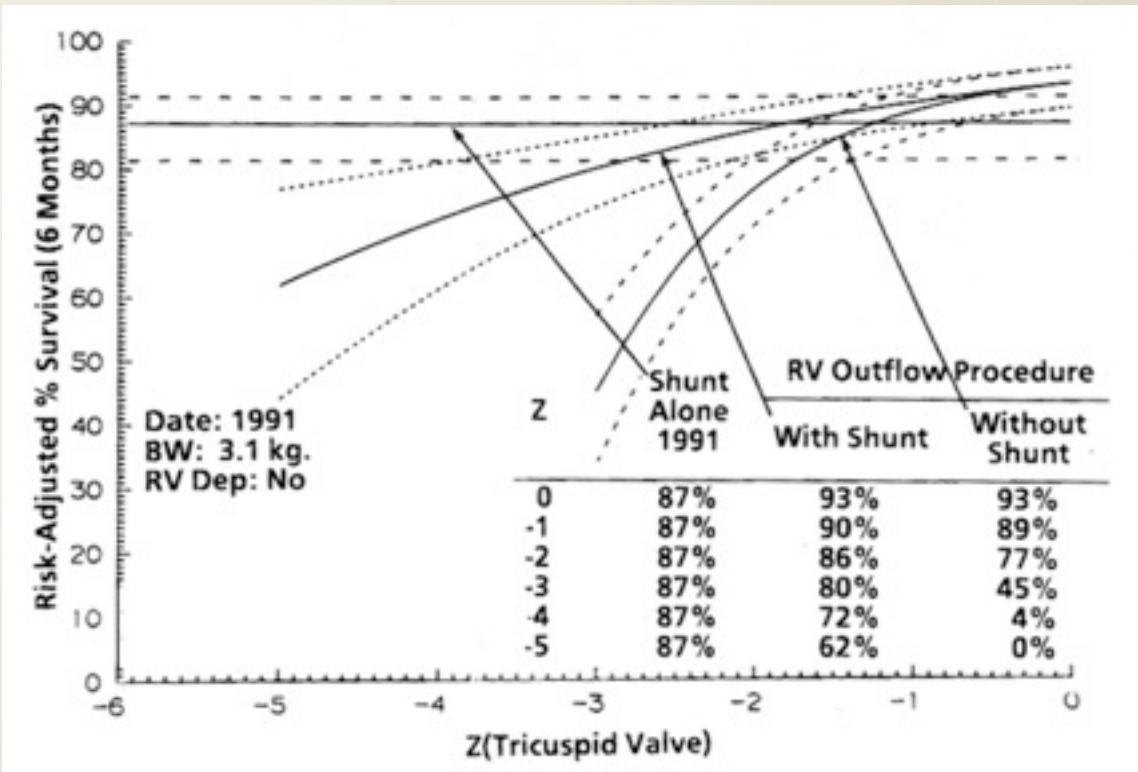
- Selection of best treatment pathway should be individualised
- Biventricular, one-and-a-half and univentricular routes
- Management protocols for neonates continues to be controversial

	Biventricular Route	Borderline	Univentricular Route
TV z-score	>-2.5	-2.5 to -5	<-5
RV Morphology	Tripartite	Bipartite	Unipartite
RVDI	>0.35	<0.35	<0.35 and muscular atresia
Presence of RV infundibulum	Yes	Small and narrow	No
RV fistulae	Nil/Minor	Minor/Major	RVDCC
Treatment at Presentation	RF perforation or closed surgical valvotomy	RF perforation or surgical valvotomy/RVOT patch and consider PDA stent/BTS	Palliative procedure-BTS +/- BAS

Chubb and Daubeney 2012

PAIVS: Congenital heart surgeons study (CHSS) algorithm

- Hanley algorithm based on TV Z score
- Many criticisms: single parameter hinge point v inflow diameter Rowlatt Z scores, over simplistic etc
- But easy to use, most enduring and widely used



THE JOURNAL OF THORACIC AND CARDIOVASCULAR SURGERY

Hanley 1993

A lifetime of specialist care

PAIVS: Prognosis

Study	Number of Patients	Median Length of Follow-Up	% RVDCC	% Biventricular Outcome	% Catheter Valvotomy	Survival at latest follow-up
Daubeney et al (2005)	168	9 years	4.2%	32%	24%	70%
Ashburn et al (2004)	408	10.3years	5%	33%	Nil	59%
Ekman-Joelsson et al (2001)	77	6 years	9% *	41%	Unknown	68%
Hanley et al (1993)	171	4 years	9%	32%	Nil	64%
Moller (2010)	1039	At 1 year	Unknown	Unknown	14%	80%

Risk factors for poor outcome

* Toronto: Fistulae, weight (lower), RVp/LVp lower, Ebstein malformation
* GOSH: TV Z score (-2.4 to -5), weight (lower)
* CHSS: TV Z score (smaller), weight (lower), RV dependence, earlier date
* UK & Ireland: RV inlet Z score (smaller), date (earlier), Ebstein malformation
* Sweden: Male (worse), birth weight (lower), muscular atresia,

Late management

- Aim to separate the circulations, abolish cyanosis, close shunts (ASD/ BT shunt), minimise gradients/ regurgitation
- Can be achieved in a biventricular, 1.5 or univentricular circulation
- Biventricular repair- late concerns:
  - Pulmonary and tricuspid stenosis and regurgitation
  - Atrial septal defect
  - RV hypoplasia- if significant then 1.5 ventricle repair
  - Atrial and ventricular arrhythmias

PAIVS: Late echo assessment

* Growth of TV and RV: assessing suitability for biventricular repair
* Ventricular function
* Functionality of shunts
* Growth of pulmonary arteries
* Direction of flow through ASD/ PFO
* Restriction of RV
* Pulmonary and tricuspid regurgitation
* RV dilatation

PAIVS: Conclusions

* Rare disease
* Considerable morphologic variation
* Echo can document most if not all morphologic features
* Echo can guide decision-making in neonatal period
* Often catheterisation/ MRI needed as well
* Decision-making should be based on assessment of all morphologic features
* Echo important for ongoing assessment