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Fetal Cardiology Education Series: Cardiac Evaluation of the Hydropic Fetus

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BEST CHILDREN'S HOSPITALS U.S.News & WORLD REPORT RANKED IN 10 SPECIALTIES 2019-20

BEST CHILDREN'S HOSPITALS U.S.News & WORLD REPORT CARDIOLOGY & HEART SURGERY 2019-20

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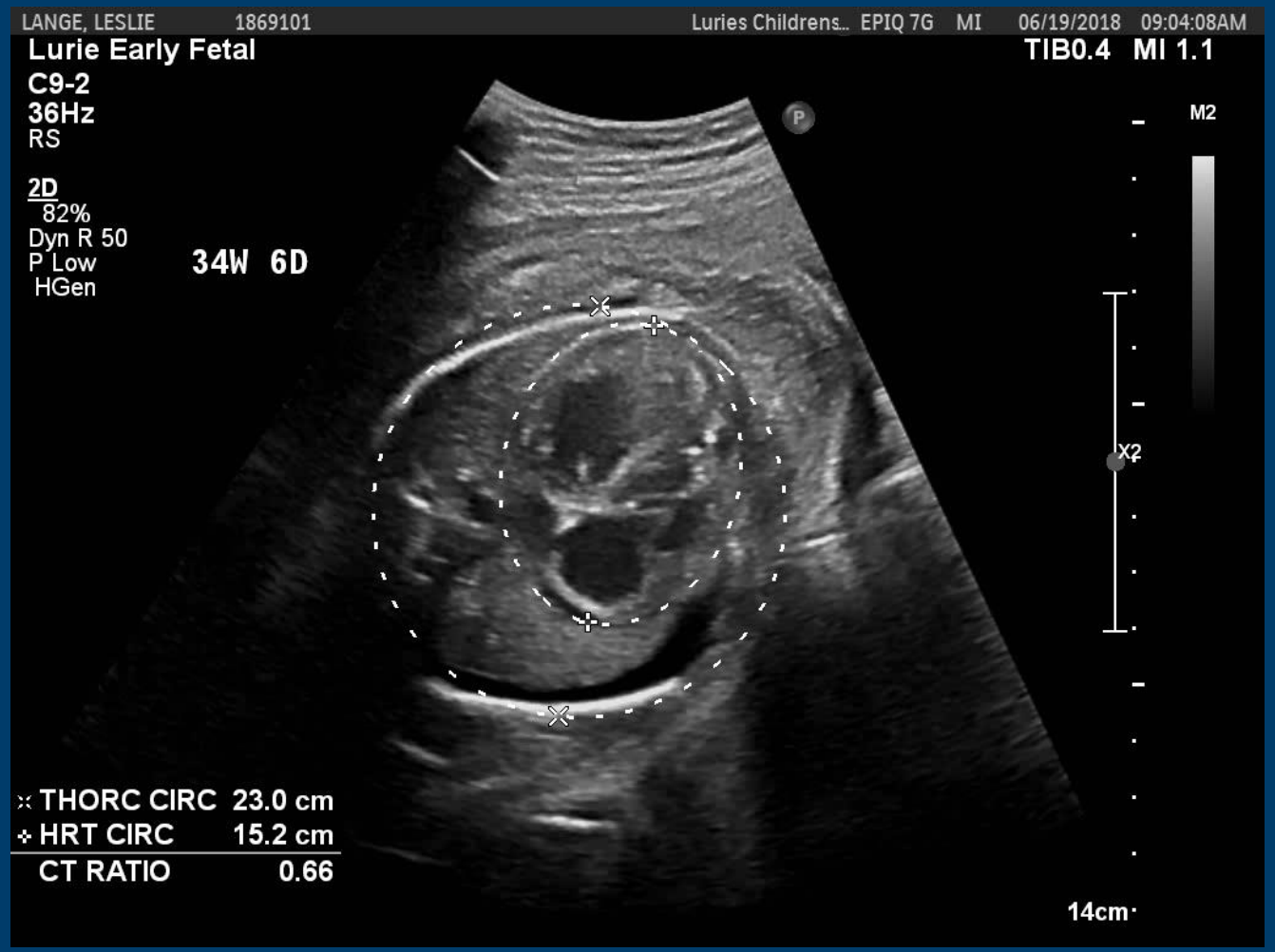
No conflicts of interest to declare

Outline

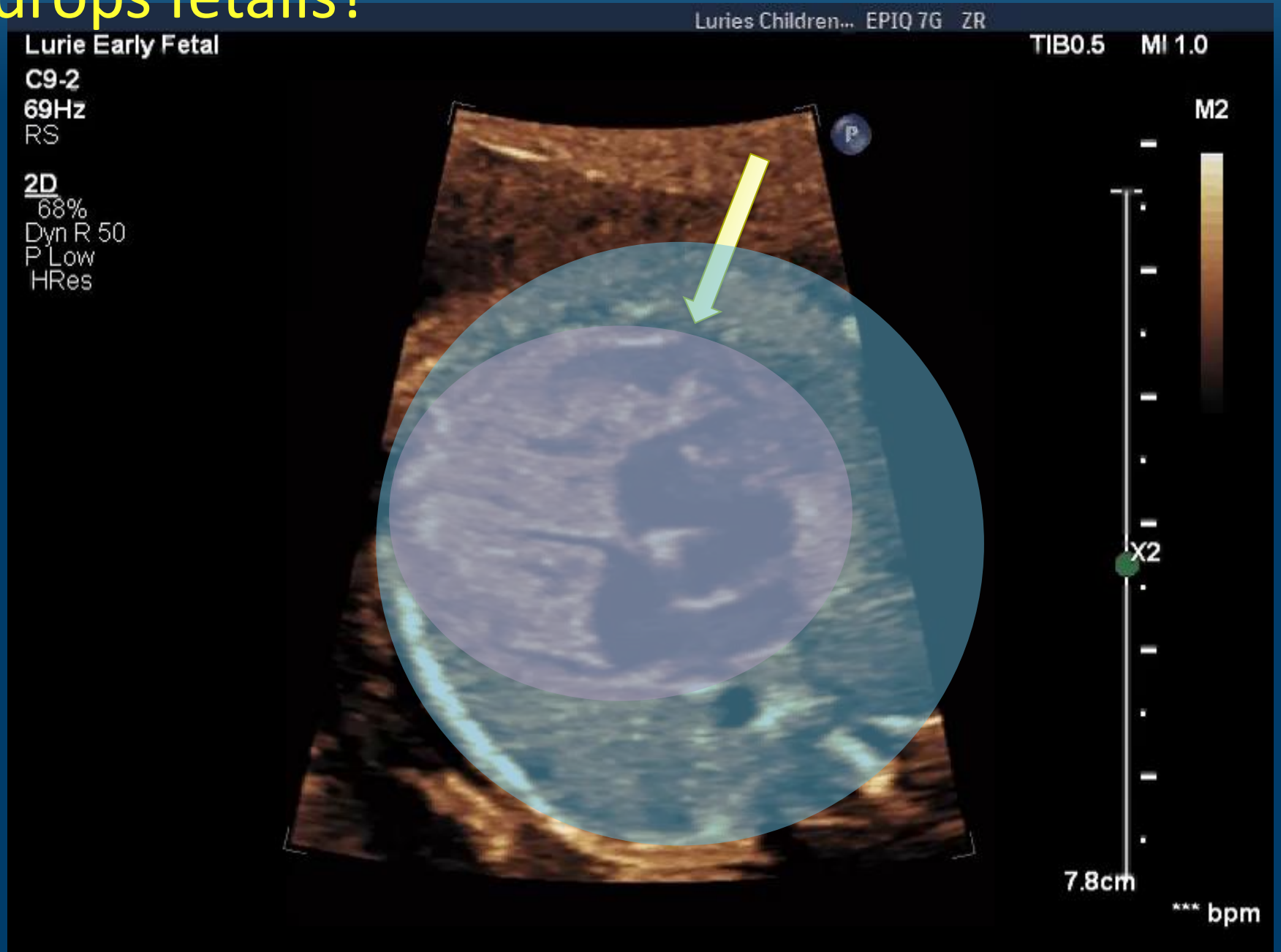
- What is hydrops fetalis?
- Why does it happen?
- Cardiac causes of hydrops
- Cardiac evaluation in patients with/at risk for hydrops
- Prognosis
- Delivery room preparation and postnatal planning

What is fetal hydrops?

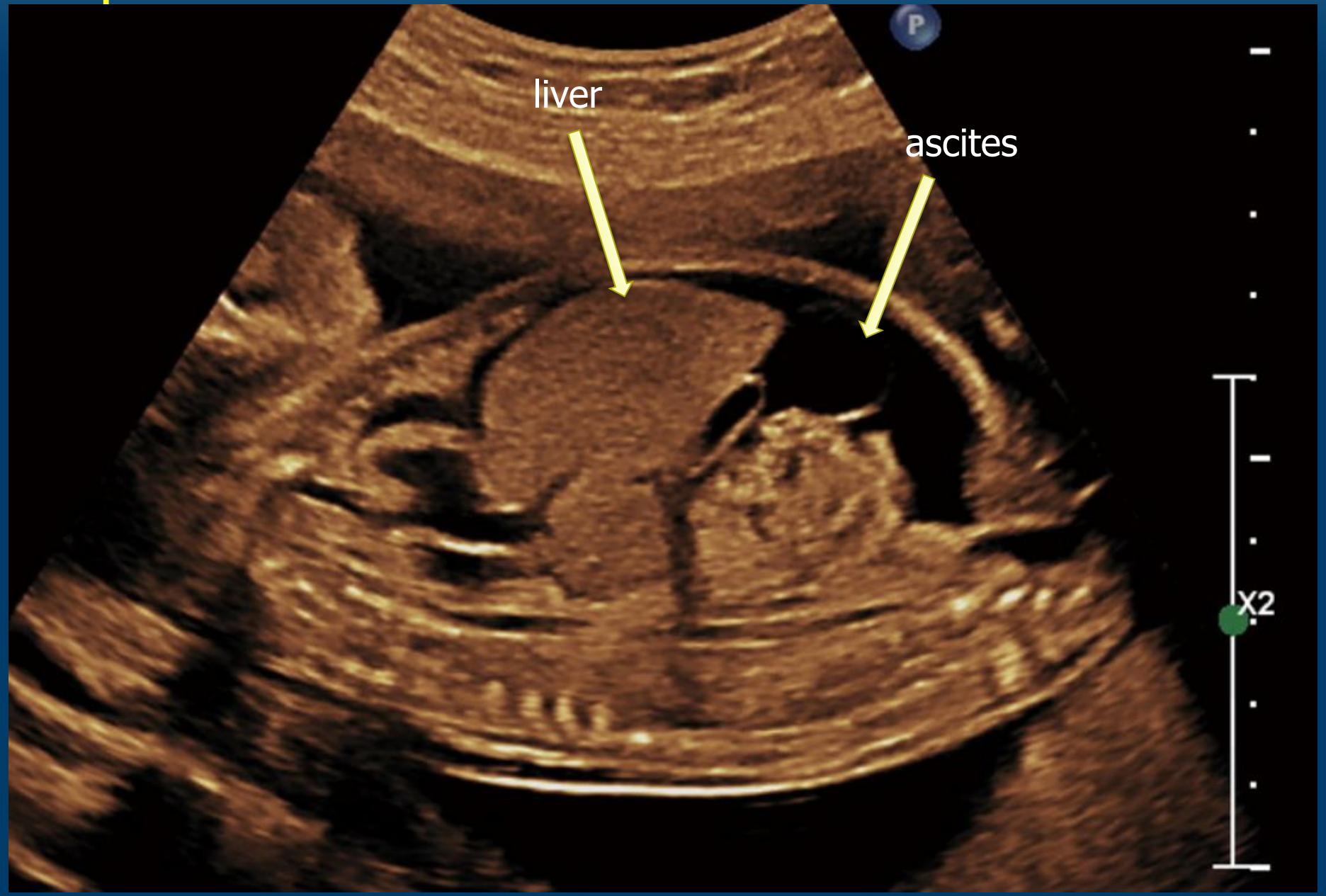
- **Hydrops** (Greek: water)
  - Accumulation of fluid within fetal soft tissue and serous cavities
  - At least 2 abnormal fluid collections
    - ascites
    - pleural effusions
    - pericardial effusion
    - Generalized skin edema (skin thickness >5 mm)
  - May also see:
    - placental thickening
    - polyhydramnios
  - High intrauterine fetal demise/perinatal mortality


LANGE, LESLIE 1869101
Luries Childrens... EPIQ 7G MI 06/19/2018 09:04:08AM
Lurie Early Fetal
TIB0.4 MI 1.1
C9-2
36Hz
RS
2D
82%
Dyn R 50
P Low
HGen
34W 6D
M2
X2
THORC CIRC 23.0 cm
HRT CIRC 15.2 cm
CT RATIO 0.66
14cm

What is hydrops fetalis?



What is hydrops fetalis?



Why we care: hydrops outcomes

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Isolated non-immune hydrops fetalis: an observational study on complete spontaneous resolution, perinatal outcome, and long-term follow-up

Sophie Neveling[1] · Alexander Johannes Knippel[2] · Peter Kozlowski[2]

Received: 3 April 2022 / Accepted: 29 July 2022

Abstract

Purpose The aim of our study was to investigate spontaneous resolution and postnatal outcome in non-immune hydrops fetalis (NIHF). We specifically studied NIHF cases that occurred without any other anomalies in the prenatal diagnostic workup, defined as isolated NIHF (iNIHF).

Methods To identify iNIHF we retrospectively classified prenatal findings of 700 NIHF singletons, diagnosed in our prenatal referral center between 1997 and 2016. We studied the occurrence of prenatal resolution in iNIHF and linked it to the perinatal outcome. We obtained long-term outcome by contacting the parents, children, and the pediatricians and listed all functional and structural anomalies and temporary logopedic, psychosocial and motoric impairments.

Results Among 70 iNIHF cases, 54 (77.1%) resolved completely prenatally. The baby-take-home rate was 98.1% in these cases. In contrast, the baby-take-home rate in the subgroup without complete resolution was 25.0%.
We achieved pediatric long-term outcome in 27 of 57 survivors (47.4%) of iNIHF with a mean follow-up period of 10.9 years. Among these 27 children, fetal hydrops had completely resolved prenatally in 26 cases and had regressed to a mild effusion in one case. In the pediatric development, two children had significant functional impairment and two children showed recurrent skin edema.

Conclusion Complete spontaneous resolution was the most common intrauterine course of iNIHF in our collective. Completely resolved iNIHF had a favorable perinatal outcome in our study. Our data on the long-term outcomes are consistent with the assumption of an increased rate of functional impairments.

Types of fetal hydrops

Immune hydrops

(erythroblastosis fetalis)

↓

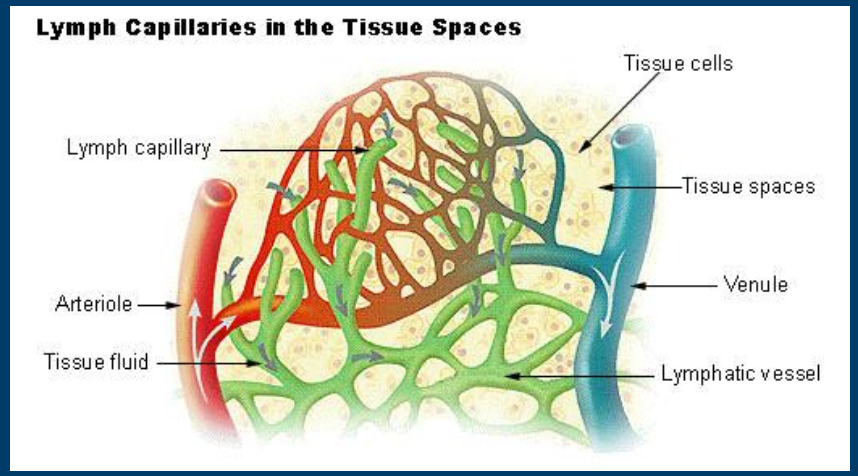
e.g., Rh+ fetus and Rh- mother with anti-Rh antibodies cross placenta

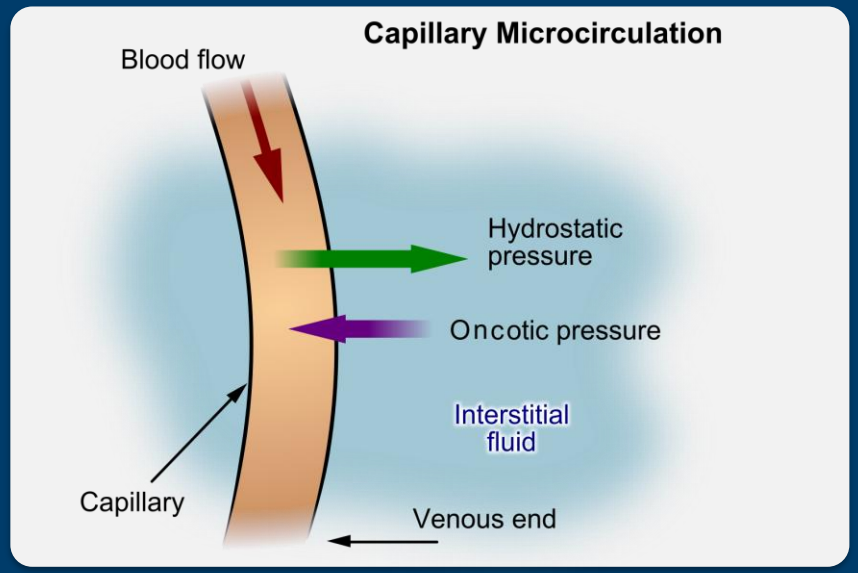
↓

fetal anemia, hepatic/renal/cardiac dysfunction

- Rho(D) immune globulin
- Immune hydrops now uncommon

Nonimmune hydrops





Non-immune hydrops

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CARDIOVASCULAR
- Arrhythmia
- Myocardiopathy
- Structural malformations (Ebstein anomaly, premature closure of the foramen ovale)
- Vascular obstruction (tumor, structural, fibroelastosis)
- Vascular malformation and hemangioma

GENETIC
- Skeletal dysplasias and myopathies
- Metabolic diseases (Gaucher, GM1 gangliosidosis, mucopolysaccharidosis)
- Autosomic diseases (Nooan, Prune belly, Fanconi)
- Chromosomal abnormalities (trisomy 21, 18, 13, Turner's syndrome)

CONGENITAL INFECTIONS
- Virus (cytomegalovirus, parvovirus B19, rubella, varicella, herpes, sintitial respiratory)
- Toxoplasmosis
- Syphilis
- Chagas disease

HEMATOLOGIC
- Nonimmune anemia
- Alpha-thalassemia
- Others (leukemia)

PLACENTAL
- Twin-twin transfusion syndrome
- Causes related to the umbilical cord

MISCELLANEOUS
- Respiratory (pulmonary sequestration, adenomatoid disease, chylothorax, tumor)
- Genitourinary (obstructive uropathy, dysplasia, cysts, thrombosis, nephrotic syndrome)
- Gastrointestinal (duodenal/jejunal atresia, anal imperforation, peritonitis)
- Neurological (encephalocele, intracranial hemorrhage, cerebral aneurysm)
- Tumoral (sacrococcygeal teratoma, neuroblastoma, hepatoblastoma)
- Multiple causes (presence of more than one associated etiopathic causes)

IDIOPATHIC
- Non-defined cause

Modified from the classification of Phibbs R, 1996.[2]

TABLE 1
Etiologies of nonimmune hydrops fetalis[6,11,12,14,75]

Cause	Cases	Mechanism
Cardiovascular	17-35%	Increased central venous pressure
Chromosomal	7-16%	Cardiac anomalies, lymphatic dysplasia, abnormal myelopoiesis
Hematologic	4-12%	Anemia, high output cardiac failure; hypoxia (alpha thalassemia)
Infectious	5-7%	Anemia, anoxia, endothelial cell damage, and increased capillary permeability
Thoracic	6%	Vena caval obstruction or increased intrathoracic pressure with impaired venous return
Twin-twin transfusion	3-10%	Hypervolemia and increased central venous pressure
Urinary tract abnormalities	2-3%	Urinary ascites; nephrotic syndrome with hypoproteinemia
Gastrointestinal	0.5-4%	Obstruction of venous return; gastrointestinal obstruction and infarction with protein loss and decreased colloid osmotic pressure
Lymphatic dysplasia	5-6%	Impaired venous return
Tumors, including chorioangiomas	2-3%	Anemia, high output cardiac failure, hypoproteinemia
Skeletal dysplasias	3-4%	Hepatomegaly, hypoproteinemia, impaired venous return
Syndromic	3-4%	Various
Inborn errors of metabolism	1-2%	Visceromegaly and obstruction of venous return, decreased erythropoiesis and anemia, and/or hypoproteinemia
Miscellaneous	3-15%	
Unknown	15-25%	

SMFM. Nonimmune hydrops fetalis. Am J Obstet Gynecol 2015.

Norton et al., 2015. Am J Obstet Gynecol

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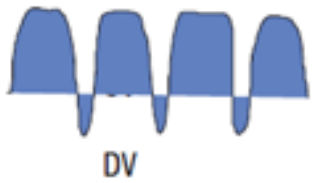
Cardiovascular Profile Score

Cardiovascular Profile Score

	Normal, 2 Points	–1 Point	–2 Points
Hydrops	None	Ascites or pleural effusion or pericardial effusion	Skin edema
Venous Doppler (Umbilical vein and ductus venosus)	 UV  DV	 UV  DV	 UV pulsations
Heart size (heart area/ chest area)	>0.20 and ≤0.35	0.35–0.50	>0.50 or <0.20
Cardiac function	Normal TV and MV RV/LV FS >0.28 Biphasic diastolic filling	Holosystolic TR or RV/LV FS <0.28	Holosystolic MR or TR dP/dt< 400 or monophasic filling
Arterial Doppler (umbilical artery)	 UA	 UA (AEDV)	 UA (REDV)

1. Hydrops

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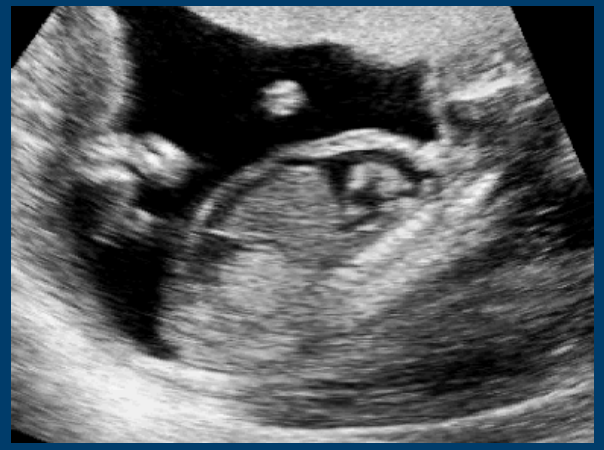
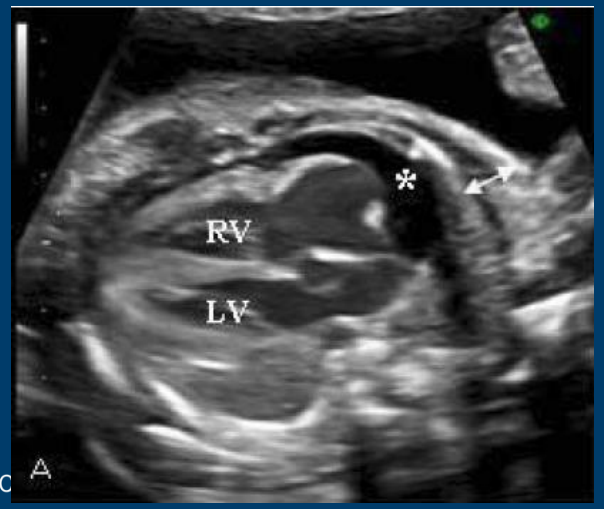
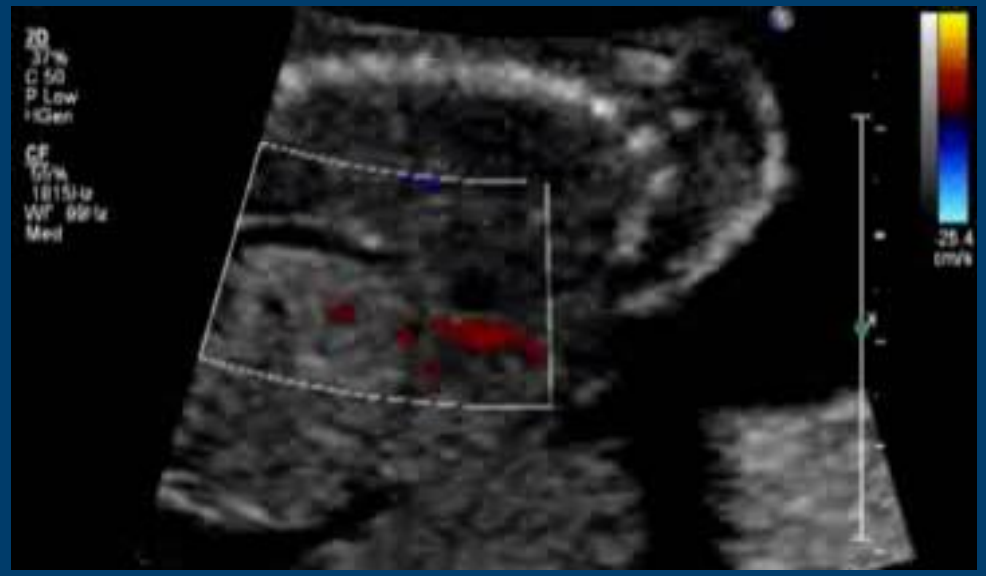
	Normal, 2 Points	–1 Point	–2 Points
Hydrops	None	Ascites or pleural effusion or pericardial effusion	Skin edema

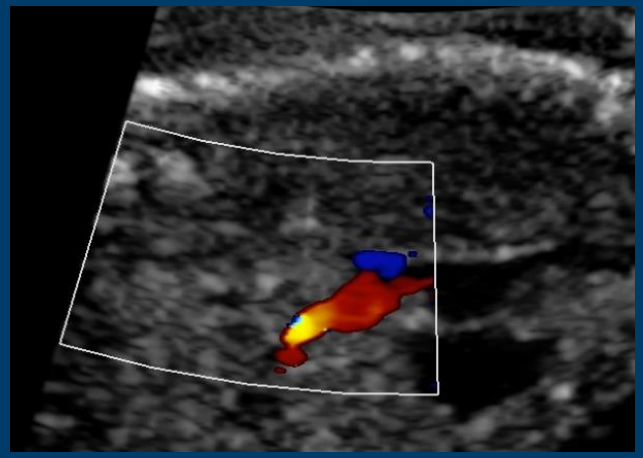


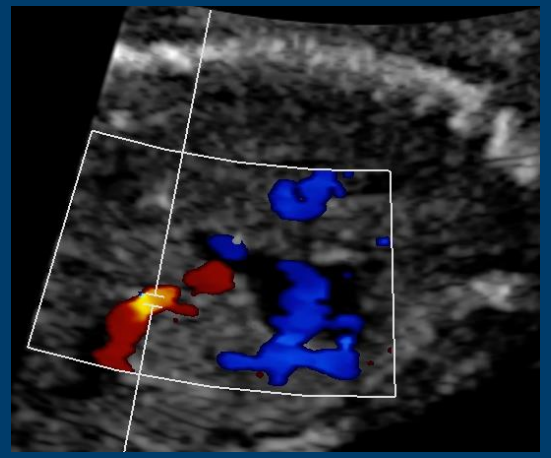


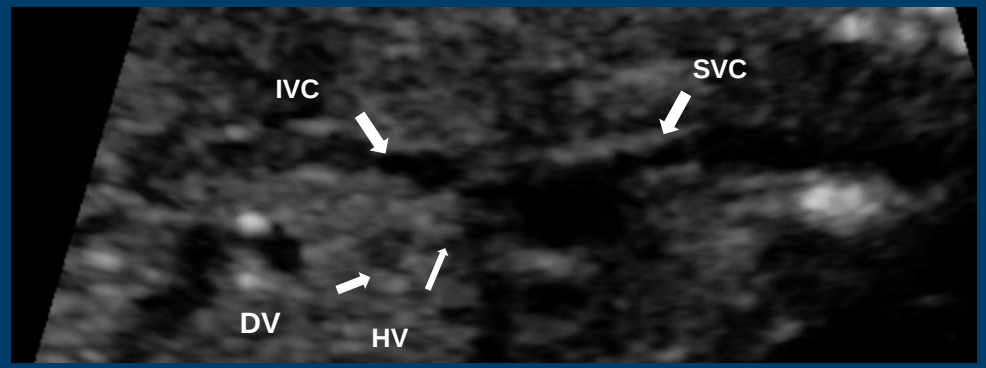
- Presence of ECF in at least 2 Cardiac Hydrops:
  - Skin edema, pleural effusion, pericardial effusion, ascites
  - Possibly polyhydramnios (due to increased urine output) and placentomegaly
- Ascites Only:
  - Non-hydrops causes of ascites (UPJ obstruction with rupture into peritoneal cavity; small bowel perforation)
- Early Hydrops:
  - Presence of fluid in only one cavity When hydrops is a known outcome in certain diseases

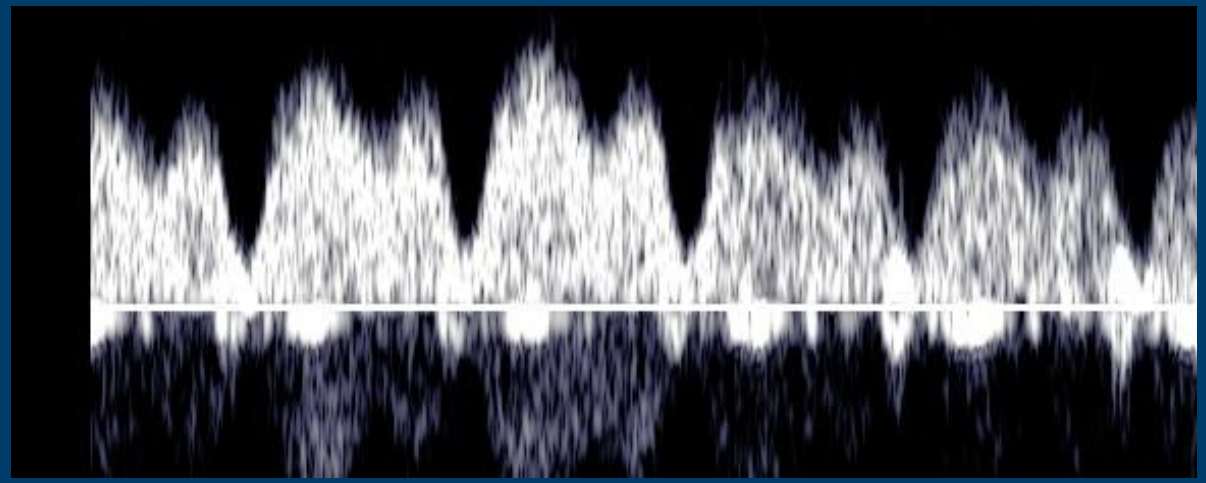
2. Venous Doppler : Ductus Venosus



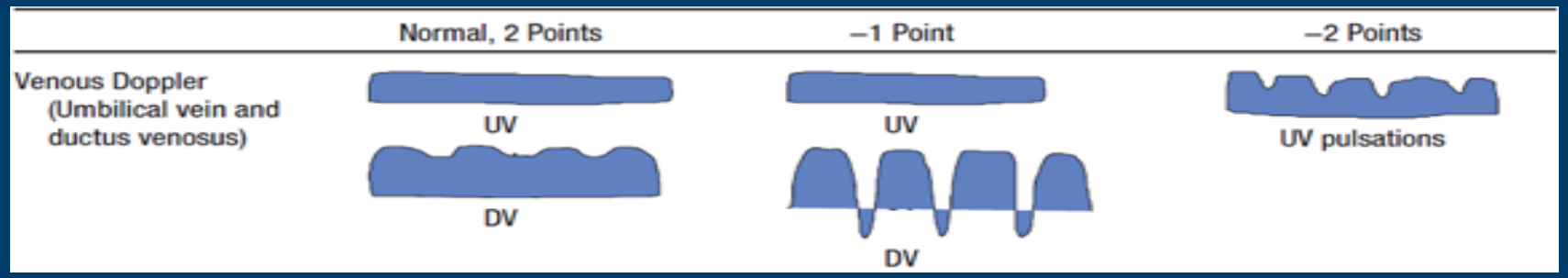


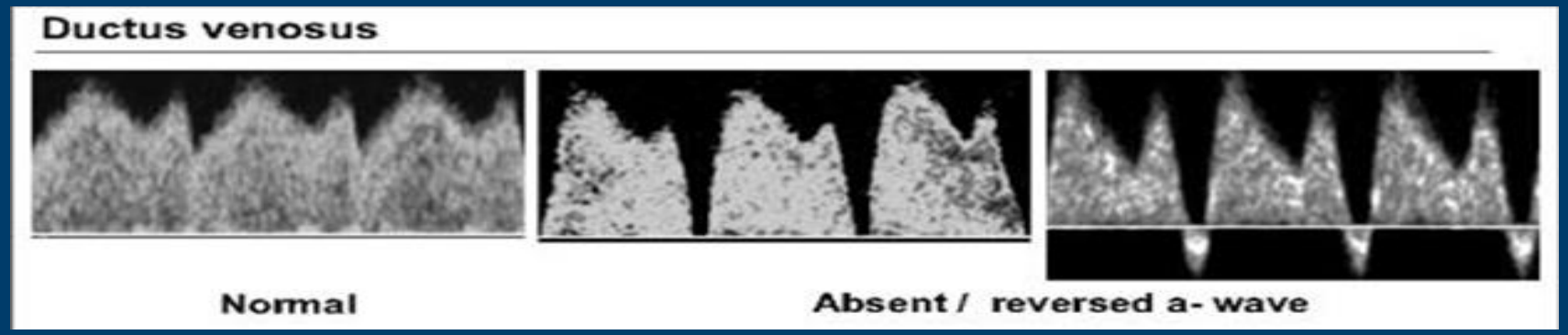






2. Venous Doppler Assessment

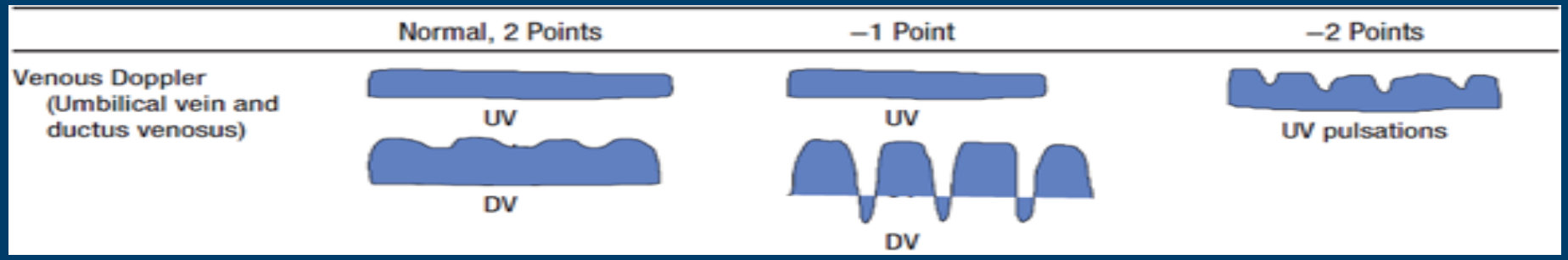


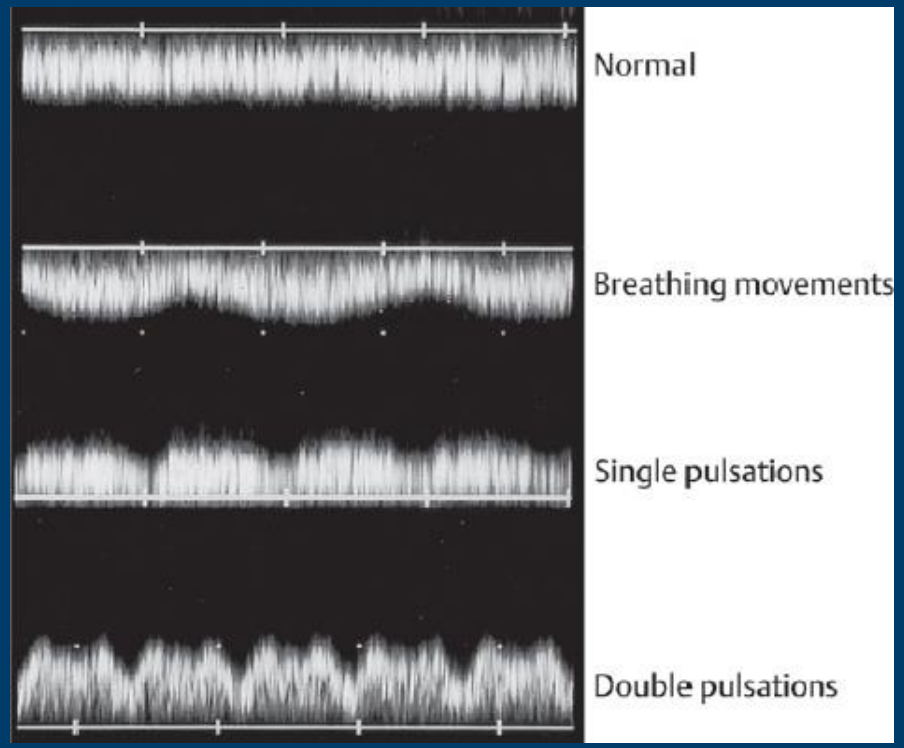


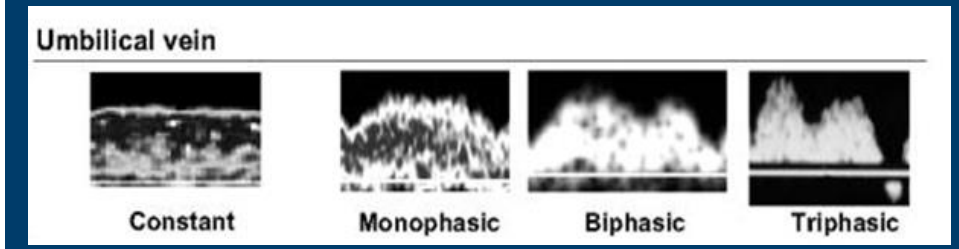
- Progressive decrease in atrial systolic forward velocities; eventually absent or reversed a-wave

- False positive findings if there is A:V desynchrony causing a wave reversal when the TV is closed

2. Venous Doppler Assessment





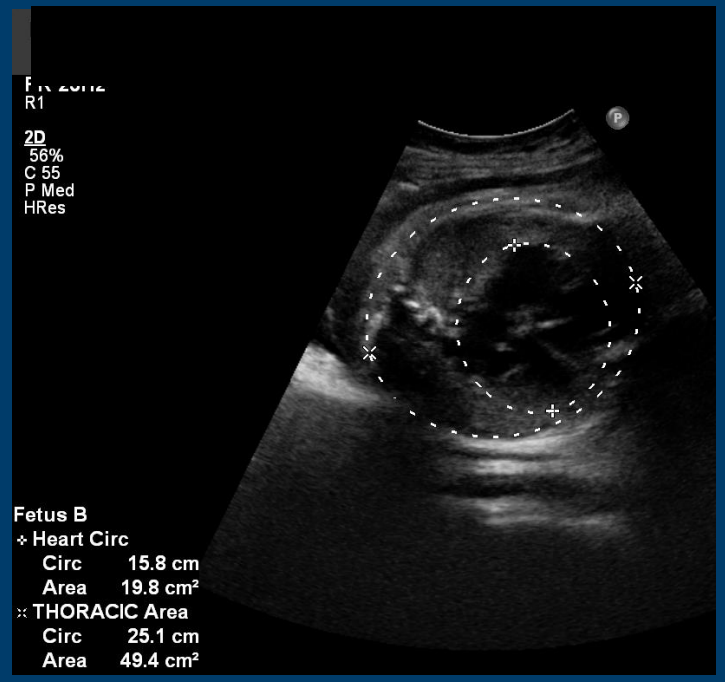


Umbilical Vein

- Increase in central venous pressure leads to progression form mono to bi to triphasic pattern

3. Heart Size

	Normal, 2 Points	–1 Point	–2 Points
Heart size (heart area/ chest area)	>0.20 and ≤0.35	0.35–0.50	>0.50 or <0.20

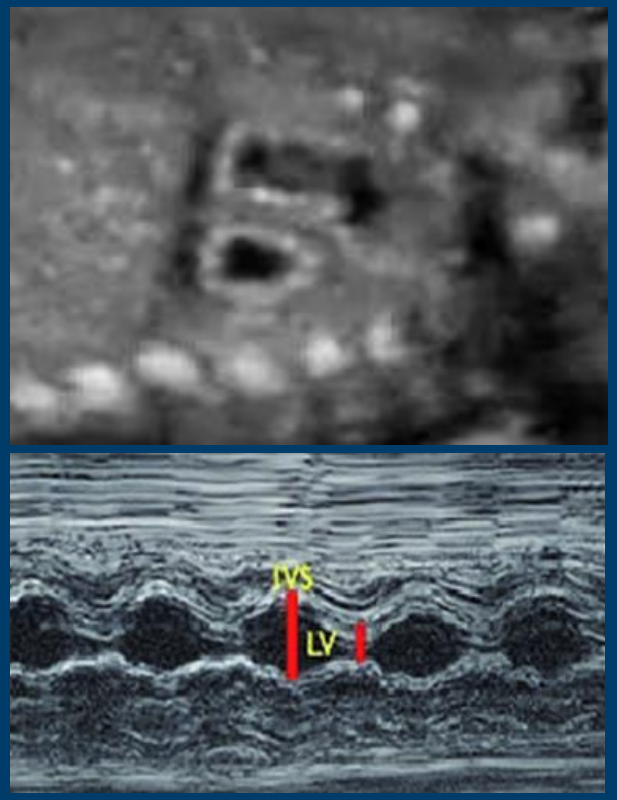


CTA = 0.41, CTC = 0.63

- Cardio Thoracic Area ratio
  - Normal < 0.35
  - 0.35-0.5 = mild cardiomegaly
  - >0.5 = severe cardiomegaly
- Cardio Thoracic Circumference ratio
  - Slowly increases through gestation
    - 11 weeks: 0.38
    - 17-20 weeks: 0.45
    - Term: 0.50
    - Normal less than 0.50-0.53

4. Cardiac Function: Fractional Shortening

	Normal, 2 Points	–1 Point	–2 Points
Cardiac function	Normal TV and MV RV/LV FS >0.28 Biphasic diastolic filling	Holosystolic TR or RV/LV FS <0.28	Holosystolic MR or TR dP/dt< 400 or monophasic filling



1. **Shortening Fraction:** M – mode of ventricles

$$SF = \frac{\text{Diastolic – systolic dimension}}{\text{Diastolic dimension}} \times 100$$

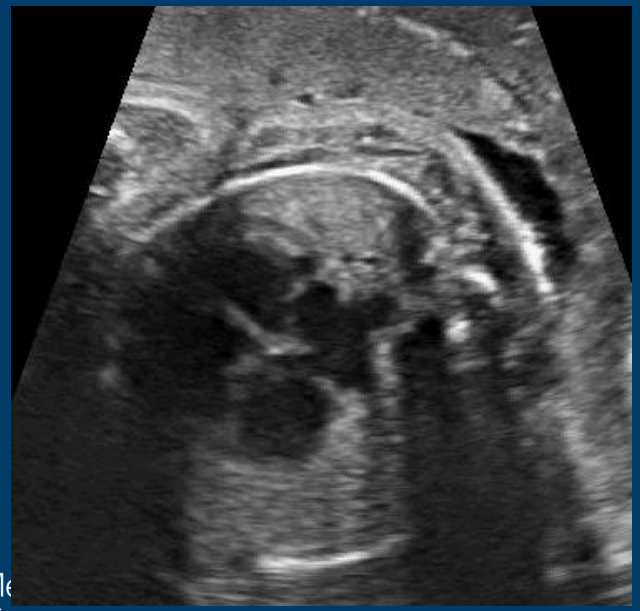
- Normal shortening fraction = 28-40%
- Normal values: http://fetal.parameterz.com/gagnon

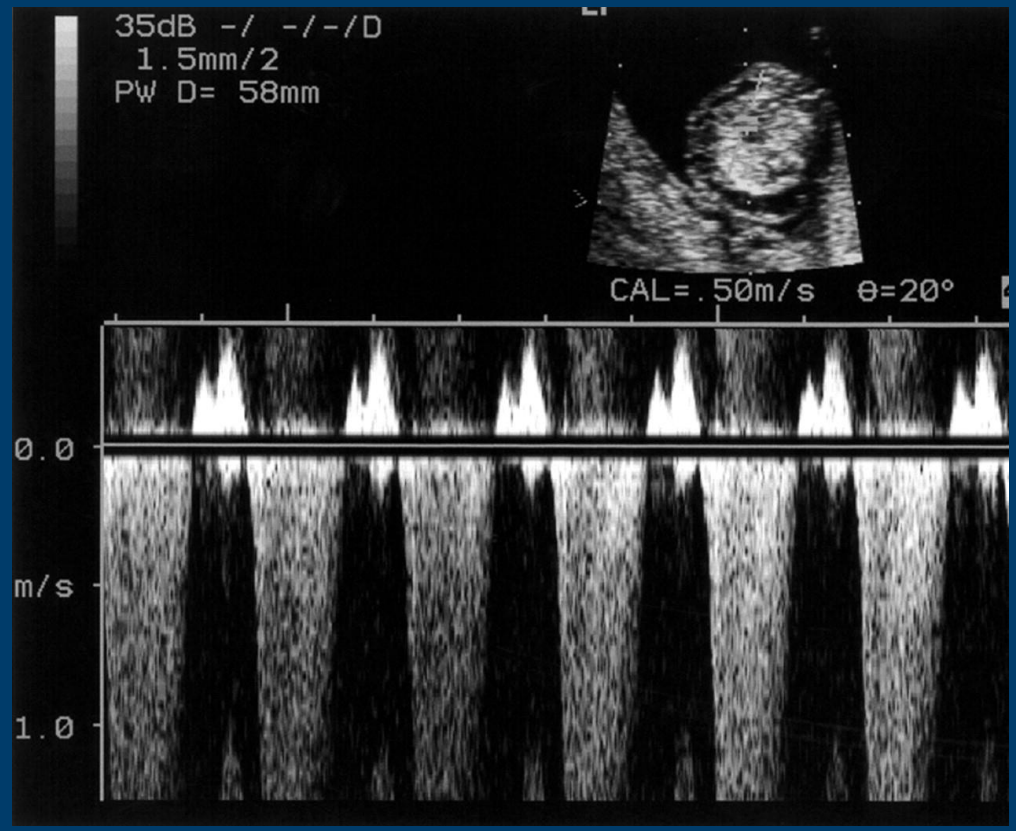
4. Cardiac Function: dP/dT

Cardiac function	Normal TV and MV RV/LV FS >0.28 Biphasic diastolic filling	Holosystolic TR or RV/LV FS <0.28	Holosystolic MR or TR dP/dt< 400 or monophasic filling

dP/dt : Doppler velocity of AV valve regurgitation signal

- Slow upstroke suggests RV dysfunction
- Abnormal if < 800 mmHg/s
- Poor fetal outcome if < 400 mmHg/s

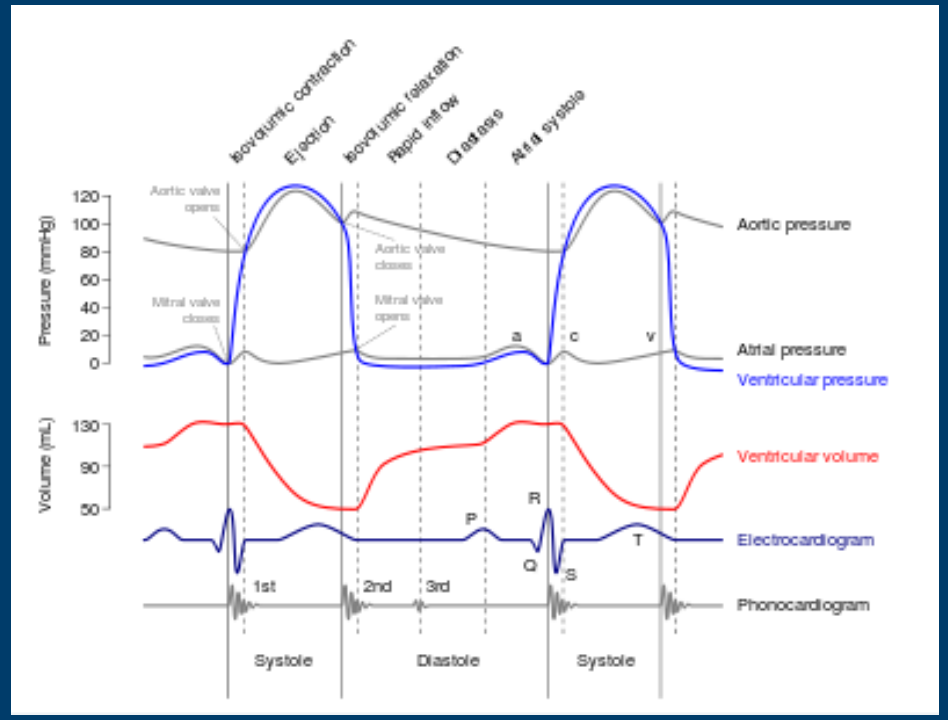


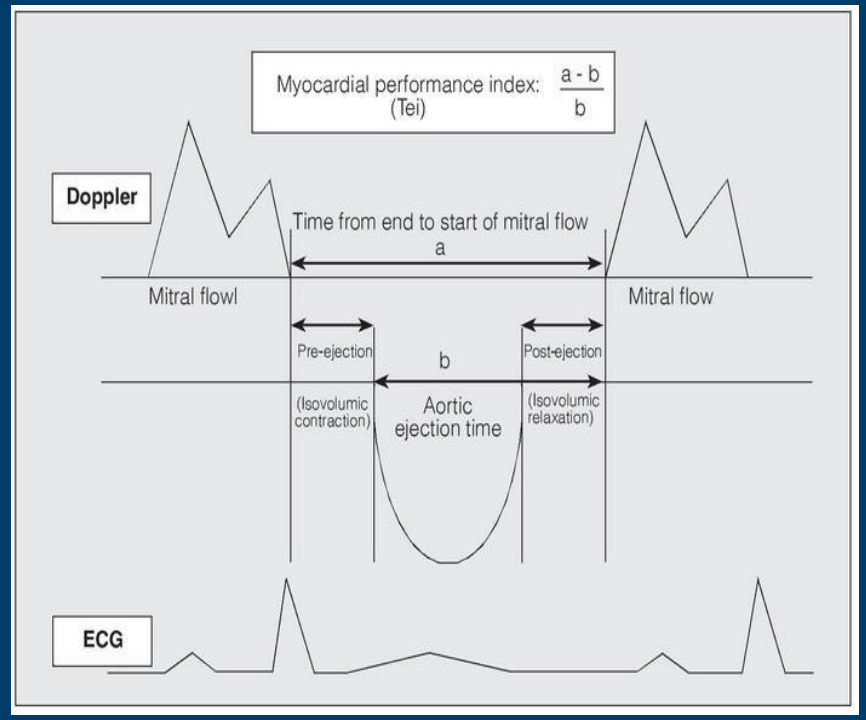


4. Cardiac Function: Myocardial Performance Index

	Normal, 2 Points	–1 Point	–2 Points
Cardiac function	Normal TV and MV RV/LV FS >0.28 Biphasic diastolic filling	Holosystolic TR or RV/LV FS <0.28	Holosystolic MR or TR dP/dt< 400 or monophasic filling

A healthy ventricle has a short IRT and ICT resulting in a low MPI

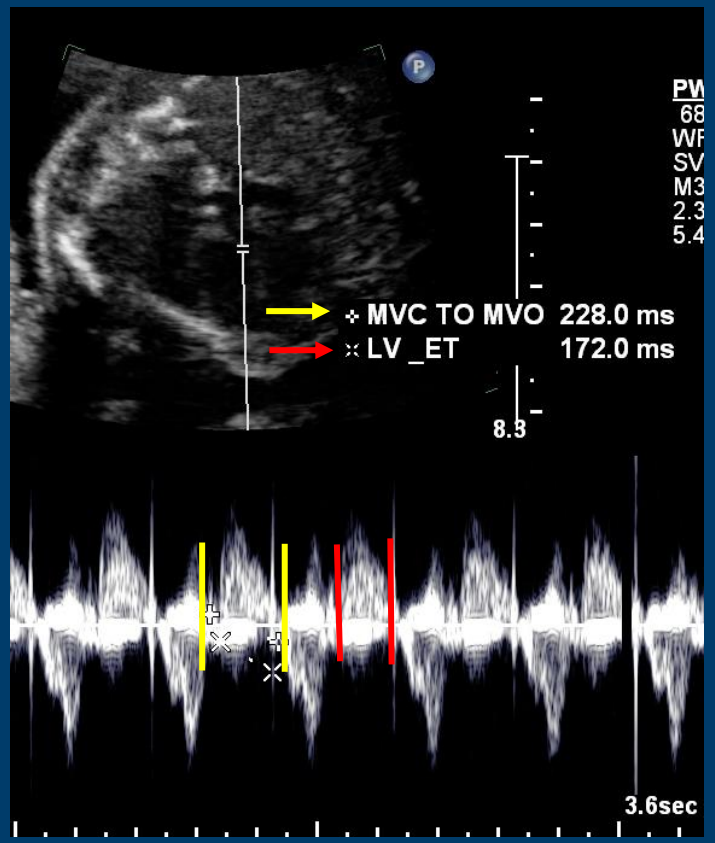




Fetal Cardiac Exam

Myocardial Performance Index

- LV MPI
  - Five chamber view
  - Doppler sample at tip of MV leaflets but angled toward the LVOT
  - Measurements made from same cardiac cycle

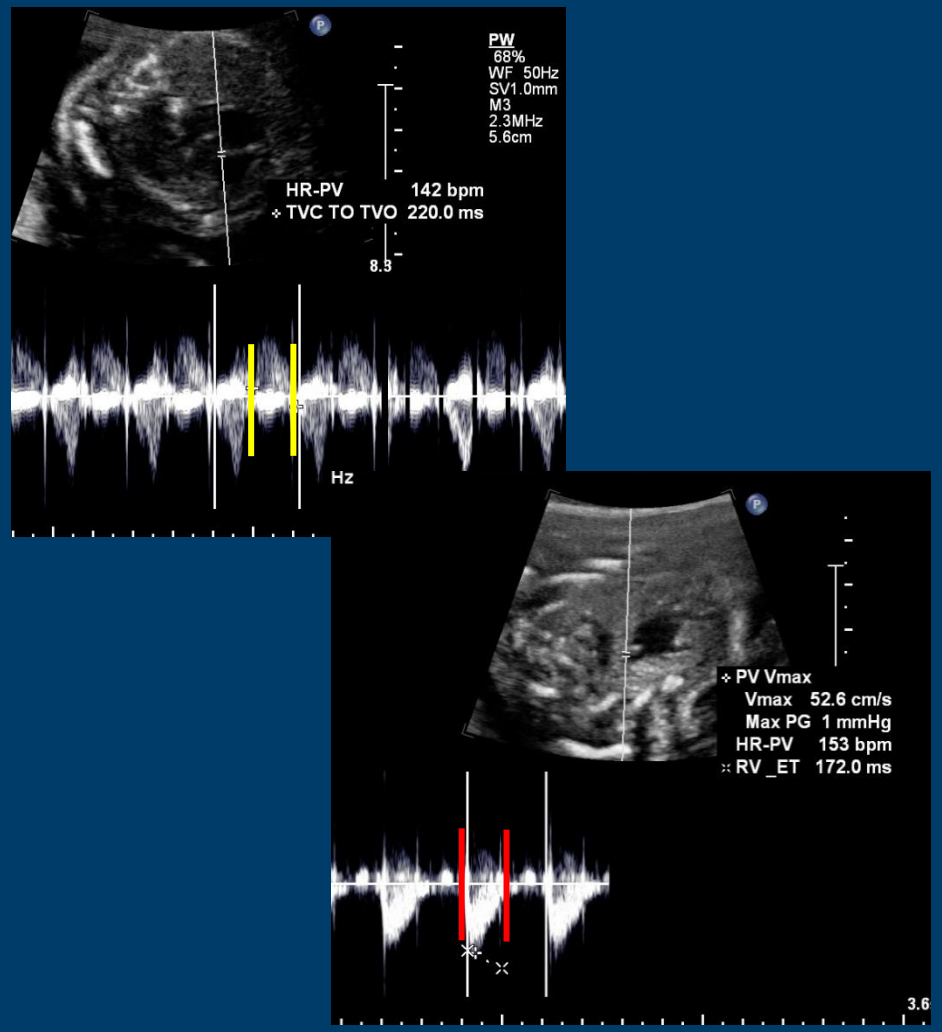


MPI = 228-172/172 = 0.32

Fetal Cardiac Exam

Myocardial Performance Index

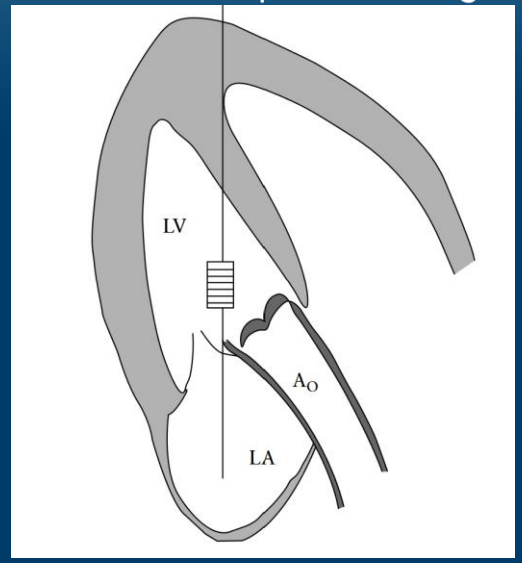
- RV MPI
  - TV and PV separated by conal tissue
  - TV inflow Doppler with cycle HR
  - PV outflow Doppler with cycle HR
  - If cycle HR of both measurements within 10 bpm, calculate MPI

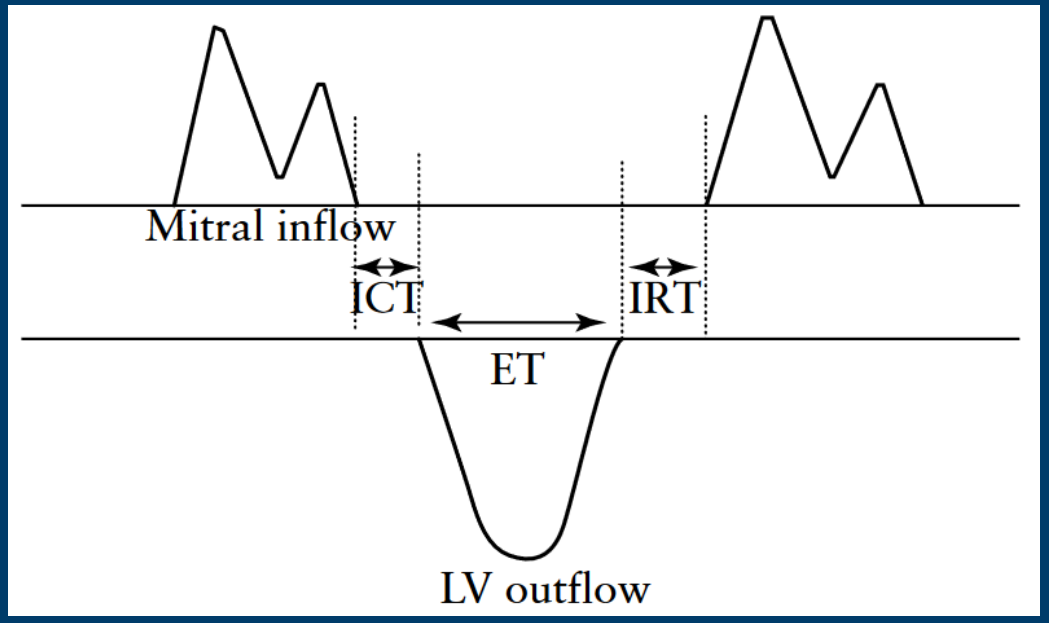


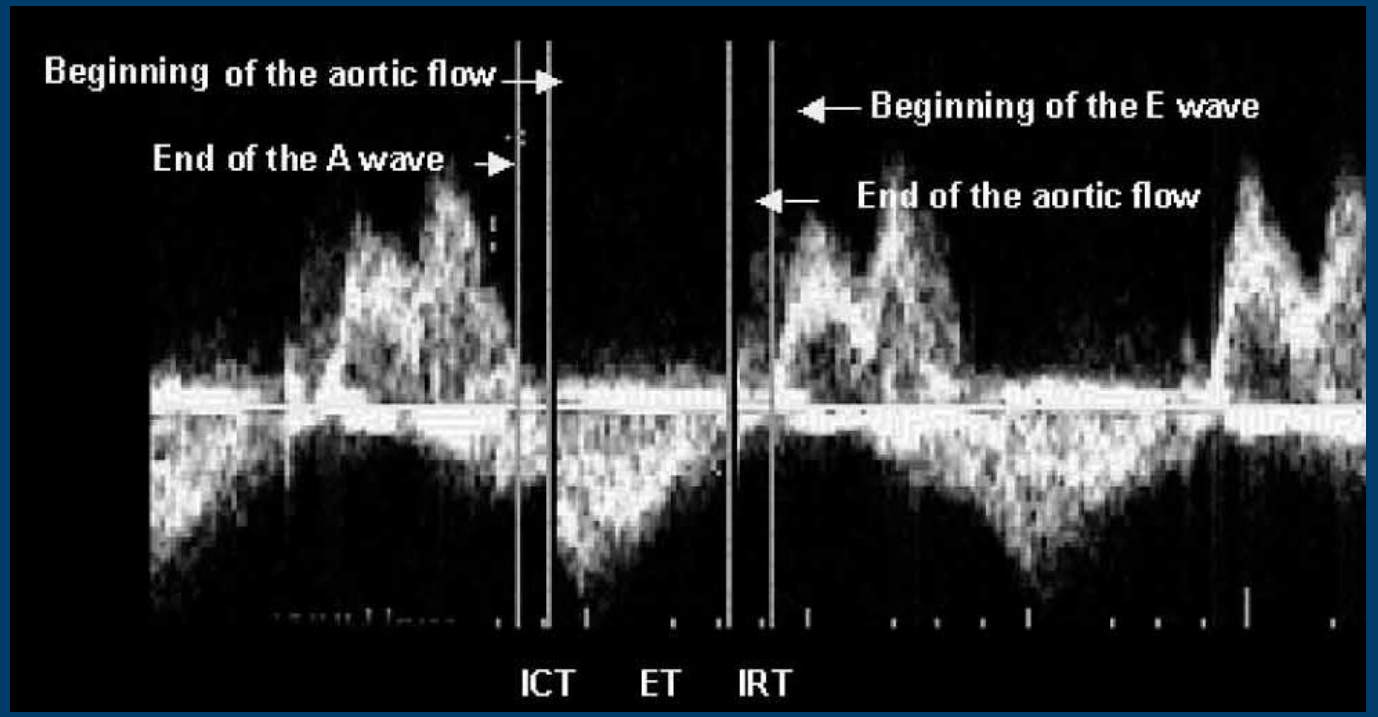
MPI = 220-172/172 =0.28

Myocardial performance index (MPI)

- MPI = Tei index = (ICT + IRT) / ET
- Reflects systolic + diastolic function
- LV: Doppler simultaneously MV inflow and LVOT
- RV: Separate Doppler TV inflow, pulmonary valve
- Normal MPI: ≈0.38 ±0.04 RV, 0.41 ±0.05 LV (higher worse)

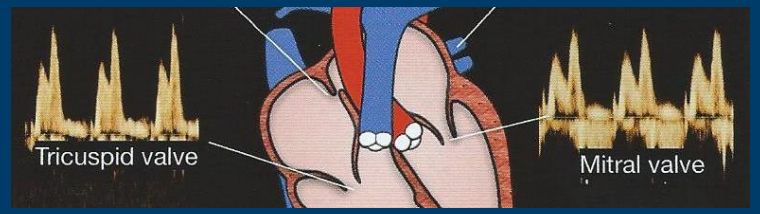


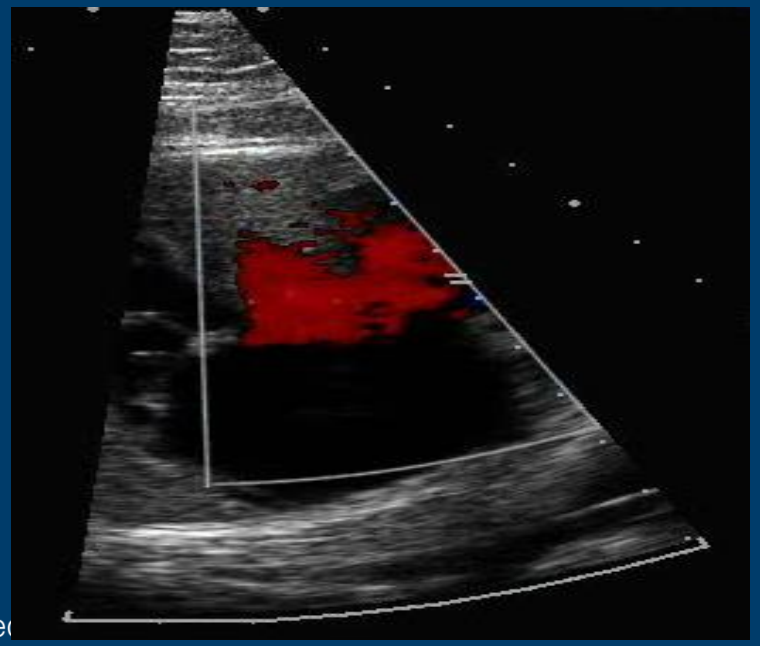




4. Cardiac Function: Valve function

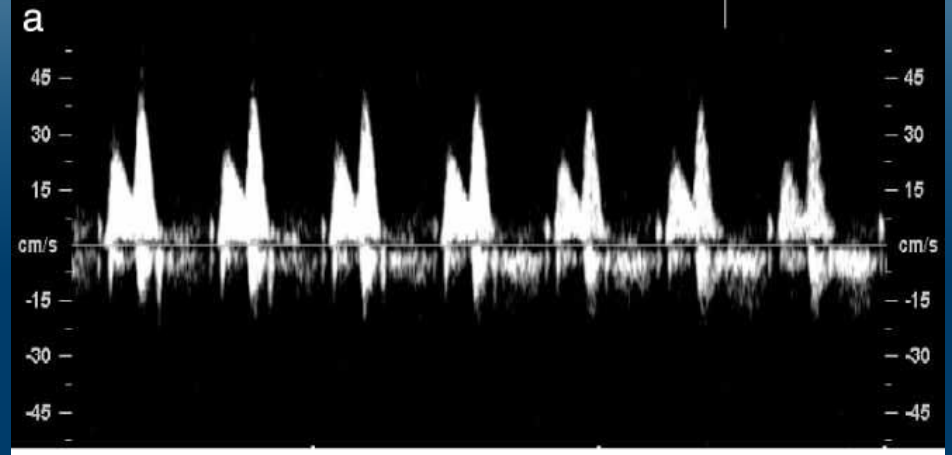
	Normal, 2 Points	−1 Point	−2 Points
Cardiac function	Normal TV and MV RV/LV FS >0.28 Biphasic diastolic filling	Holosystolic TR or RV/LV FS <0.28	Holosystolic MR or TR dP/dt< 400 or monophasic filling

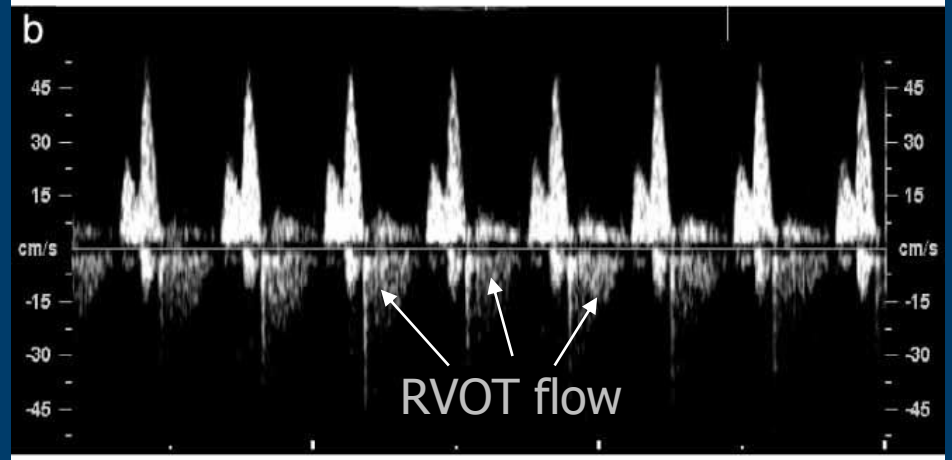


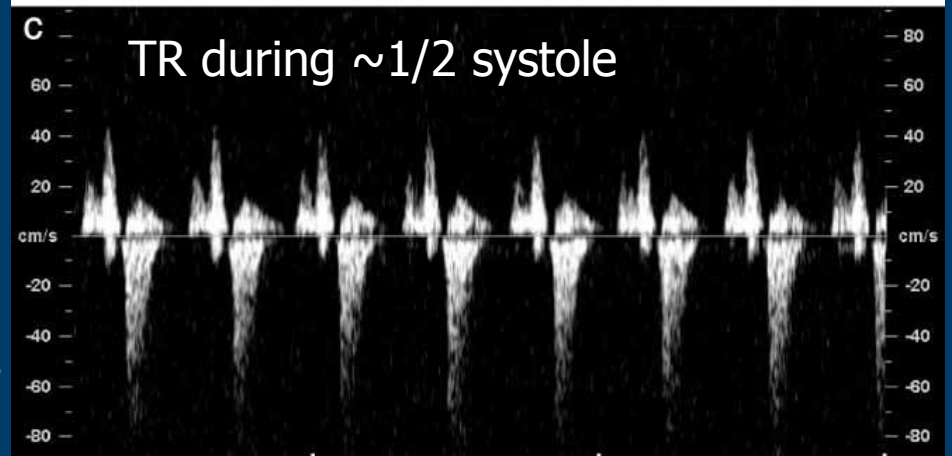


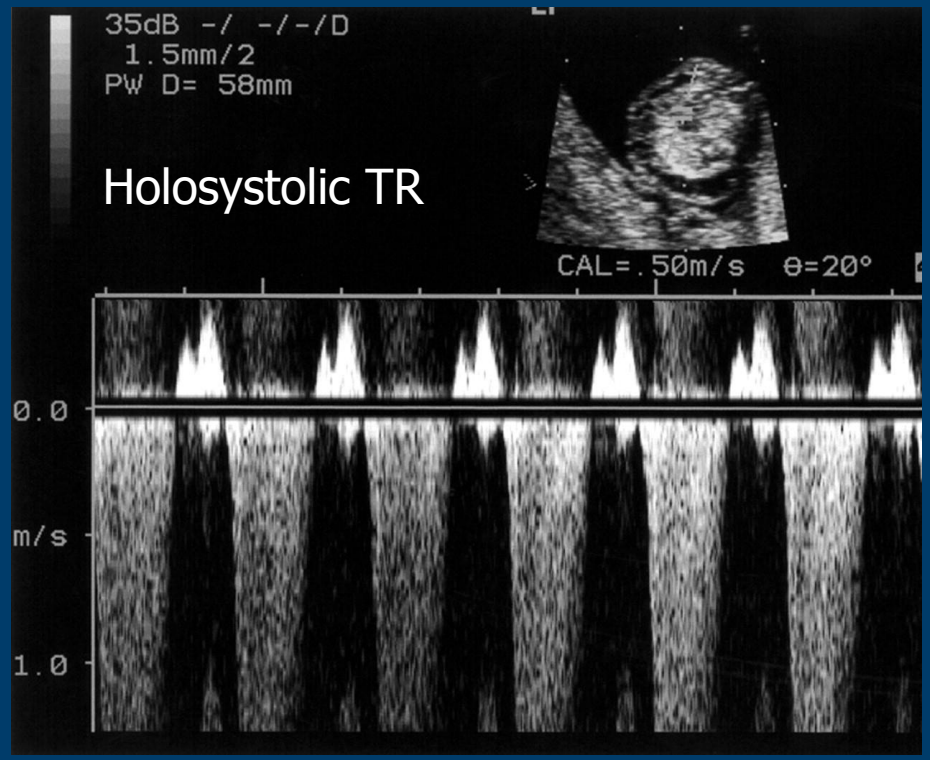
- Normal: Biphasic inflow, no regurgitation
- Holosystolic tricuspid regurgitation is always abnormal
- Regurgitation of other valves is usually a sign of advanced failure

Tricuspid valve regurgitation









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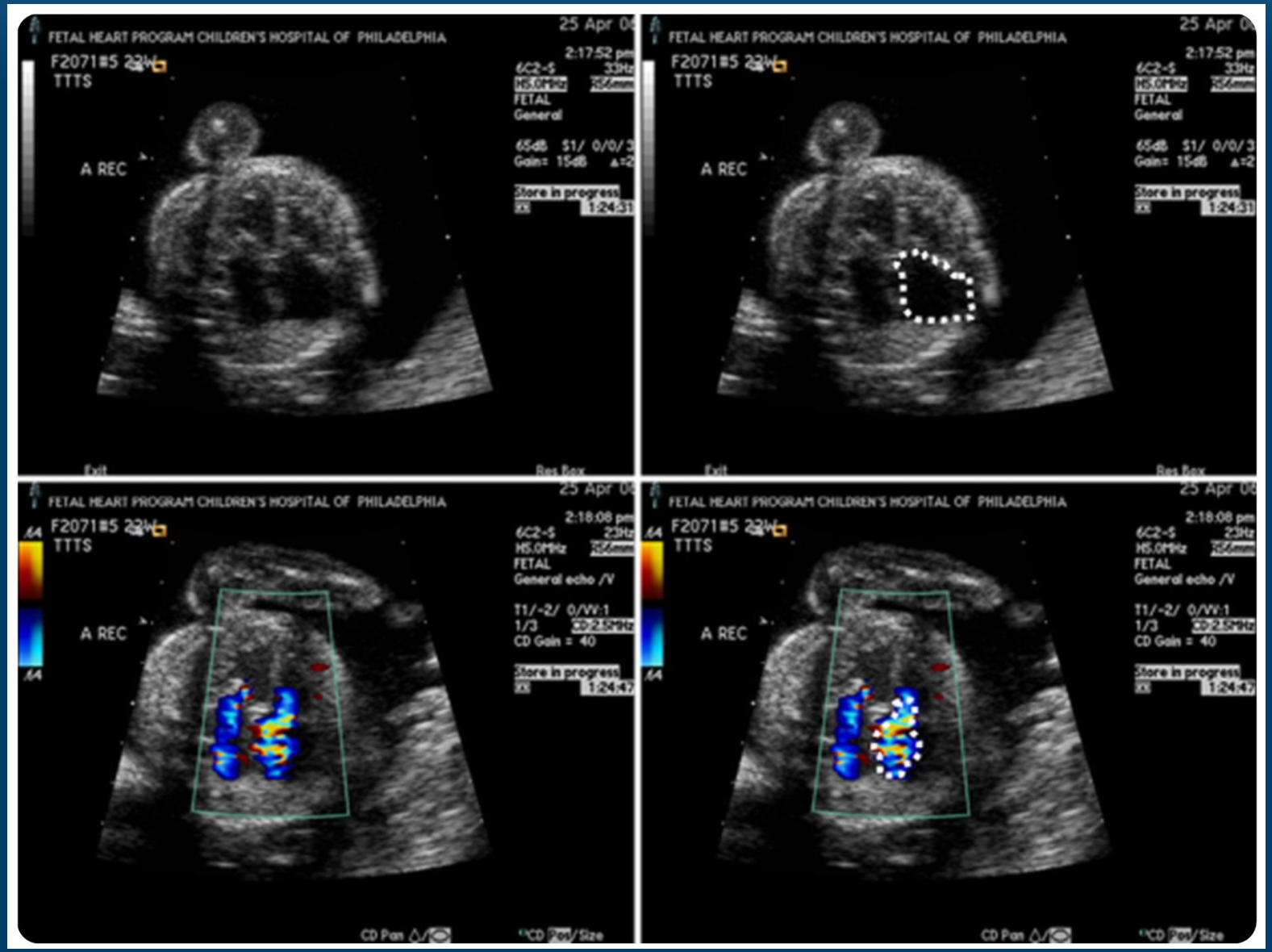
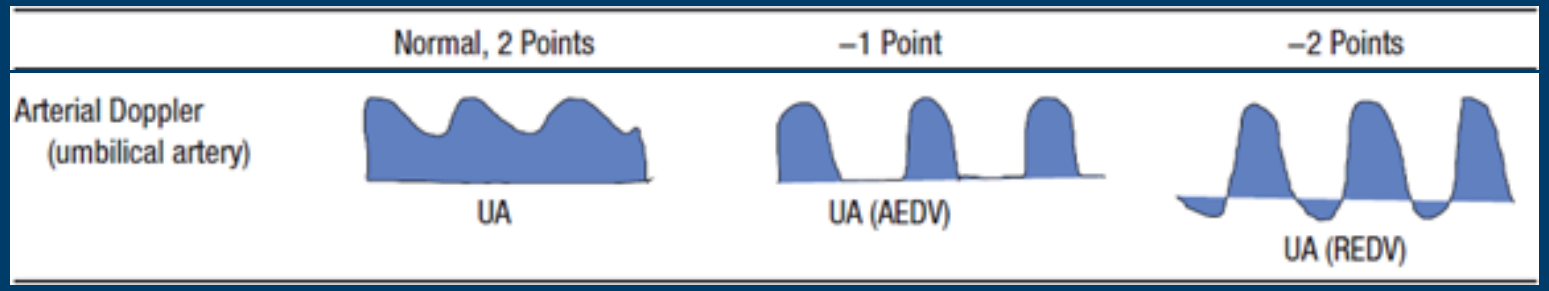
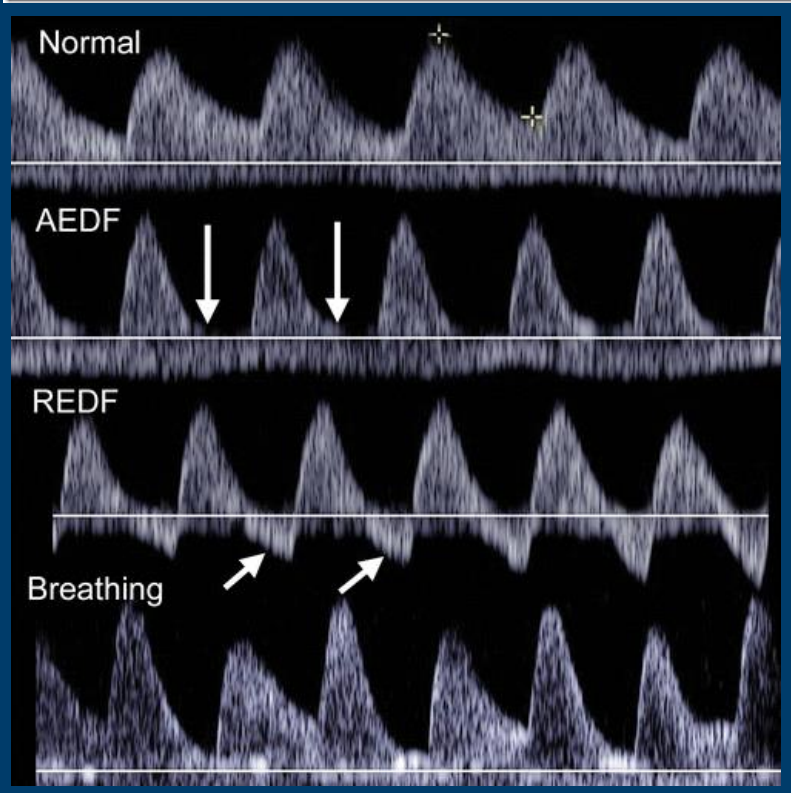
- Tricuspid and mitral valve regurgitation are graded as none (0 points), mild (1 point) when the regurgitant jet area is ≤25% of the atrial area, or more than mild (2 points) when the regurgitant jet area is >25% of the atrial area (Figure 2).



Rychik J, et al. Am J Obstet Gynecol. 2007

5. Arterial Doppler Assessment





Umbilical Artery

- Absent end diastolic flow or reversal of diastolic flow is abnormal

Predicting Fetal Outcomes by CVPS

J Matern Fetal Neonatal Med. 2006 Jul;19(7):407-13.

A cardiovascular profile score in the surveillance of fetal hydrops.

Hofstaetter C[1], Hansmann M, Eik-Nes SH, Huhta JC, Luther SL.

Author information

Abstract

OBJECTIVE: To assess the value of a cardiovascular profile score in the surveillance of fetal hydrops.

METHODS: In a retrospective study, 102 hydropic fetuses were examined between 15 and 37 completed weeks of gestation with ultrasonographic assessment of hydrops, heart size, and cardiac function, and arterial umbilical and venous Doppler sonography of the ductus venosus (DV) and the umbilical vein (UV). A cardiovascular profile score (CVPS) was constructed by attributing 2 points for normal and taking away 1 or 2 points for abnormal findings in each category. The score of the final examination prior to treatment, delivery, or fetal demise was compared to the fetal outcome in these 102 fetuses after exclusion of terminated pregnancies. The scores of the first and last examinations were compared in 40 fetuses and the relationship between these scores and the evolution of fetal hydrops and fetal outcome was assessed.

RESULTS: Twenty-one pregnancies were terminated (21%). Fifty-four of the remaining 81 hydropic fetuses survived (67%) and perinatal death (PNM) occurred in 27 fetuses (33%). The median CVPS was 6.0 (IQR 4.75-8.00) for all fetuses, with a median of 6.0 (IQR 5.00-6.00) in fetuses who died in the perinatal period compared to a median of 7.0 (IQR 4.00-8.00) in those who survived ($p < 0.035$). All fetuses in this study had a 'severe' form of hydrops with skin edema. The best predictor for adverse outcome was the venous Doppler sonography of UV and DV, in particular umbilical venous pulsations. Among fetuses included in the longitudinal arm of the study, the survival rate was 40% and the PNM was 60%, after exclusion of terminated pregnancies. CVPS increased by a median of 1 (IQR 0.00-2.00) point in the last exam for those fetuses that lived, whereas among those fetuses that died, the CVPS decreased by a median 1.5 (IQR 0.25-2.75) points ($p < 0.001$).

CONCLUSIONS: The fetal cardiovascular profile score can be used in the surveillance of hydropic fetuses for prediction of the presence of congestive heart failure and as an aid for predicting fetal outcome.

PMID: 16923695 DOI: 10.1080/14767050600682446

Cardiovascular profile score

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Abstract 18132: Prenatal Evaluation of the Fetal Cardiovascular Profile Score in a Multicenter Study of Ebstein Anomaly/Tricuspid Valve Dysplasia

Conclusions: In this study, fetuses with EA/TVD with CVP scores <7 on the first or last fetal echo were less likely to survive. The CVP score may be a valuable tool to identify fetuses with EA/TVD at risk of poor outcome for whom novel therapeutic approaches might be warranted.

Cardiovascular profile and biophysical profile scores predict short-term prognosis in infants with congenital heart defect

Takekazu Miyoshi ✉, Shinji Katsuragi, Reiko Neki, Ken-ichi Kurosaki, Isao Shiraishi, Michikazu Nakai, Kunihiro Nishimura, Jun Yoshimatsu, Tomoaki Ikeda

First published: 11 April 2019 | https://doi.org/10.1111/jog.13970 | Citations: 2

A total of 202 patients with CHD were analyzed. Perinatal and infant deaths occurred in 16 (7.9%) and 10 cases (5.0%), respectively. Infants with the last CVP score ≤ 5 had 18.7-fold higher perinatal mortality than those with a last CVP score > 5 ($P < 0.01$). Infants with a last BP score ≤ 6 had 18.7-fold higher perinatal mortality than those with a last BP score > 6 ($P < 0.01$). Infants with a CVP score decrease *in utero* had 4.5-fold higher infant mortality than those with an increase or no change ($P < 0.01$). Multivariate analysis showed that single-ventricle physiology, pre-term birth at <37 weeks of gestation, last CVP score ≤ 5, and last BP score ≤ 6 were independent predictors of perinatal mortality. Single-ventricle physiology and a CVP score decrease were independent predictors of infant mortality.

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Cardiovascular profile score

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Initial Fetal Cardiovascular Profile Score Predicts Recipient Twin Outcome in Twin-Twin Transfusion Syndrome

Amy D. Shah, M.D.[1,2], **William L. Border, M.D.**[1,2], **Timothy M. Crombleholme, M.D.**[1], and **Erik C. Michelfelder, M.D.**[1,2]

1*Fetal Care Center of Cincinnati, Divisions of Pediatric Cardiology, General, Thoracic, and Fetal Surgery, Cincinnati Children's, Hospital and the University of Cincinnati College of Medicine*

2*The Fetal Heart Center, Divisions of Pediatric Cardiology, General, Thoracic, and Fetal Surgery, Cincinnati Children's, Hospital and the University of Cincinnati College of Medicine*

Abstract

Objective—To assess the relationship between cardiomyopathy and recipient twin (RT) outcome in twin-twin transfusion syndrome (TTTS).

Methods—Fetal echocardiography and outcomes data in 62 consecutive pregnancies with TTTS was reviewed. The primary outcome was neonatal RT survival. Severity of RT cardiomyopathy at presentation was assessed by the Cardiovascular Profile Score (CVPS). RT outcome and odds of survival were compared between groups, stratified by CVPS.

Results—Overall neonatal survival for all fetuses was 61% (76/124). RT survival was 58% (36/62). Grouped by CVPS, RT survival was greater (50%) for those with a CVPS ≥ 9 and even higher (74%) for a CVPS =10. Amongst components of the CVPS, atrioventricular valve regurgitation (AVVR) was associated with negative RT outcome. Other factors at presentation were not predictive of RT outcome.

Conclusions—Normal CVPS in the RT in TTTS is predictive of improved survival compared to RT with abnormal CVPS, even RT with minor deductions. Standard clinical staging did not predict outcome. Cardiac assessment by CVPS may improve clinical decision making and timing of fetal interventions.

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Delivery Planning in Fetal Hydrops

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Delivery Preparation & Resource Planning

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Code Rainbow:
Improving the transfer process for newborn with critical CHD

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- The Problem (2018):
  - longer than desired transfer time
  - suboptimal communication
  - Suboptimal preparations to do cardiac intervention after arrival at CCU

- QI project "SMART" aim:
  - To reduce the transfer time, defined as the duration between the time of birth and time of arrival to CCU, by 20% by 12/31/19 in neonates prenatally diagnosed with critical CHD with anticipated hemodynamic instability soon after birth.

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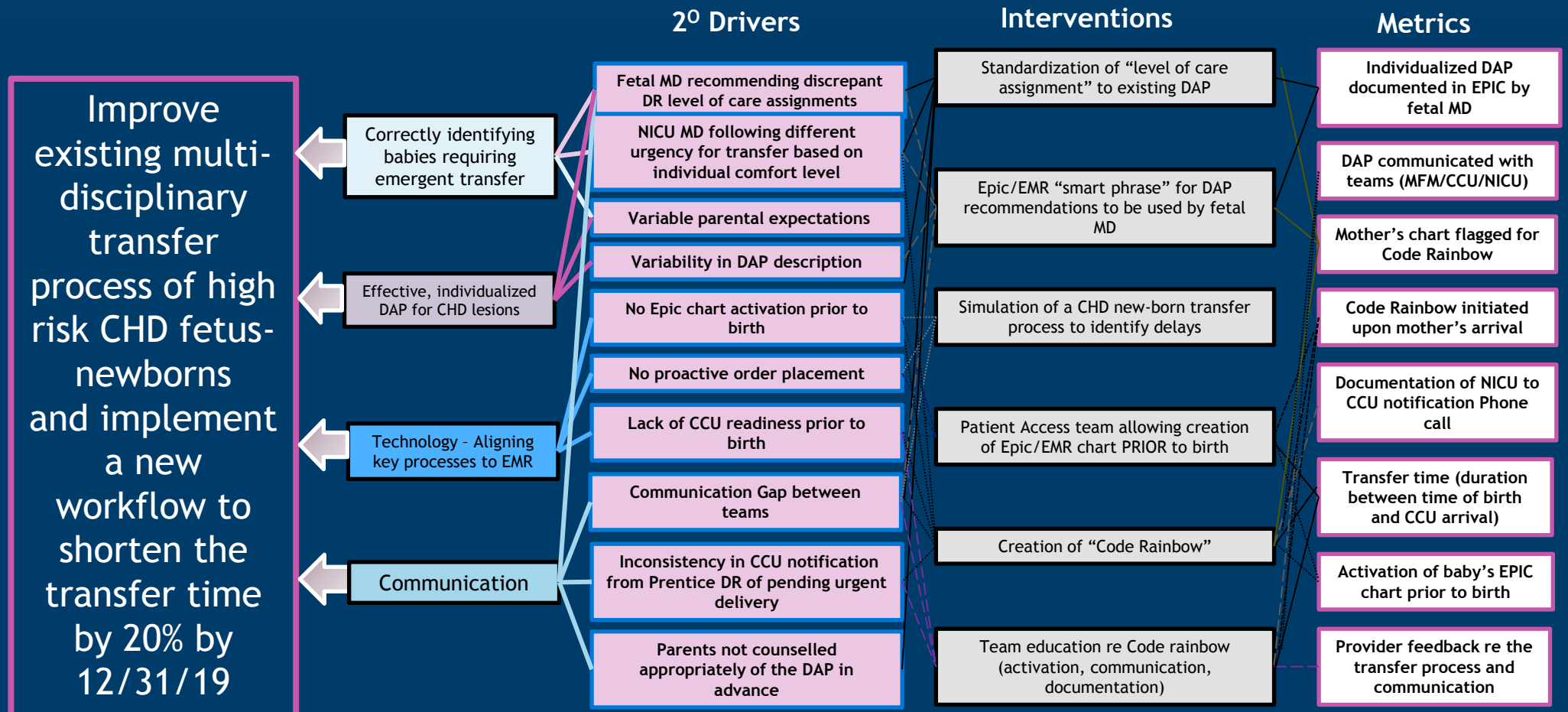
Key Driver Diagram



DR = Delivery room
DAP = Delivery Action Plan
EMR = Electronic Medical Records System (Epic)

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PDSA Ramp: "Code Rainbow"

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Changes That Result in Improvement:

- Reduced transfer time
- Activation of EMR chart prior to birth for resource preparation
- Streamlined communication between teams

PDSA #4: Nov 2019
Provider education on "Code rainbow"
Measure: # providers captured

PDSA #1: December 2018
Stakeholder meeting–Implementation of critical CHD criteria
Measure: # Providers captured

DATA

A P S D

A P S D

A P S D

Hunches
Theories
Ideas

PDSA #3: July 2019
Roll out of "Code Rainbow" workflow
Measure: Transfer time

PDSA #2: April 2019
Simulations to find reasons for delays
Measure: transfer time

Problem:

- Longer than desired transfer time
- Longer than desired resource preparation time
- Suboptimal communication

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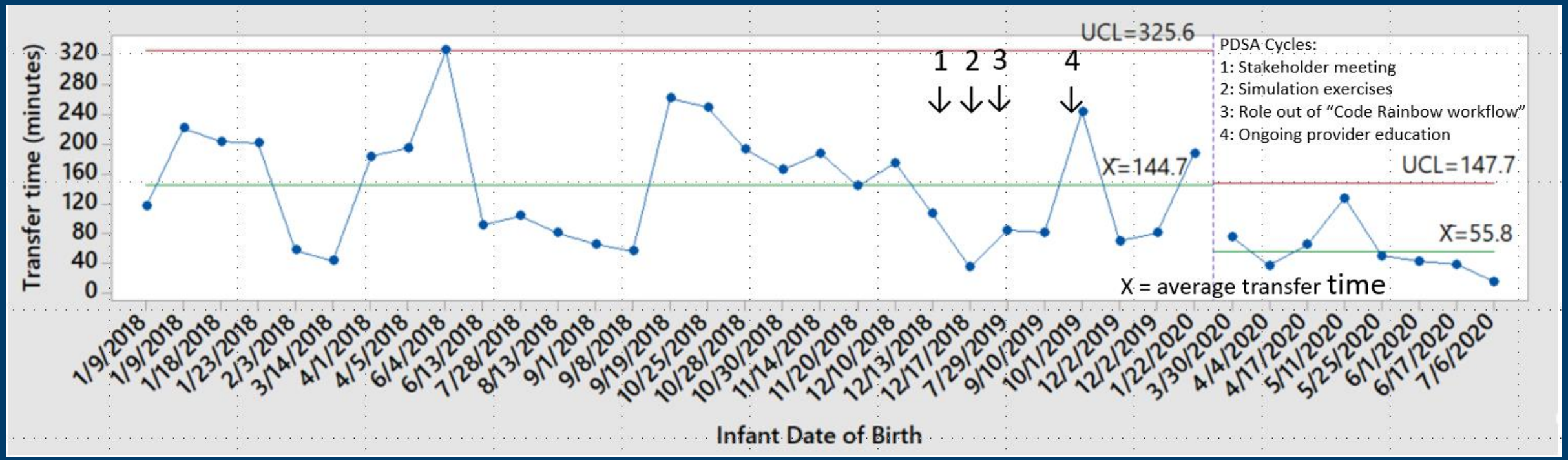
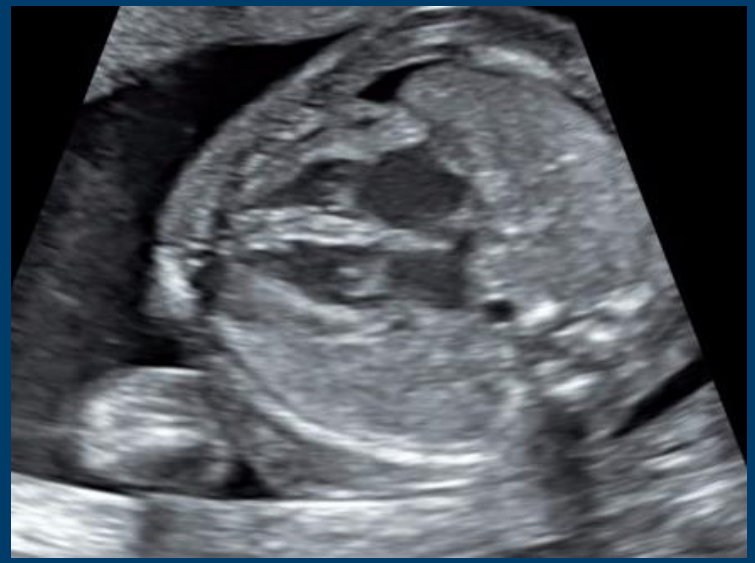

Transfer time (minutes)
320
280
240
200
160
120
80
40
0
UCL=325.6
X=144.7
1 2 3
4
PDSA Cycles:
1: Stakeholder meeting
2: Simulation exercises
3: Role out of "Code Rainbow workflow"
4: Ongoing provider education
UCL=147.7
X=55.8
X = average transfer time
1/9/2018
1/9/2018
1/18/2018
1/23/2018
2/3/2018
3/14/2018
4/1/2018
4/5/2018
6/4/2018
6/13/2018
7/28/2018
8/13/2018
9/1/2018
9/8/2018
9/19/2018
10/25/2018
10/28/2018
10/30/2018
11/14/2018
11/20/2018
12/10/2018
12/13/2018
12/17/2018
7/29/2019
9/10/2019
10/1/2019
12/2/2019
12/2/2019
1/22/2020
3/30/2020
4/4/2020
4/17/2020
5/11/2020
5/25/2020
6/1/2020
6/17/2020
7/6/2020
Infant Date of Birth

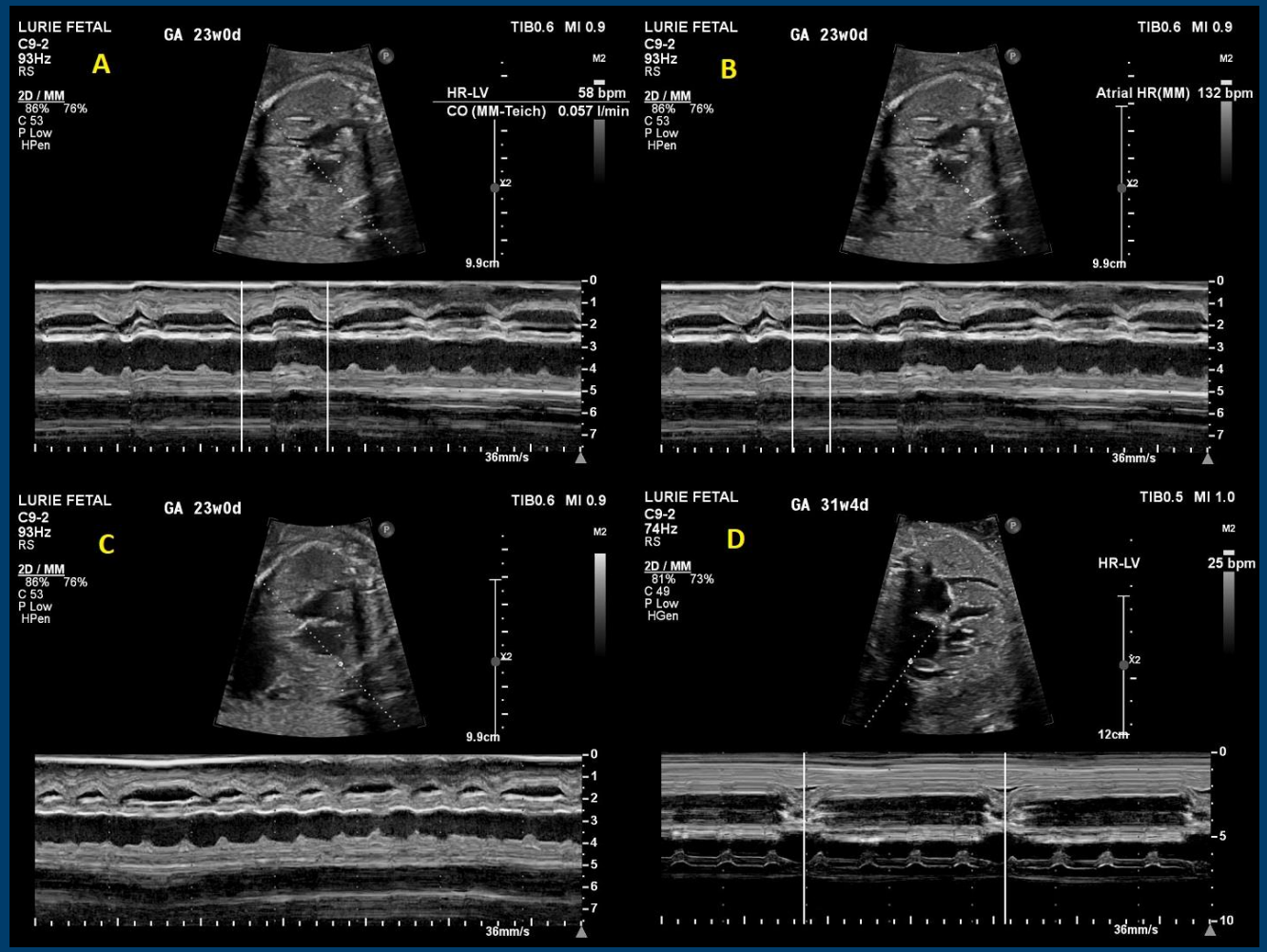
Hydrops: why does it happen?

- Cardiac etiologies
  - Usually: **diastolic dysfunction** (elevated filling pressure) → high RA pressure
  - Arrhythmia: tachyarrhythmia, bradyarrhythmia
  - Structural abnormality: Ebstein's anomaly, premature constriction of the ductus arteriosus, restrictive atrial septum, cardiac tumors
  - Functional abnormality: myocarditis, cardiomyopathy, myocardial infarction, arterial calcification/calcinsosis
  - High-output heart failure: absent ductus venosus, tumor, AV malformation

- Twin-twin transfusion syndrome
  - Placental vascular abnormality due to abnormal vascular connections in monochorionic twin gestation

Bradyarrhythmia: Complete Heart Block





Complete Heart Block Workflow

High Risk CHD – Direct Emergent Transfer from
Prentice Delivery Room to Lurie Children's

Ann & Robert H. Lurie Children's Hospital of Chicago

CLINICAL INDICATION

Babies expected to have hemodynamic instability or low cardiac output state with placental separation due to complete heart block or prolonged bradycardia. (Fetal HR <80 bpm)

CLINICAL TEAM

Team members to be present, or prepared, at time of delivery:

- NICU Resuscitation Team
- MFM OR Team
- CV OR Team and CV Anesthesia Team
- CV Surgery Team (prepared for epicardial pacemaker leads if heart rate after birth indicates need for it)
- EP Team (for pacemaker/HR management)
- CICU Team
- Fetal Cardiology RN Coordinator (if clinically indicated to facilitate communication at time of delivery and transfer)

Arrival to L & D

- Activate CODE RAINBOW to have EPIC chart created prior to birth
- CICU to notify appropriate team members, as listed in clinical teams, that patient has arrived to L & D
- C/S should not start until the cardiac team preparation is completed

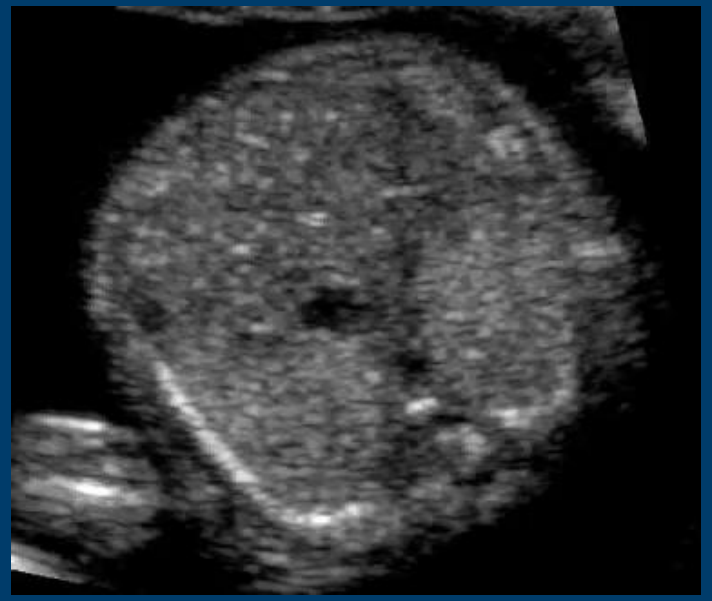
Delivery

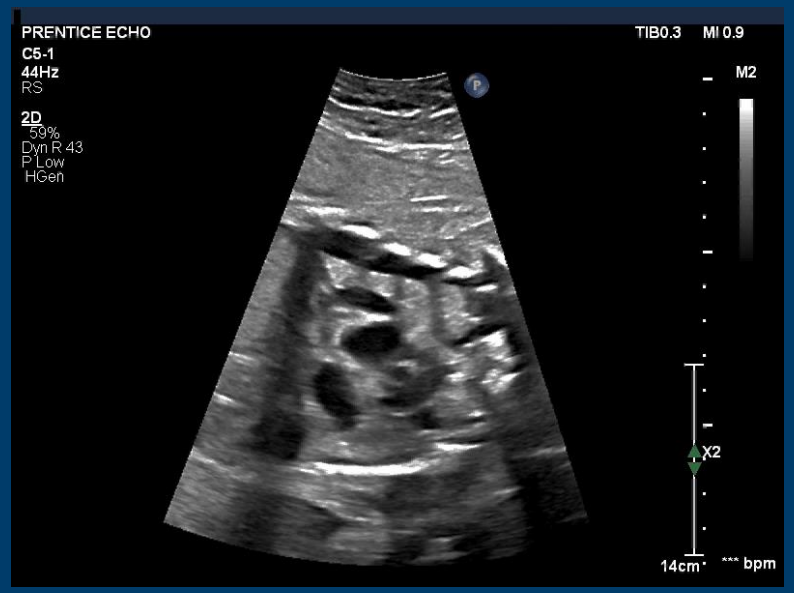
- NICU Resuscitation team to attend delivery
- Anticipate bradycardia (FHR < 100 bpm), consider intubation and sedation/paralytic if necessary
- If there is poor perfusion, HR <60 bpm AND clinical instability, consider epinephrine bolus and infusion
- Place lines and emergent transfer to CICU for bedside epicardial lead placement
- If hemodynamically stable, pt. may bond/breastfeed prior to admission to PWH NICU for stabilization / lines and then transfer to CICU.

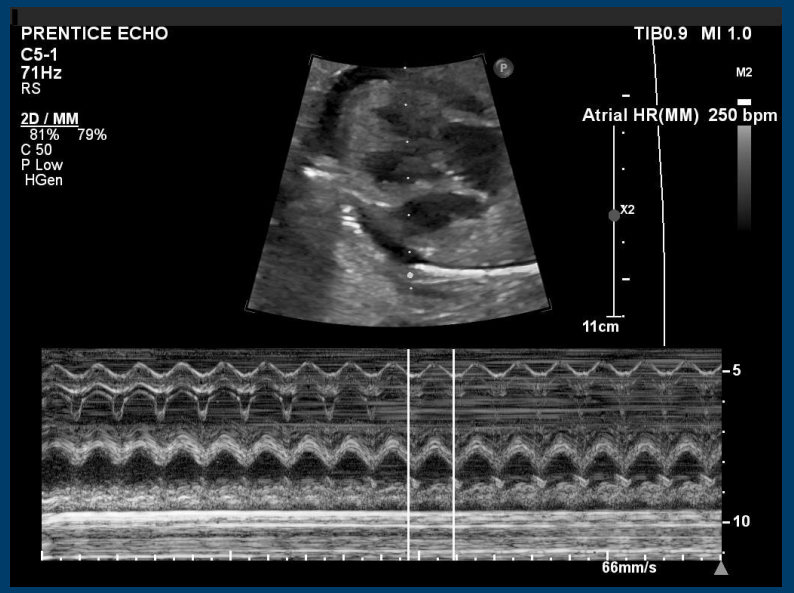
Transfer

- Anticipate urgent transfer to LC CCU for further management.
- CICU will communicate with EP/CVS/CV anesthesia/OR teams re updates.

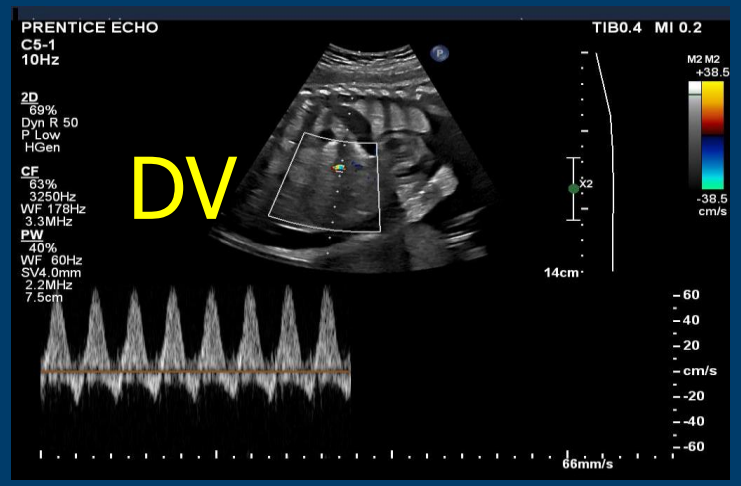
Tachy-arrhythmia : Supra Ventricular Tachycardia

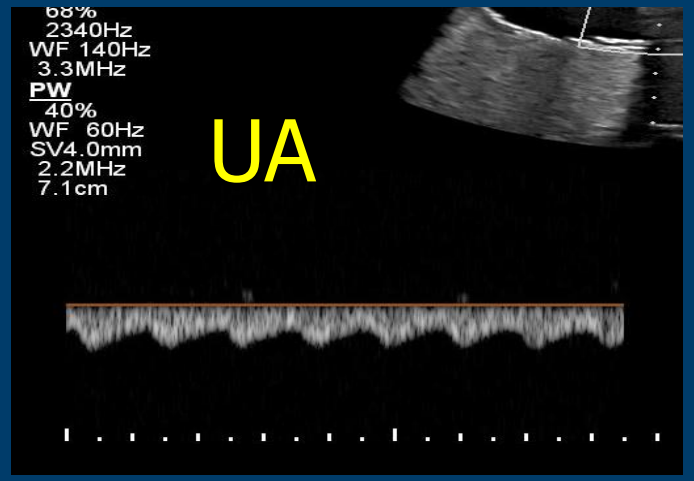


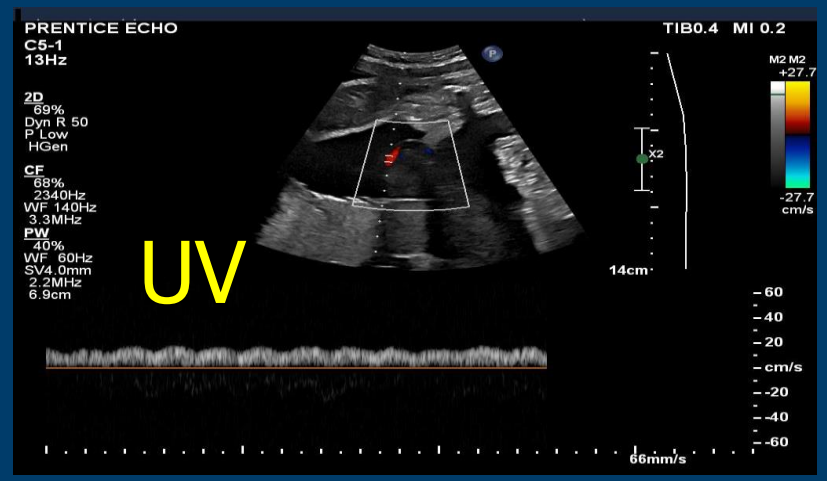




CVP = 4/10







Category	*Score 0 (normal)*	*Score −1*	*Score −2*
Hydrops fetalis	None	Ascites or pleural effusion or pericardial effusion	Skin edema
Venous Doppler	Normal	Flow reversal in ductus venosus tracing	Umbilical venous pulsations
Heart size (heart area/chest area)	≥ 0.2 and < 0.35	0.35–0.50	> 0.5 or < 0.20
Cardiac function	Normal TV and MV, RV/LV shortening fraction > 0.28 and biphasic diastolic ventricular filling	Holosystolic TR or RV/LV shortening fraction < 0.28	Holosystolic MR or dP/dt < 400 or monophasic filling
Arterial Doppler	Normal UA tracing	UA D-wave reaches baseline	UA diastolic flow reversal

Score is 10 if there are no abnormal signs, and there are 1-point or 2-point deductions from total score depending on extent of cardiovascular abnormalities noted for each category[9]. dP/dt, rate of rise of left ventricular pressure; LV, left ventricular; MR, mitral regurgitation; MV, mitral valve; RV, right ventricular; TR, tricuspid regurgitation; TV, tricuspid valve; UA, umbilical artery.

High-output cardiac failure – Tumors & AV malformations

- **Sacrococcygeal teratoma**
  - Tumor near the sacrum with random differentiation into multiple tissues

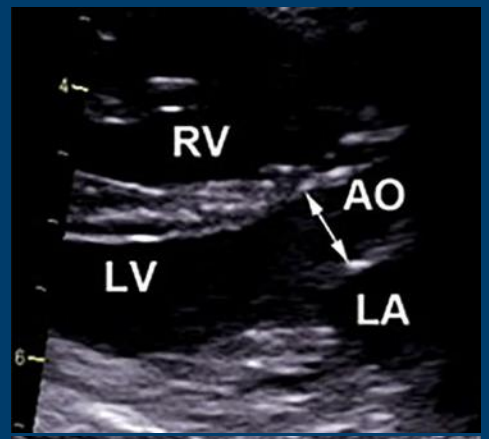
- **Cerebral arteriovenous (AV) malformations**
  - Vein of Galen malformation
  - Associations: superior sinus venosus defect, coarctation
  - Dilation of superior vena cava, "runoff" from aorta into carotid arteries

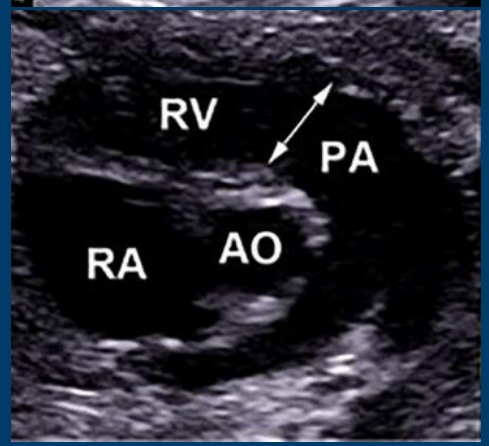
- **Placental chorioangioma**
  - Benign angioma of the placenta
  - Most small (< 5 cm)
  - When large, may act as A-V shunt
  - Preeclampsia, preterm labor, placental abruption and polyhydramnios

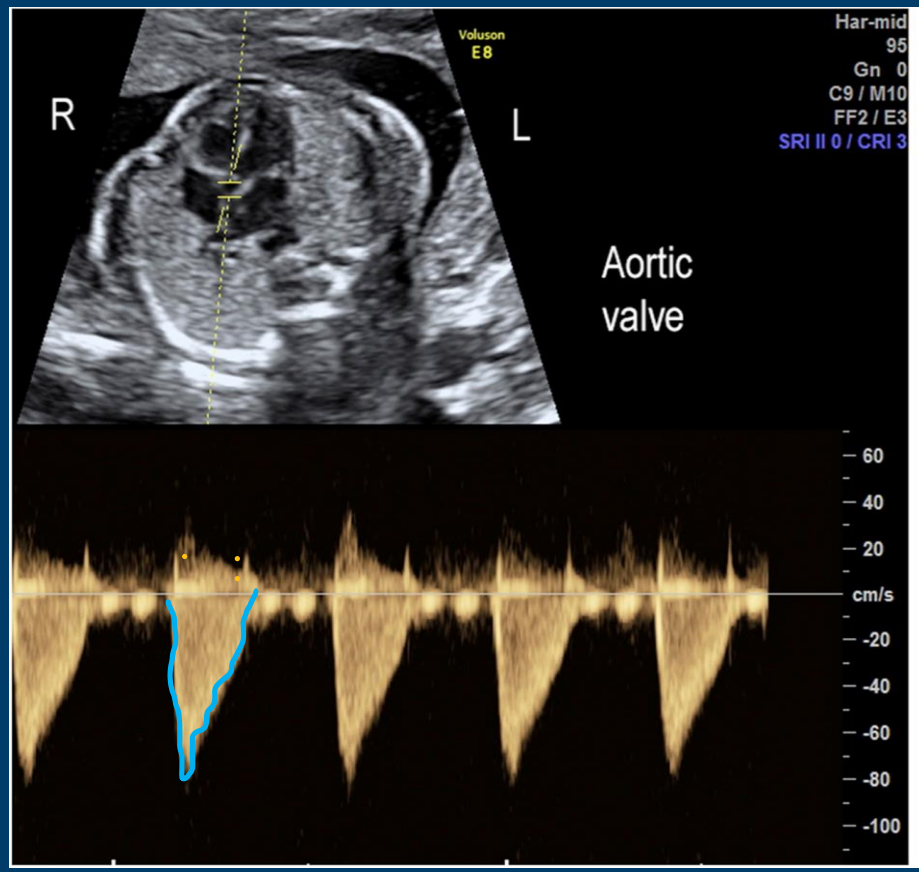
- **Pulmonary AV malformation (rare), Agenesis of ductus venosus**

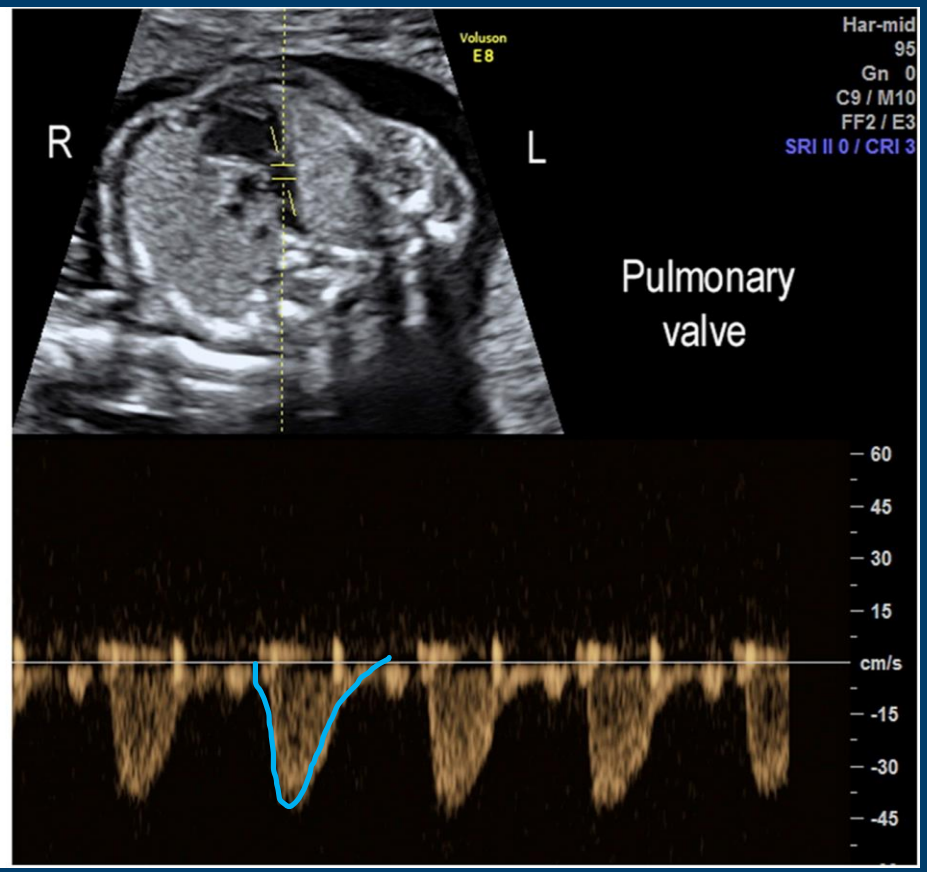
High output cardiac failure: Combined cardiac output

- LV output = aortic valve time-velocity integral * AoV area * heart rate
- RV output = pulmonary valve time-velocity integral * PV area * heart rate
- CCO = LV output + RV output; CCOi = CCO / estimated fetal weight
- Calculator/normal values: http://fetal.parameterz.com/cco









High-output cardiac failure

Ultrasound Obstet Gynecol 2013; 41: 54–58
Published online in Wiley Online Library (wileyonlinelibrary.com). DOI: 10.1002/uog.12309

Estimated cardiac output and cardiovascular profile score in fetuses with high cardiac output lesions

C. J. STATILE, J. F. CNOTA, S. GOMIEN, A. DIVANOVIC, T. CROMBLEHOLME and E. MICHELFELDER

Cincinnati Children's Hospital Medical Center, Cincinnati, OH, USA

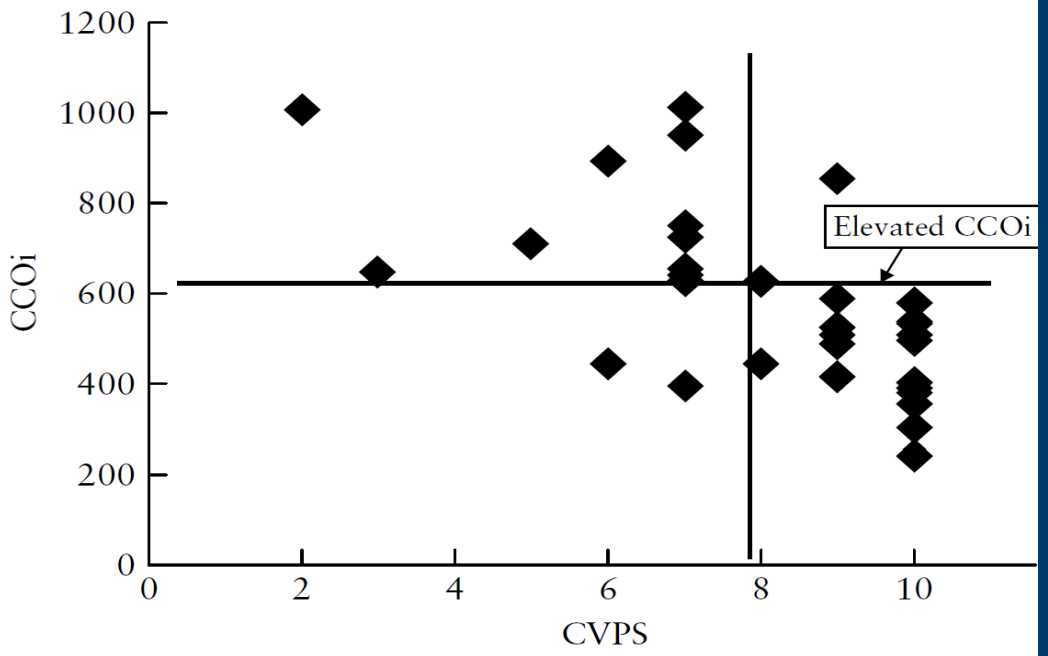


Figure 2 Scatterplot of indexed combined cardiac output (CCOi) against cardiovascular profile score (CVPS) for individual fetuses (filled diamonds). Significant cardiovascular compromise was defined as CVPS < 8 (vertical line). Abnormal CCOi was defined as > 625 mL/min/kg[14] (horizontal line).

Twin-twin transfusion syndrome (TTTS)

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- Placental vasculopathy
- Monochorionic multiple pregnancies (≈10%)

- **Donor**:
  - Oligohydramnios

- **Recipient**:
  - Polyhydramnios
  - Initially increased output
  - Cardiomegaly, AV valve regurgitation, cardiomyopathy
  - Risk of developing pulmonary stenosis/RVOT obstruction
  - Fetal demise

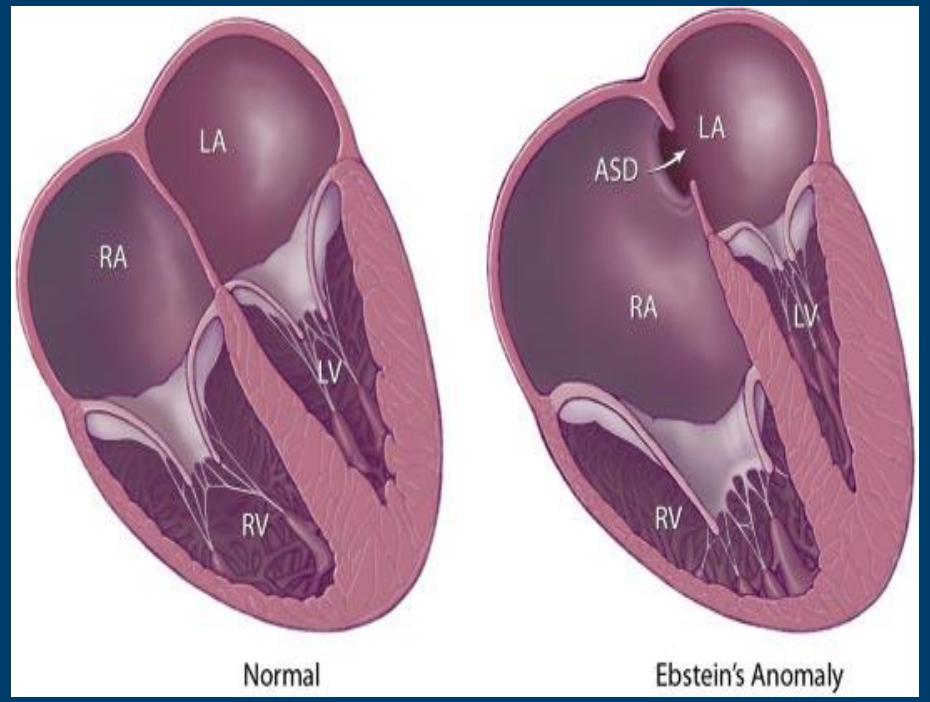
Twin-twin transfusion syndrome (TTTS)

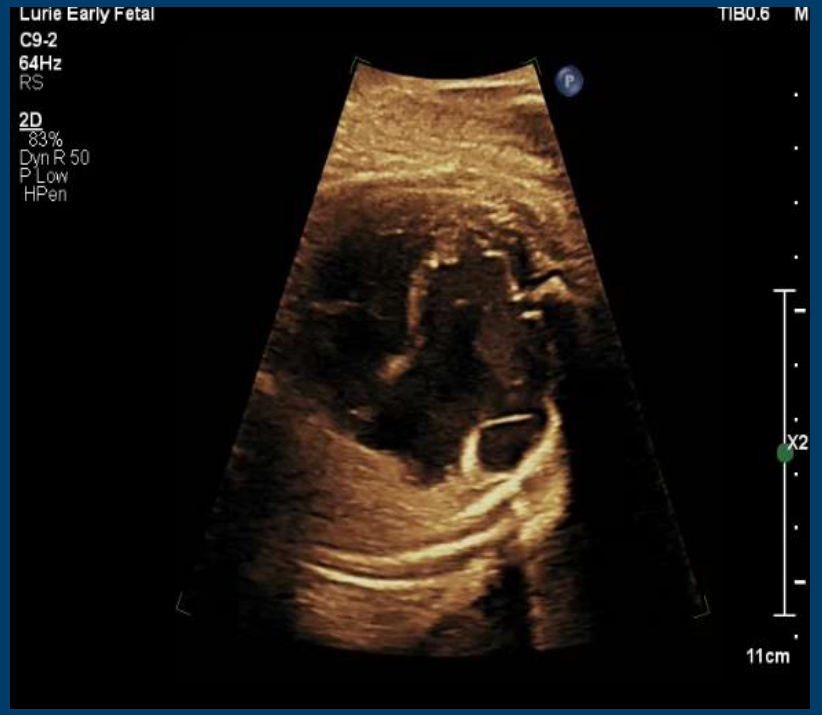
Ann & Robert H. Lurie Children's Hospital of Chicago®

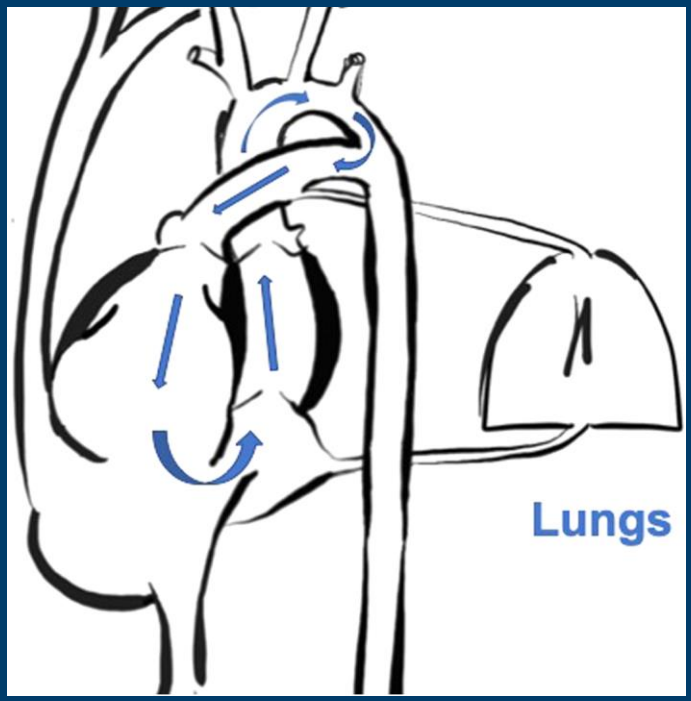
- **Pathophysiology**
  - Initially increased output
  - Polyhydramnios
  - Cardiomegaly, ventricular hypertrophy
  - AV valve regurgitation
  - Right heart → left heart systolic/diastolic function
  - Abnormal Dopplers
  - Effusions/hydrops
  - Fetal demise

Fetal lie	Fetal presentation is transverse.
Umbilical artery	Normal.
Umbilical vein	Normal.
Ductus venosus	Normal.
LV MPI (Tei)	0.34.
RV MPI (Tei)	0.33.
LV hypertophy	No left ventricular hypertrophy.
RV hypertrophy	No right ventricular hypertrophy.
Mitral regurgitation	No.
Tricuspid regurgitation	No.
Pulmonary reguitation	No.
Pericardial effusion	No.
Pleural effusion	No.
Ascites	No evidence of ascites.
Hydrops	No hydrops.
Demise	

Tricuspid Regurgitation







1. Ebstein's anomaly of the tricuspid valve
2. Tricuspid valve dysplasia
3. Pulmonary atresia with intact ventricular septum resulting in significant tricuspid regurgitation

Tricuspid regurgitation (TR)

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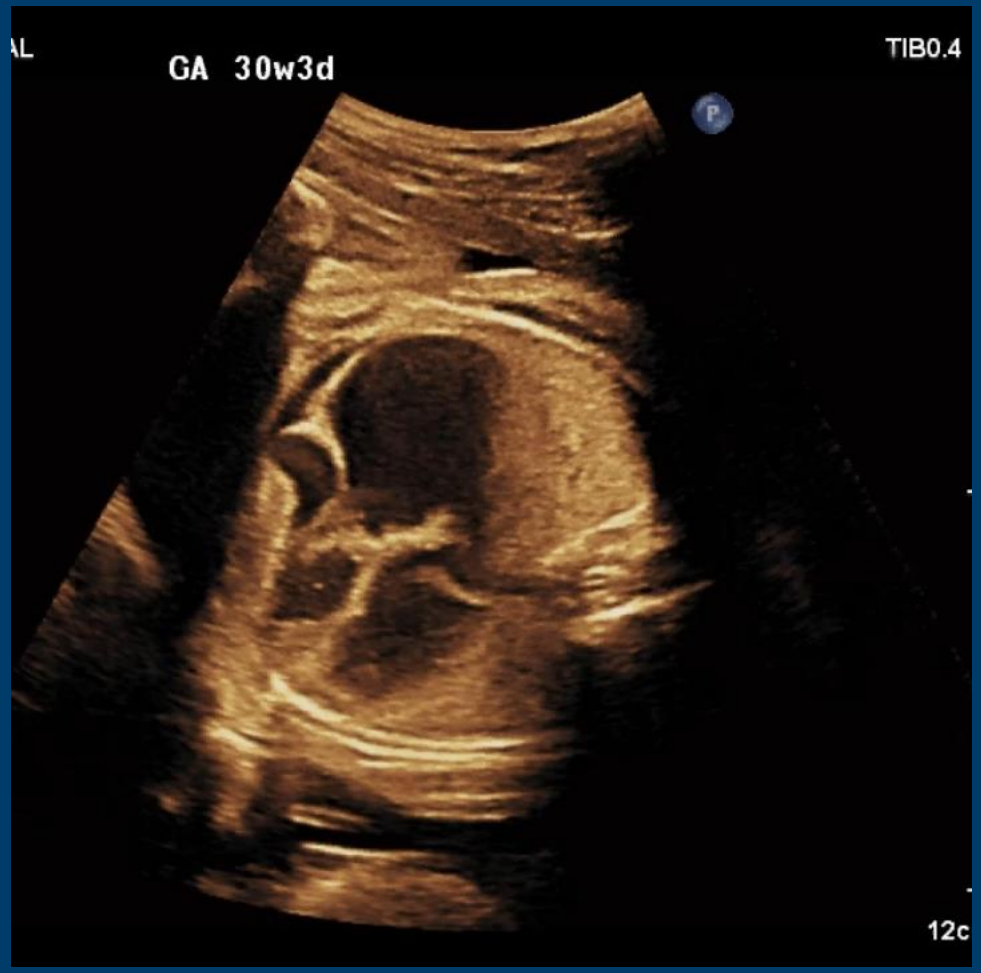
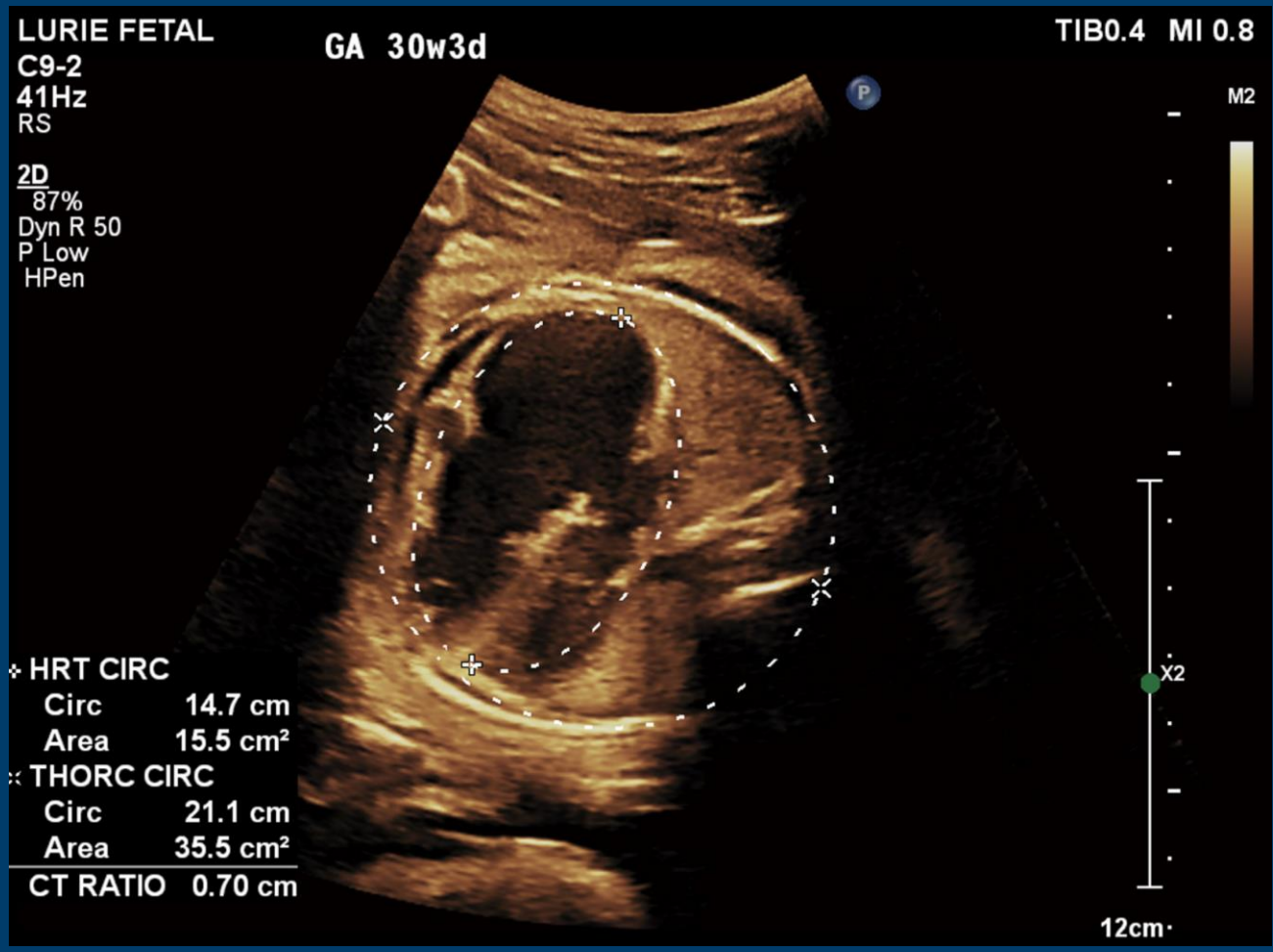
- **Pulmonary atresia, intact ventricular septum with TR**





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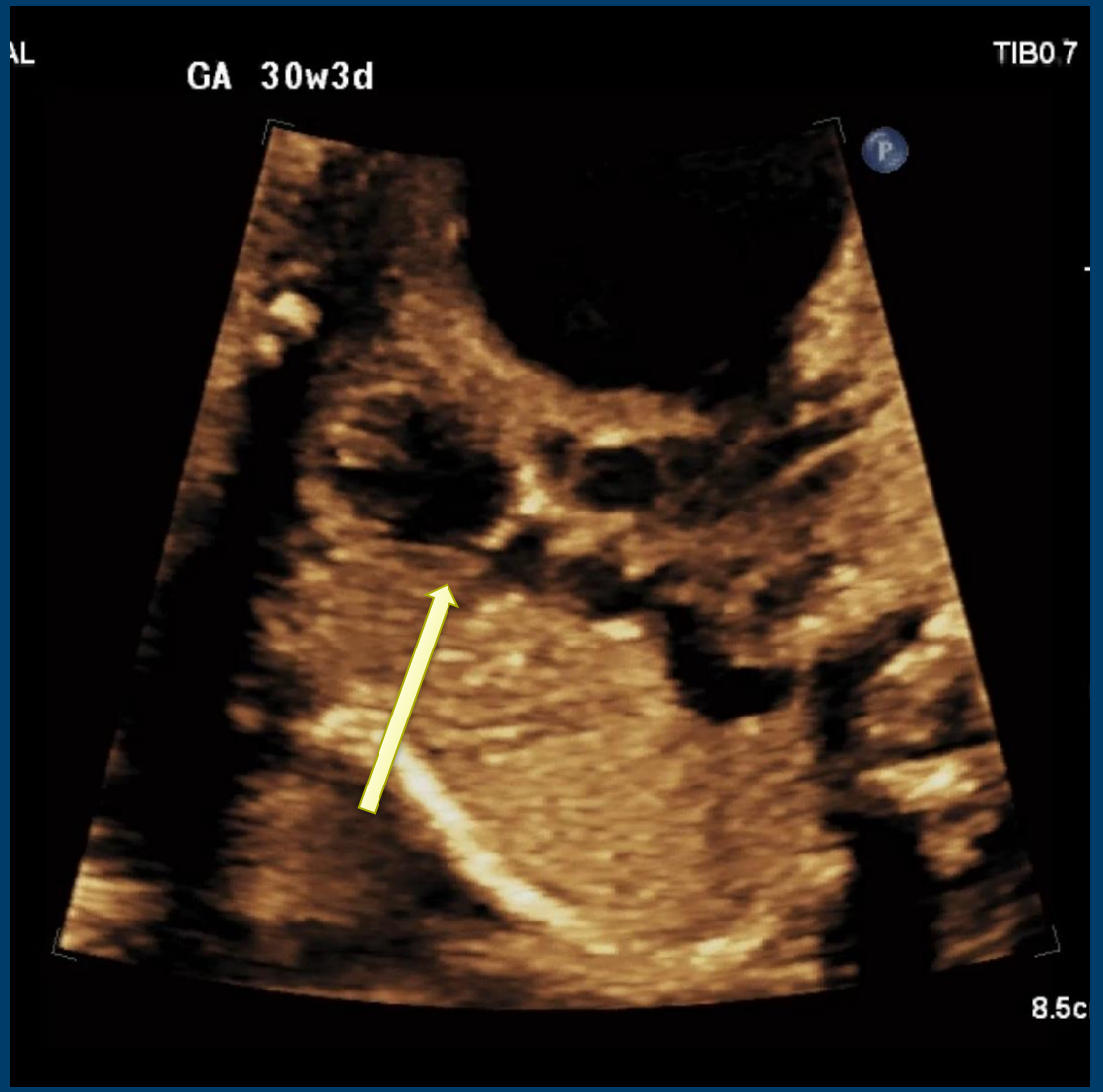
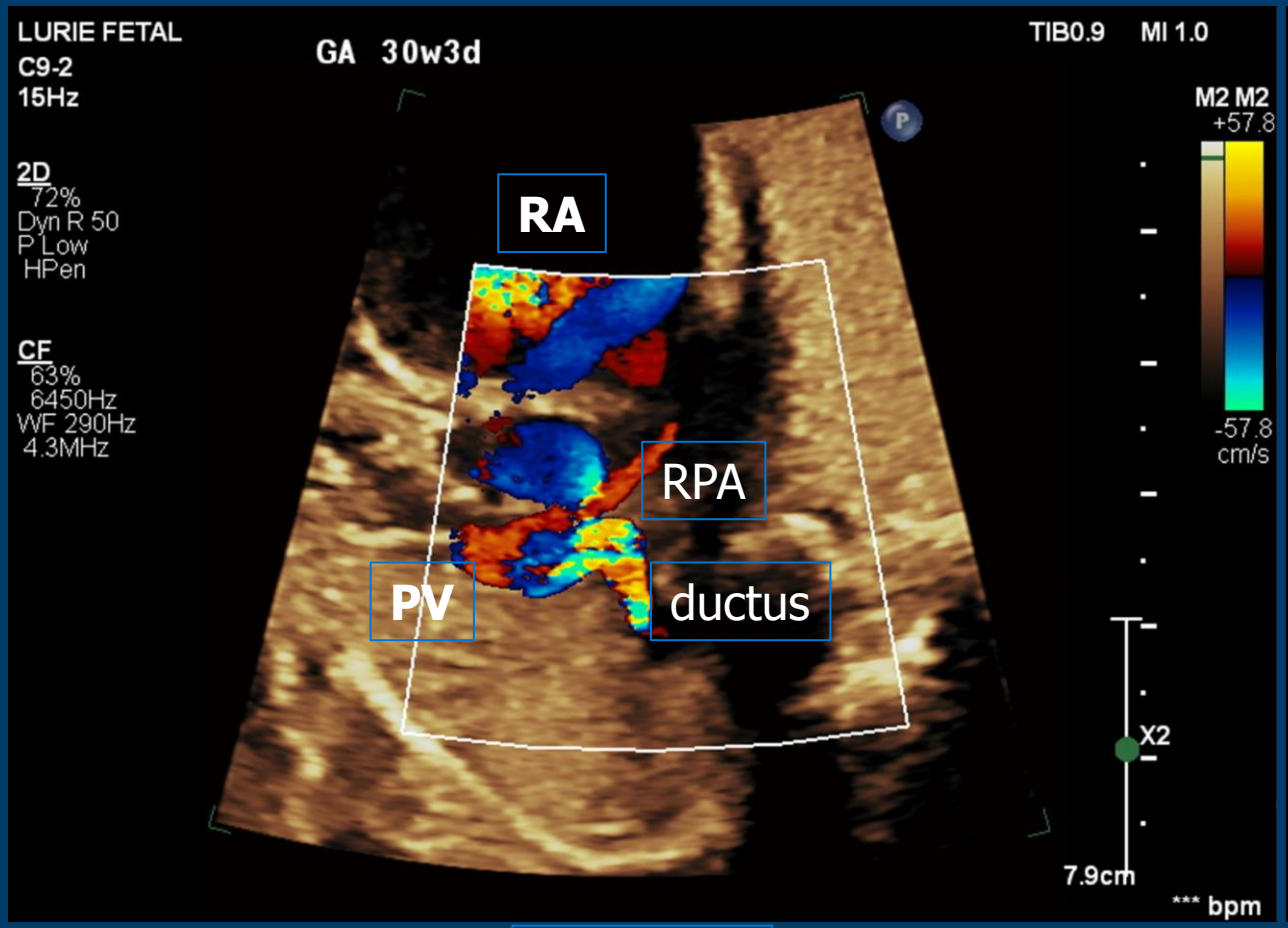
Tricuspid regurgitation (TR)

- **Pulmonary atresia, intact ventricular septum with TR**

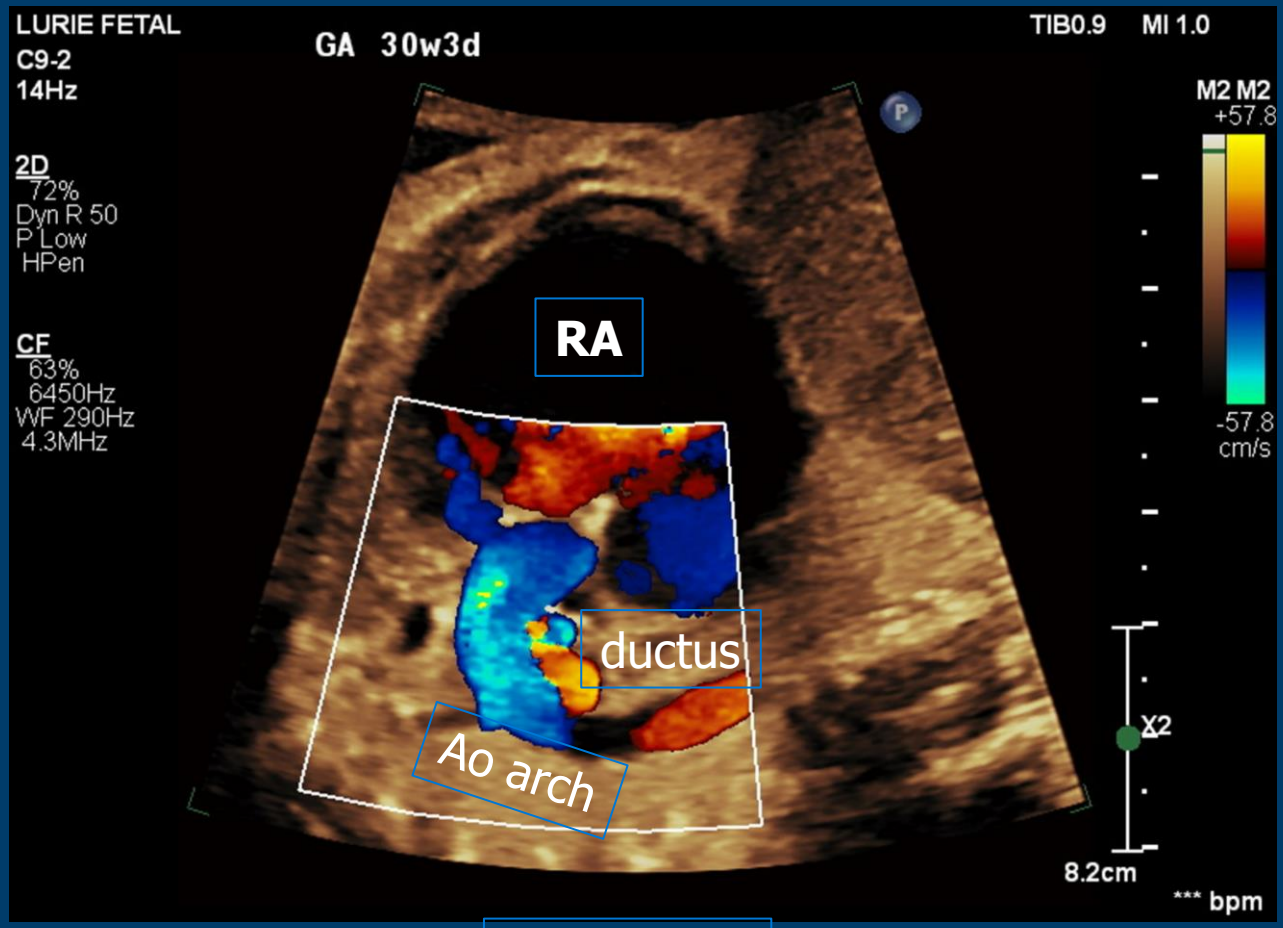


Tricuspid regurgitation (TR)

- **Pulmonary atresia, intact ventricular septum with TR**

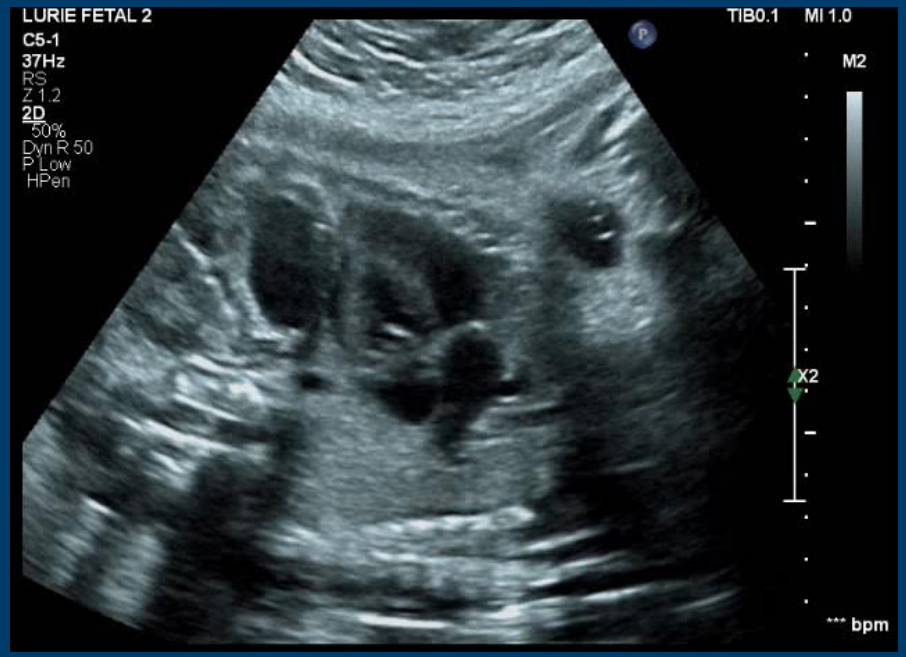


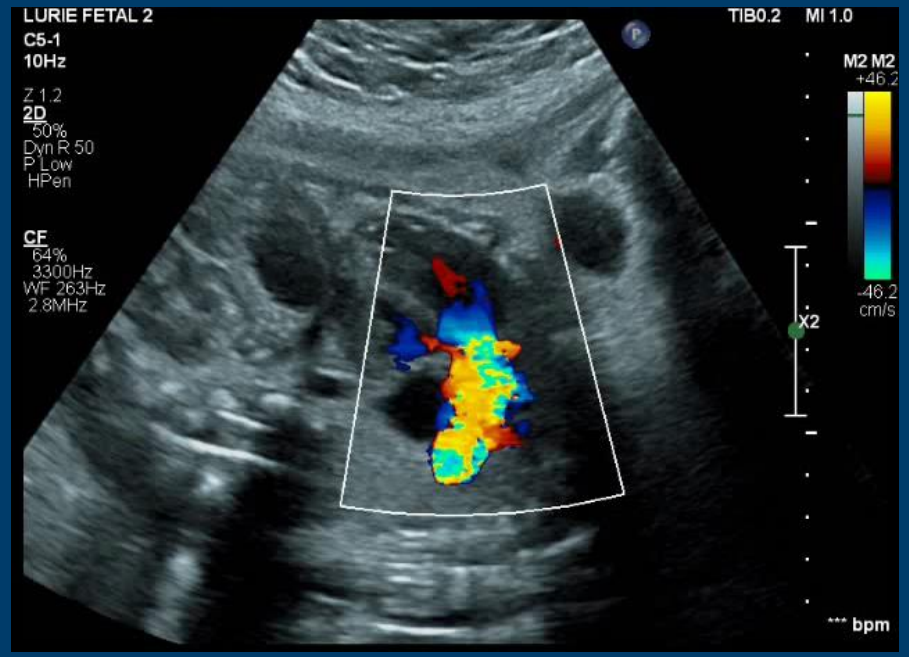
transverse

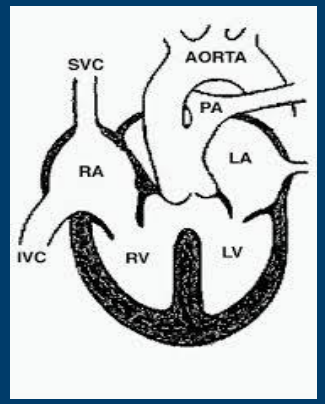


parasagittal

Truncus arteriosus with truncal valve stenosis







Prenatal Predictors of Perinatal Mortality in Truncus Arteriosus: A Fetal Heart Society Collaborative Study

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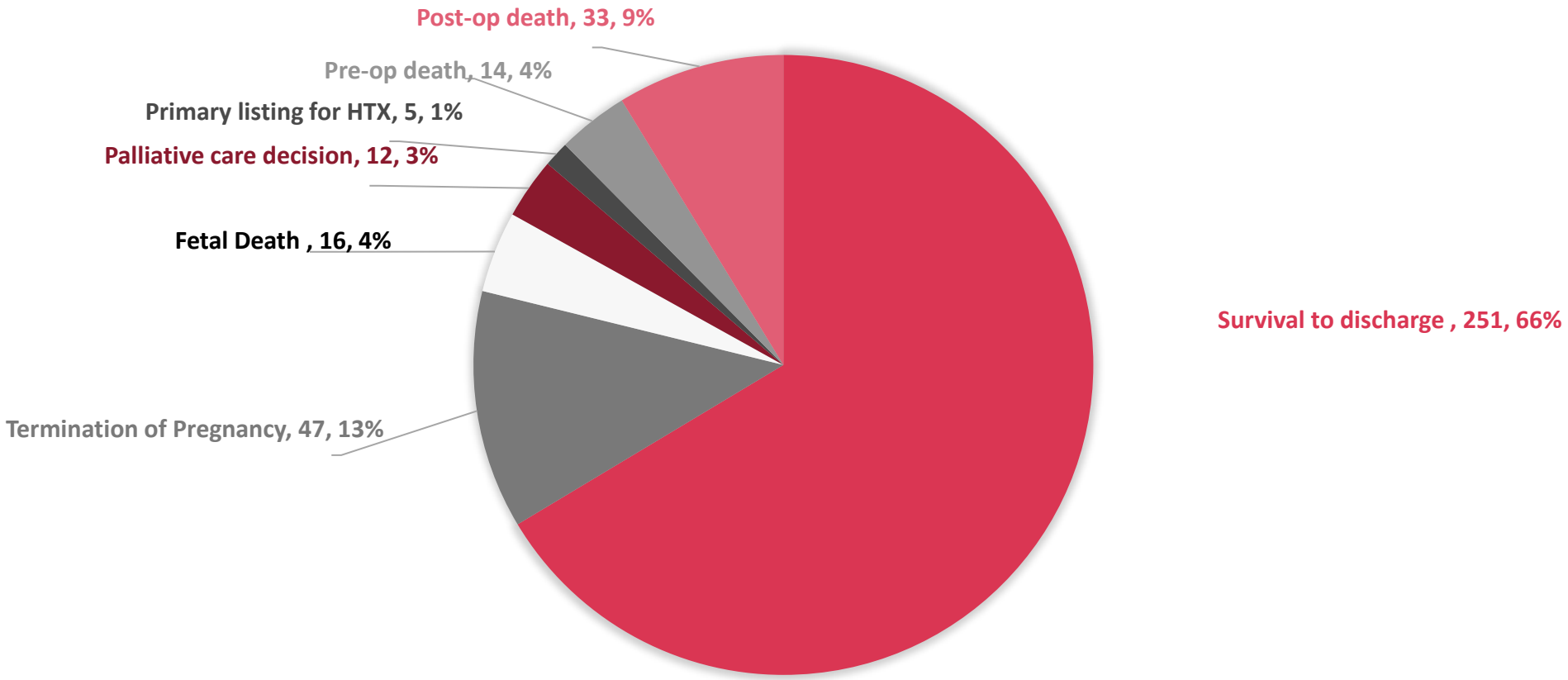
Pregnancy Outcomes in Prenatally Diagnosed TA



Fetal Heart Society®

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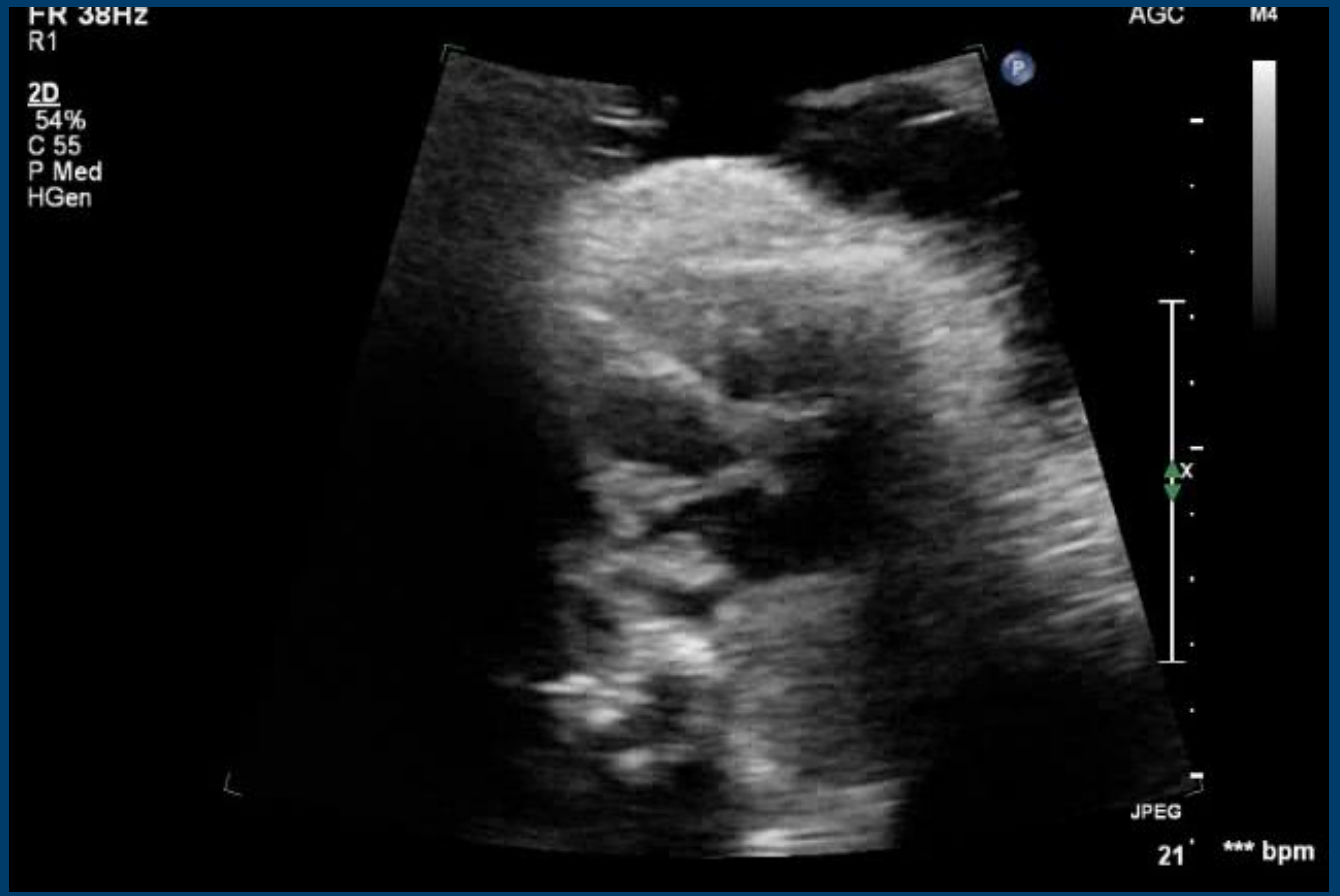
Cardiovascular Profile Score Associated with Perinatal Mortality

		Pre-Operative Mortality			Pre- Hospital Discharge Mortality		
	Overall N = 319	No mortality N = 282 (88.4%)	Death N = 37 (11.6%)	p-value	No Mortality N = 253 (79.3%)	Death N = 66 (20.7%)	p-value
Pericardial Effusion	16 (5%)	9 (3%)	7 (19%)	**0.000**	8 (3%)	8 (12%)	**0.012**
Skin Edema	4 (1%)	1 (1%)	3 (8%)	**0.000**	1 (1%)	3 (5%)	**0.025**
Pleural Effusion	3 (1%)	1 (1%)	2 (5%)	**0.006**	1 (1%)	2 (3%)	0.109
Ascites	4 (1%)	2 (1%)	2 (5%)	**0.039**	2 (1%)	2 (3%)	0.298
Cardiomegaly	58 (18%)	48 (17%)	10 (27%)	0.206	43 (17%)	15 (23%)	0.555
Abnormal Umbilical Vein Doppler	6 (2%)	2 (1%)	4 (11%)	**0.000**	2 (1%)	4 (6%)	**0.015**
Abnormal Umbilical Artery Doppler	7 (2%)	3 (1%)	4 (11%)	**0.001**	3 (1%)	4 (6%)	**0.049**
Abnormal Ductus venosus Doppler	20 (6%)	11 (4%)	9 (24%)	**0.000**	10 (4%)	10 (15%)	**0.004**
Overall CVP Score (Median (IQR))	9 (8, 10)	9 (8, 10)	9 (7.5, 10)	0.117	10 (8, 10)	9 (8, 10)	0.174
CVP </= 7	22 (7%)	17 (6%)	5 (13%)	0.19	16 (6%)	6 (9%)	0.668

Fetal Cardiomyopathy



Key takeaway points

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- Fetal echocardiogram should be performed if hydrops or effusion is identified
  - 15 – 25% with cardiac abnormalities
- Common cardiac/CV etiologies (hydrops/pericardial effusion):
  - fetal arrhythmia, structural abnormalities (Ebstein's, outflow tract obstruction with shunting restriction, arterial duct constriction, agenesis of ductus venosus); rarely: cardiac tumor
- High-output heart failure can be seen with anemia, vascular tumors, AVM and twin-twin transfusion syndrome
- Use of quantifiable measures (CVP, CCO<others) allow for serial monitoring

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