



Prenatal diagnosis of DORV

Case presentation

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Utrecht Congenital Heart Sessions

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Key elements
when analysing
prenatal DORV

Anatomy

VSD position and its
relationship to the great
vessel

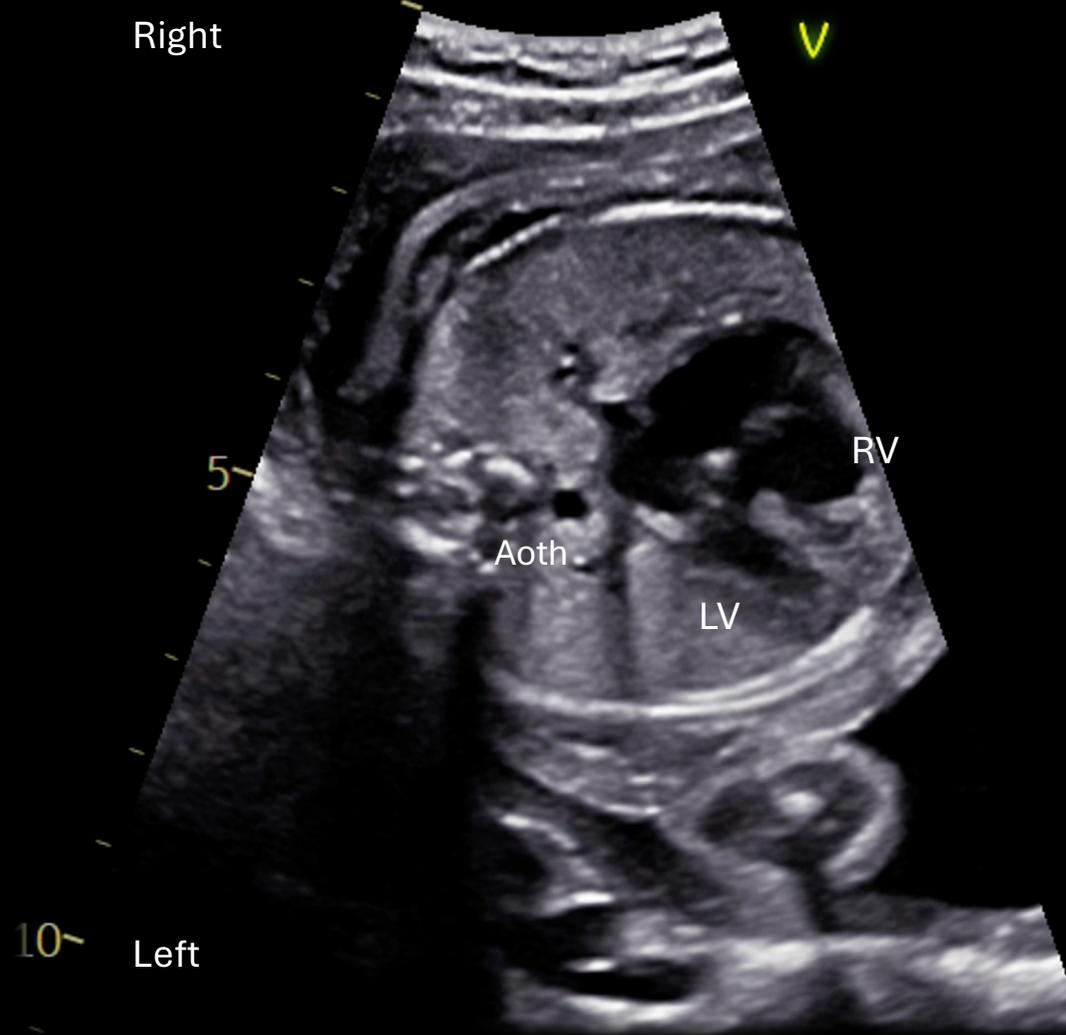
Right ventricular
outflow tract obstruction

Prediction of postnatal
surgical pathway

Clinical history

- 30 year old woman, 2nd pregnancy, no family history of CHD
- First trimester: NT 1.2 mm, normal serum markers
- **2nd trimester anatomical scan : suspicion of CHD**
 - EFW 426 g (30% Intergrowth21)
 - no extracardiac abnormalities
- Genetic testing normal (CGH array)

First echocardiography at 22 GA



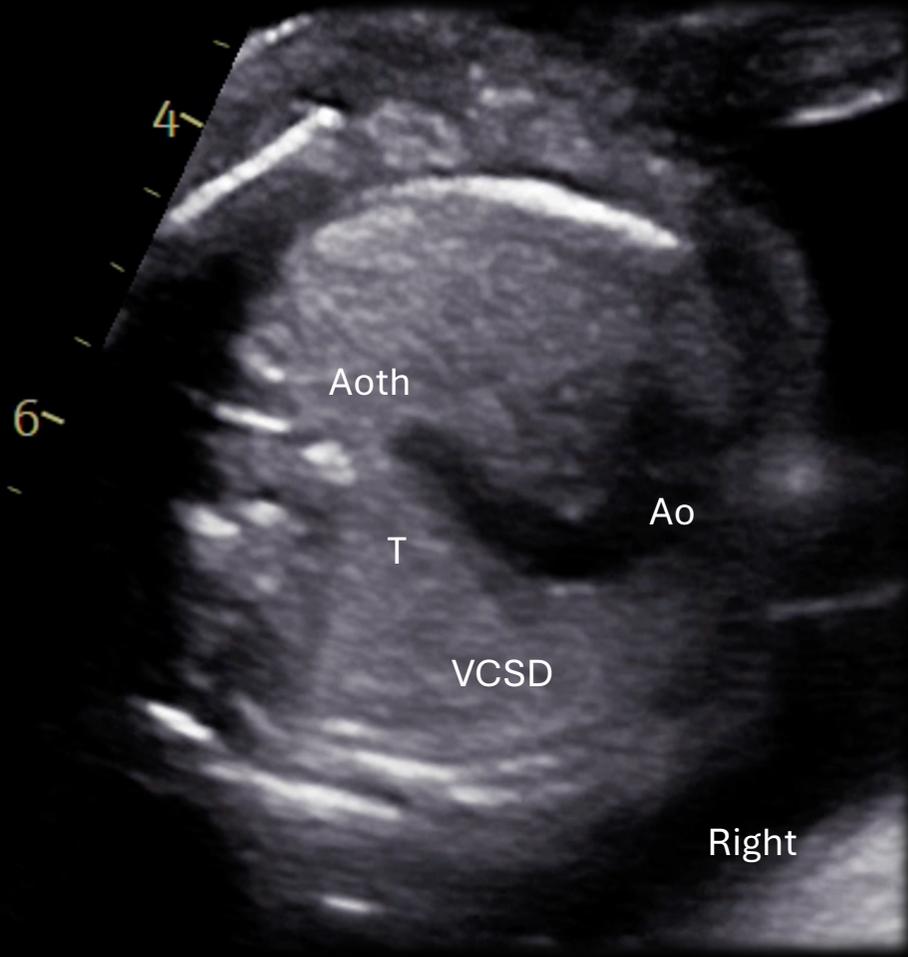
Aorta: 4.5 mm (Z Score 3.74*)

PA: 2.5 mm (Z Score -3.25*)

Pulmonary branches

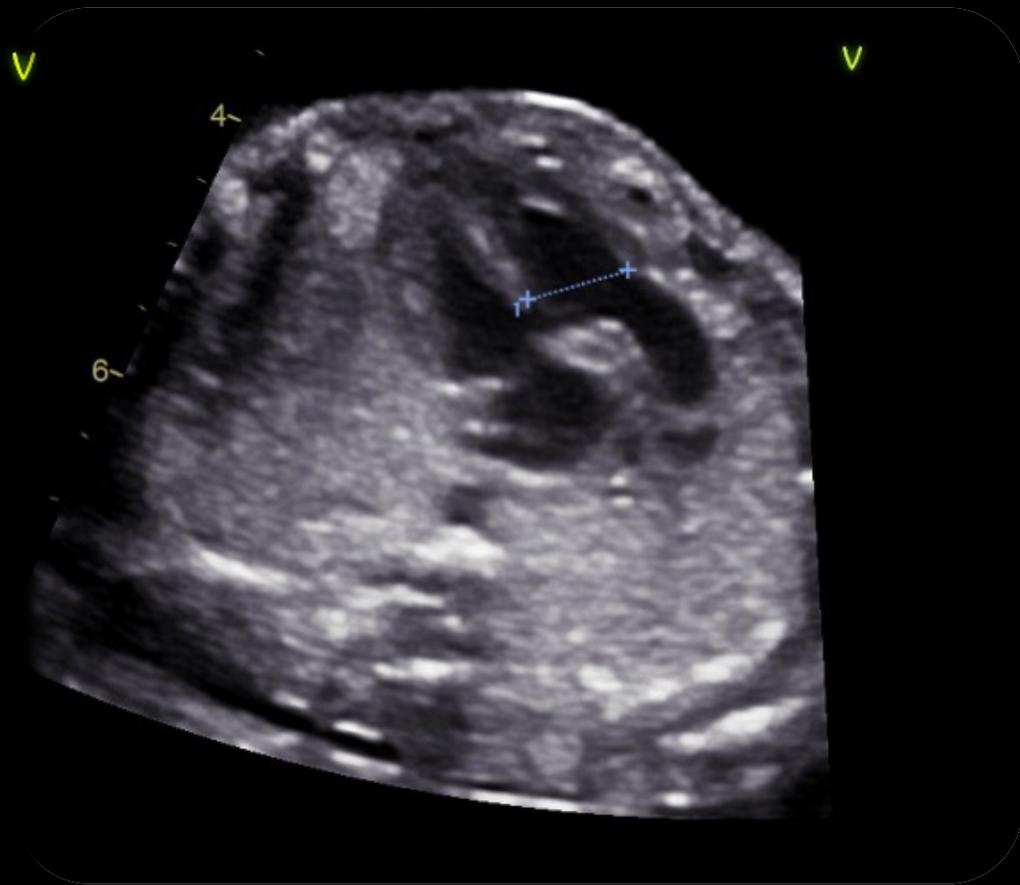
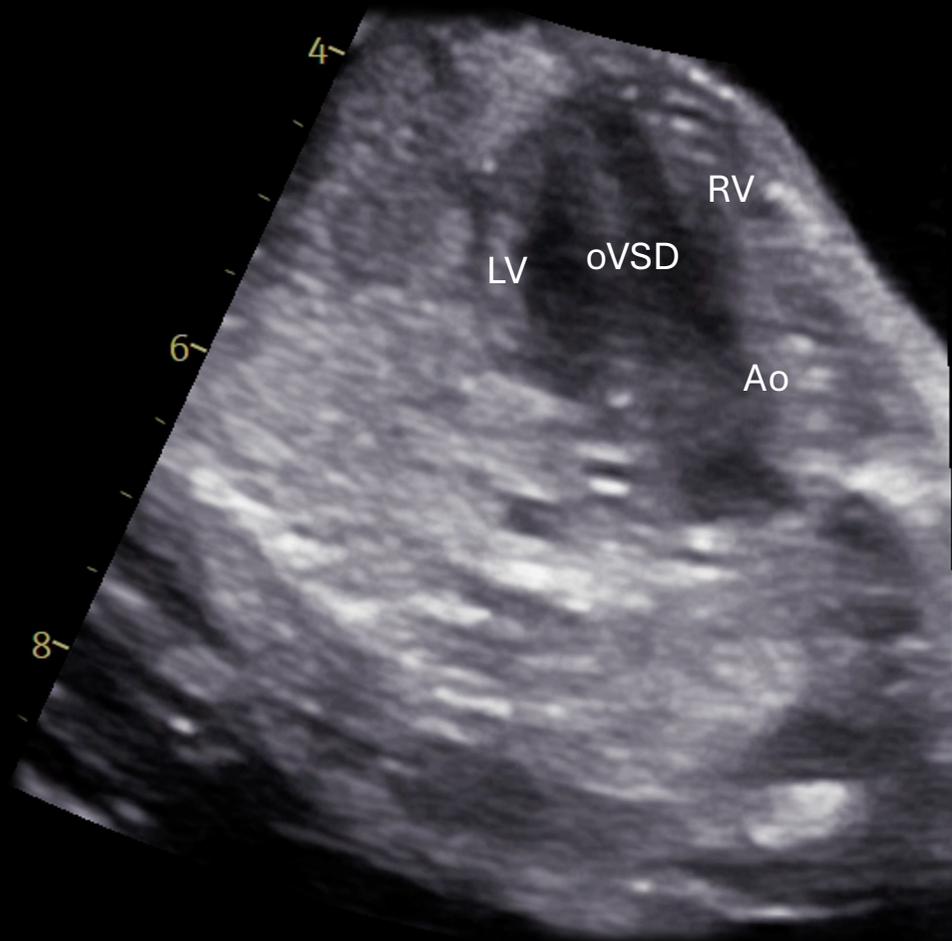
RPA: 2 mm

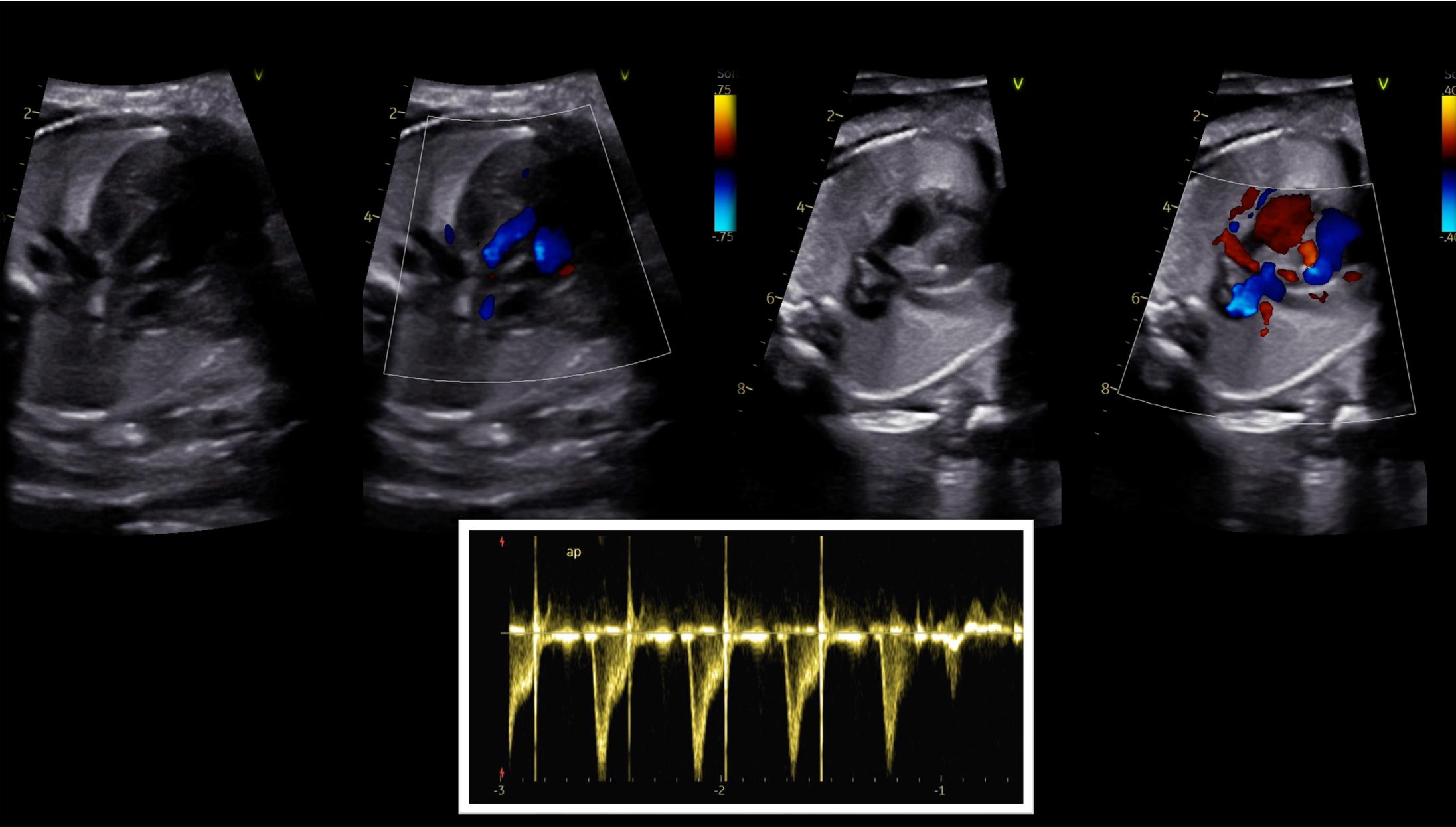
LPA: 1.7 mm



Three vessel view

*Krishnan JASE 2016





Follow-up echocardiography at 28 GA



JLB
C9-2
79Hz
RS

2D
61%
R Dyn 52
P Moy
HRés

TIB1.3 MI 1.3

M5

Left

Right



L

7.7cm
x3
*** bpm



L

JLB
C9-2
19Hz

TIB0.3 MI 1.0

2D
58%
R Dyn 50
P Bas
HRés
Coul
63%
2600Hz
FP 169Hz
2.6MHz

M2
+38.5
-38.5
cm/s

Left



X3

8.4cm

Right

*** bpm

L

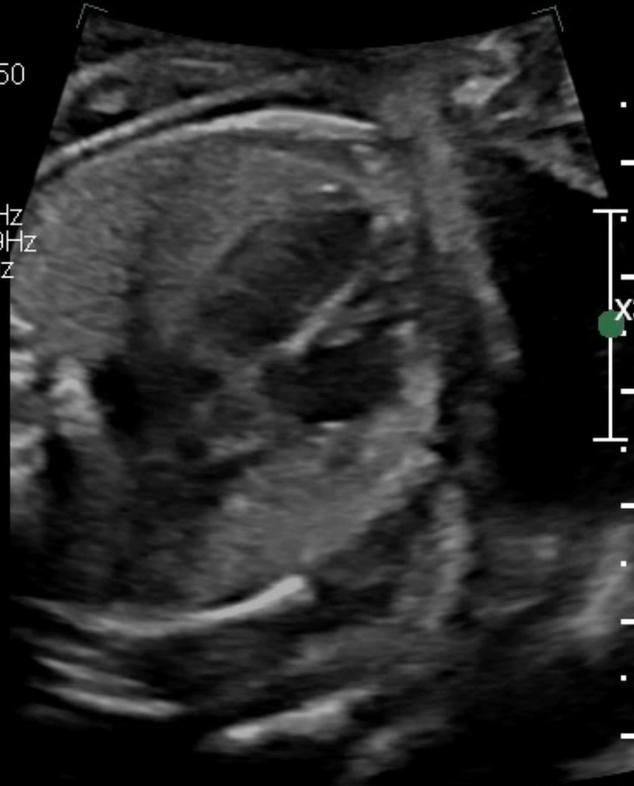
JLB
C9-2
15Hz

TIB0.3 MI 1.0

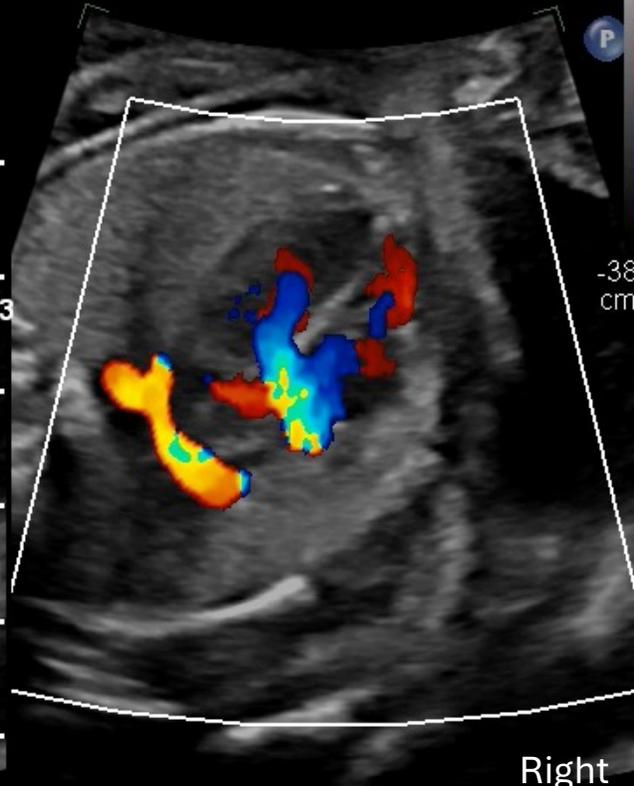
2D
58%
R Dyn 50
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63%
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M2
+38.5
Left
P
-38.5
cm/s

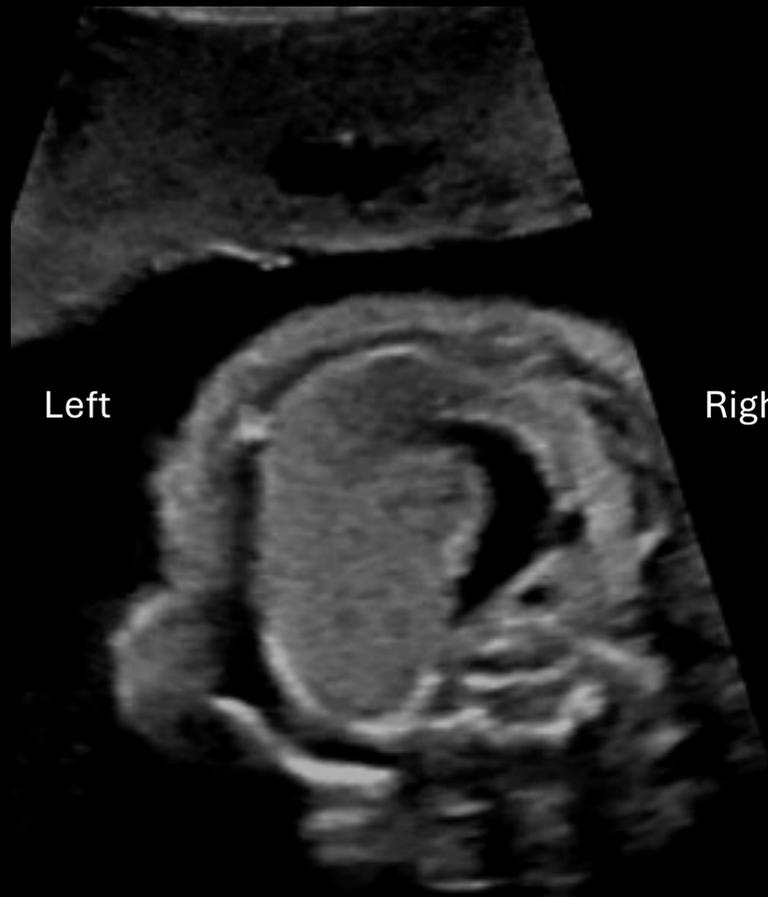


8.6cm



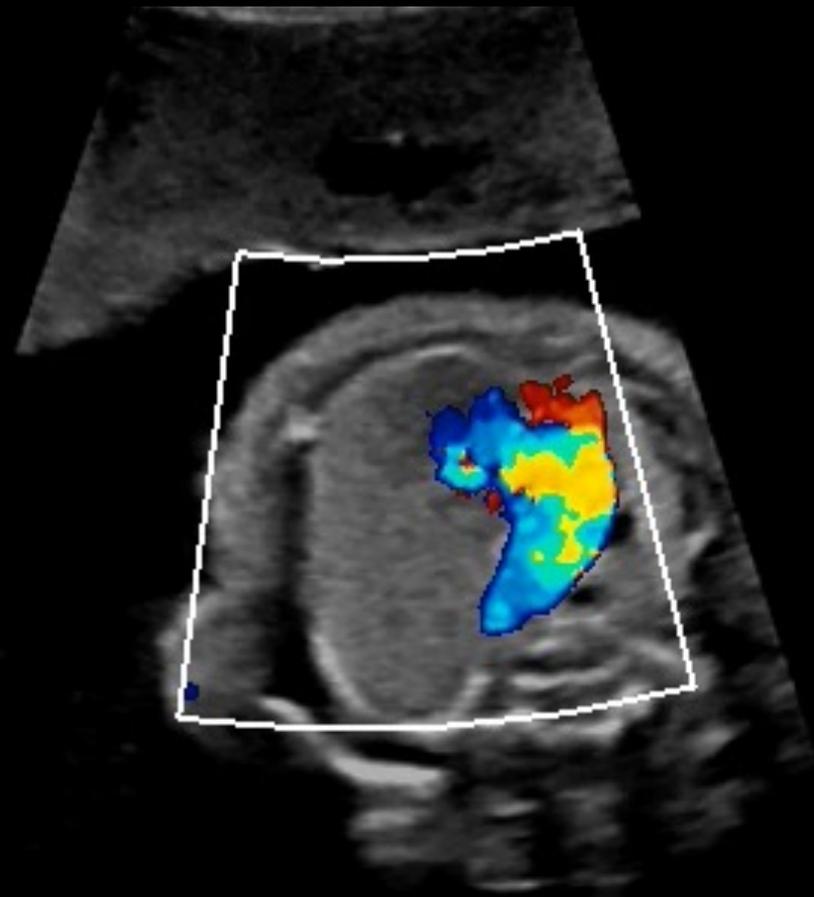
Right

*** bpm



Right

X3

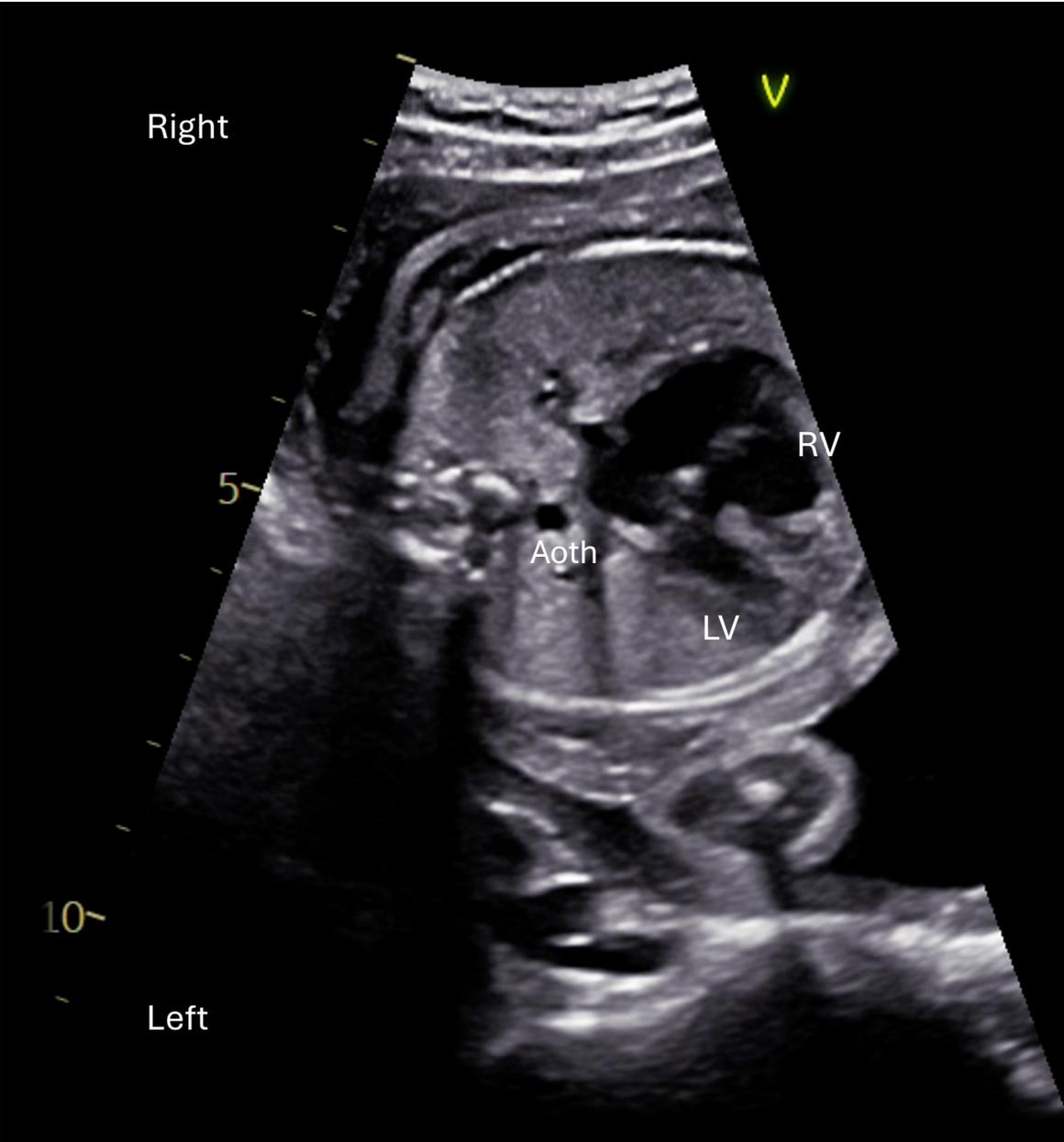


The VSD is ...

- in subaortic position.
- in subpulmonary position.
- doubly committed.
- remote or non committed.
- I don't know yet.

The
great
vessels
are

- in a normal spatial relationship.
- malposed with right anterior aorta and left posterior PA.
- malposed side-by-side.
- I don't know yet.



Anticipated neonatal tolerance ?

- 1 The baby will be desaturated < 90% (TGA physiology).
2. The baby will have normal oxygen saturation (pink TOF physiology).
3. It is too early to make a prediction on neonatal tolerance
4. Prostaglandine will be obligatory in the neonatal period.

When counseling
the parents, the
probable
postnatal surgical
approach will
be....

- 1. Neonatal switch procedure with VSD closure and LVOT relief
- 2. No neonatal intervention in absence of cyanosis but REV or Rastelli procedure > 8 kg
- 3. No neonatal intervention in absence of cyanosis but IVR > 6 kg with RVOT relief
- 4. Prediction of postnatal pathway is not possible at this time of pregnancy.

Prenatal diagnosis

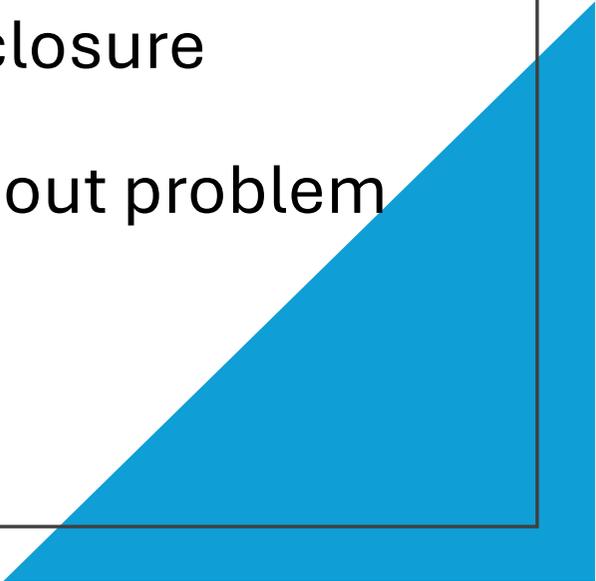
- Double outlet right ventricle
- Levocardia, normal heart axis, normal heart size
- Single outlet VSD, subaortic position
- Malposed great vessels
- Large anterior right aorta, smaller posterior left pulmonary artery
- Normal cardiac function
- No valve regurgitation
- No retrograde flow in the DA



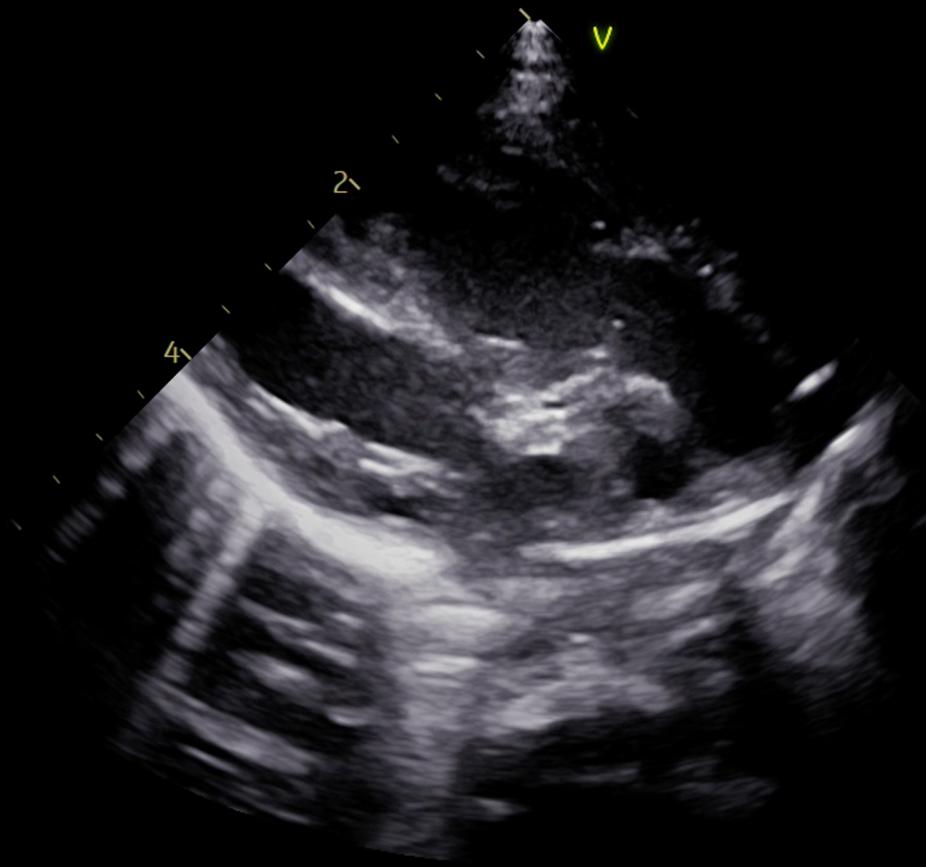
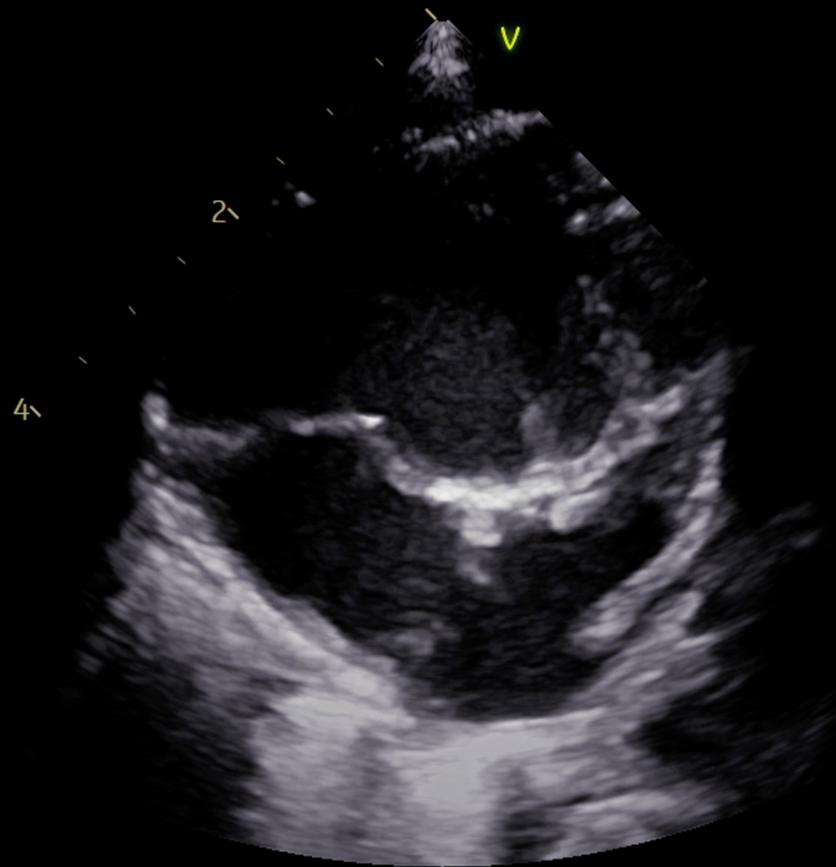
Postnatal history

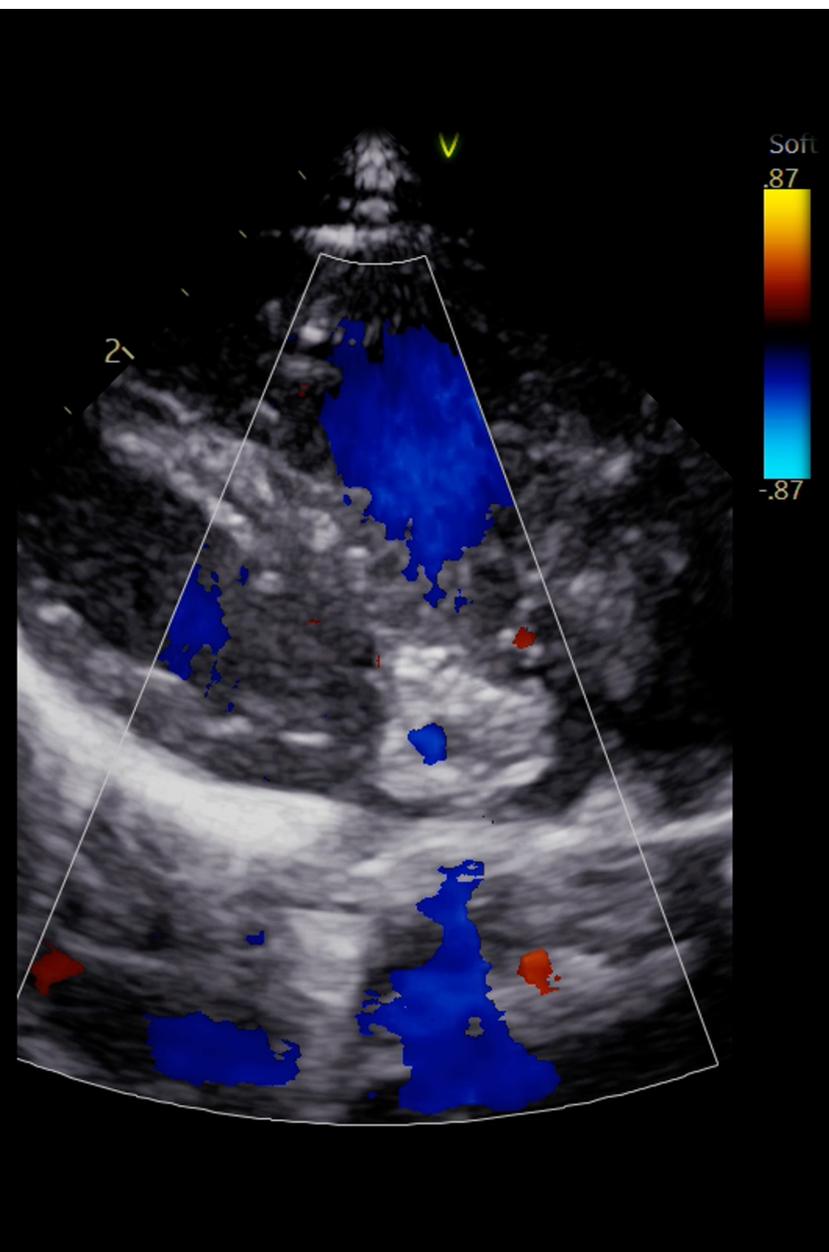
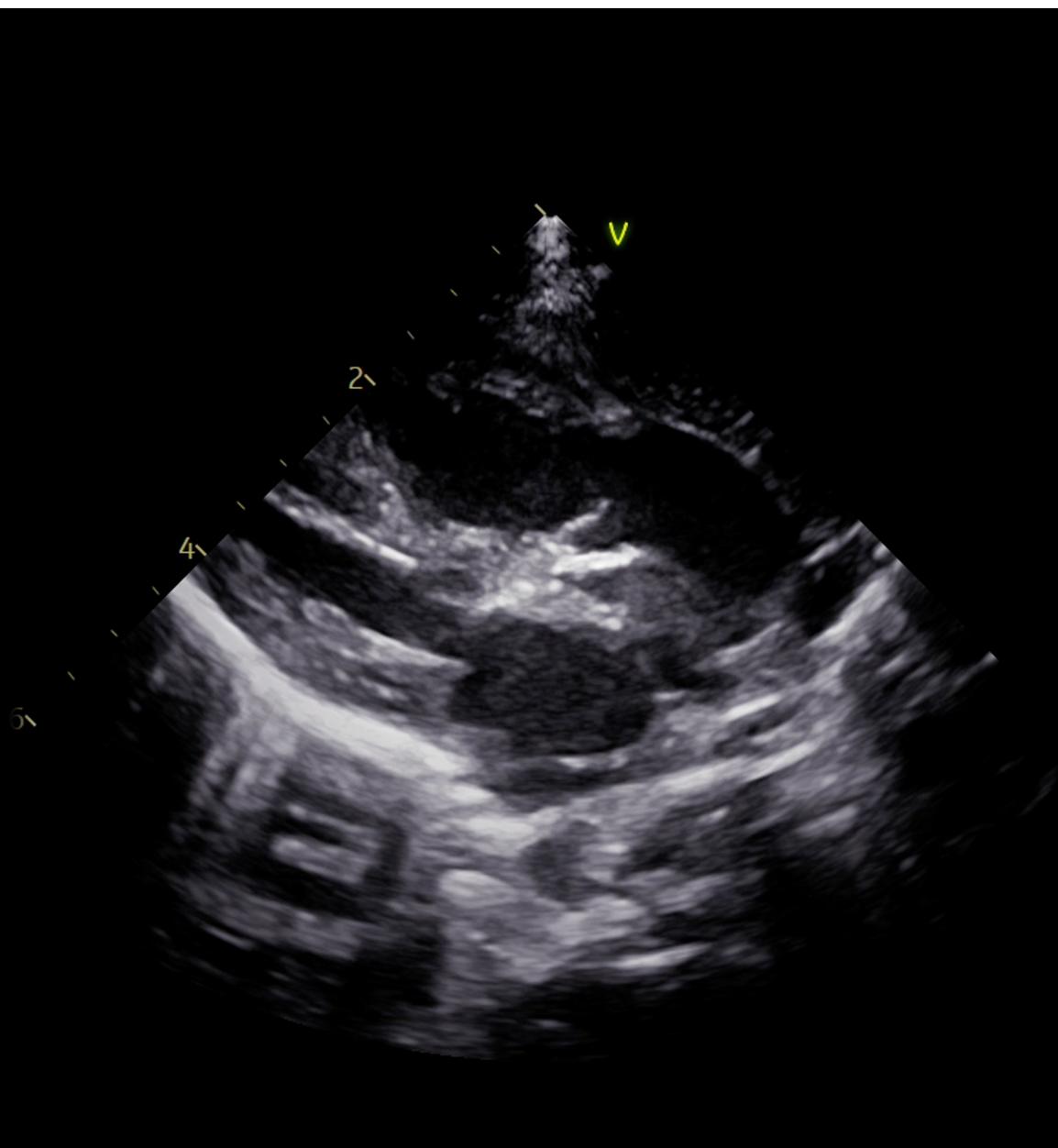
36+6 GA, birthweight 2.4 kg

Postnatal clinical history

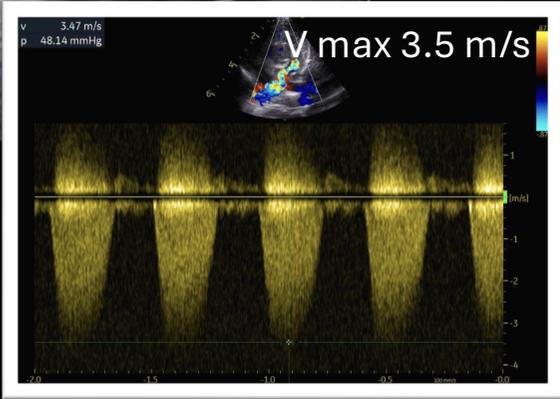
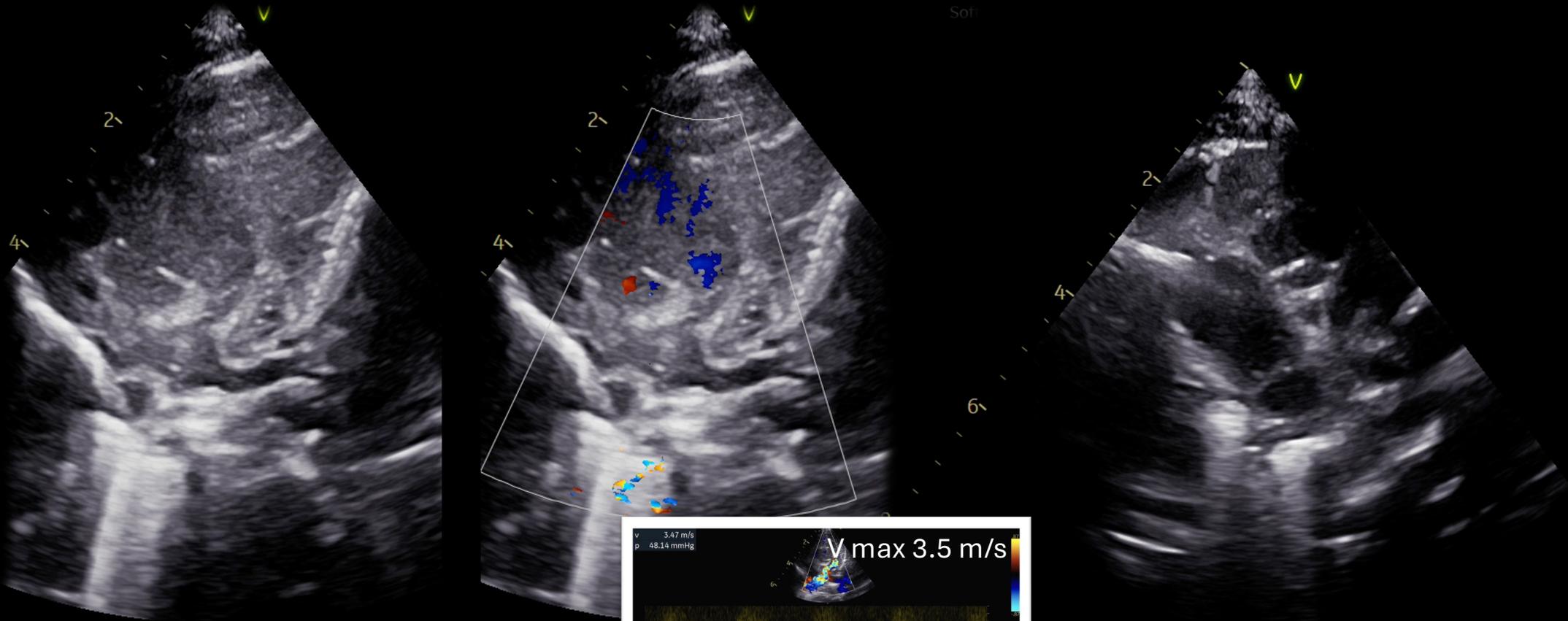
- Spontaneous near term labor with good postnatal adaptation
 - Normal oxygen saturation (97%) after DA closure
 - No feeding difficulties, gaining weight without problem
 - No spells, no desaturation
- 

Postnatal echocardiography





Pulmonary annulus: 5.5 mm



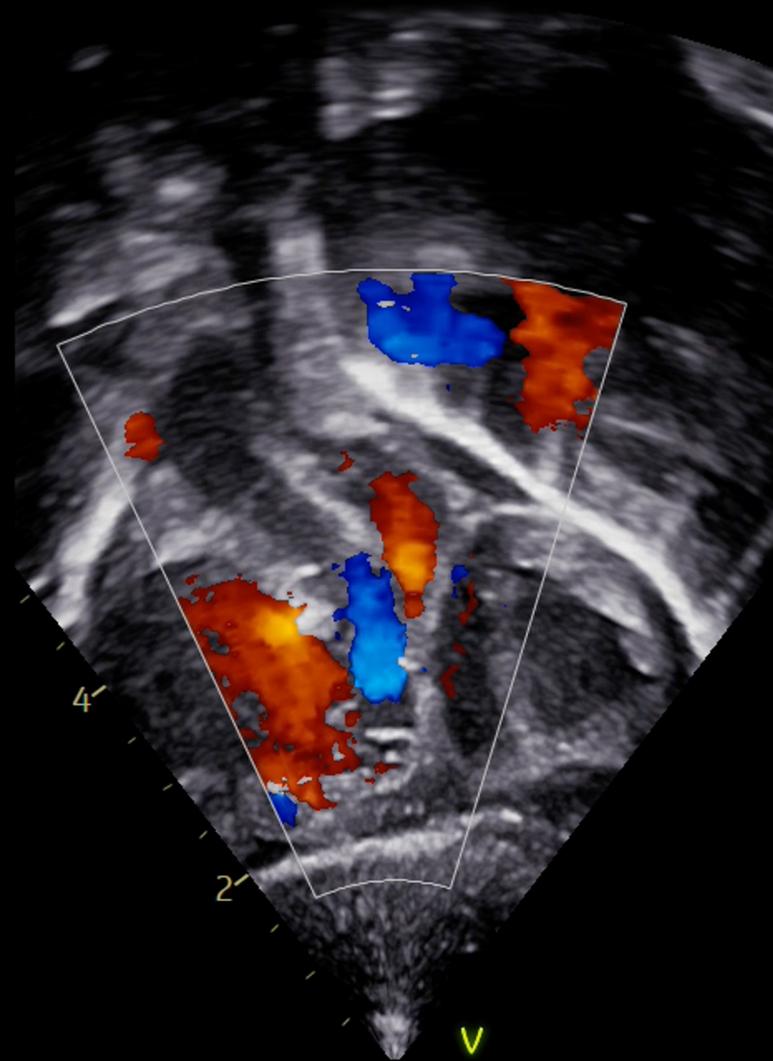
1D



Soft

87

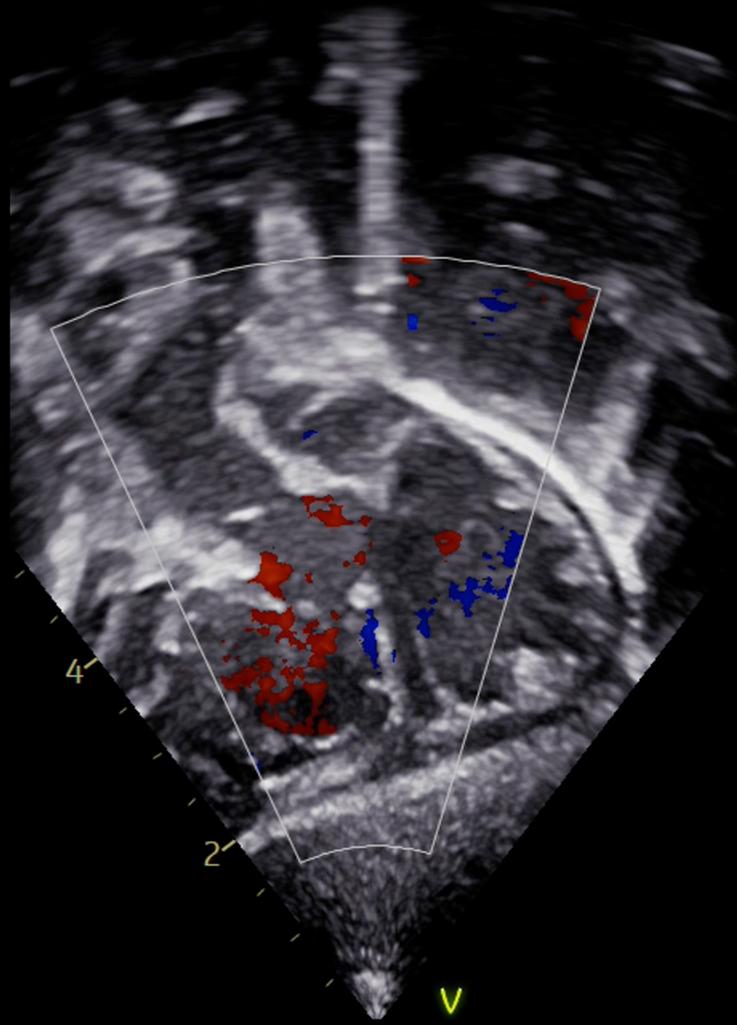
-87



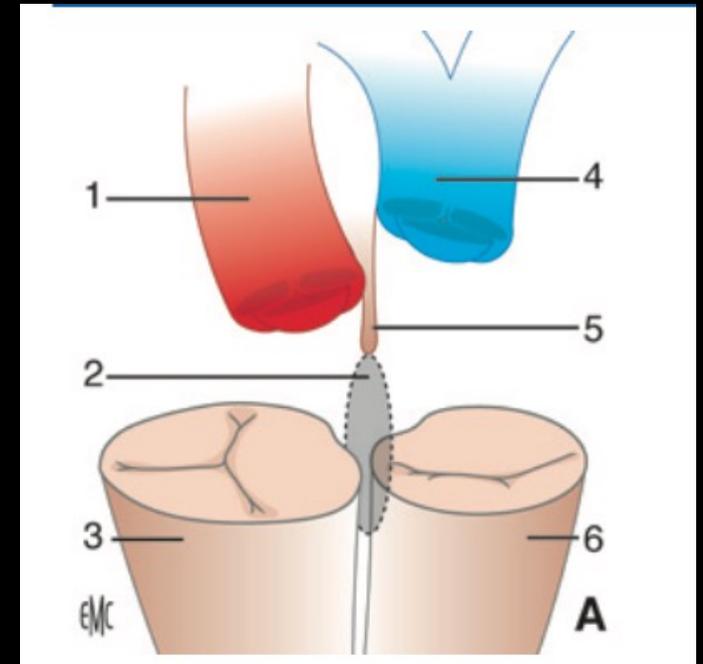
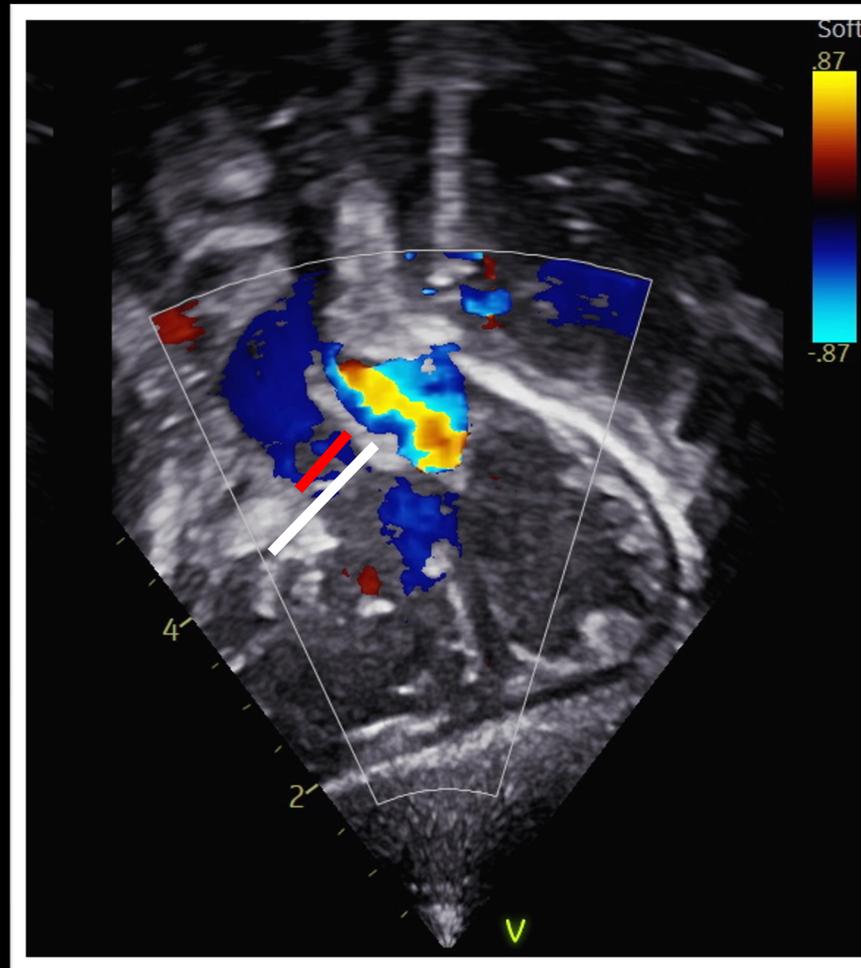
ID



Soft
.87
-.87



Pulmonary-tricuspid distance 13 mm **aortic diameter: 8-10 mm**



What
surgical
approach
will you
suggest ?

- Neonatal arterial switch procedure with VSD closure and LVOT relief
- REV or Rastelli > 8 kg
- IVR with RVOT resection > 6 kg
- We will decide on definite surgical strategy in 6 months.

Multidisciplinary decision

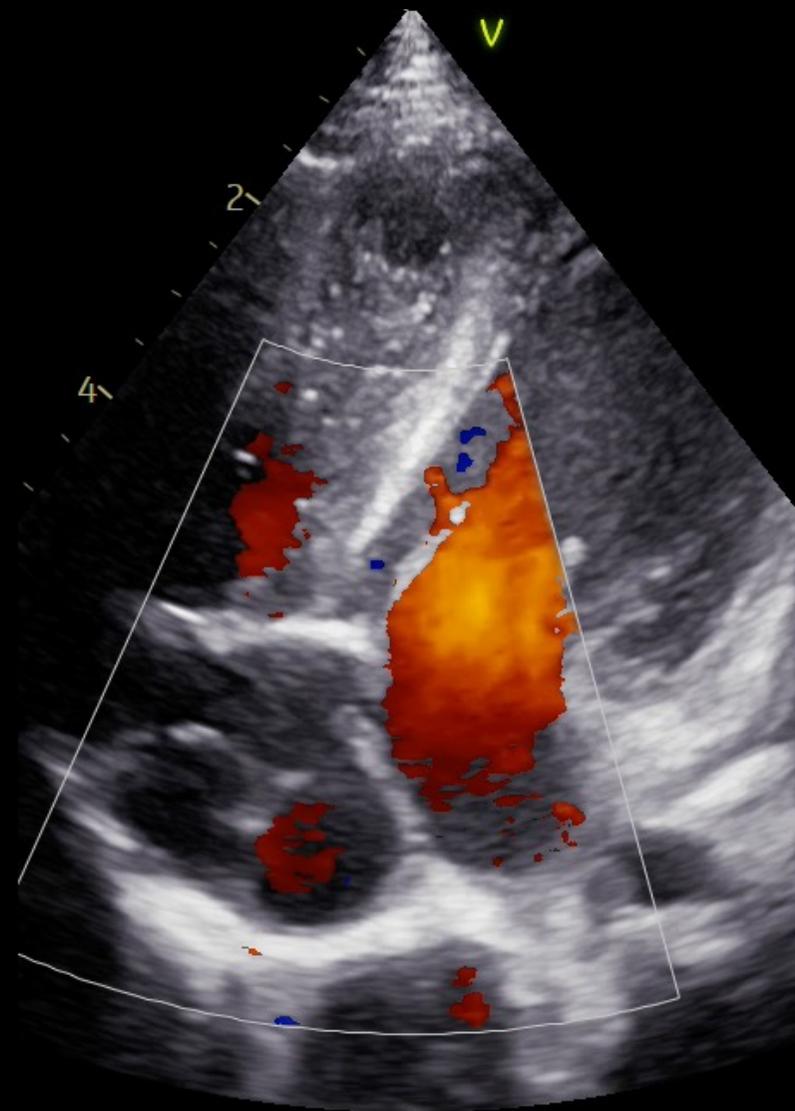
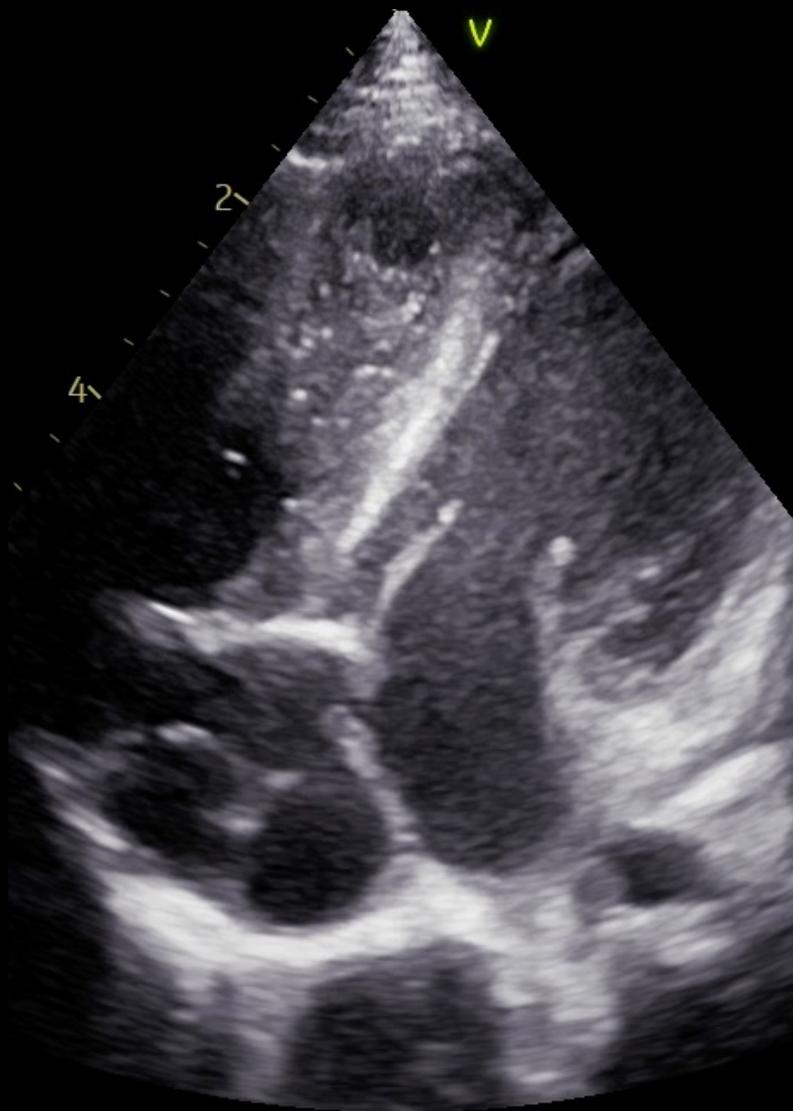
Confirmation of prenatal diagnosis :
DORV-physiologic pink TOF type
with anatomically transposed great
vessels

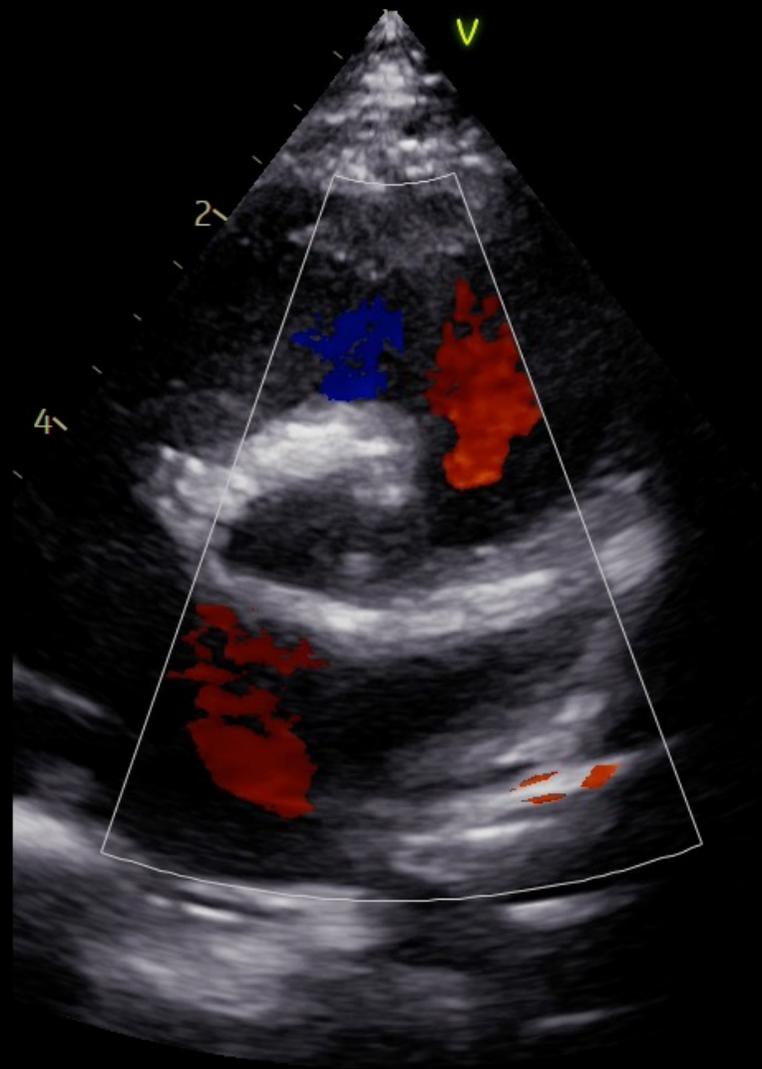
Suggested surgical pathway:

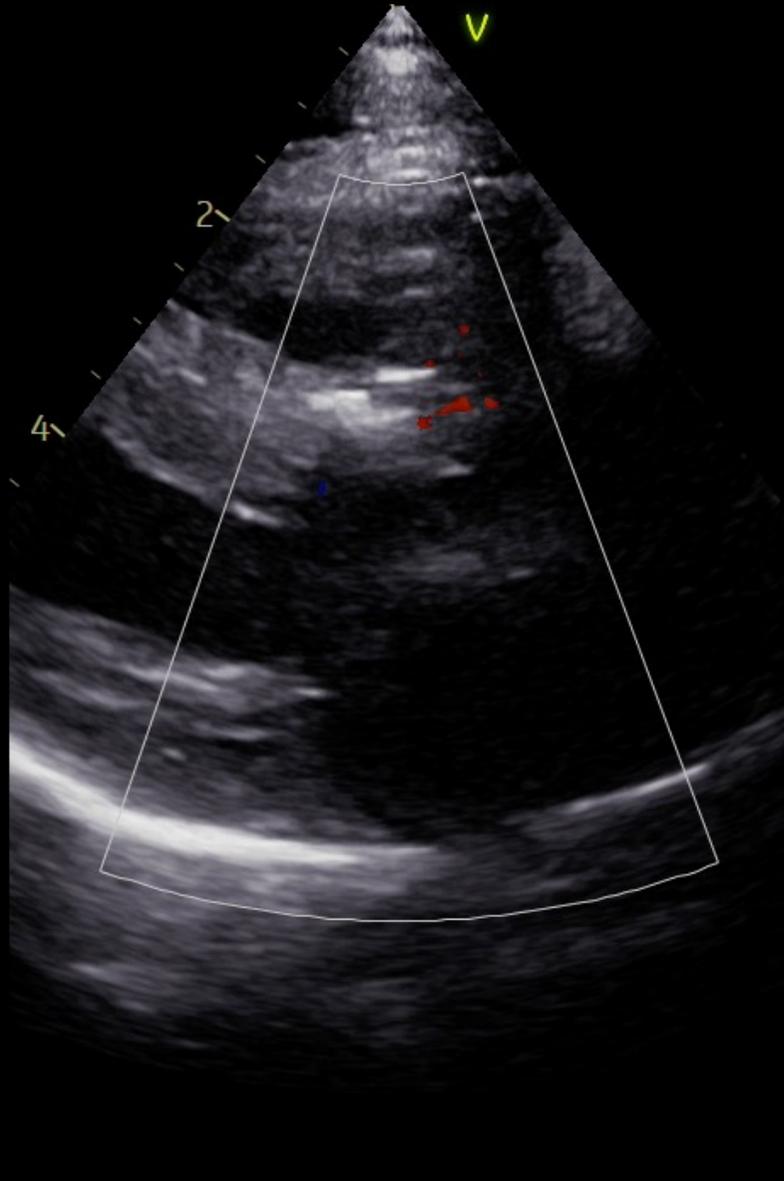
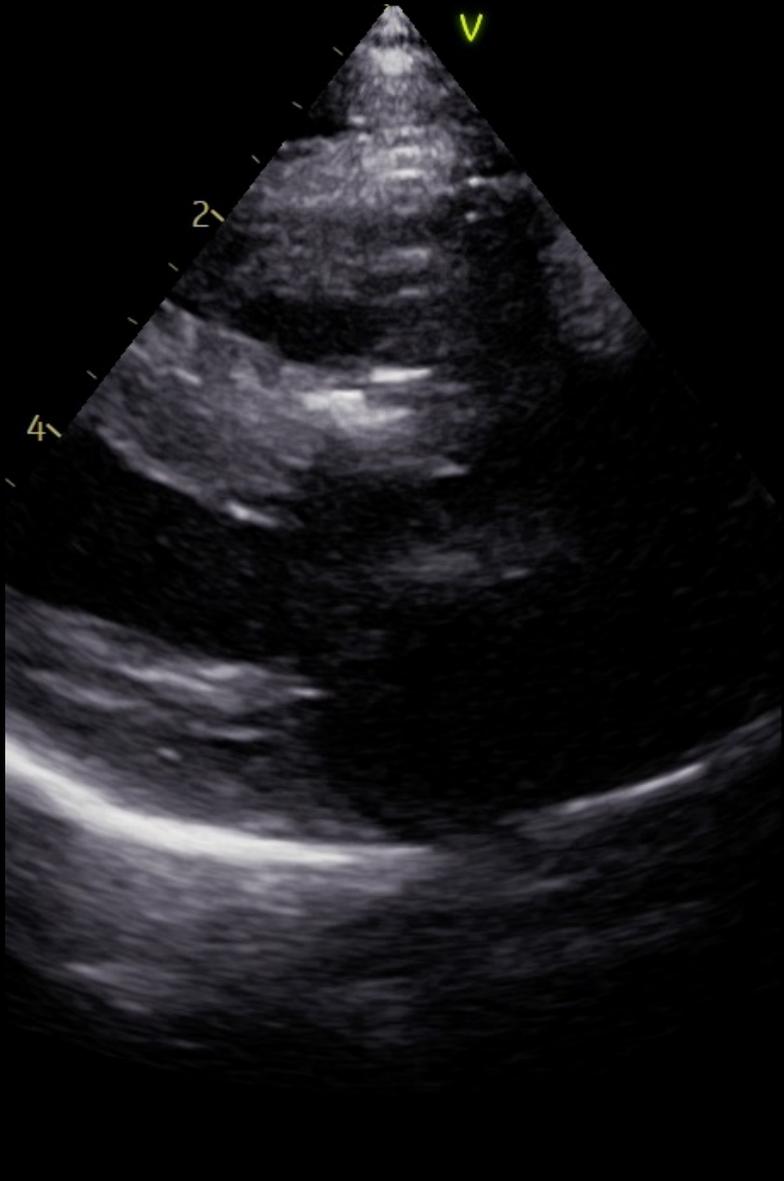
- IVR at > 6 kg with RVOT resection

Postoperative result

Surgery at 7 months, weight 6.1 kg, major complication: pace-maker







Prague



Thank you for your attention

