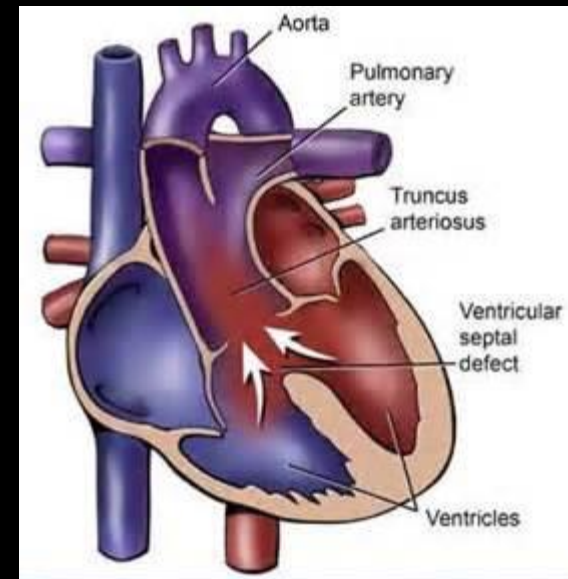
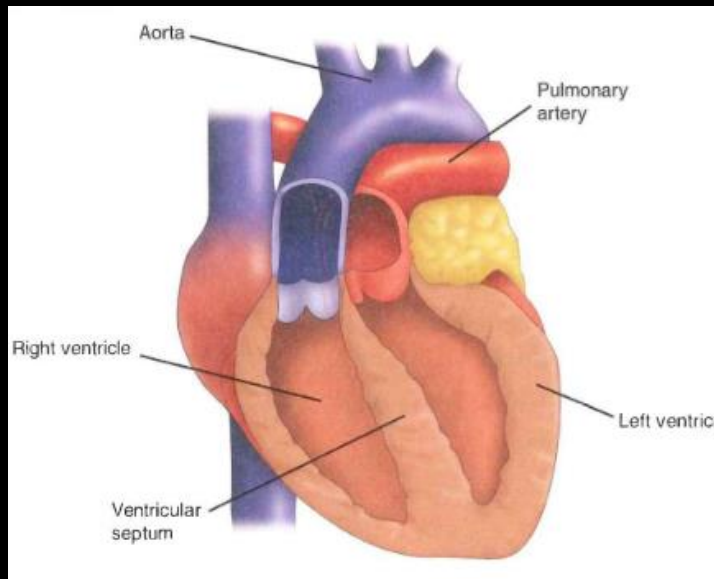
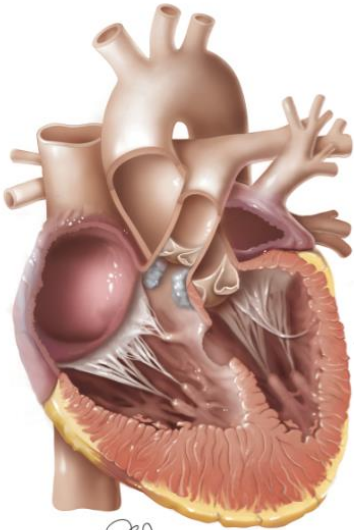


Key Points in Prenatal Imaging: TOF, TGA, and Truncus Arteriosus



Mary T. Donofrio MD, FAAP, FACC, FASE
Professor of Pediatrics
Director of the Fetal Heart Program
Director of the Critical Care Delivery Program
Children's National Health System
Washington DC

Prevalence of Serious CHD

♥ Nationwide Inpatient Sample (NIS) database

- n=9,696,908 births over 10 years (1999-2008)

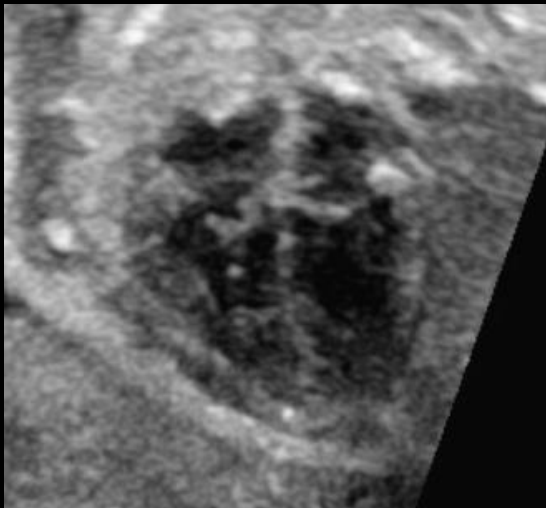
♥ Prevalence/100,000

- Serious CHD- 147
- TOF- 34.7
- TGA- 21.9
- TA- 20.7

CHD	1999–2000 n (%)	2001–2002 n (%)	2003–2004 n (%)	2005–2006 n (%)	2007–2008 n (%)	p value
Tetralogy of Fallot	684 (38.1)	622 (37.8)	608 (34.7)	588 (35.6)	582 (34.1)	0.001 ^a
Truncus arteriosus	402 (22.6)	372 (21.2)	382 (20.7)	364 (19.6)	328 (18.1)	0.02 ^a
d-TGA	368 (20.7)	402 (22.6)	396 (21.9)	392 (20.4)	496 (20.4)	0.3
Double-outlet right ventricle	306 (17.2)	332 (18.6)	296 (17.4)	310 (17.1)	388 (18.0)	0.5
HLHS	244 (13.8)	226 (12.6)	212 (11.7)	188 (9.8)	194 (8.8)	0.001 ^a
Pulmonary atresia	216 (12.1)	196 (10.9)	184 (10.2)	176 (9.5)	192 (9.6)	0.01 ^a
Tricuspid atresia	188 (10.6)	176 (9.8)	198 (10.3)	202 (10.4)	242 (9.9)	0.1
Interrupted aortic arch	184 (10.4)	212 (9.8)	196 (10.1)	188 (9.9)	264 (10.4)	0.09
Ebstein anomaly	132 (7.4)	142 (7.9)	152 (7.7)	198 (7.1)	202 (.2)	0.07
TAPVR	102 (5.7)	110 (6.1)	106 (5.9)	138 (6.1)	198 (5.7)	0.2
Single ventricle	92 (5.2)	96 (5.4)	92 (5.5)	96 (5.9)	122 (5.2)	0.4
L-TGA	74 (4.2)	82 (4.6)	81 (4.3)	84 (4.4)	106 (4.9)	0.2
Total CHD	2,992 (168.9)	2,958 (161.5)	2,844 (157.1)	2,864 (138.7)	2,634 (129.3)	0.03 ^a
Live births	1,772,012	1,786,442	1,804,946	1,923,734	2,409,774	

Objectives

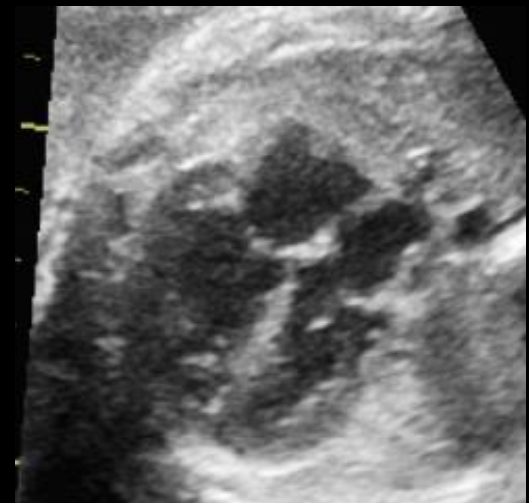
- ♥ Define the features of TOF, TGA, and Truncus
 - Diagnosis
 - Additional Details
- ♥ The challenge



TOF



TGA



Truncus

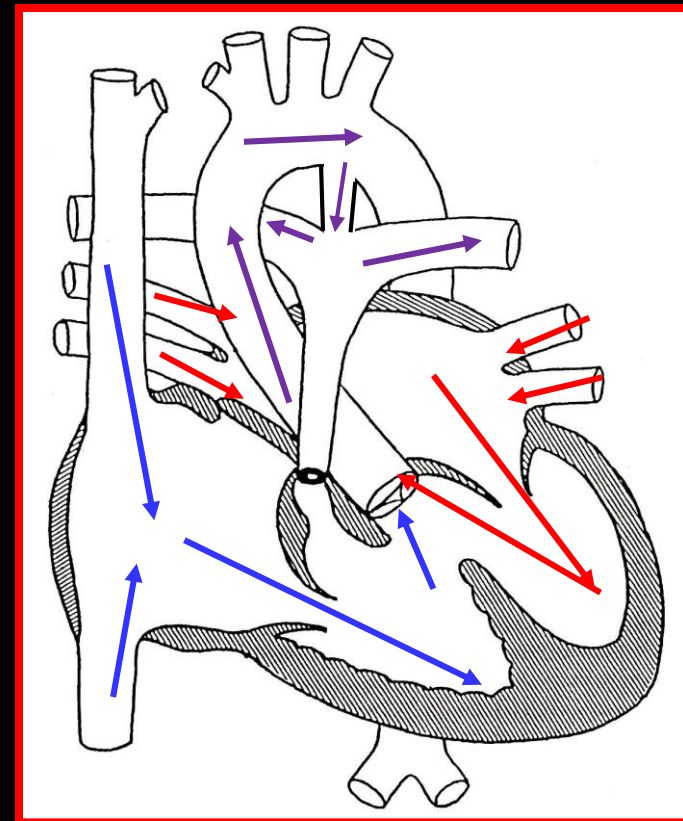
Tetralogy of Fallot

♥ Defining features

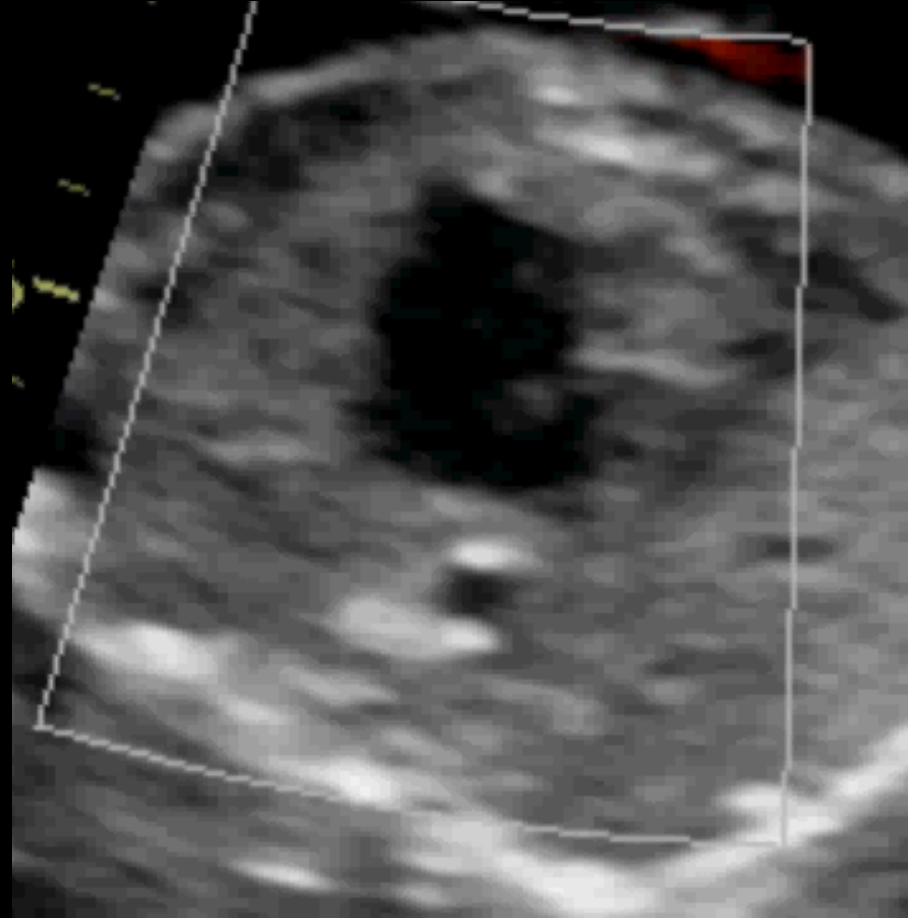
- Anterior malalignment of the conal septum
 - Subpulmonary/pulmonary obstruction
- Overriding aorta
- VSD
- (RVH)

♥ Details

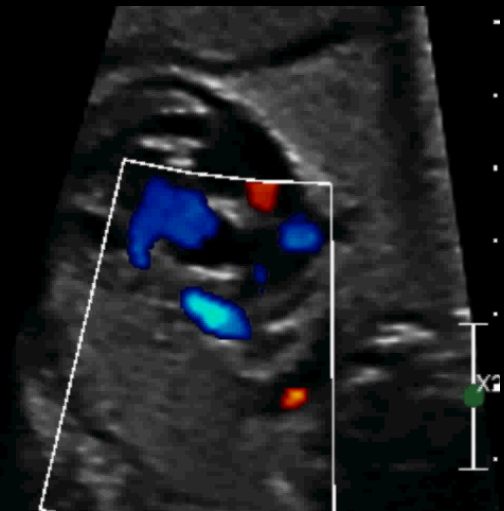
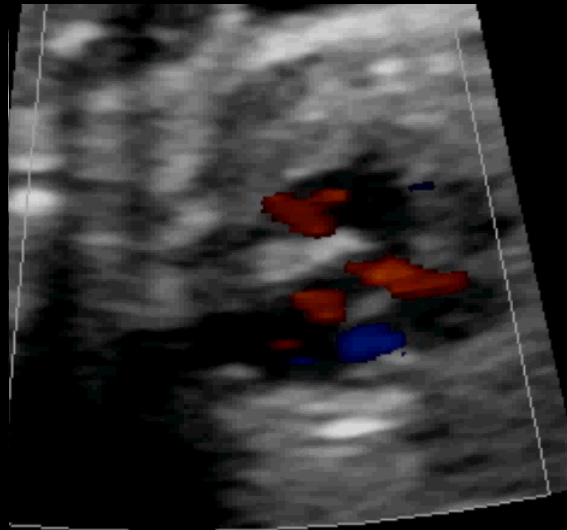
- Degree of pulmonary obstruction
 - Ductal dependent?
- Arch sidedness
- More complex disease
 - TOF with AV canal
 - TOF/PA with collaterals
 - TOF with absent PV



Tetralogy of Fallot



Tetralogy of Fallot



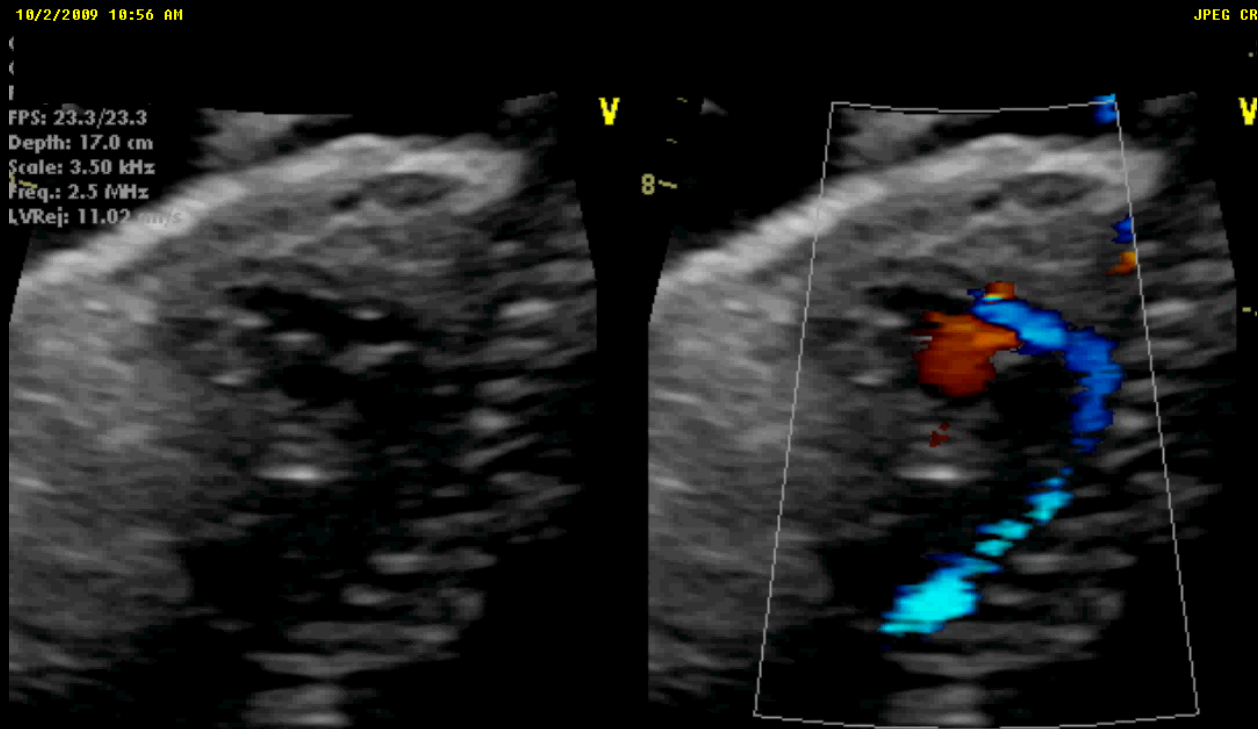
Tetralogy of Fallot

Pink vs. Blue

- ♥ Fetal predictors for need for neonatal repair
 - 33 with TOF/ 6 DORV

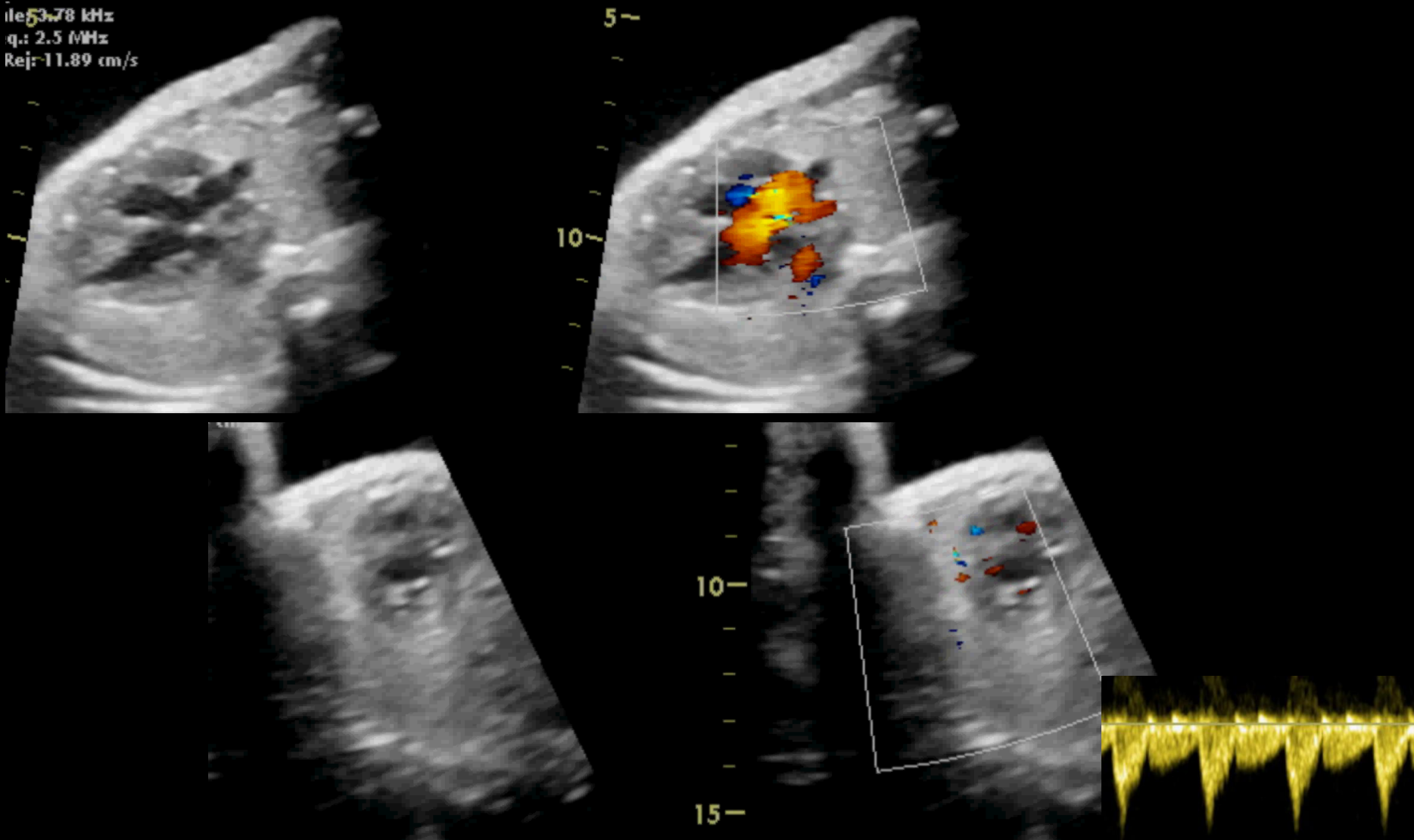
<i>Parameter</i>	<i>Sensitivity (%)</i>	<i>Specificity (%)</i>
PV-Z-score < -3 or PV/AoV ratio < 0.6		
Early-gestation (< 24 weeks)	100	50
Mid-gestation (24-32 weeks)	100	48
Final echo (mean age, 34 weeks)	92	50
Ductal flow pattern		
Early-gestation (< 24 weeks)	75	100
Mid-gestation (24-32 weeks)	88	94
Final echo (mean age, 34 weeks)	100	95

Tetralogy of Fallot and the Ductus Arteriosus

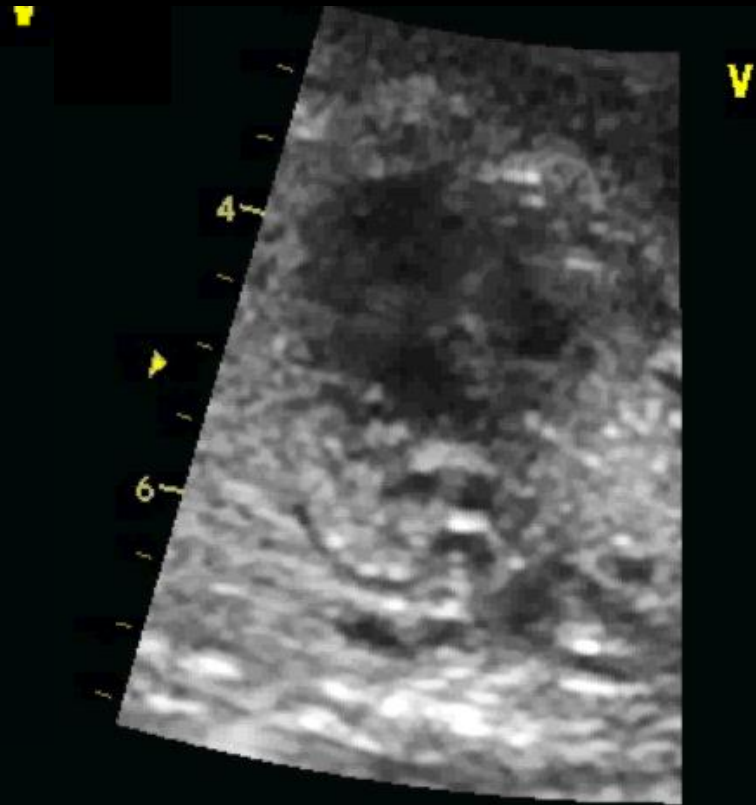
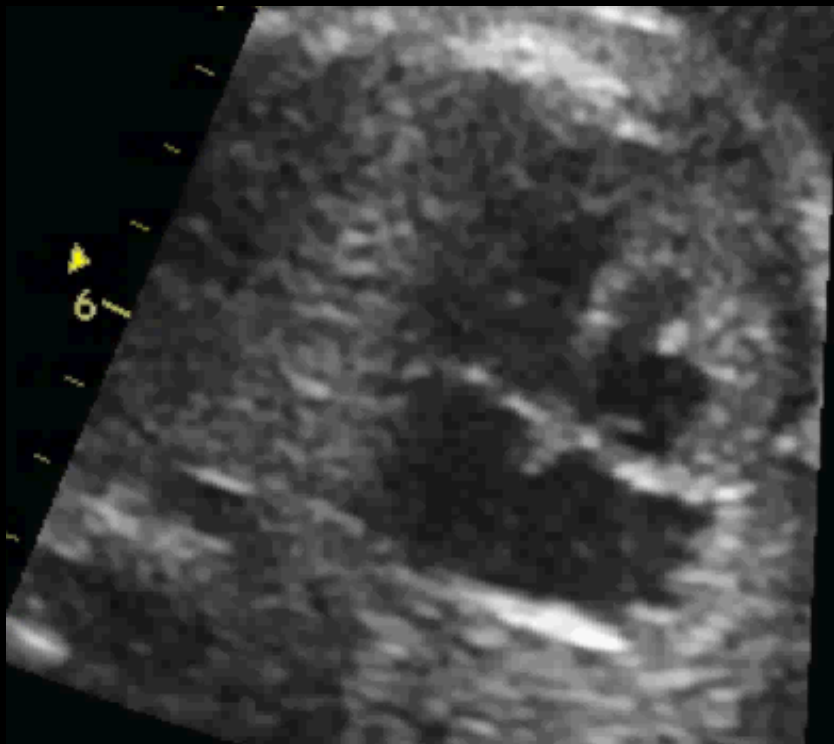


Tetralogy of Fallot and the Ductus Arteriosus

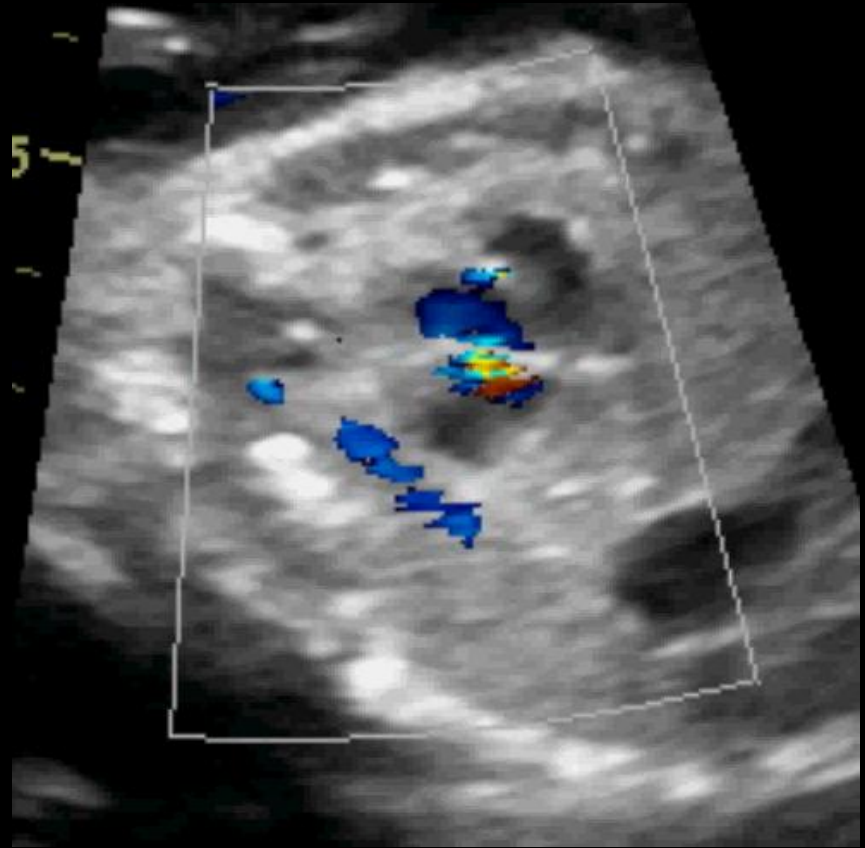
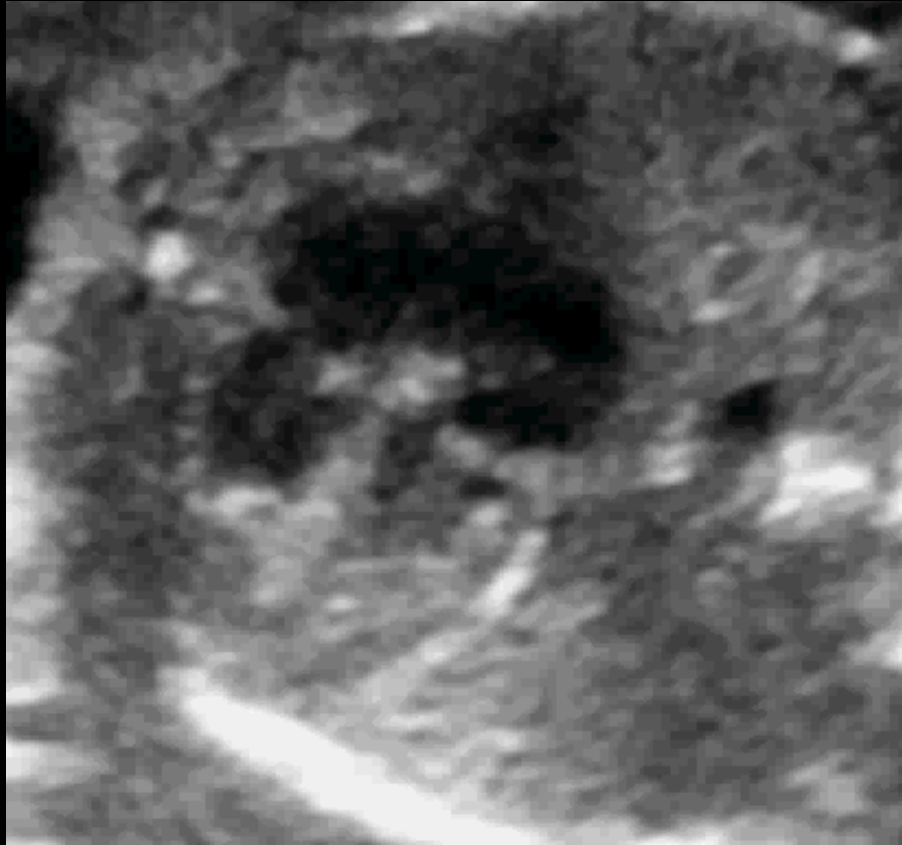
ile 53.78 kHz
q.: 2.5 MHz
Rej.: 11.89 cm/s



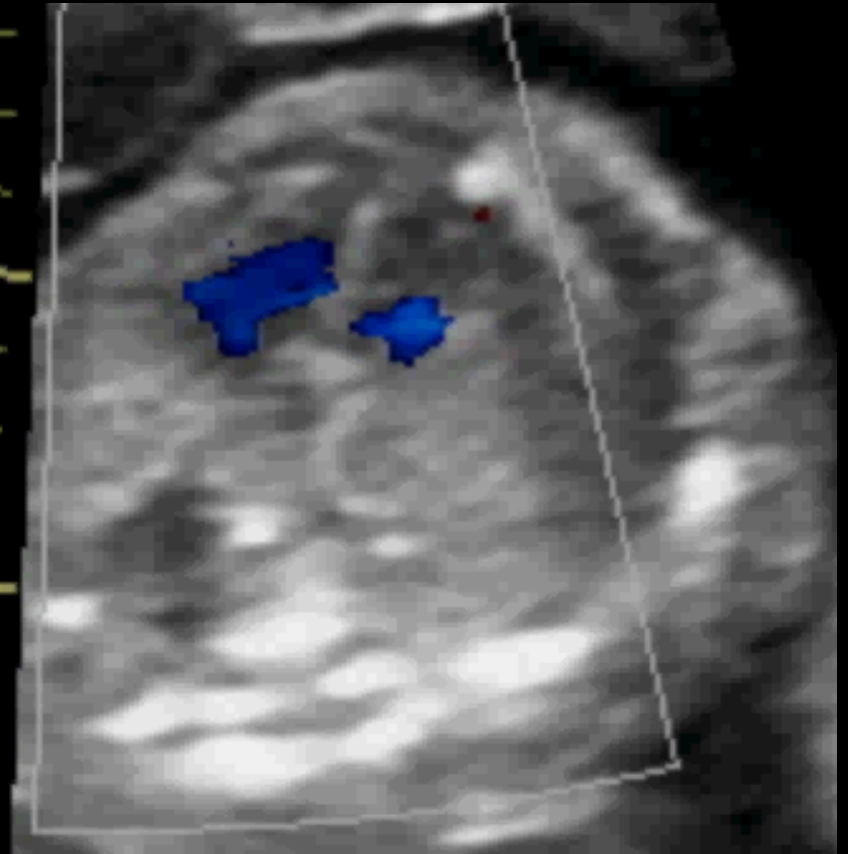
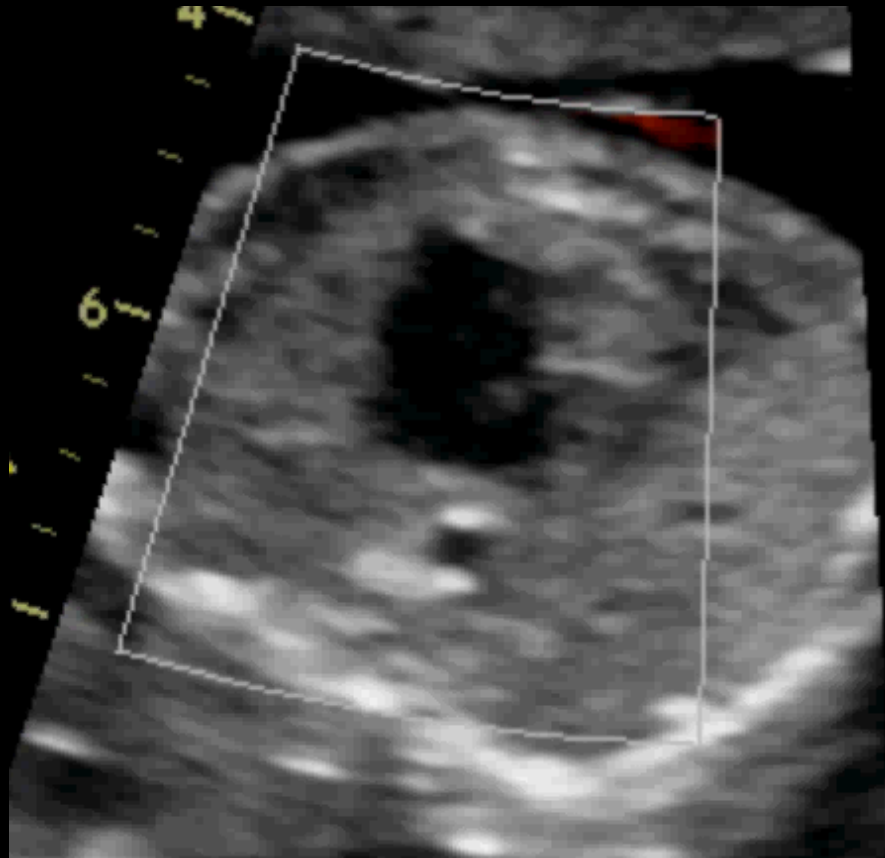
Tetralogy of Fallot and the Ductus Arteriosus



Tetralogy with Pulmonary Atresia

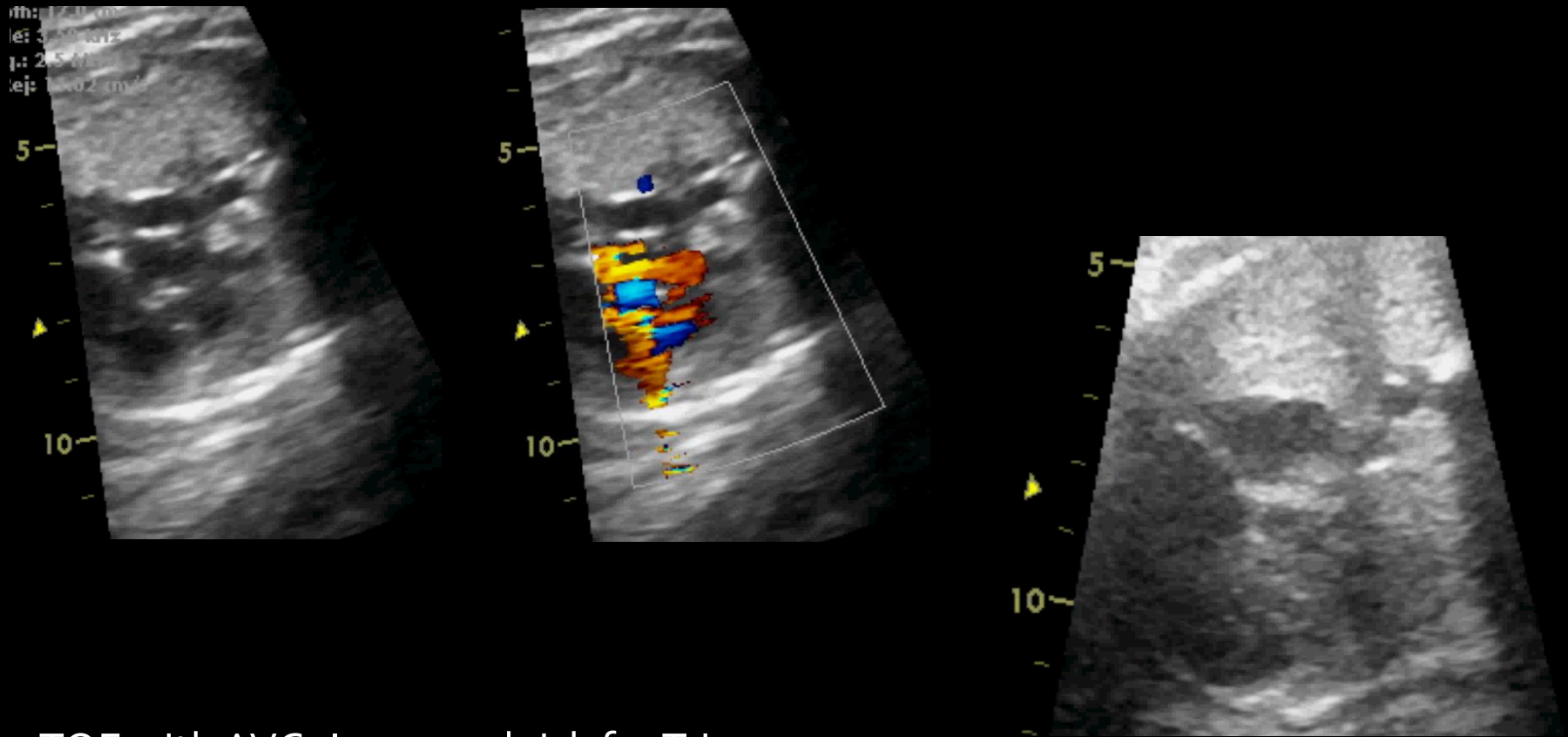


Tetralogy of Fallot and the Aortic Arch



Right arch- Increased risk for DiGeorge

Tetralogy with AV Canal



TOF with AVC- Increased risk for Trisomy 21

Tetralogy of Fallot with Absent Pulmonary Valve

♥ Defining features

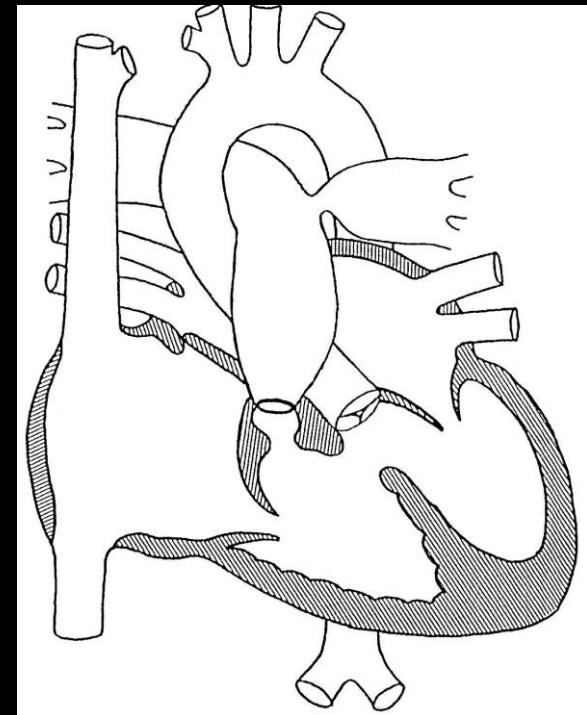
- Anterior malalignment of the conal septum, VSD, overriding aorta
- Dysplastic PV with severe insufficiency
- Marked dilation of the PAs

♥ Clinical presentation

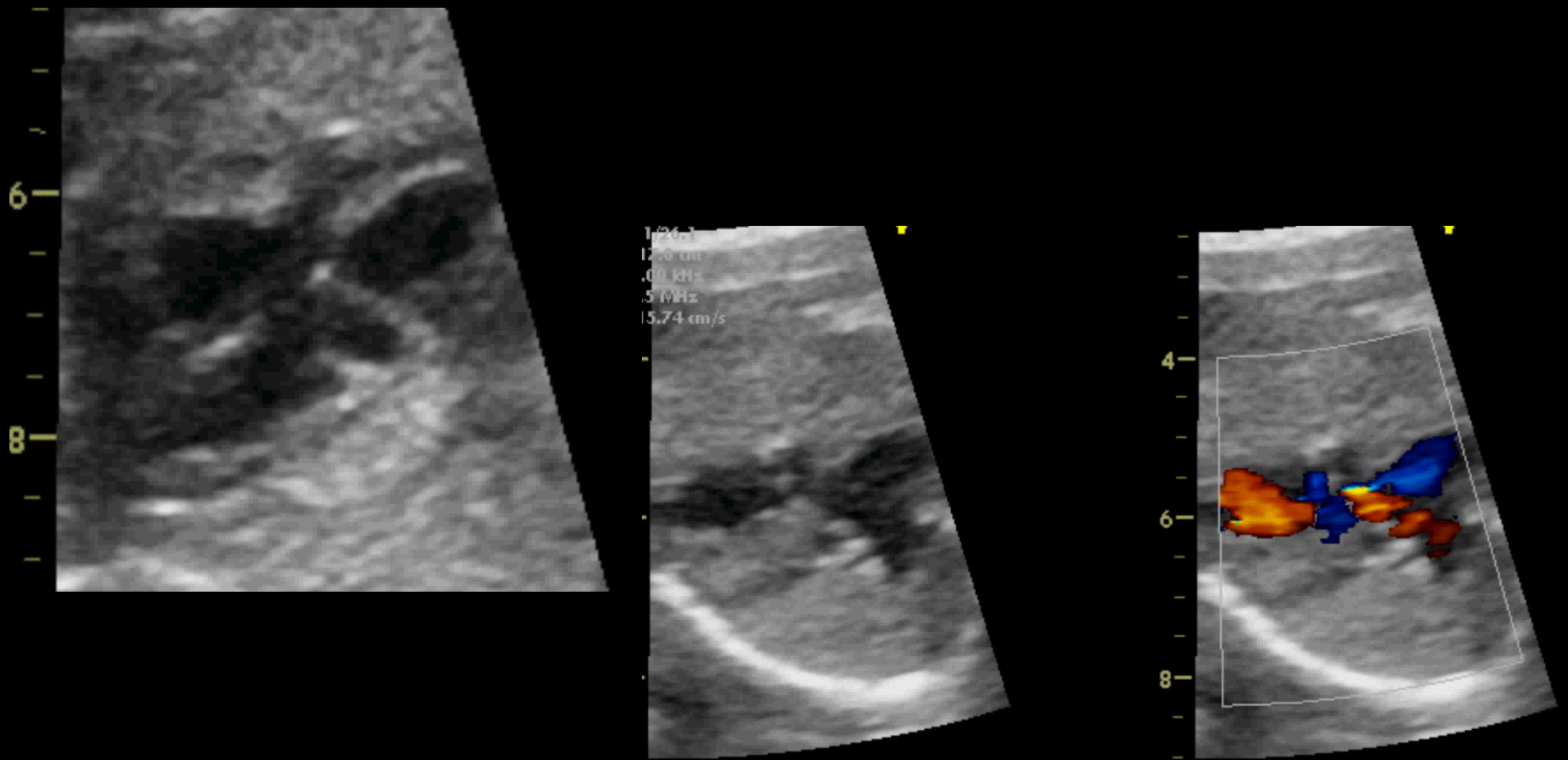
- Respiratory distress at birth (40-50%)
 - Tracheobronchial malacia, lung abnormalities
- No respiratory symptoms
 - Straightforward TOF clinical course

♥ Fetal/neonatal mortality- 40-70%

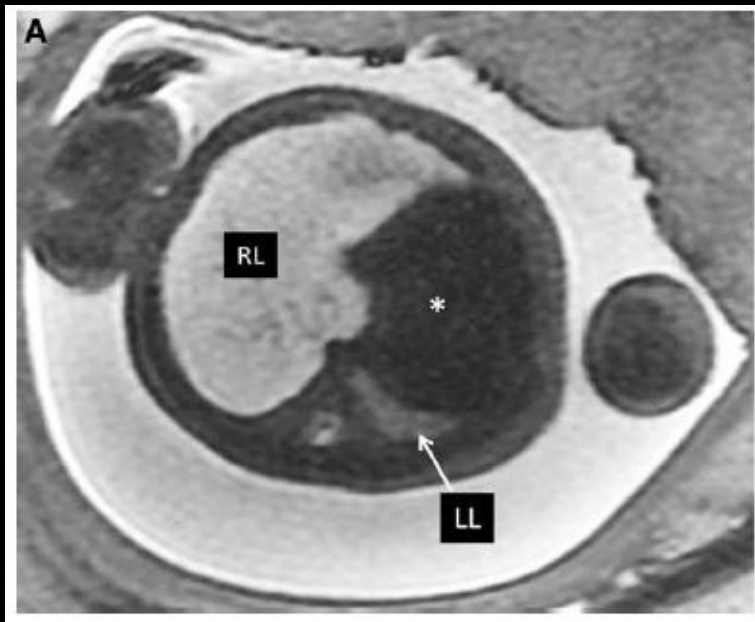
- Morbidity/mortality predictors
 - Associated malformations/chromosome abnormalities
 - Ventricular dysfunction, hydrops fetalis



Tetralogy of Fallot with Absent Pulmonary Valve



Tetralogy with Absent Valve Lung Pathology

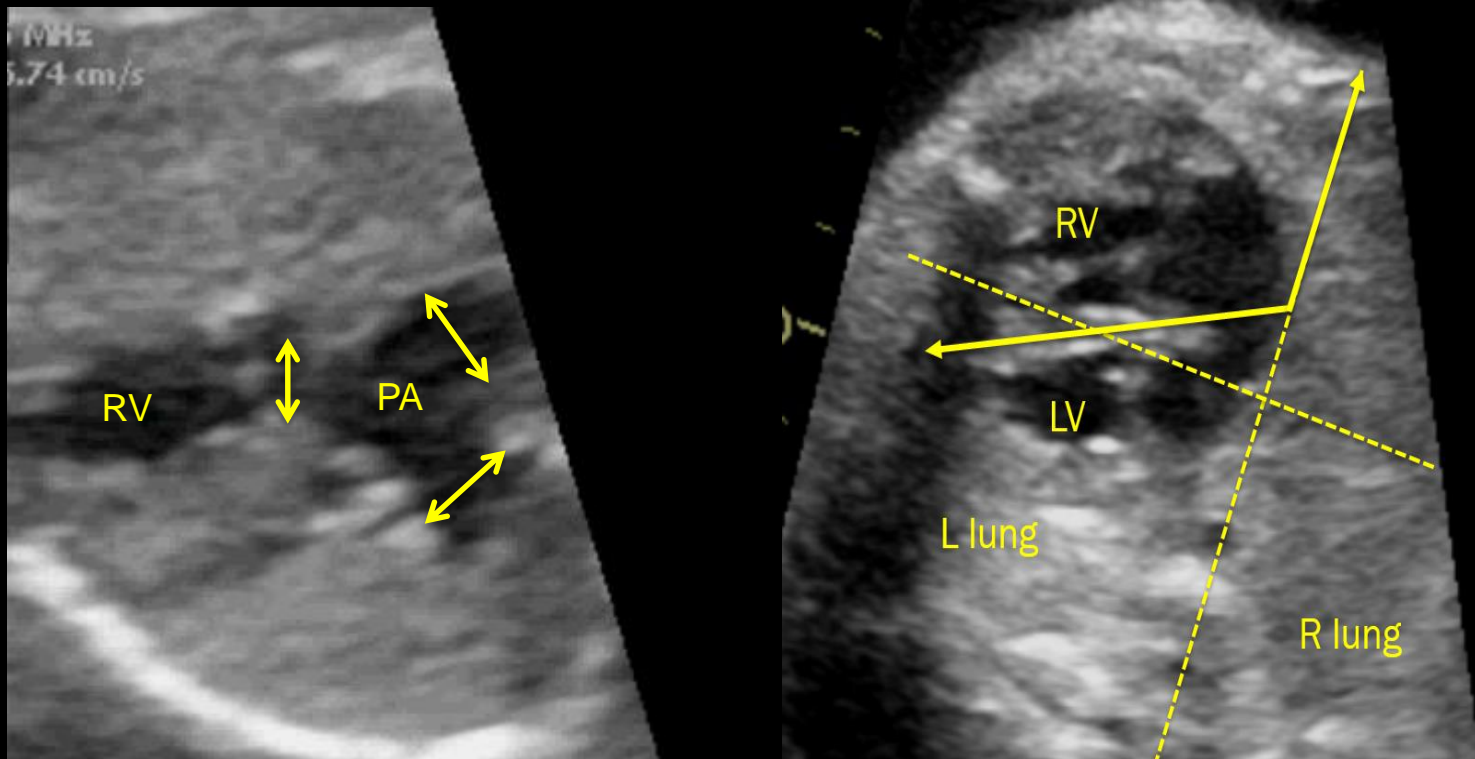


Fetal TOF/APV







Multicenter Collaborative Study

♥ Methods

- Retrospective analysis over 10 years, 2002-2012
- 78 patients, 19 institutions

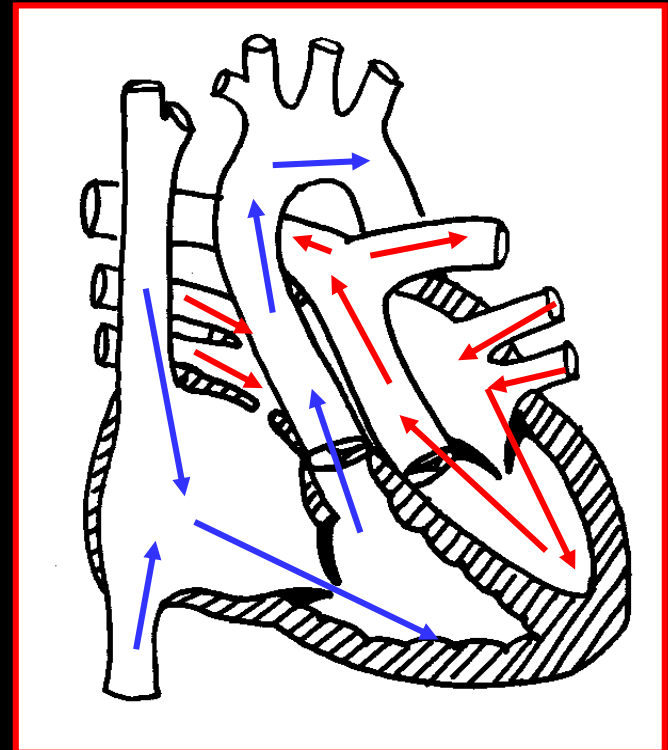


Fetal Findings Associated with Early Mortality

Echo Finding	Fetal or Postnatal Death (n=26)	Survived (n=42)	P-Value
Pulmonary valve diameter (median, cm)	0.57	0.50	0.09
Main pulmonary artery diameter (median, cm)	1.25	1.10	0.47
Right pulmonary artery diameter (median, cm)	1.00	1.10	0.73
Left pulmonary artery diameter (median, cm)	1.10	1.00	0.09
 Mediastinal shift	17 (65%)	16 (38%)	0.03
Abnormal cardiac axis (<30° or >75°)	13 (of 25; 52%)	14 (33%)	0.10
 Moderate or severe RV dilation	19 (73%)	13 (31%)	0.001
 LV dilation	6 (23%)	2 (5%)	0.05
 Cardiothoracic area ratio	0.37	0.33	0.01
 RV dysfunction	12 (46%)	3 (7%)	<0.001
 LV dysfunction	7 (27%)	2 (5%)	0.02

Transposition of the Great Arteries

- ♥ Defining features
 - Transposed great arteries
- ♥ Details
 - Foramen ovale
 - Ductus arteriosus
 - More complex disease
 - VSD
 - PS
 - AS with coarctation
 - L-TGA



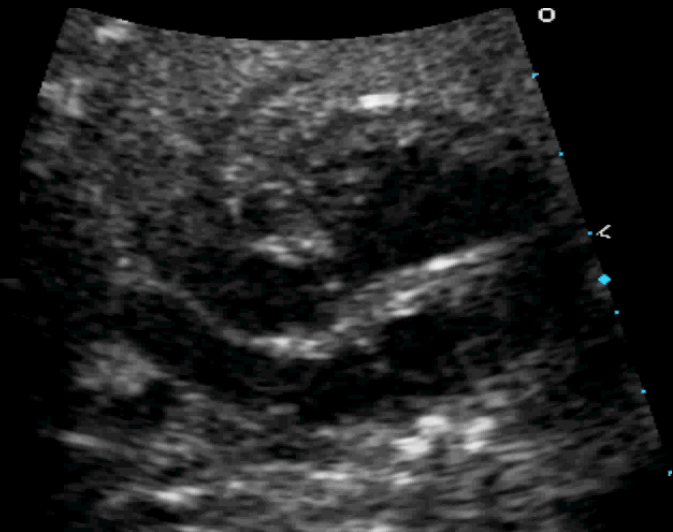
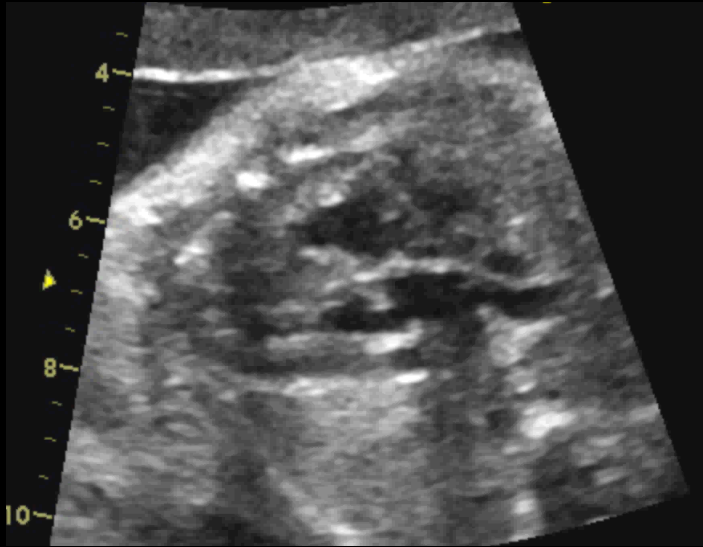
D-TGA: Diagnosis

♥ Diagnostic clues

- Abnormal long axis (or 5-chamber view)
 - Lateral branching artery originating from the LV
- Parallel course of the ductal and aortic arches
 - Lack of crossing outflow tracts
- Abnormal 3 Vessel view
 - Ascending aorta reaches more anterior than the pulmonary artery
 - 2 vessels seen instead of 3 (transverse arch and SVC)



D-TGA: Outflow Tracts

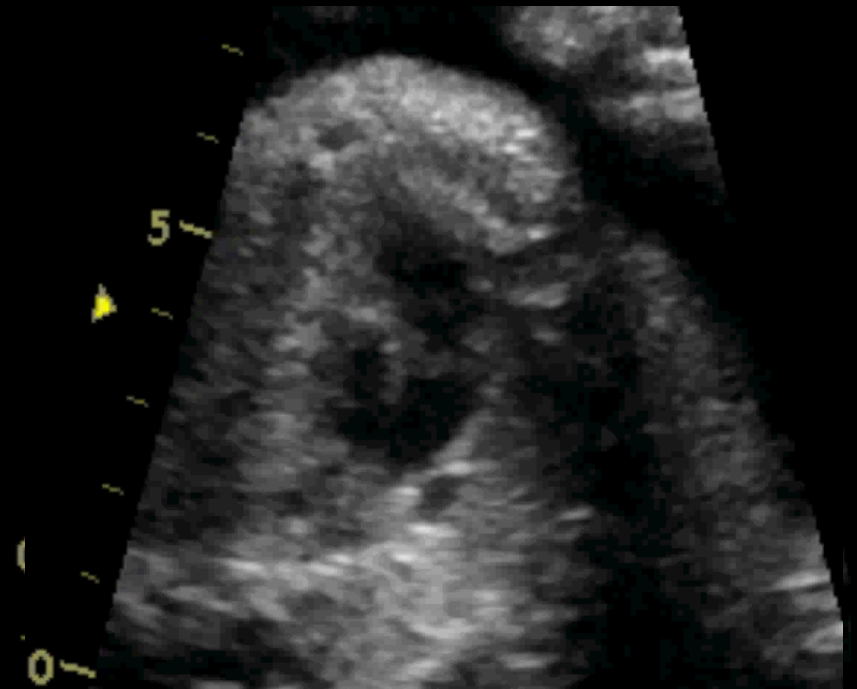


D-TGA: Three Vessel View

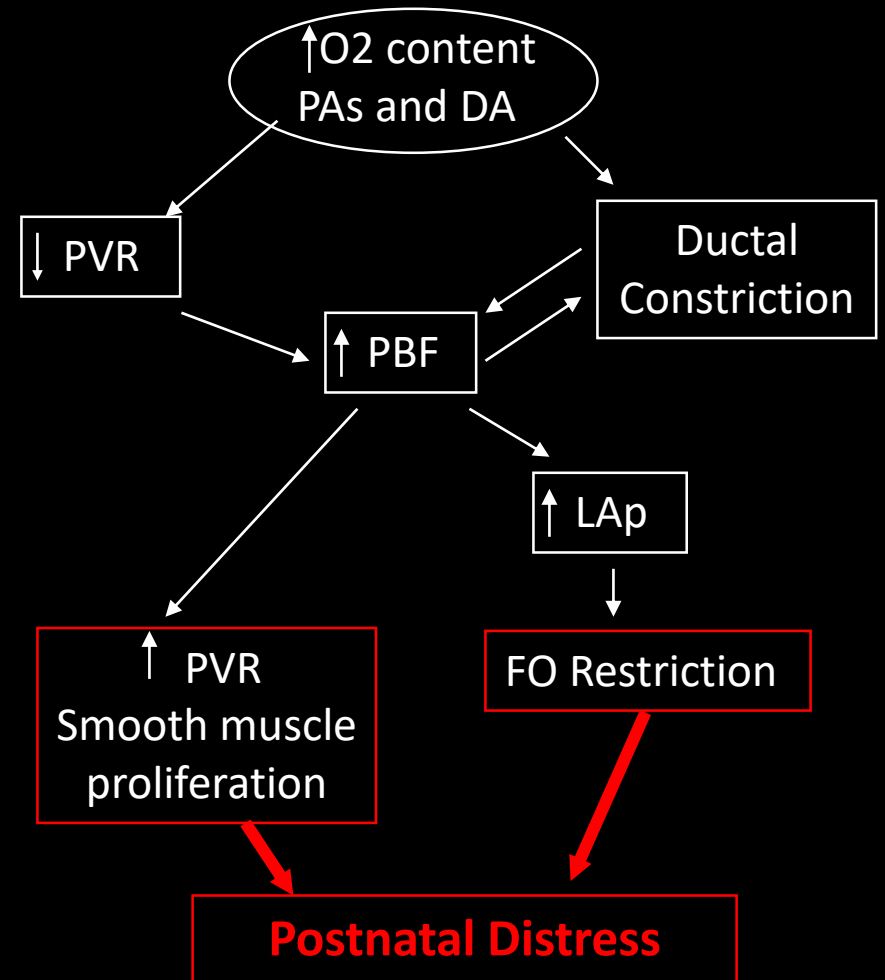
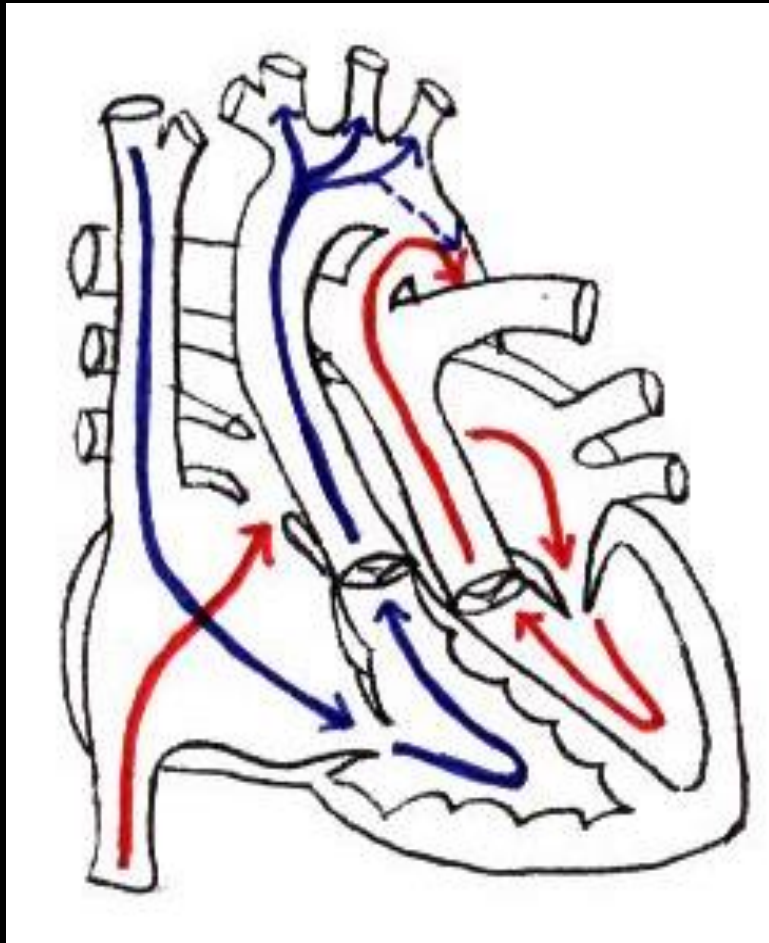
Normal



TGA- 2 Vessels



Fetal Circulation in TGA

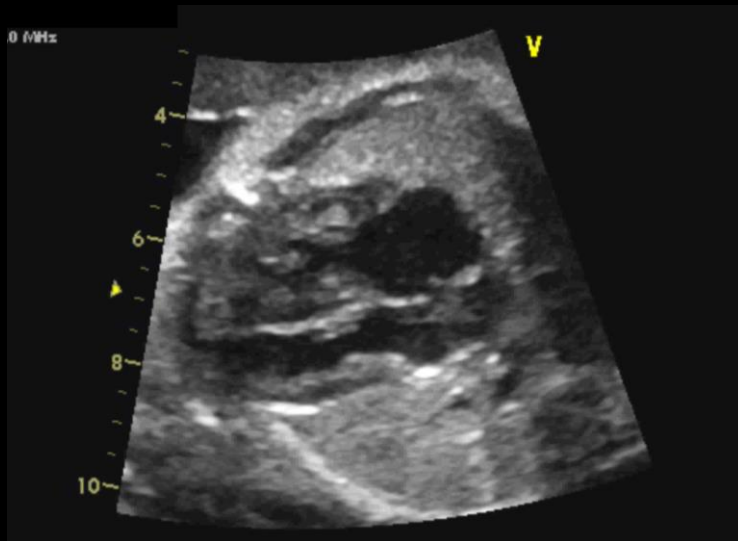


Transposition of the Great Arteries

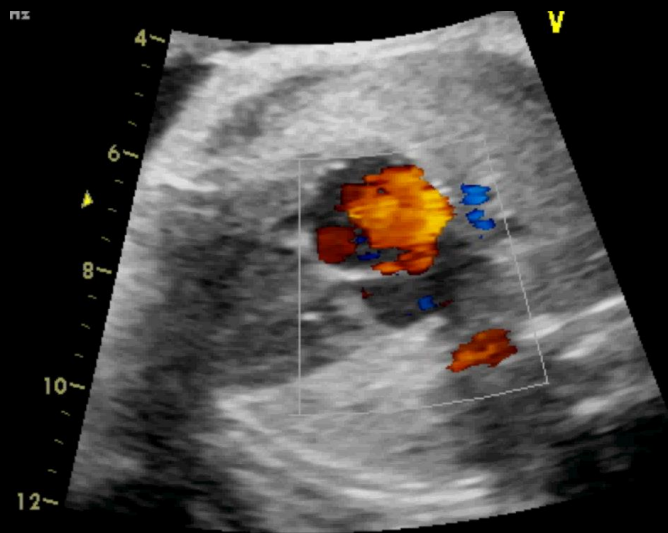
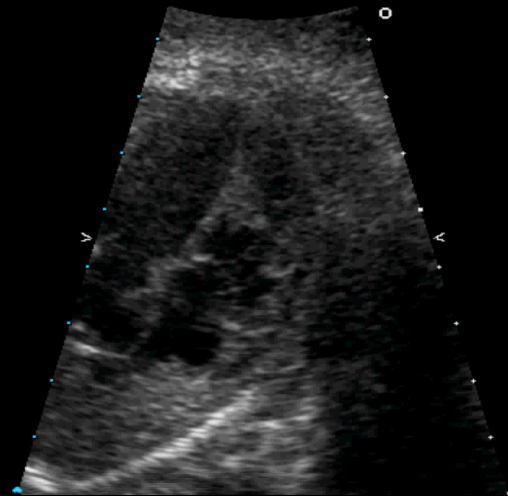
Prediction of Need for BAS

- ♥ Foramen ovale abnormalities
 - Septum primum – abnormal if bulged $>50\%$ to the LA
 - Angle of septum primum – abnormal if $<30\%$ septum
 - Motion- abnormal if lack of swinging motion
 - Hypermobility
- ♥ Ductus arteriosus abnormalities
 - Abnormal size
 - Abnormal flow (continuous high velocity, bidirectional, reversed)
- ♥ Prediction of neonatal emergency
 - Sensitivity approximately 50-60%
 - Specificity up to 100%

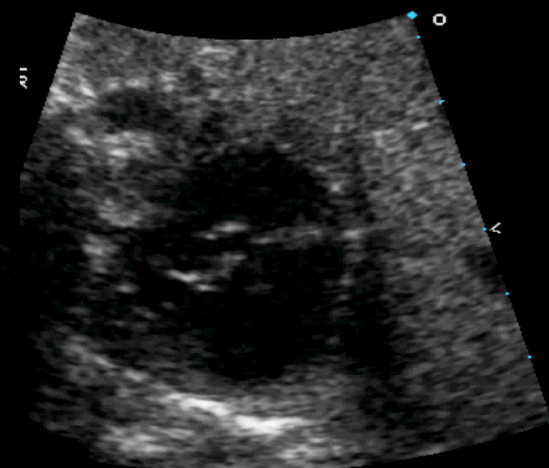
TGA: The Foramen Ovale



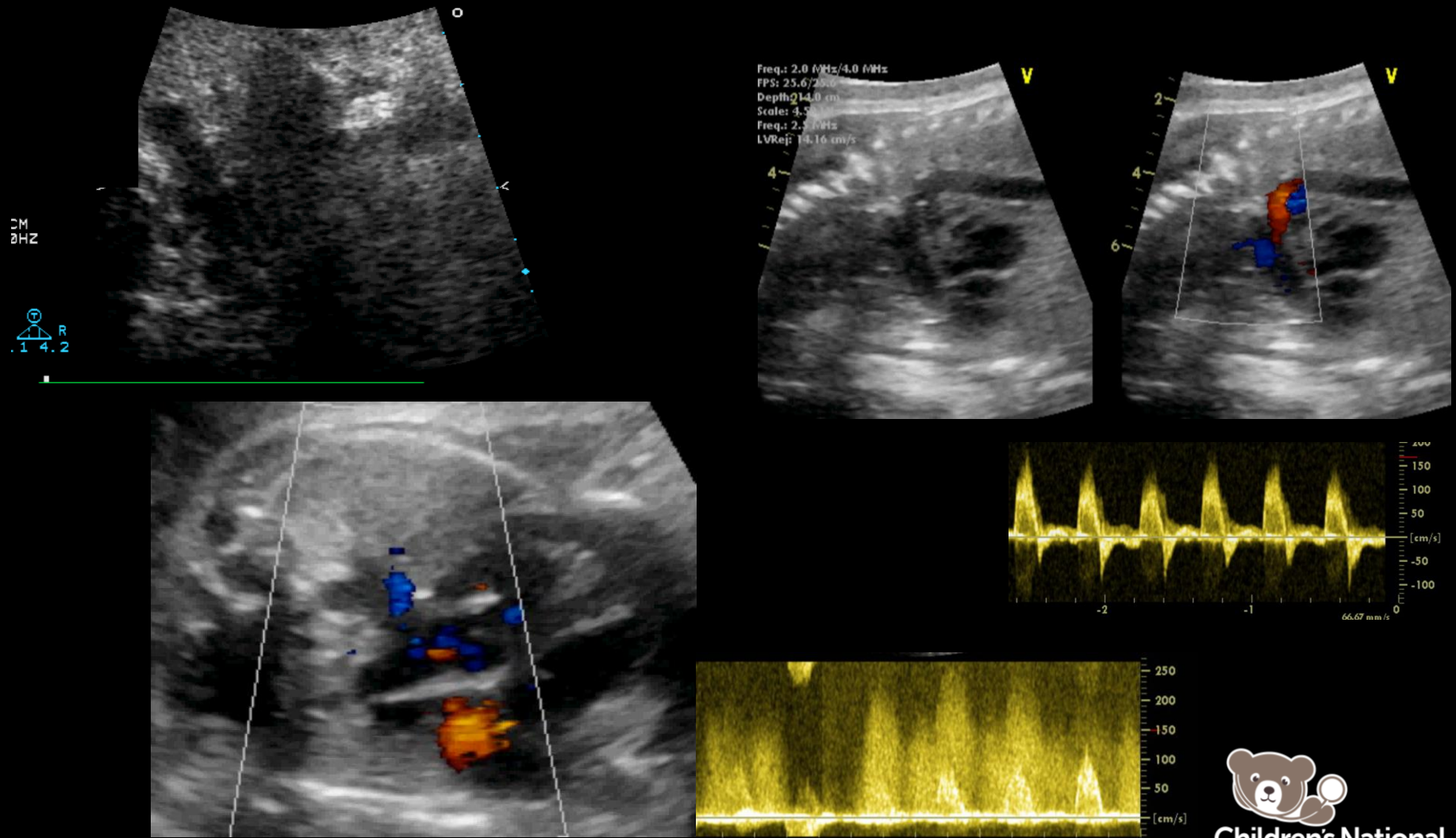
14CM
69HZ



DM
0HZ



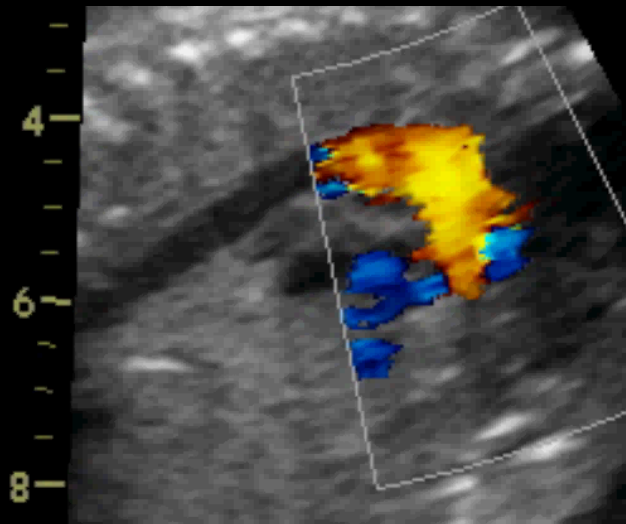
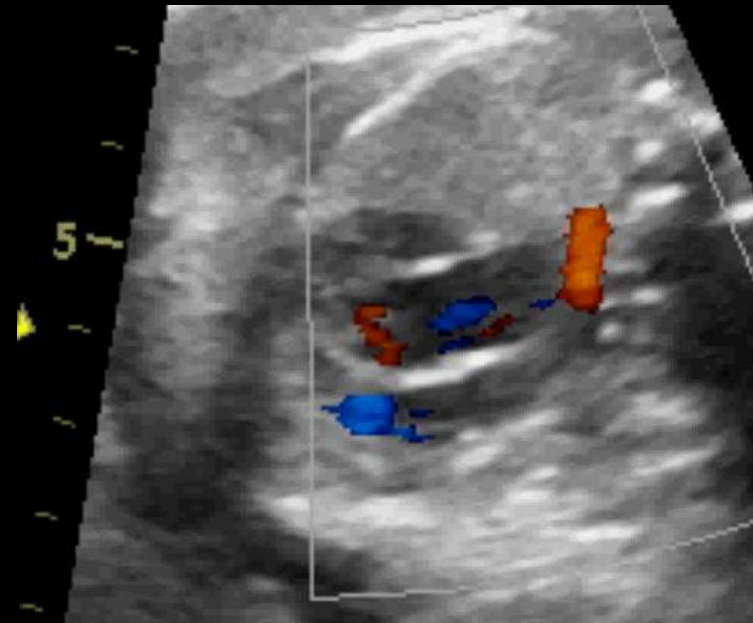
TGA: The Ductus Arteriosus



D-TGA: Intact FO/No DA



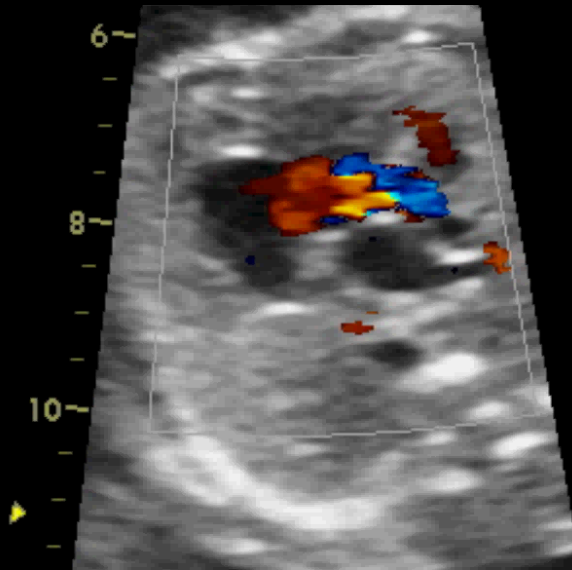
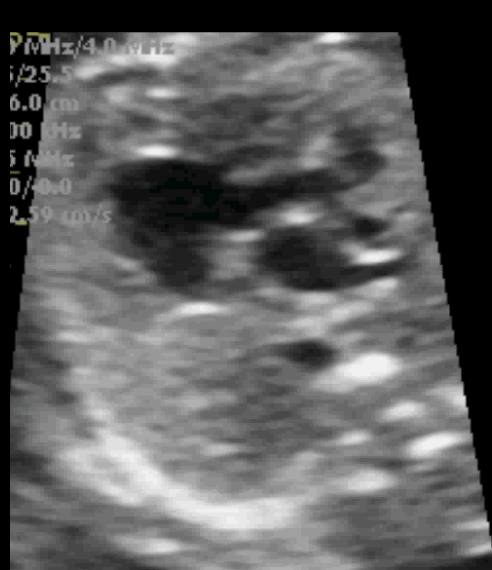
Associated Defects: TGA with VSD/ Outflow Obstruction



L-TGA

♥ Ventricular inversion with TGA

- Associations
 - AV block
 - Other CHD including VSD, PS, AS, SV



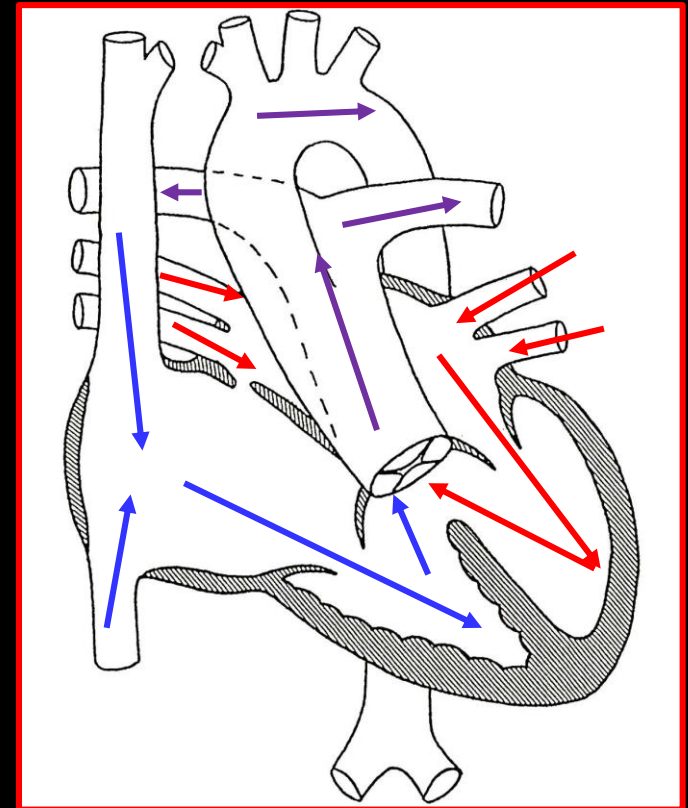
Truncus Arteriosus

♥ Defining features

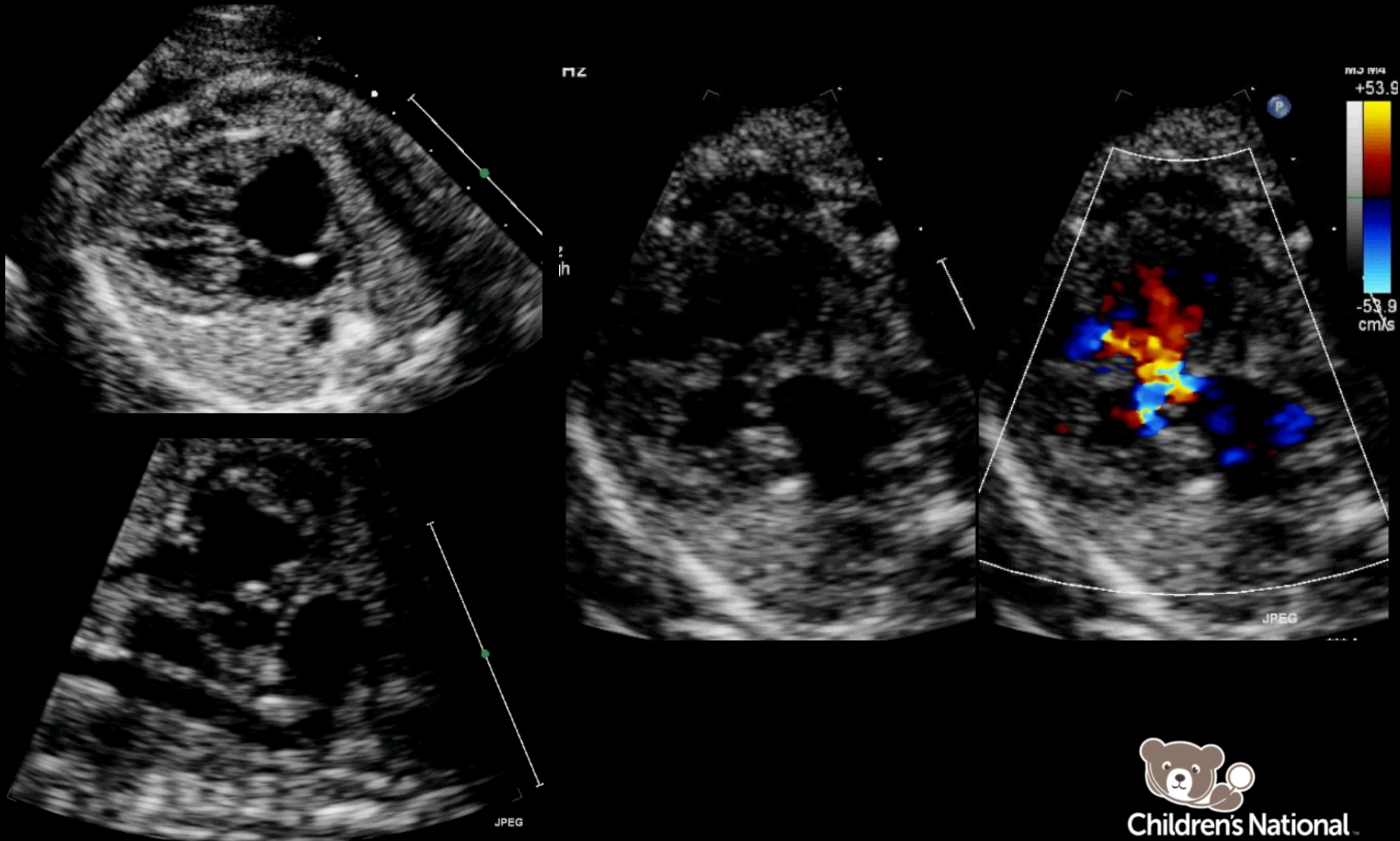
- Overriding aorta
- VSD
- No pulmonary valve, PAs from arch
 - Distinguish from TOF/PA

♥ Details

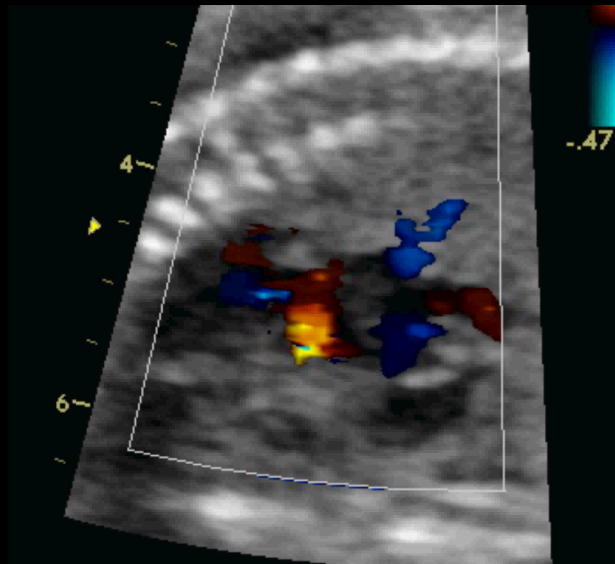
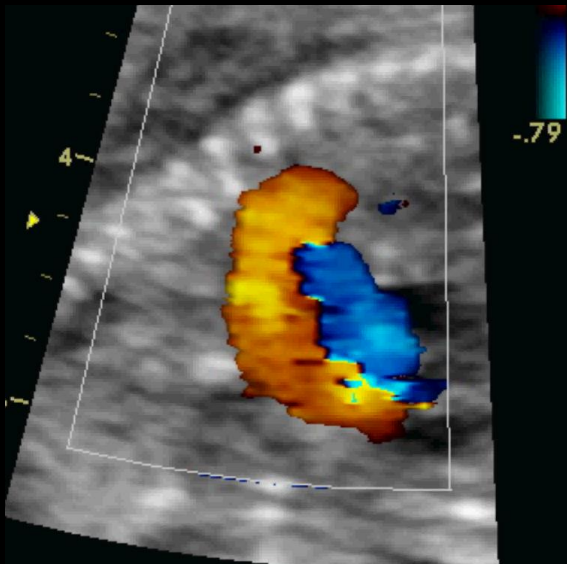
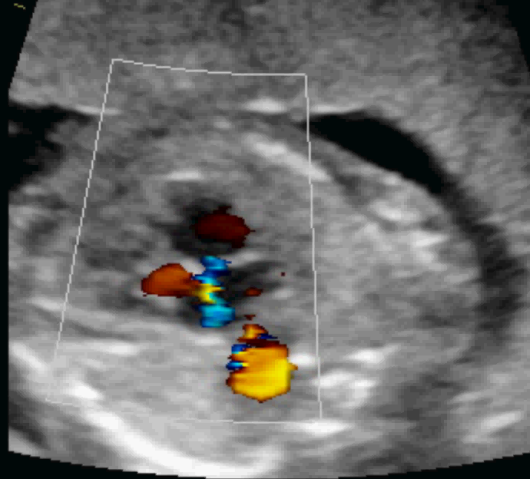
- Severity of truncal stenosis or insufficiency
- Associated interrupted aortic arch



Truncus Arteriosus



Truncus Arteriosus with Interrupted Aortic Arch



Conclusion

- ♥ Key Points in Prenatal Imaging for Tetralogy of Fallot, TGA, and Truncus
 - Normal 4-chamber view
 - Abnormal outflow tracts
 - Look for associated findings

