Prenatal Evaluation of the Fetus with Pulmonary Valve Stenosis

Children's Mercy Fetal Cardiology Education Series

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Disclosures

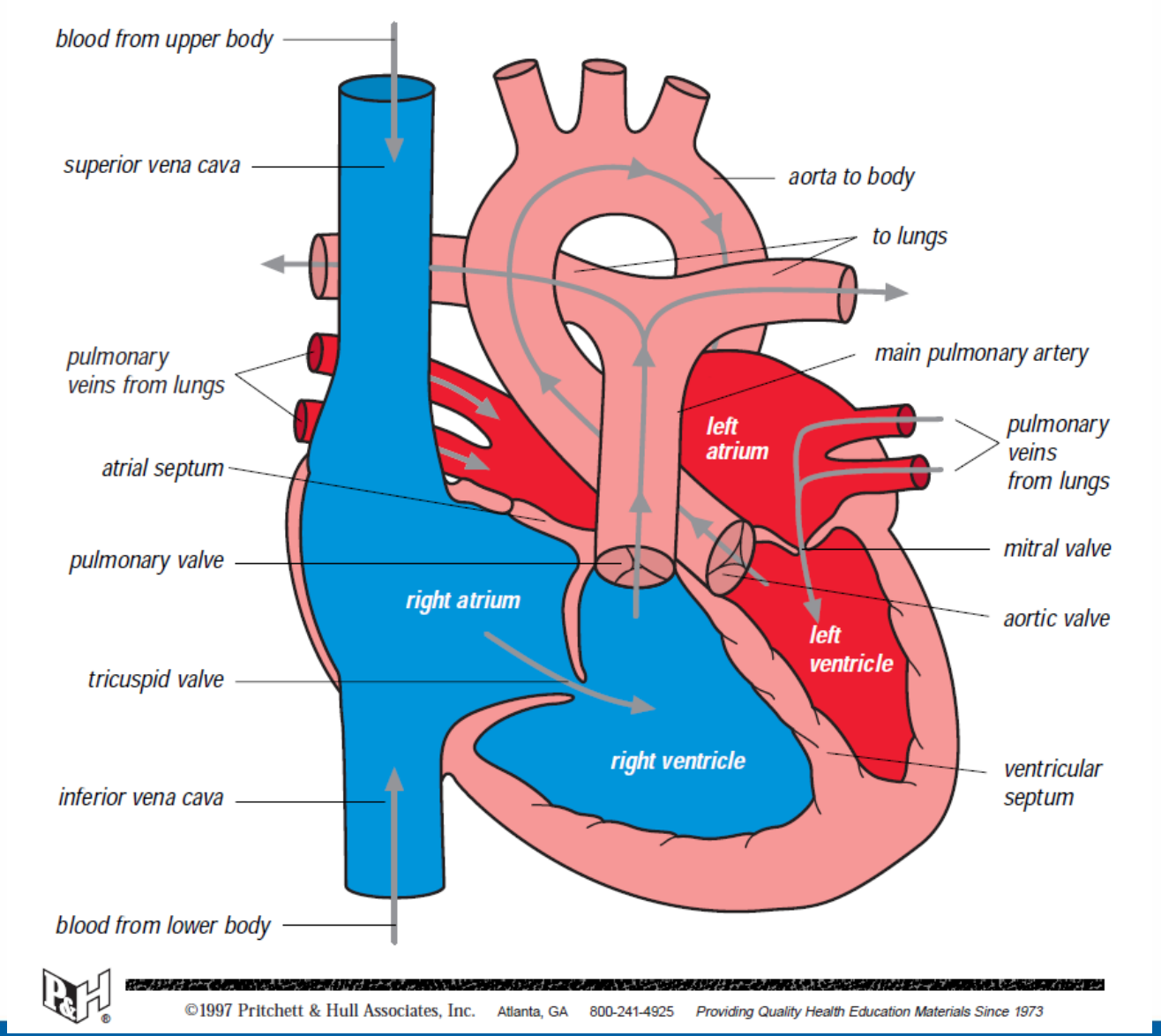
- No disclosures

Rychik, Jack. Zhiyun, Tian. *Fetal Cardiovascular Imaging: A Disease-Based Approach.* Elsevier Saunders, 2012.

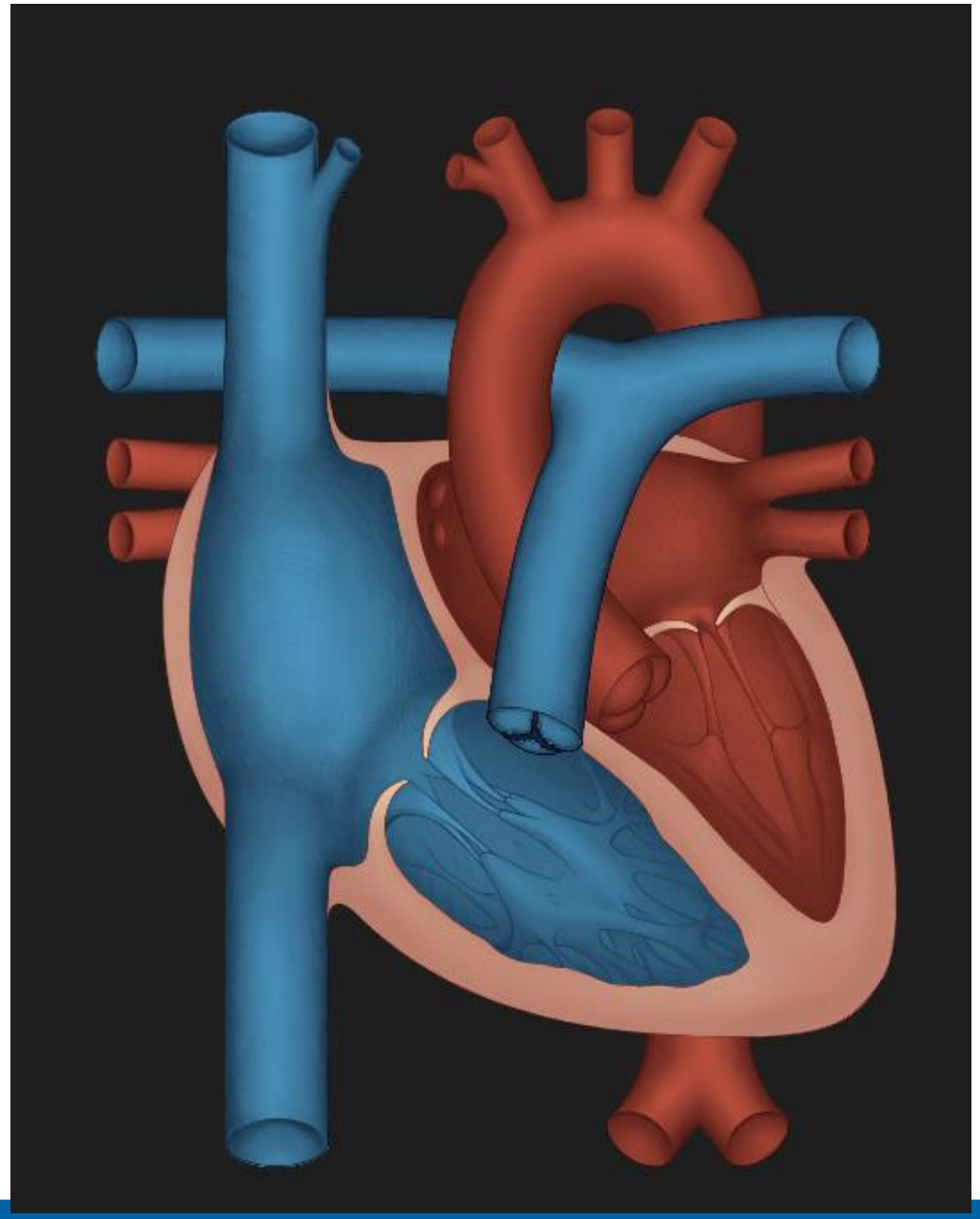
Objectives

- Understand the basic anatomical considerations of pulmonary valve stenosis (PS)
- Define fetal imaging goals for PS
- Recognize aspects of PS anatomy on shared PS fetal imaging cases
- Discuss prenatal and postnatal management and outcomes

Normal Heart



Anatomy of PS



Anatomy of PS

- Subvalvar, valvar, supravalvar
- Valvar: 6 anatomic subtypes!
  - Doming 42%
  - Unicommissural 16%
  - Bicuspid 10%
  - Tricuspid 6%
  - Hypoplastic annulus 6%
  - Dysplastic 19%
- Dysplastic: markedly thickened, redundant leaflets (Noonan syndrome)

Anatomy of PS

- Post-stenotic dilation of the main pulmonary artery (high velocity flow jet through the narrowed pulmonary valve)
- Tricuspid regurgitation (high RV pressure)
- Dilated right atrium (elevated RV filling pressures)
- RV hypoplasia
- Coronary sinusoids (absence of significant TR, connections between the hypertensive RV and developing coronary circulation; more common in pulmonary atresia)

Associated Lesions

- Tetralogy of Fallot
- Double outlet right ventricle or d-transposition of the great arteries
- Ventricular septal defect
- Tricuspid atresia or stenosis
- Ebstein
- Congenitally corrected transposition of the great arteries

Frequency and Genetics

- Isolated PS: 7-12% of patients with CHD
- Including associated lesions: PS occurs in 25-30% of patients
- Associated genetic syndromes:
  - Neurofibromatosis
  - LEOPARD syndrome
  - Noonan syndrome
  - William syndrome (supravalvar PS)

Prenatal Evaluation

- PV Doppler: Peak velocity of >1 m/sec across PV is abnormal
- Flow within the ductus arteriosus (may be reversed)
- RV size, hypertrophy, function
- TV size and function, tricuspid regurgitation
- Main and branch pulmonary arteries

Fetal Echo Features of PS

Key Echocardiographic Features

- Size of the pulmonary annulus in comparison with the aortic annulus.
- Laminar or turbulent flow across pulmonic valve?
- Peak systolic velocity across the pulmonic valve.
- Morphology of the pulmonary valve.
- Size of the tricuspid annulus compared with the mitral annulus.
- Degree of tricuspid regurgitation.
- Right ventricle pressure estimate based on tricuspid regurgitation peak velocity.
- Right atrial size.
- Right ventricle size and function.
- Degree of right ventricular hypertrophy.
- Direction of flow within the ductus arteriosus.
- Size of the branch pulmonary arteries.
- Size and appearance of the foramen ovale.
- Evidence for elevated central venous pressure as marked by reversal of flow with atrial contraction in the ductus venosus.
- Evidence for elevated central venous pressure as marked by pulsations within the inferior vena cava.
- Presence of any ventriculocoronary "sinusoid" connections.

Fetal Imaging Essentials

Imaging Essentials and Important Points

- Mild pulmonary stenosis can be missed on fetal echocardiography.
- The pulmonary valve can be imaged in multiple planes. Examples of excellent views include the short axis at the level of the great vessels and long-axis angling anteriorly toward the right ventricular outflow tract. From the four-chamber view, anterior angling just beyond visualization of the left ventricular outflow tract will reveal the right ventricular outflow tract. This view provides an opportunity for optimal Doppler sampling across the pulmonary valve.
- In pulmonary stenosis, Doppler interrogation across the pulmonary valve will reveal "turbulent" flow and loss of laminar flow as well as a peak velocity that is greater than 1 m/sec.
- Both the pulmonary annulus and the tricuspid annulus are larger than the aorta and mitral valve in the normal fetus. In pulmonary stenosis, however, the pulmonary annulus may be smaller than normal and should be measured and plotted as gestation progresses.
- The degree of associated tricuspid valve hypoplasia reflects the degree of right ventricular hypoplasia that is present. In many cases, the tricuspid annulus will be small, but of adequate size, typically at a gestational age Z-score of greater than −3 (no smaller than 3 standard deviations from the expected mean for gestational age).
- A general sense and qualitative impression of the cavity size of the right ventricle should be made in every fetus with pulmonic stenosis. Actual measurement of cavity volume can be extremely challenging because of the geometric nature of the right ventricle and its hypertrophied state in pulmonary stenosis. This does not lend itself to accurate formulaic equations that can be applied. The best indirect gauge of right ventricular capacity is measurement of the tricuspid valve annulus.
- Doppler evaluation of tricuspid valve inflow, ductus venosus, and umbilical vein flow in the fetus can help assess right ventricular compliance and predict who may have significant diastolic dysfunction after birth, with right-to-left shunting at the foramen ovale and consequential cyanosis. Features such as (1) single-peak inflow across the tricuspid valve, (2) absence or reversal of forward flow with atrial contraction in the ductus venosus, or (3) venous pulsations in the umbilical vein reflect altered right ventricular compliance and should be noted.
- Signs of hydrops fetalis in association with pulmonary stenosis are troubling. Abnormal Doppler flow patterns as described previously have not been demonstrated to progress to hydrops in the fetus with pulmonic stenosis and do not predict impending hydrops because diastolic dysfunction may remain stable throughout fetal life without any significant clinical manifestations until birth. However, the combination of a restrictive foramen ovale with the venous Doppler changes described previously may place the fetus at risk for impediment to forward flow and may impair cardiac output. Such a fetus is at substantial risk for hydrops.
- Severity of tricuspid regurgitation, if present, should be monitored carefully. Severe tricuspid regurgitation is a risk factor for development of hydrops.
- The tricuspid valve should be inspected carefully when there is tricuspid regurgitation in the presence of pulmonary stenosis. Abnormalities such as Ebstein's anomaly or dysplasia of the tricuspid valve can be seen. Careful attention should be paid to the septal leaflet of the tricuspid valve as a mild Ebstein anomaly can be missed.

Prenatal Management

- Mild to moderate PS: well tolerated in utero
- Severe: need for neonatal intervention
- Serial evaluation: PS may progress over the pregnancy
- Most fetuses tolerate a vaginal delivery
- Prostaglandin recommendation
  - Inadequate antegrade PV flow
  - Reversal of flow in the ductus arteriosus

Fetal PS Intervention

Pro (critical PS)

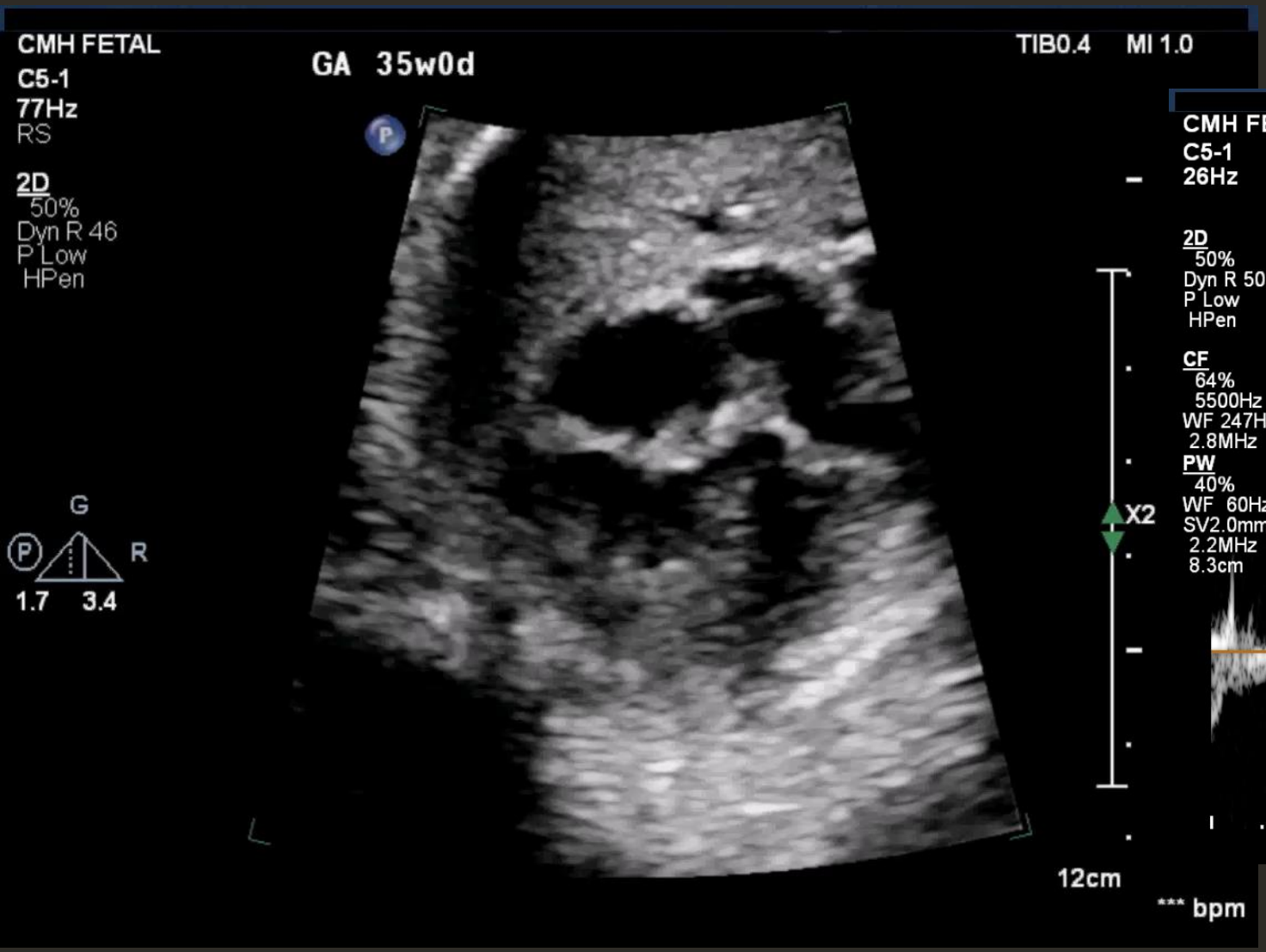
- Prevent fetal hydrops
- Ensure biventricular circulation
- Proposed indications:
  - Impending hydrops
  - Decreased LV function
  - Marked flow reversal in DV
  - IVC pulsations
  - Restrictive PFO

Con

- Most fetuses with PS have an adequately sized RV and achieve biventricular circulation
- Procedural risk to mother and fetus

PS Cases

PS Case 1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50%
Dyn R 46
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HPen
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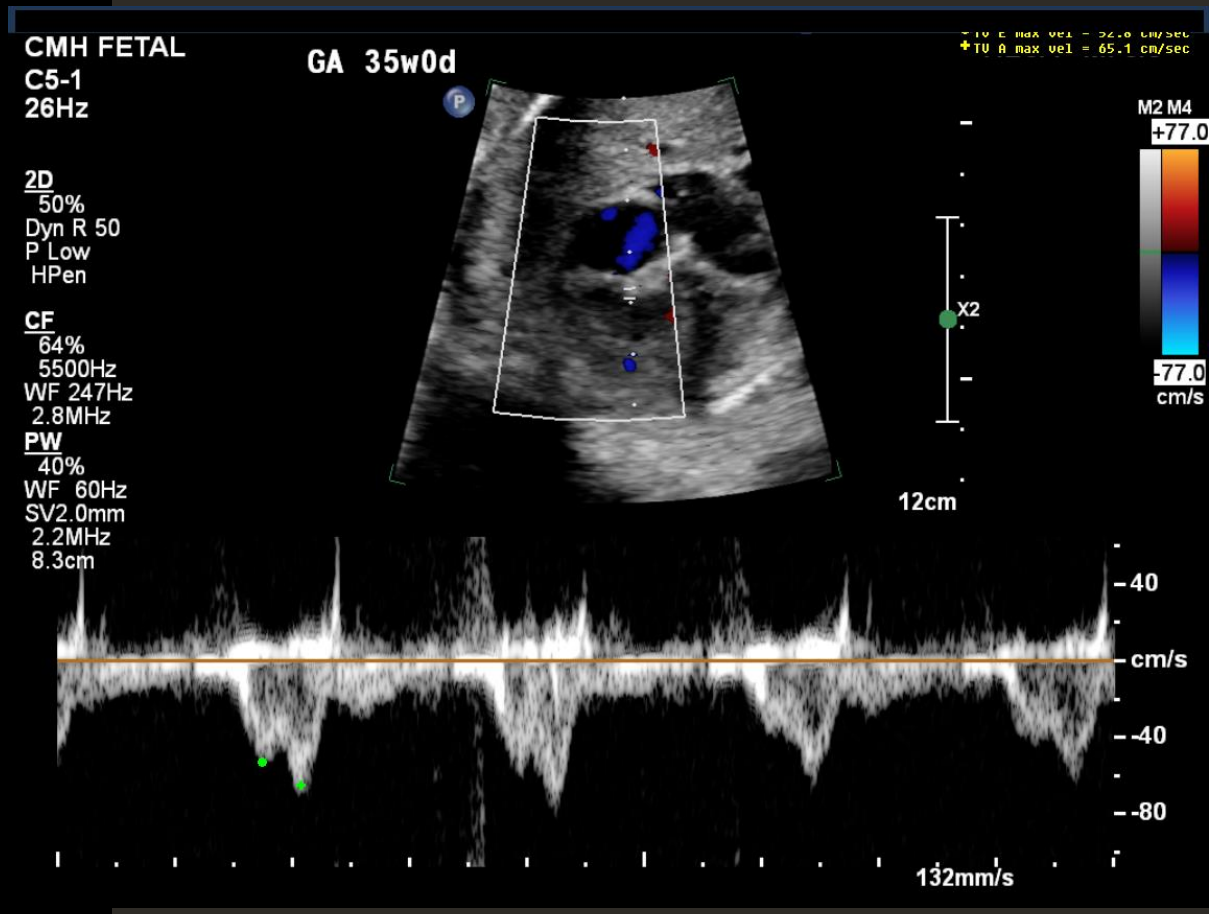
G
P
R
1.7 3.4
X2
12cm
*** bpm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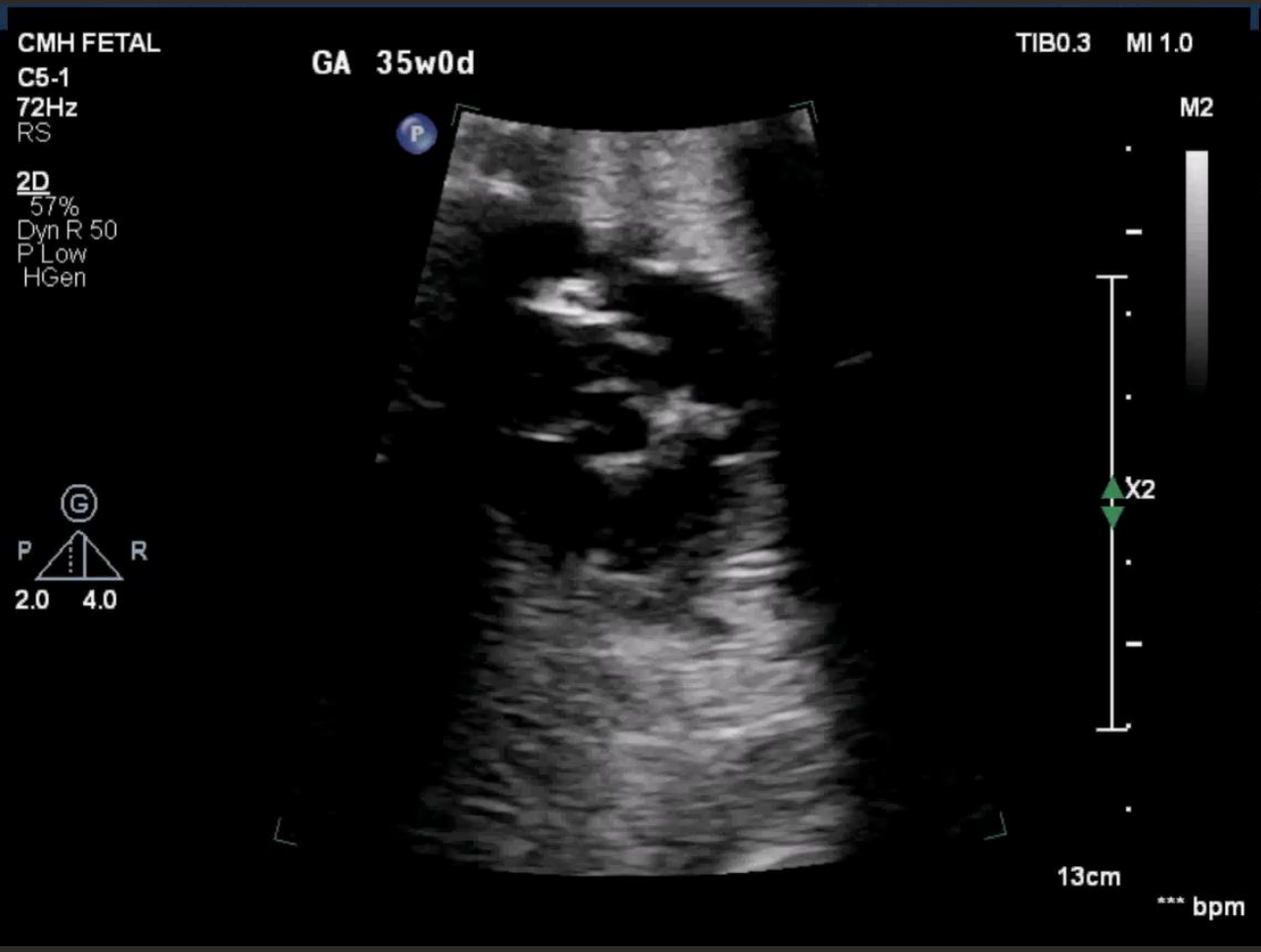
TV A max vel = 65.1 cm/sec
M2 M4
+77.0
-77.0
cm/s
2D
50%
Dyn R 50
P Low
HPen
CF
64%
5500Hz
WF 247Hz
2.8MHz
PW
40%
WF 60Hz
SV2.0mm
2.2MHz
8.3cm
X2
12cm
40
cm/s
-40
-80
132mm/s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Dyn R 50
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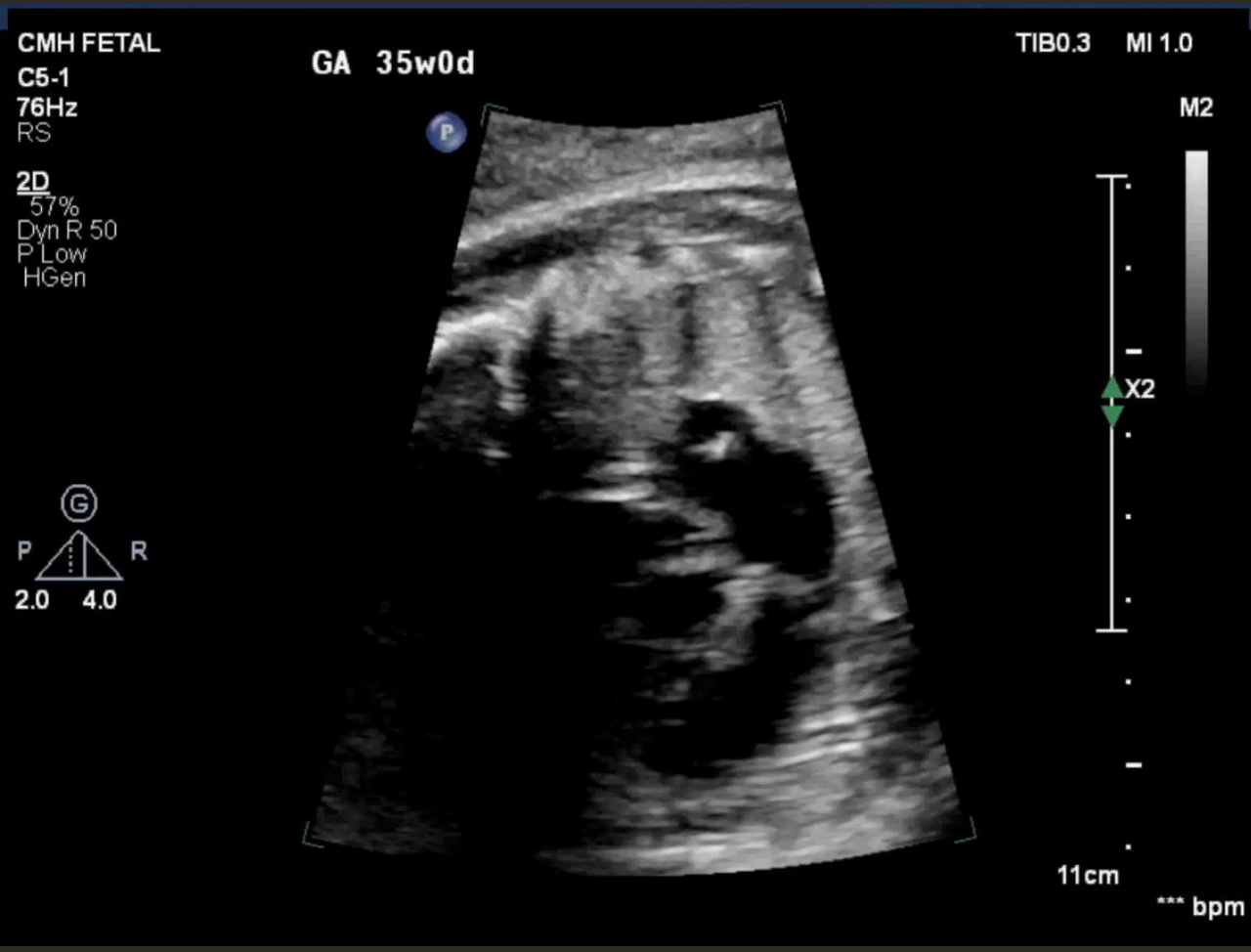
M2
X2
13cm
*** bpm
P
R
2.0 4.0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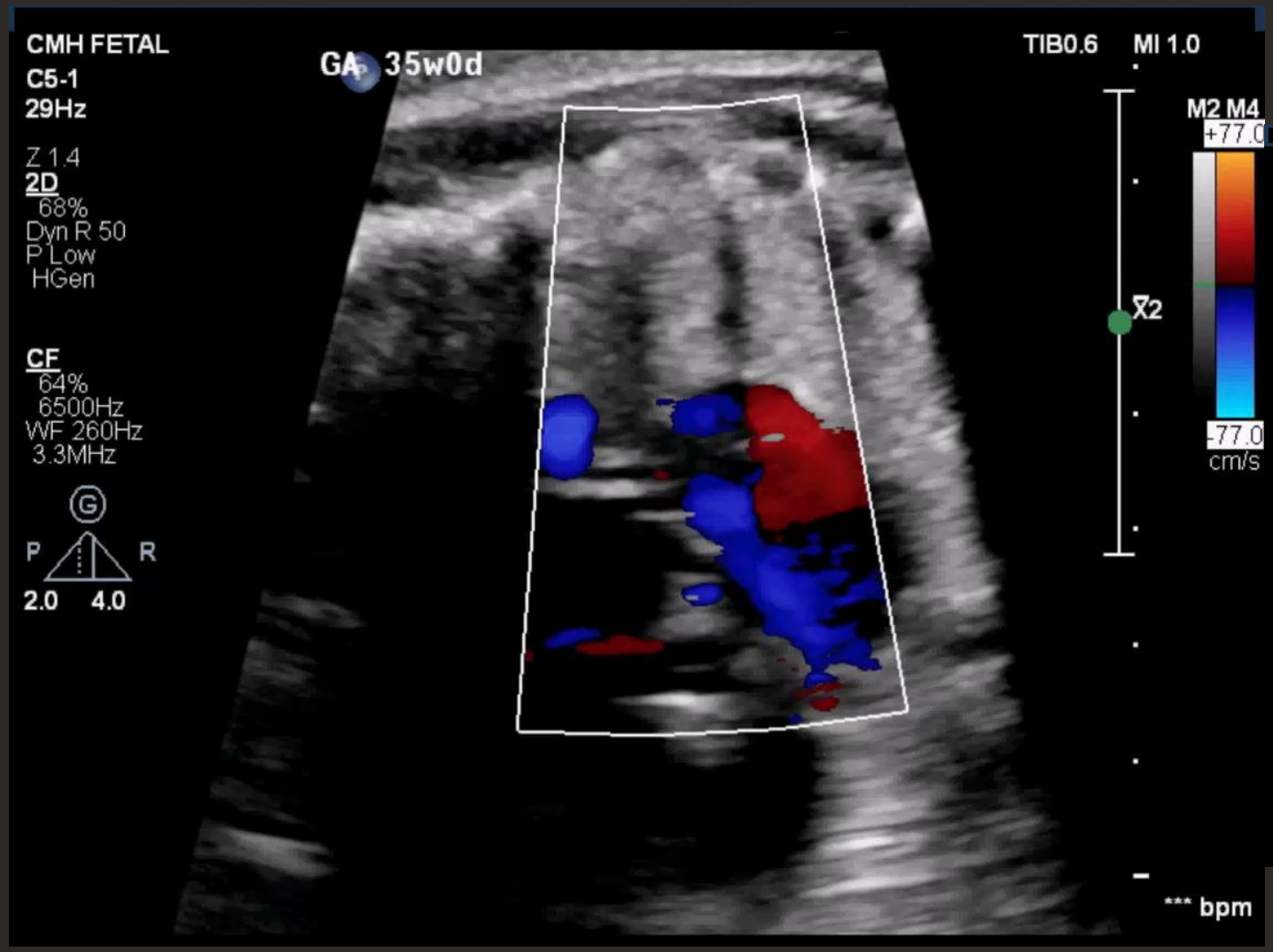
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WF 260Hz
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4.0
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TIB0.6
MI 1.0
M2 M4
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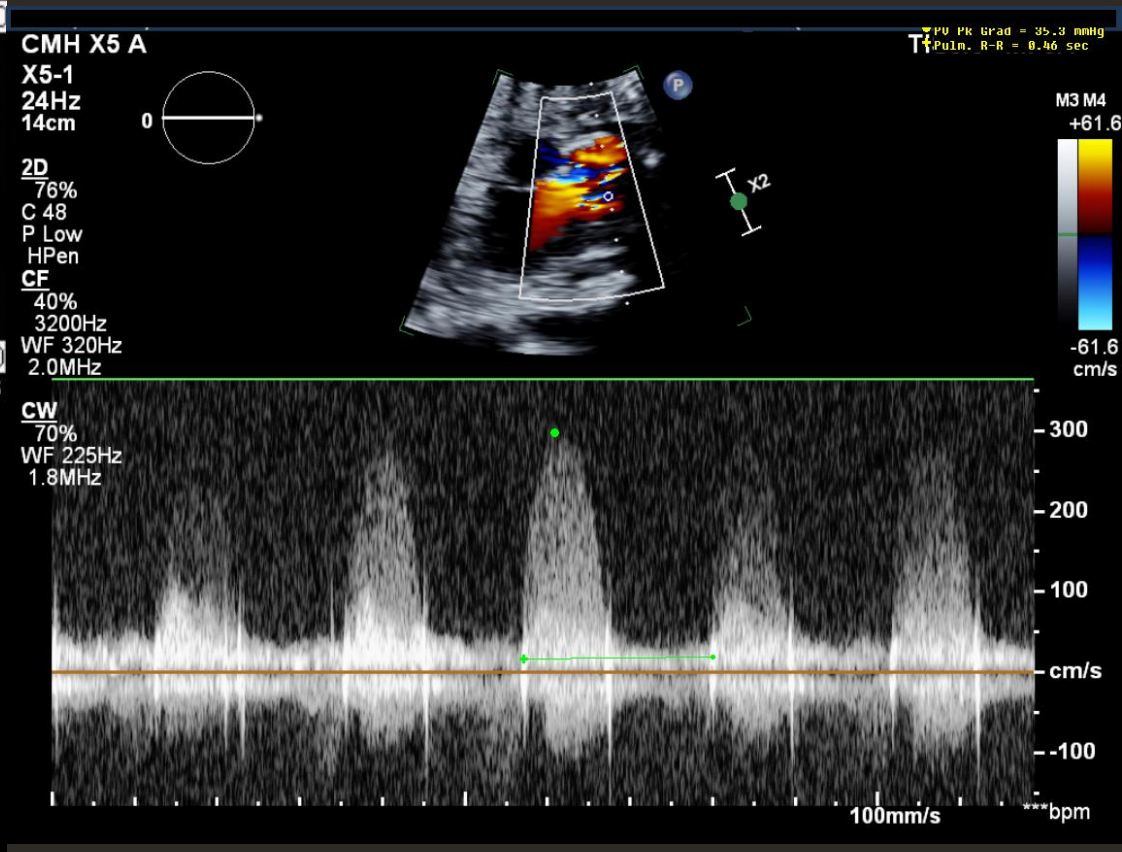
X2
-77.0
cm/s
*** bpm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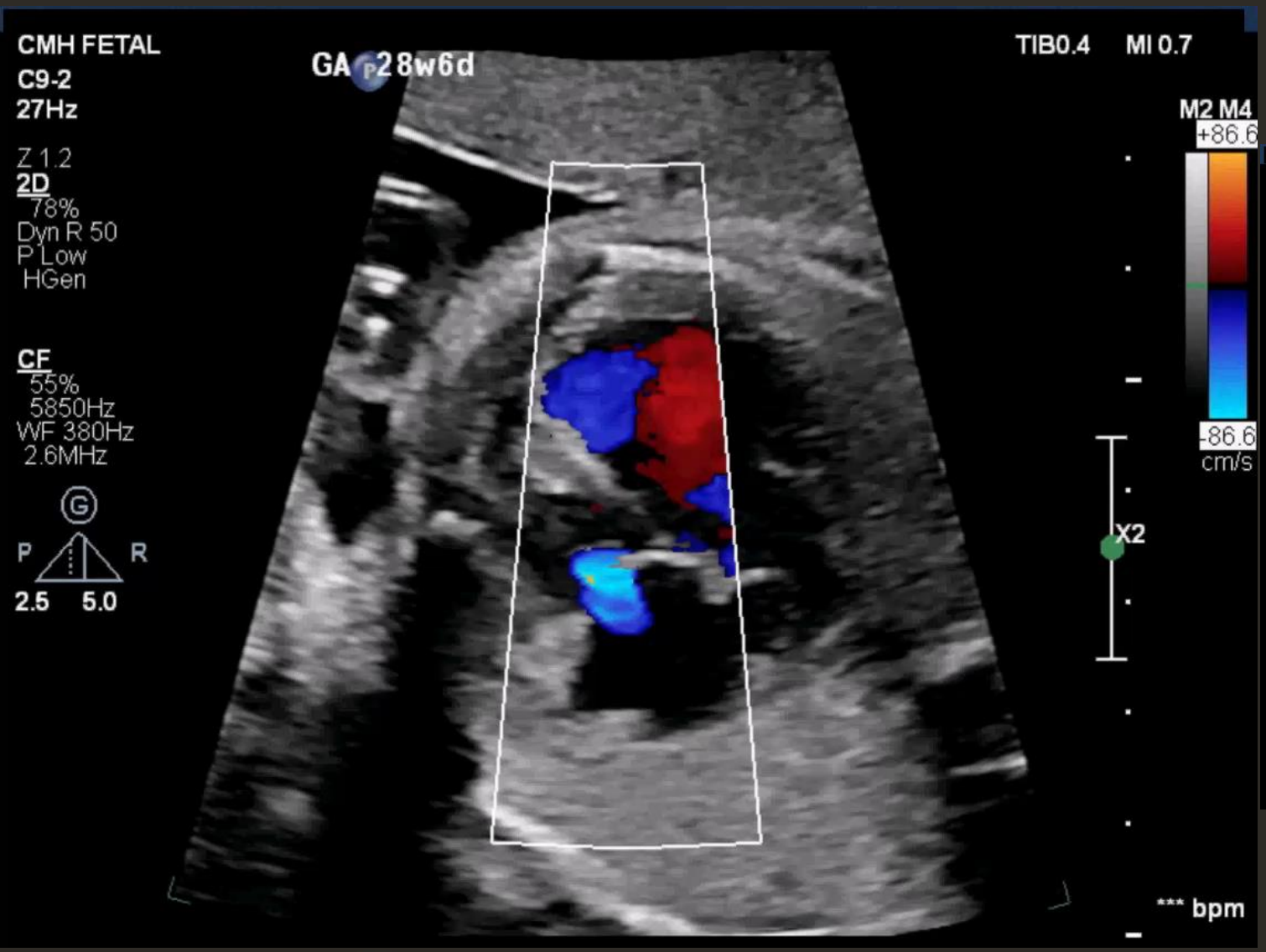
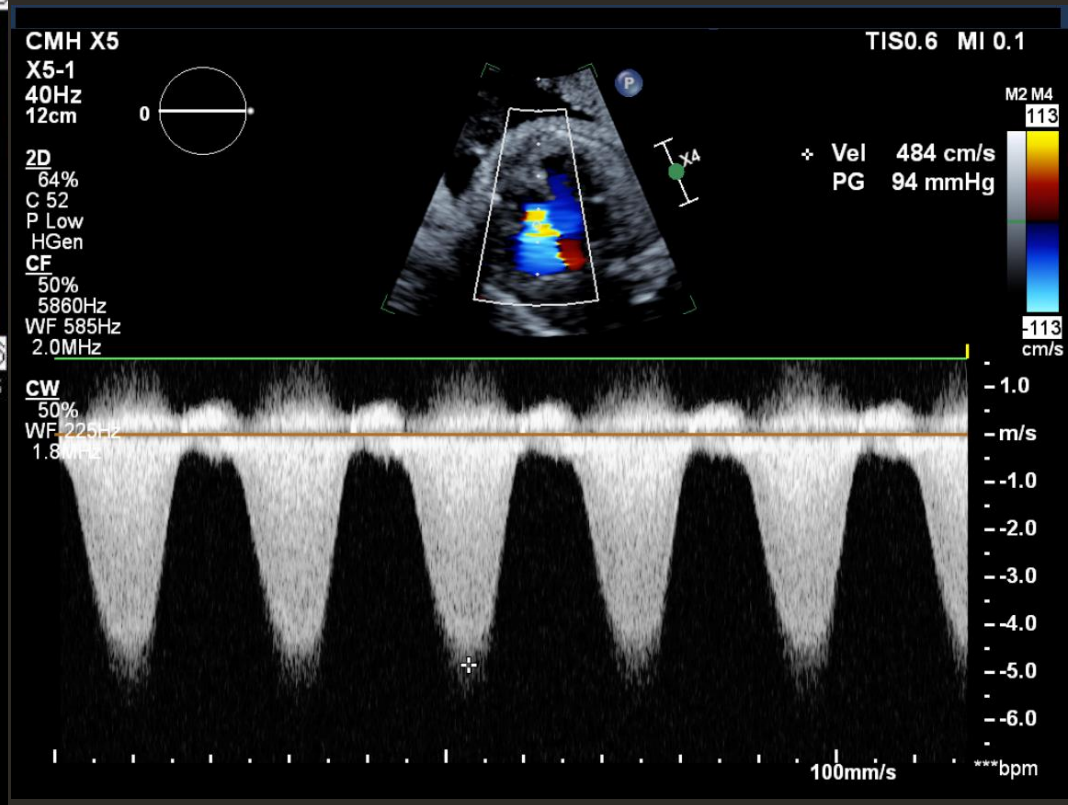
24Hz
14cm
0
2D
76%
C 48
P Low
HPen
CF
40%
3200Hz
WF 320Hz
2.0MHz
PV PK Grad = 35.3 mmHg
Pulm. R-R = 0.46 sec
M3 M4
+61.6
X2
-61.6
cm/s
CW
70%
WF 225Hz
1.8MHz
300
200
100
cm/s
-100
100mm/s
***bpm

PS Case 2





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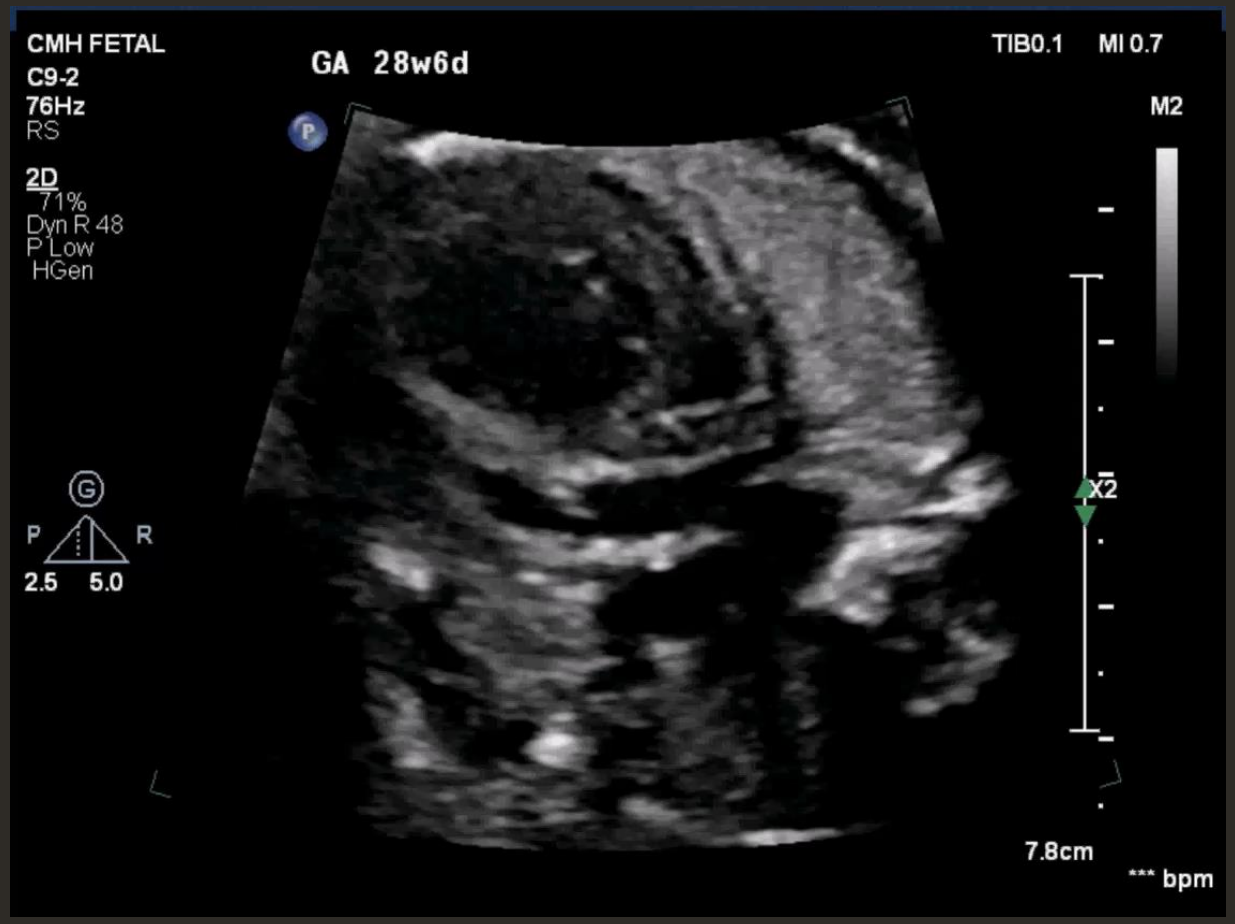
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76Hz
RS
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71%
Dyn R 48
P Low
HGen
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TIB0.1 MI 0.7
M2
X2
7.8cm
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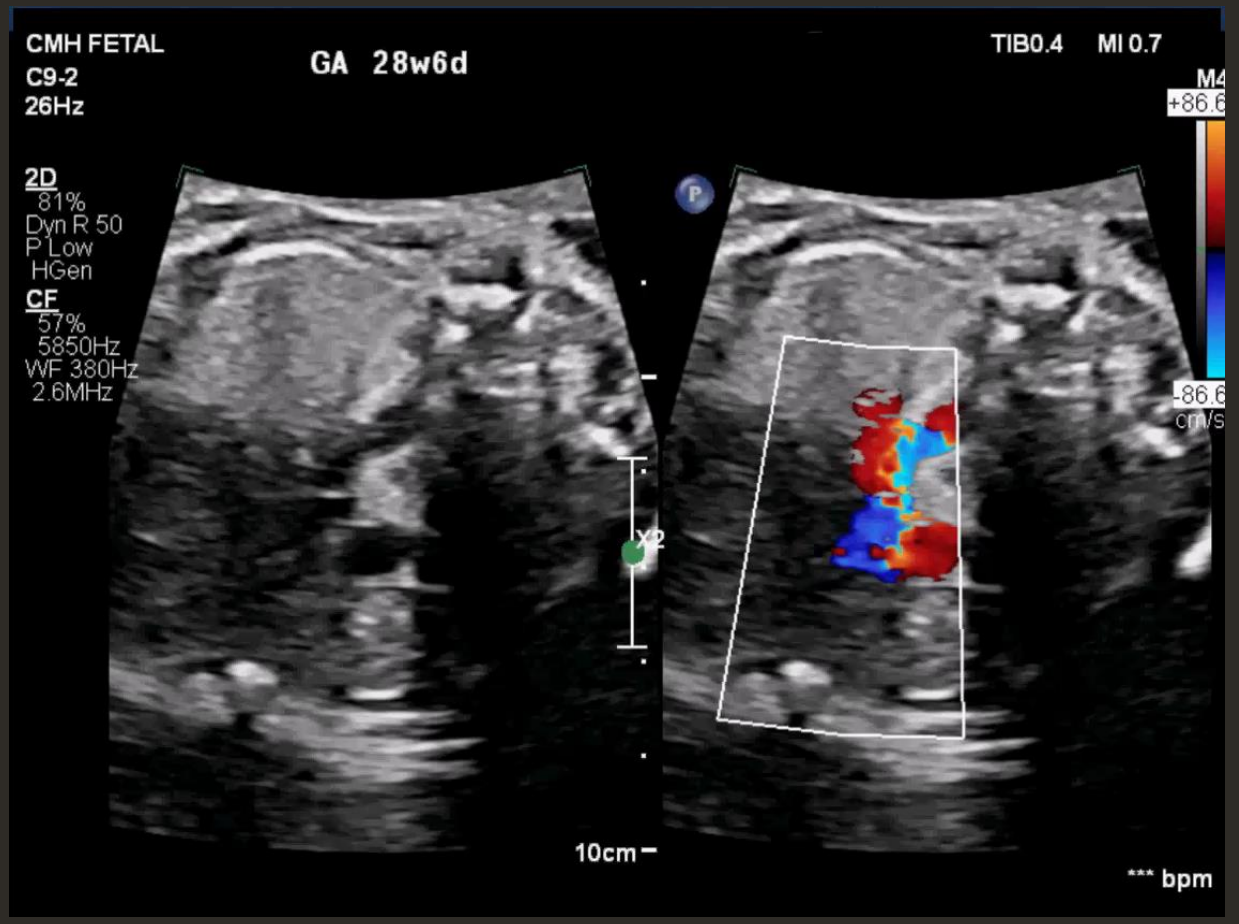
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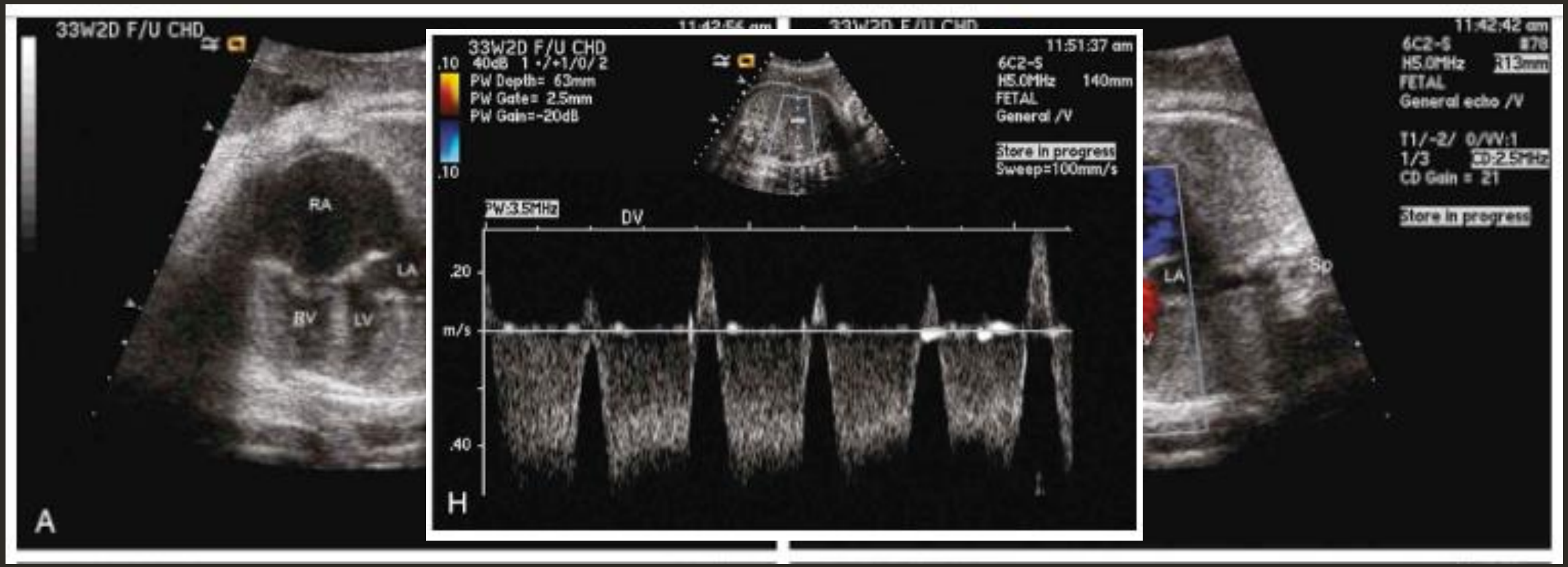
2D
81%
Dyn R 50
P Low
HGen
CF
57%
5850Hz
WF 380Hz
2.6MHz
GA 28w6d
TIB0.4 MI 0.7
M4
+86.6
-86.6
cm/s
X2
10cm
*** bpm

PS Cases

Rychik, Jack. Zhiyun, Tian. *Fetal Cardiovascular Imaging: A Disease-Based Approach.* Elsevier Saunders, 2012.



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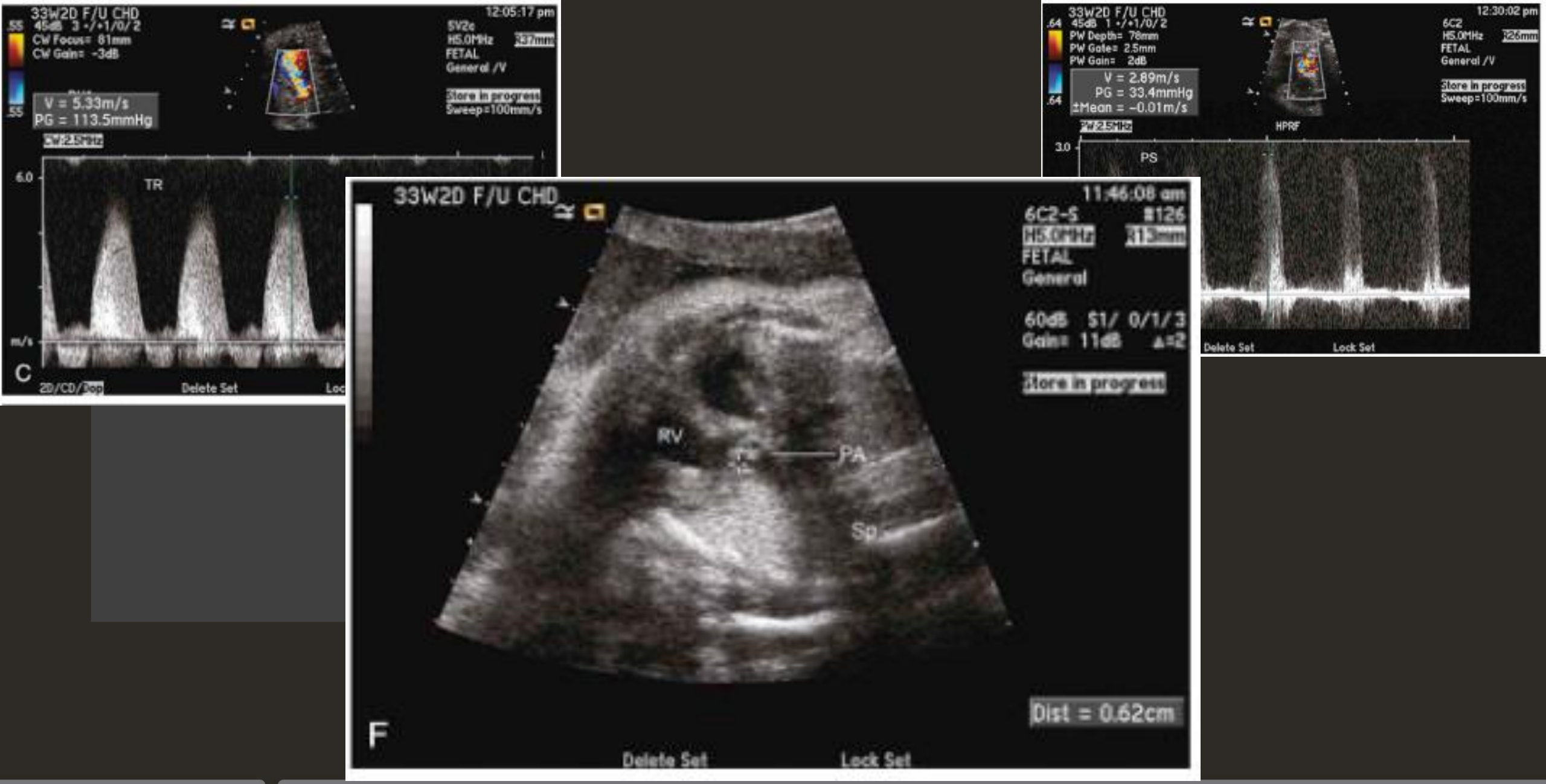

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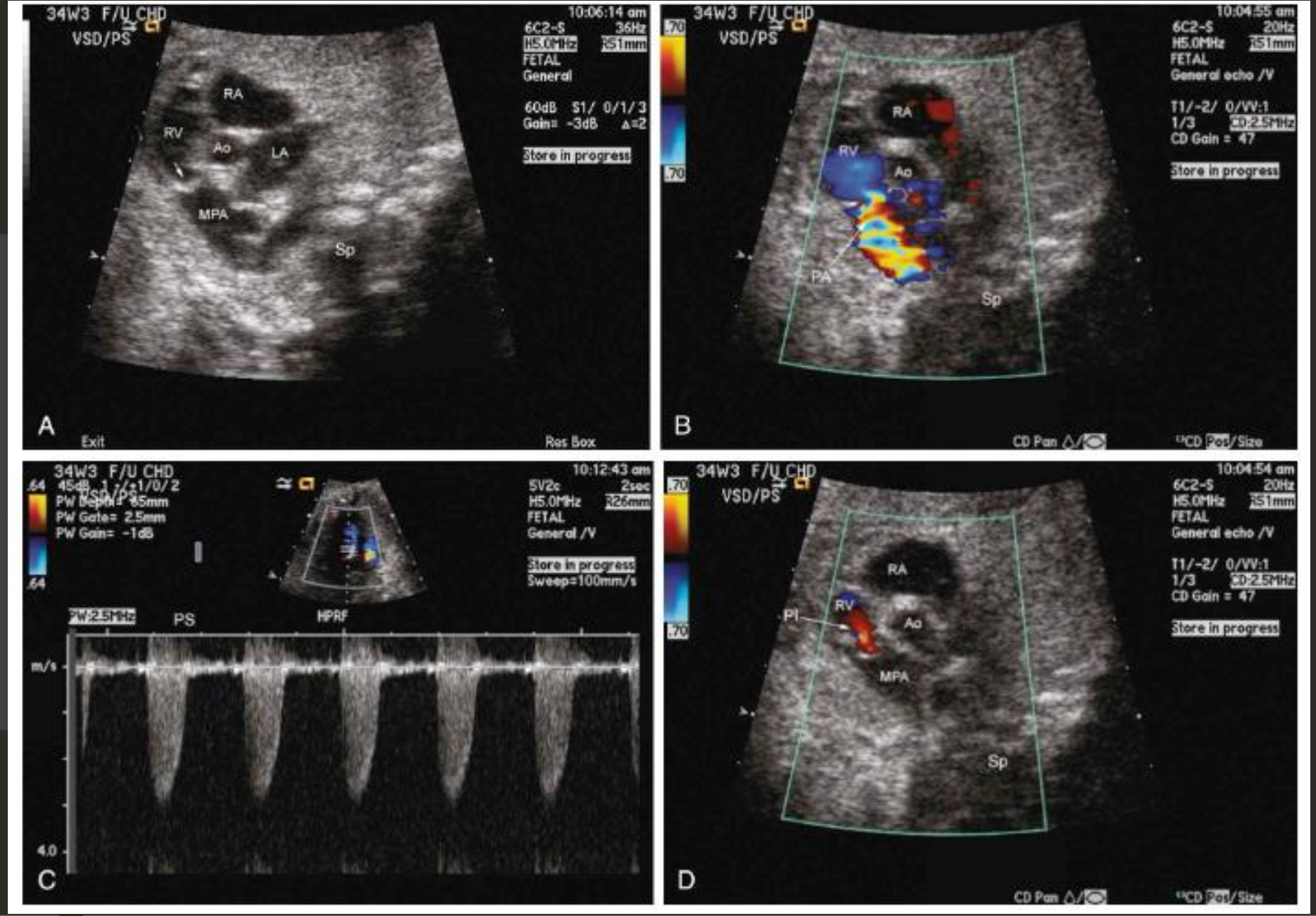

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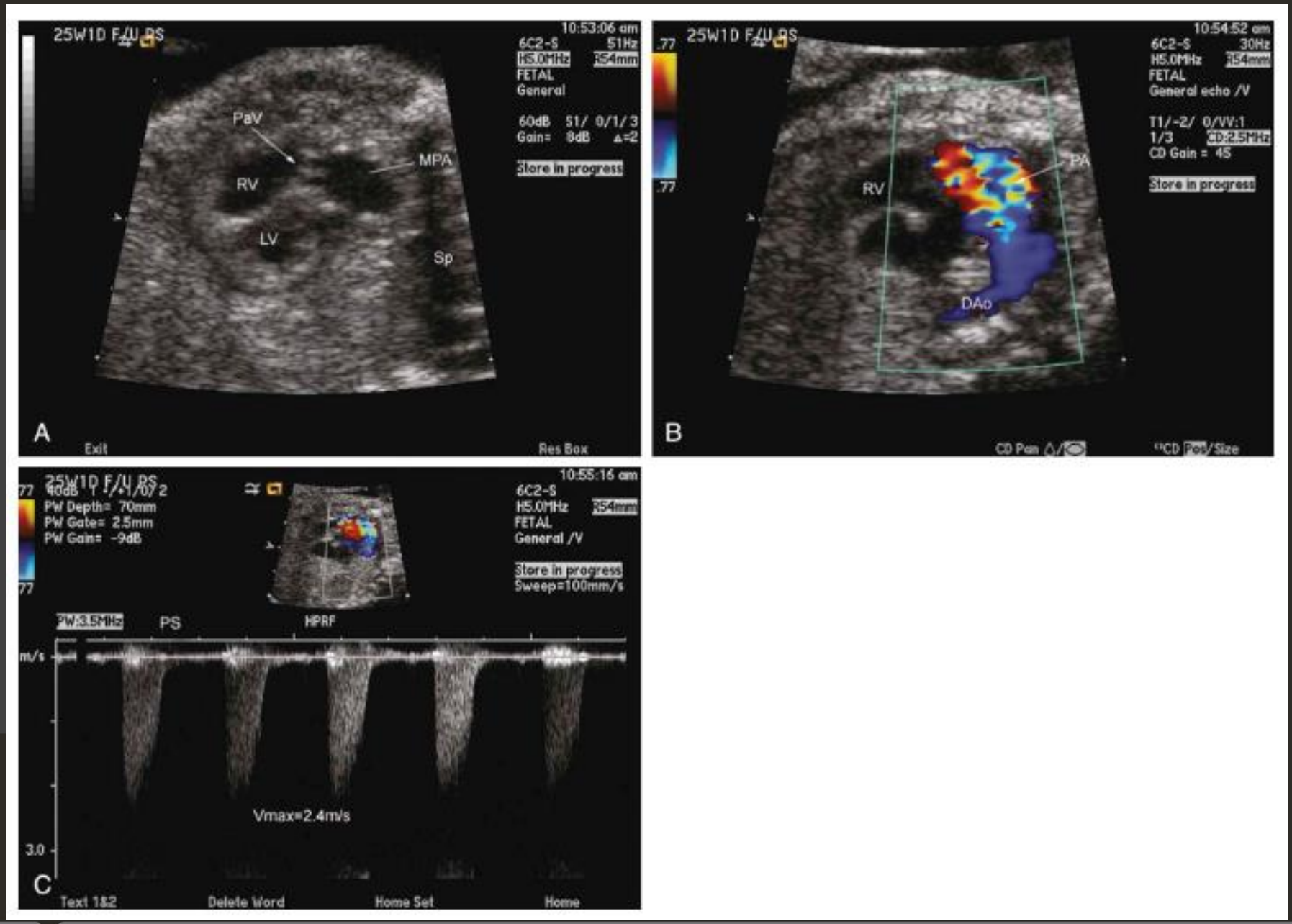

Rychik, Jack. Zhiyun, Tian. *Fetal Cardiovascular Imaging: A Disease-Based Approach.* Elsevier Saunders, 2012.

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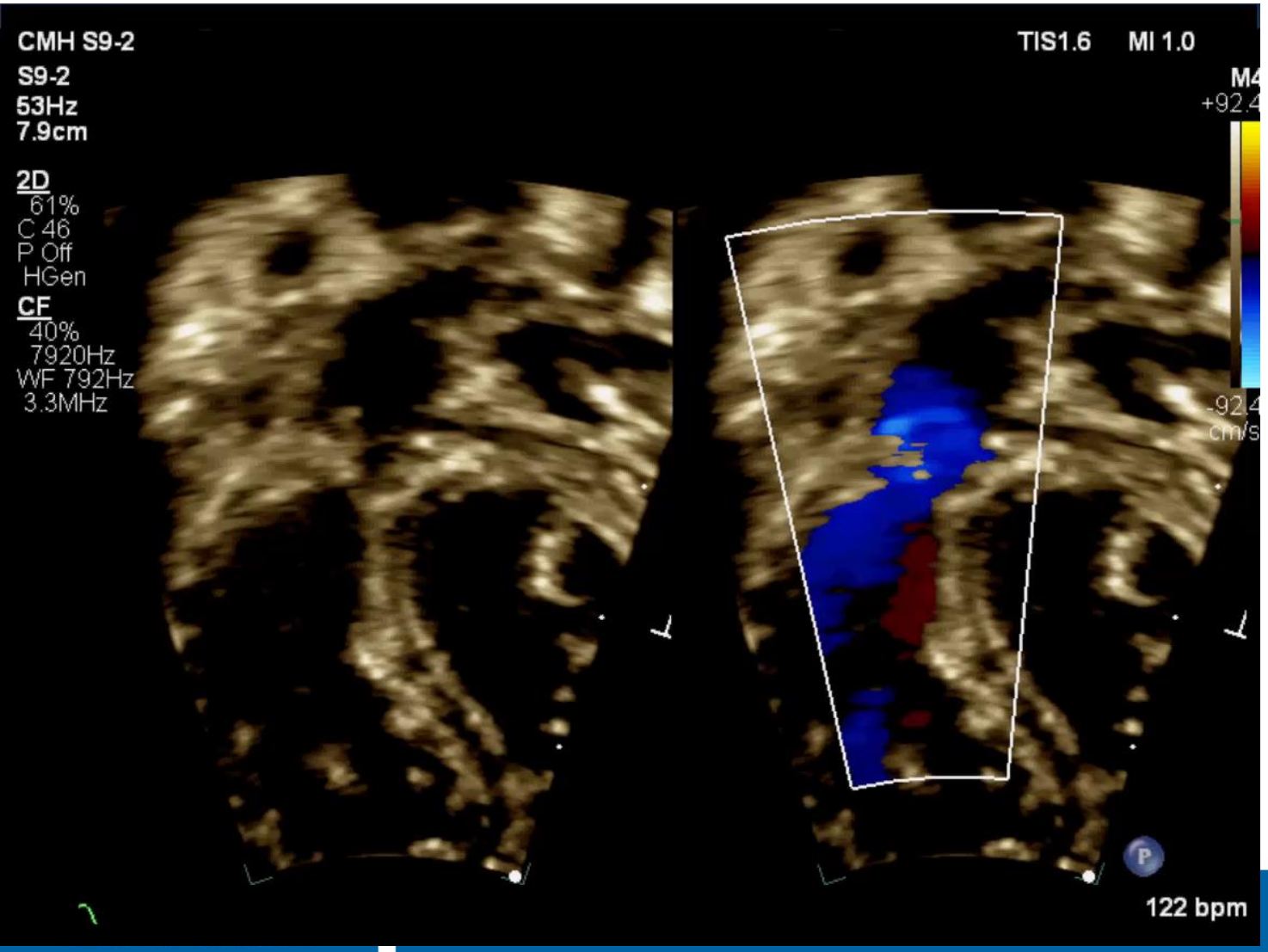
Delivery Planning

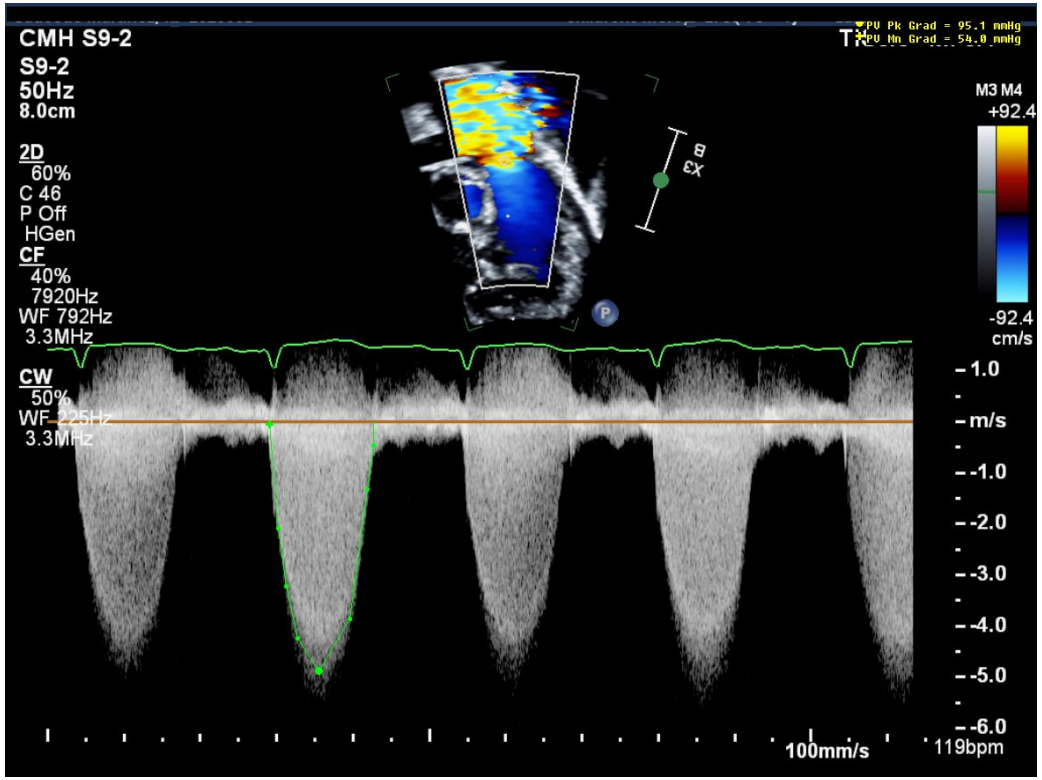
- Most fetuses tolerate a vaginal delivery
- C-section
  - Critical PS and impending hydrops
  - LV dysfunction
  - Restrictive PFO
- Prostaglandin recommendation
  - Inadequate antegrade PV flow
  - Reversal of flow in the ductus arteriosus

Postnatal Management

- Depends on the severity of the obstruction
- Mild to moderate PS: newborns discharged home with outpatient cardiology follow-up
- Severe and/or critical PS
  - Cyanotic newborns
  - Prostaglandin
  - Neonatal intervention: most often catheter intervention with pulmonary balloon valvuloplasty (subvalvar or hypoplastic annulus – surgical intervention)

Postnatal Echo





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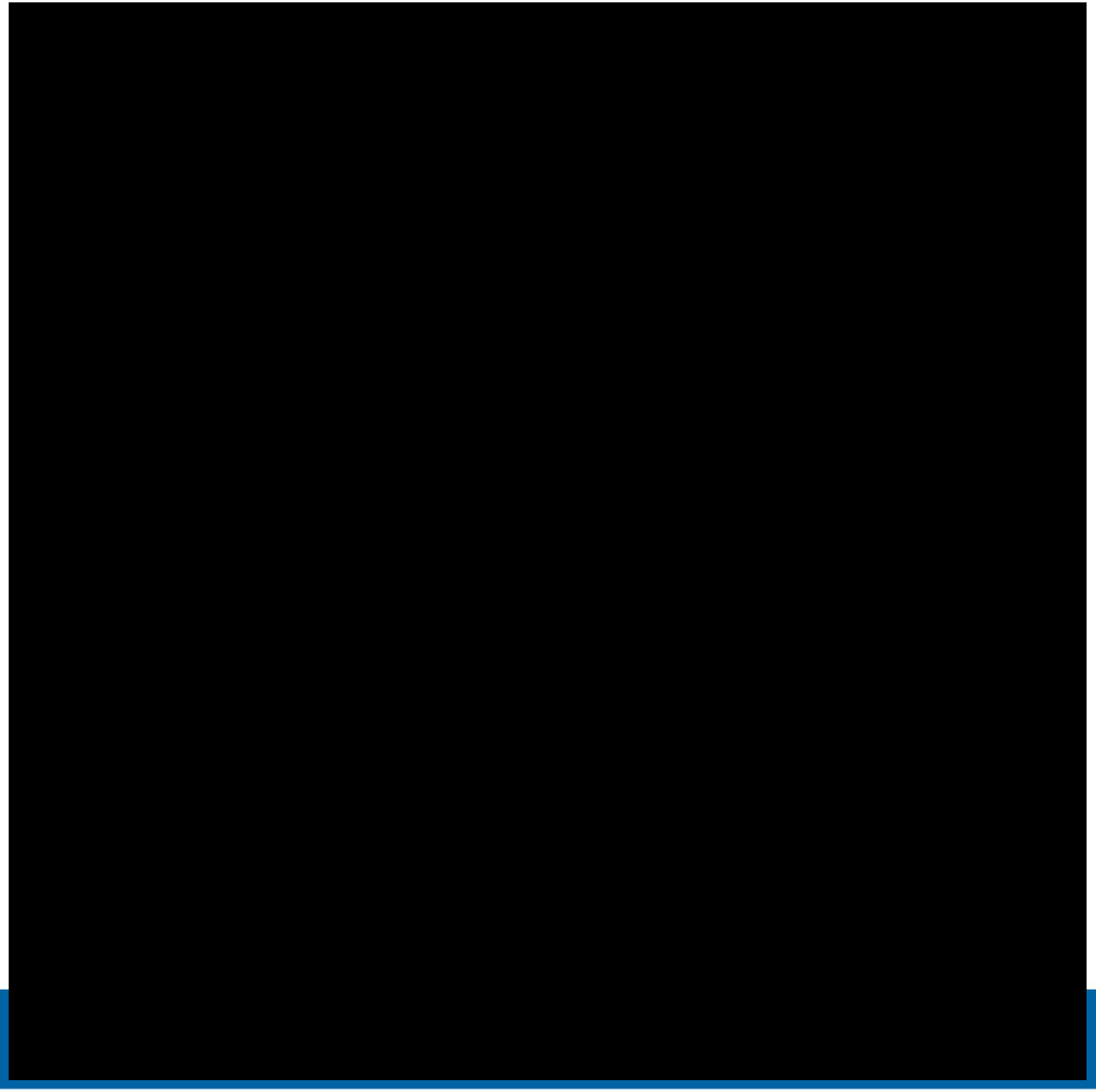
Pulmonary Balloon Valvuloplasty



PS Outcomes

- Effective balloon dilation in over 90% of patients
- Surgical intervention is uncommon
- Isolated PS: outcomes are excellent!
- Possible pulmonary valve replacement in the future
- Natural history studies have not included prenatal findings so predictions for a fetus with PS is challenging

ASE2022 HYBRID
June 10-13, 2022 • Seattle, WA

Prenatal Predictors for Pulmonary Balloon Valvuloplasty in the Newborn

Jennifer A. Johnson[1], Timothy Canavan[2], Tarek Alsaied[1], Lisa Howley[3], Hayley S. Hancock[4], Divya Suthar[5], Elisa Marcuccio[6], Taylor M. Trussell[6], Bettina Cuneo[7]

UPMC Children's Hospital of Pittsburgh, Pittsburgh, PA[1]; UPMC Magee Womens Hospital, Pittsburgh, PA[2]; The Children's Heart Clinic/Children's Minnesota, Minneapolis, MN[3]; Children's Mercy Kansas City, Kansas City, MO[4]; Children's Healthcare of Atlanta, Atlanta, GA[5]; Cincinnati Children's Hospital Medical Center, Cincinnati, OH[6]; Children's Hospital Colorado, Denver, CO[7]

METHOD

- A 6-center, retrospective case series of singleton fetuses identified between 2010-2020 with IPS
- Fetal and newborn echocardiogram characteristics were abstracted and correlated with the need for inpatient newborn PBV
- Pulmonary valve gradients, the presence of pulmonary insufficiency, right ventricular function and flow characteristics in the ductus arteriosus were collected
- Logistic regression, student t-test and Chi-Square testing with a p-value of ≤ 0.05 considered statistically significant

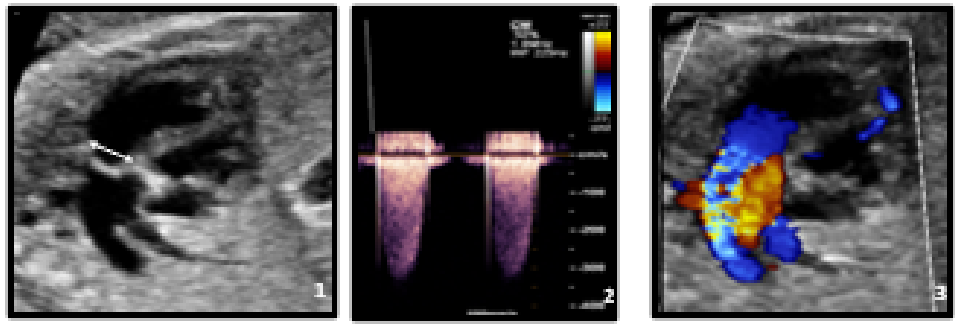


Figure 1: Right ventricular outflow pulmonary valve measurement. Figure 2: Pulmonary valve Doppler > 3 m/sec. Figure 3: Antegrade flow in ductus arteriosus

Conclusions:

- Newborns requiring inpatient pulmonary balloon valvuloplasty were more likely to have:
  - Left to right or bidirectional ductus arteriosus flow
  - 3rd trimester fetal echo peak PV velocity >3.0 m/sec
- PV velocity increases from 1st fetal echo to 3rd trimester fetal echo
- High 1st fetal echo PV velocity
- 3rd trimester fetal echo is good predictor of initial postnatal PV velocity

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PS Outcomes

PRENATAL DIAGNOSIS
Prenat Diagn 2011; **31**: 372–379.
Published online 20 January 2011 in Wiley Online Library
(wileyonlinelibrary.com) **DOI:** 10.1002/pd.2698

Pulmonary atresia/critical stenosis with intact ventricular septum: prediction of outcome in the second trimester of pregnancy

E. Gómez-Montes[1], I. Herraiz[1], A. Mendoza[2], L. Albert[2], J. M. Hernández-García[1] and A. Galindo[1]*

[1]*Fetal Medicine Unit, Department of Obstetrics and Gynaecology, Hospital Universitario "12 de Octubre", Madrid, Spain*
[2]*Department of Pediatrics, Pediatric Heart Institute, Hospital Universitario "12 de Octubre", Madrid, Spain*

Results Twelve fetuses had a BV outcome and four had non-BV repair. Cut-off values yielding the best results for a non-BV outcome were a tricuspid valve/mitral valve ratio ≤0.83, a pulmonary valve/aortic valve ratio ≤0.75, tricuspid inflow duration/cardiac cycle length ≤36.5%, and a right ventricle/left ventricle length ratio ≤0.64. If 3/4 markers are present, this predicts a non-BV outcome with sensitivity of 100% and specificity of 92%, and both are 100% if all the four criteria are fulfilled.

PS Outcomes

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Congenital Heart Disease

Morphologic and Functional Predictors of Eventual Circulation in the Fetus With Pulmonary Atresia or Critical Pulmonary Stenosis With Intact Septum

Helena M. Gardiner, PHD, MD, FRCP,*† Cristian Belmar, MD,* Gerald Tulzer, MD, PHD,‡
Anna Barlow, BS,*† Lucia Pasquini, MD,* Julene S. Carvalho, PHD, FRCPCH,†§
Piers E. F. Daubeney, MRCP,† Michael L. Rigby, FRCP,† Fabiana Gordon, PHD,‖
Elena Kulinskaya, PHD,‖ Rodney C. Franklin, FRCP†

London, United Kingdom; and Linz, Austria

PS Outcomes

Original Paper

Fetal Diagnosis and Therapy

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DOI: 10.1159/000357429

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Prenatal Tricuspid Valve Size as a Predictor of Postnatal Outcome in Patients with Severe Pulmonary Stenosis or Pulmonary Atresia with Intact Ventricular Septum

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Anita J. Moon-Grady[a, b]

Departments of [a]Pediatrics and [b]Fetal Treatment Center, University of California, San Francisco, Calif., USA

Thank you!

Texts

1. Allen, Hugh D., et al. *Moss and Adams' Heart Disease in Infants, Children, and Adolescents: Including the Fetus and Young Adult*. Wolters Kluwer Health, Lippincott Williams & Wilkins, 2013.
2. Park, Myung K., *Park's Pediatric Cardiology for Practitioners*. Mosby, 2014
3. Rychik, Jack. Zhiyun, Tian. *Fetal Cardiovascular Imaging: A Disease-Based Approach*. Elsevier Saunders, 2012.