Utrecht Sessions 2023 on Hypoplastic Left Heart Syndrome

Case presentation

Active follow-up for the best of the patient: a fetal patient with tight AS.

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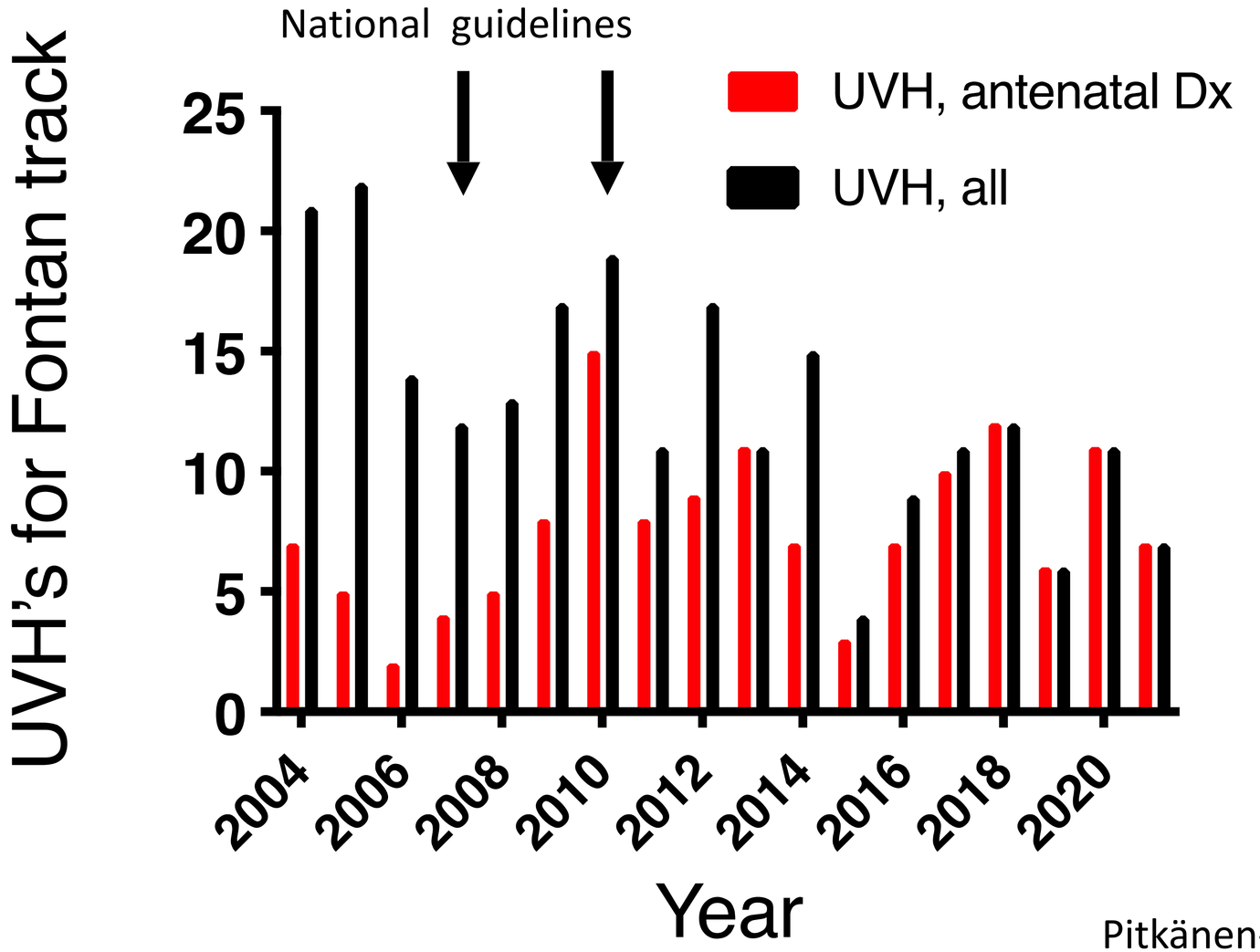
Antenatal rate of diagnosis of single-ventricle heart patients taken for palliation surgery



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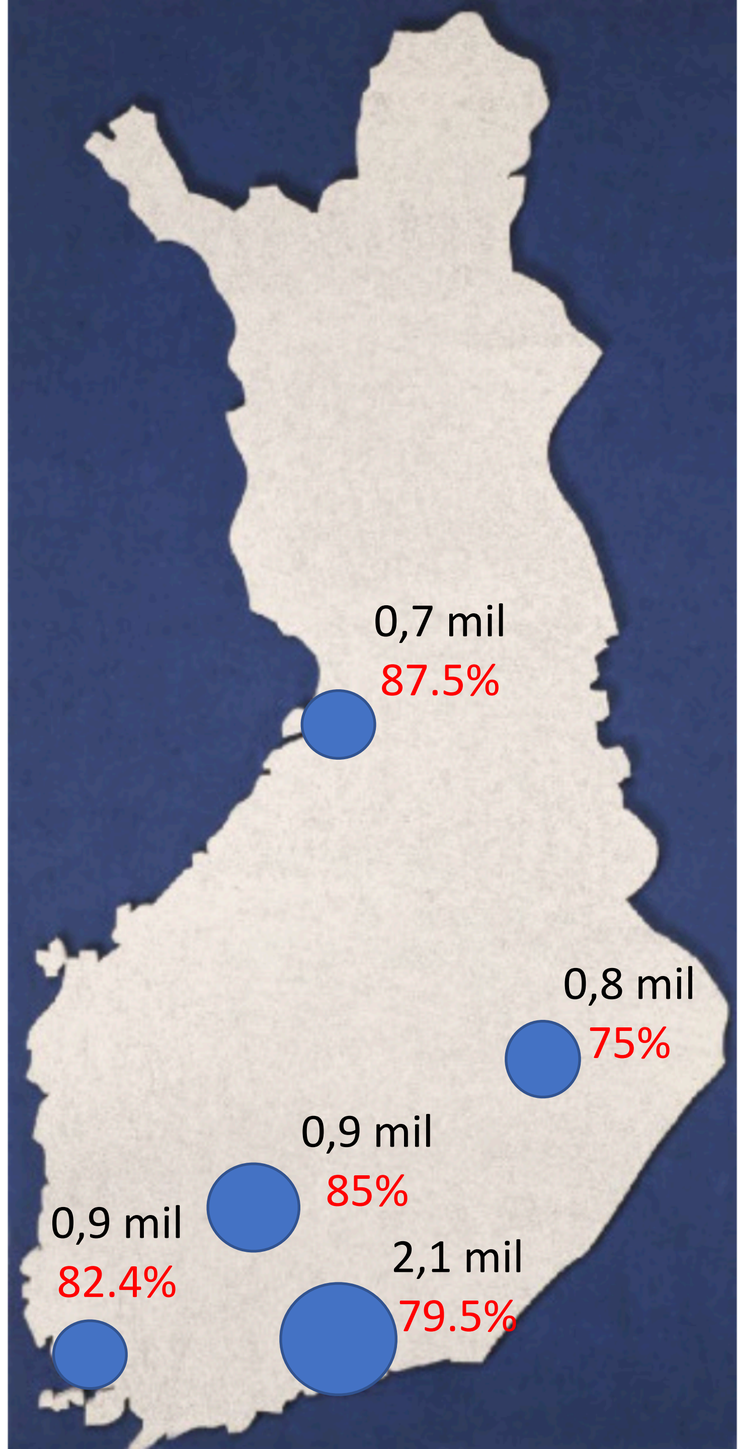
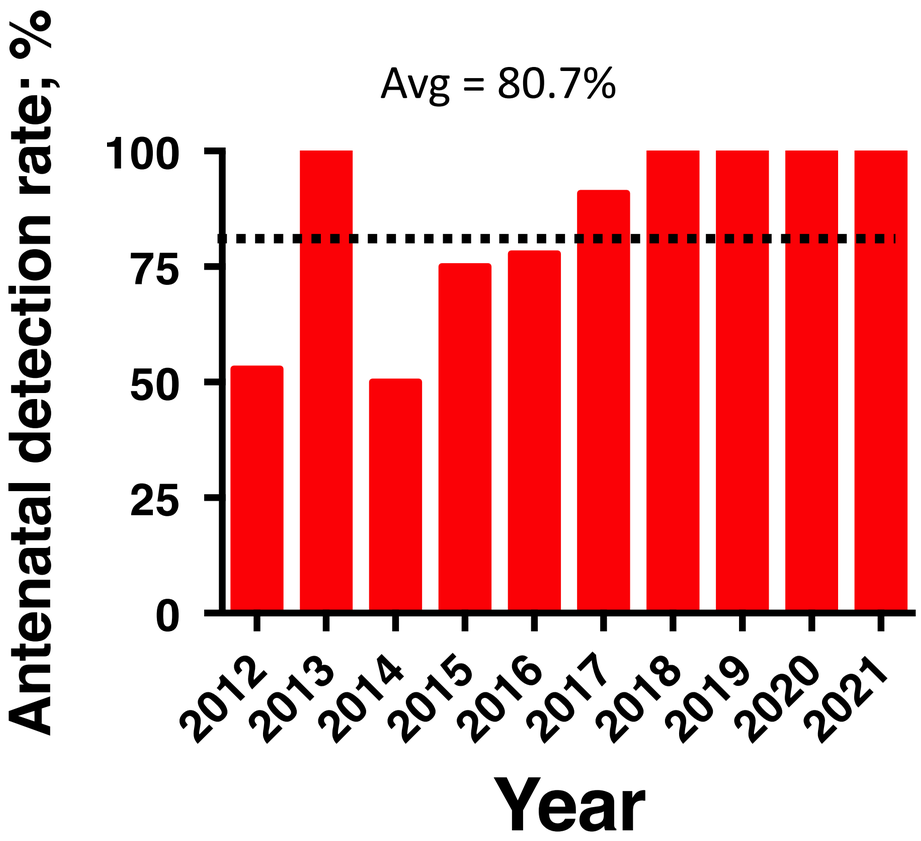
Antenatal detection rate of actively treated UVH during 2012-2021





N=104

2012-2016	**57**
2017-2021	47

All treated & Antenatal cases*/100 000

Helsinki	Tampere	Kuopio	Oulu	Turku
1,85	2,22	1,48	2,19	1,95
1,47	1,88	1,11	1,92	1,61

* Within 10 years

Pitkänen-Argillander OM, personal, 2022

Case presentation:

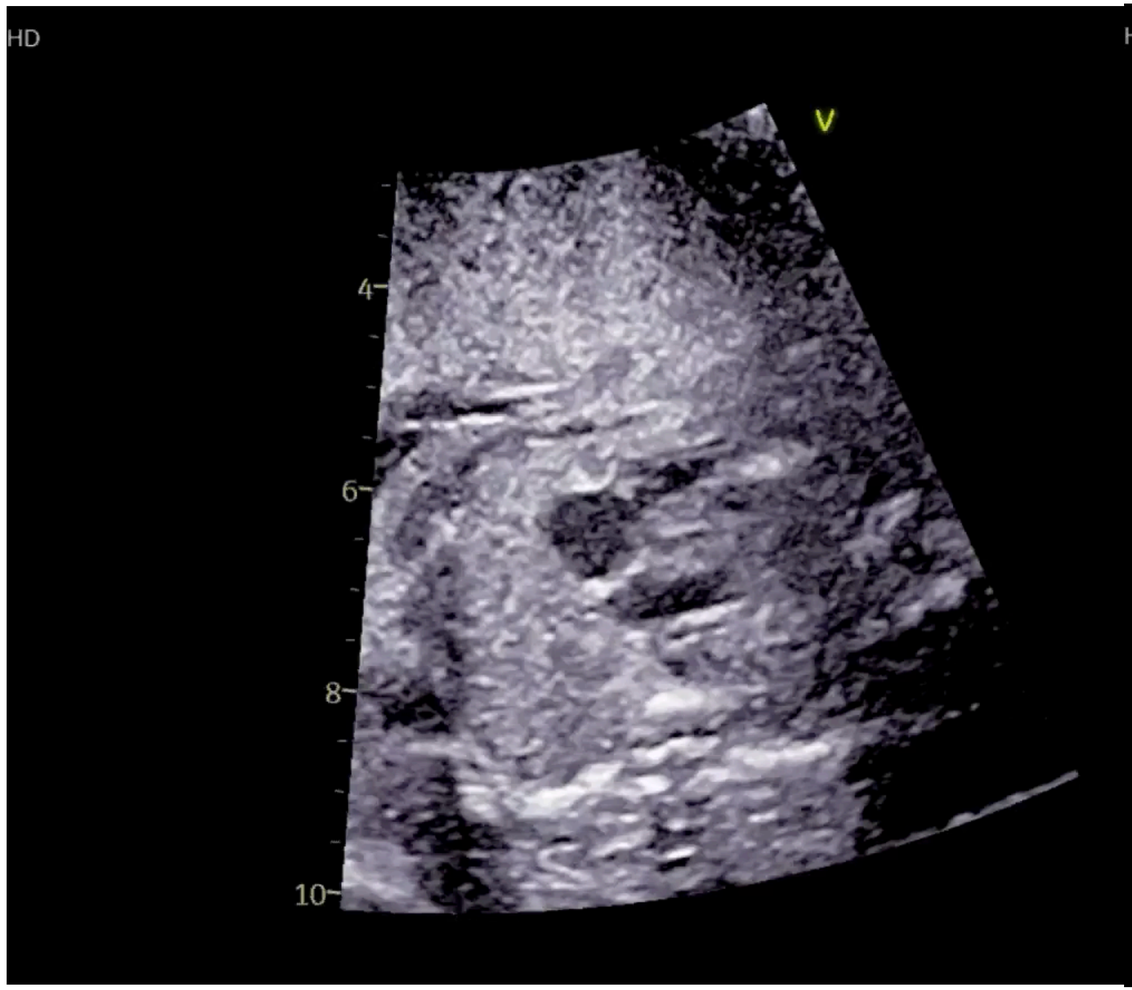
Family “M” expecting a fetal patient with tight AS - retrospective thoughts from our intentions to modify natural course.

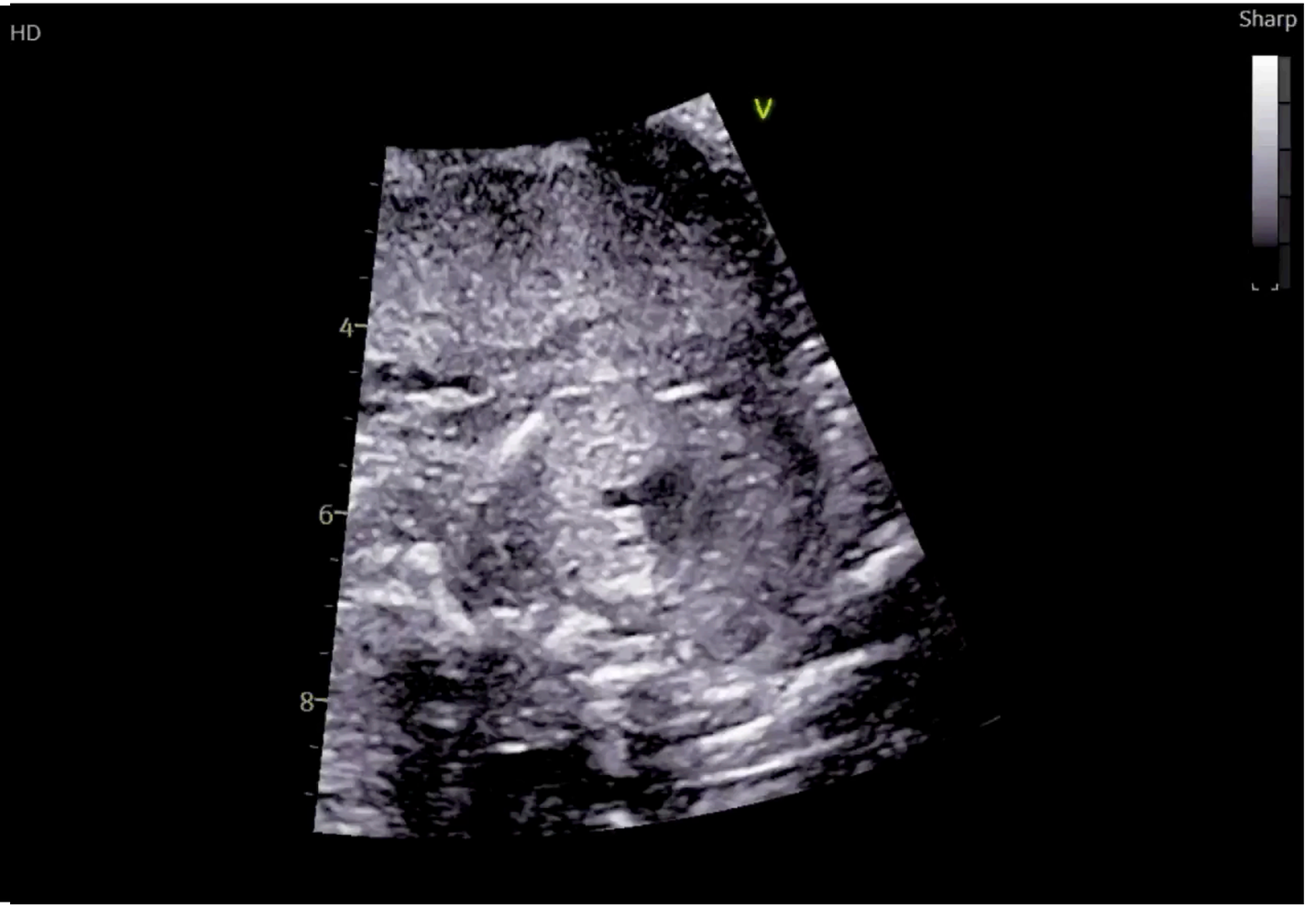
Referral from a rural central hospital

H22+1, suspected TOF

- Healthy obese mother, 160 cm/99 kg, G8P7, 37 years of age.
- Couple with deep religious conviction.
- (Instead of TOF) Fetal tight AS detected with normally contracting symmetric ventricles reaching the apex, no EFE, competent valves, PFO shunting R-L, retrograde AoA-flow up to the third neck vessel. Suspicion of a VSD (later not detected).
- Counseling: probable 2V strategy, though progression of disease described likely. AoV-valvuloplasty and Ross/Konno options discussed, as well.
- TOP not an alternative to the couple, active therapy desired should even HLHS develop.
- Possibilities for fetal valvuloplasty in Linz were probed.
- Finnish Health Insurance Office contacted in advance to inquire coverage of costs of care & travel (the couple had never been on an aeroplane).

Gestational age h22+1





In light of these “screening” images:

Was the initial obstetrician’s suspicion of tetralogy of Fallot understandable?

- o Yes
- o No

Gestational age h22+1

Initial fetal echocardiogram @ h22+1

Predicting evolving anatomy in our AS-case

Criterion	BiV probable	Criterion	HLHS probable
No aortic atresia	+	Critical aortic stenosis / atresia	+ / -
Aortic valve Z score > -3.5	+ (Z -1.3, 2.8 mm)	Reversal of flow in aortic arch	+
Mitral valve Z score > - 2	+ (Z +0.5, 6.8 mm)	LV length Z score < -2	- (Z +1.3, 19 mm)
LV long axis Z score > 0	+	MV Z score < -3	- (Z +0.5)
LV short axis Z score > 0	+	Monophasic MV inflow	-
AoV gradient > 20 mmHg	+ (turbulent flow)	Left to right FO shunting	-
MR gradient > 20 mmHg	No regurgitation	No obvious subvalvar obstruction	+
		Bidirectional pulmonary venous waveforms	-
		LV function capable of generating 10 mmHg AoV or 15 mm Hg MR	- (indirect deduction)

Hoping for 2V outcome

Consultation with the Children’s Heart Center colleagues in Linz

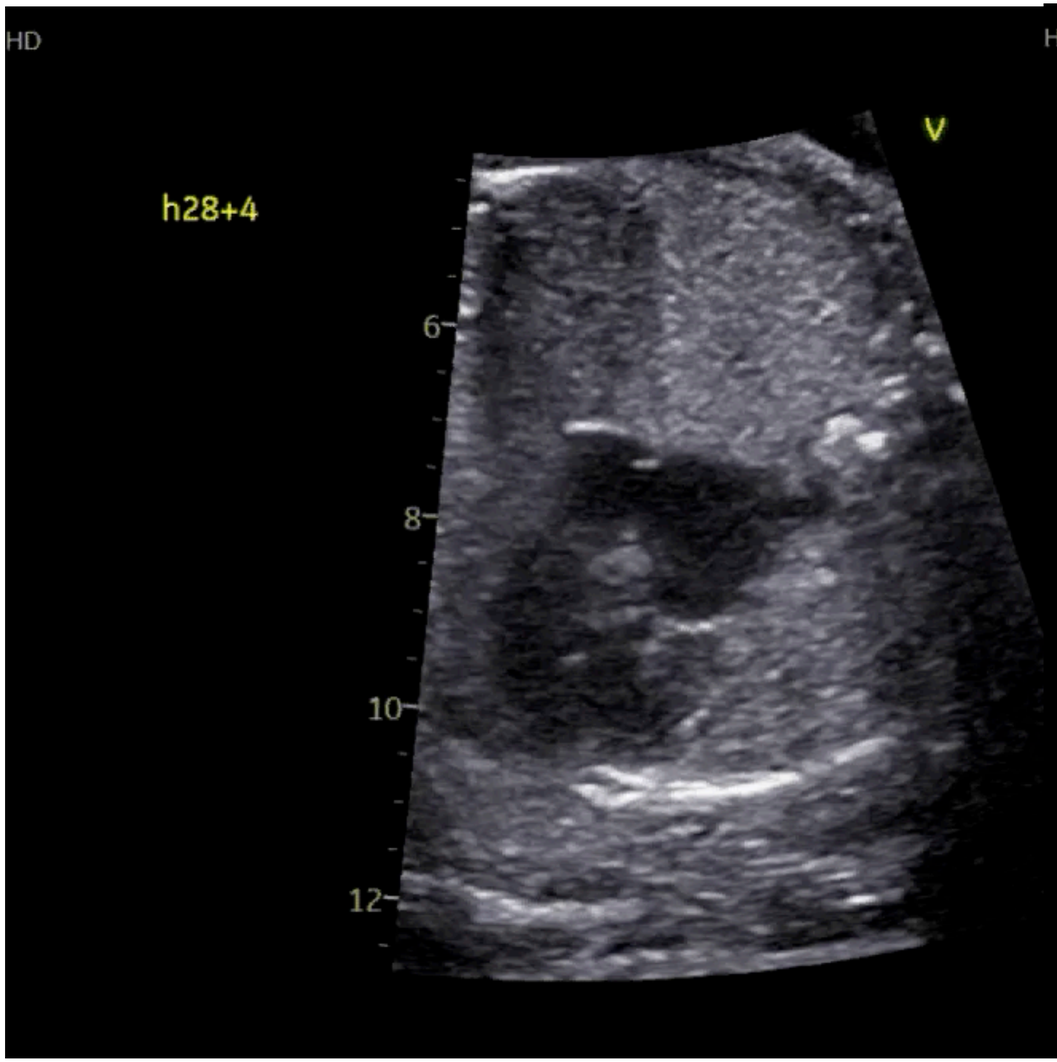
- Conclusion after the first fetal echocardiogram:
  - Tight valvar aortic stenosis
  - Normal size LV/RV
  - LV/RV function preserved
  - No EFE
- No indication for fetal AoV-intervention at this stage, close follow-up required

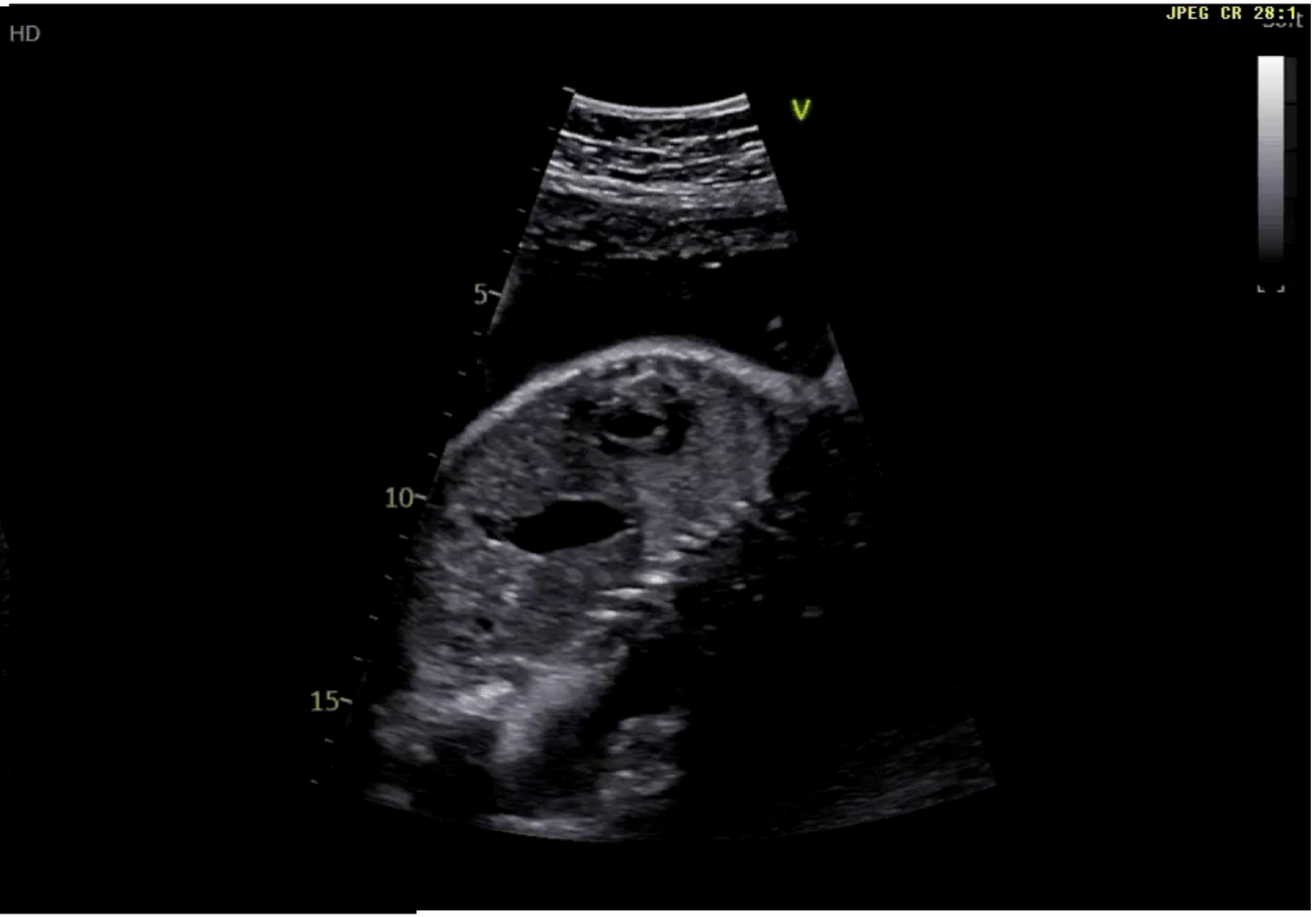
Follow-up echo at 27+4

Predicting anatomy

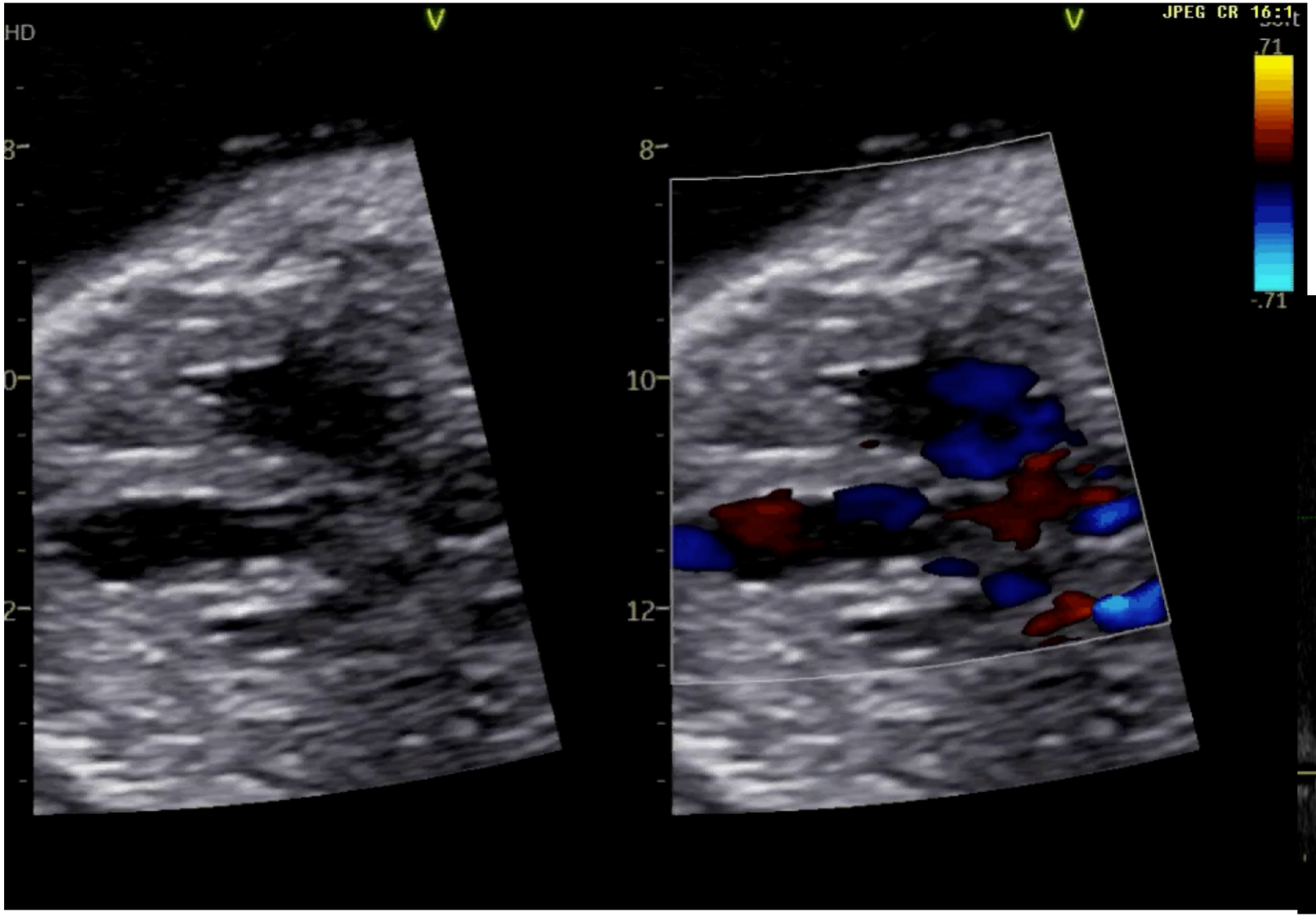
Criterion	BiV probable	Criterion	HLHS probable
Aortic valve Z score > -3.5	**+ (Z -0.6, 4 mm)**	Critical aortic stenosis or atresia	**+**
Mitral valve Z score > - 2	**+ (Z -1.2, 7 mm)**	Reversal of flow in aortic arch	**+**
LV long axis Z score > 0	**+**	LV length Z score < -2	**- (Z + 1.6, 26 mm)**
LV short axis Z score > 0	**+**	MV Z score < -3	**-**
AoV gradient > 20 mmHg	**+ (50 mmHg)**	Monophasic MV inflow	**+**
MR gradient > 20 mmHg	**+ (> 80 mmHg)**	Left to right FO shunting	**+**
Threshold score	**6/6**	No obvious LVOT Obstruction	**+**
		Bidirectional pulmonary venous waveforms	**-**
		LV function capable of generating ≤10 mmHg AoV or <15 mm Hg MR	**-**
		Threshold score	**5 / 9**

Gestational age h27+3

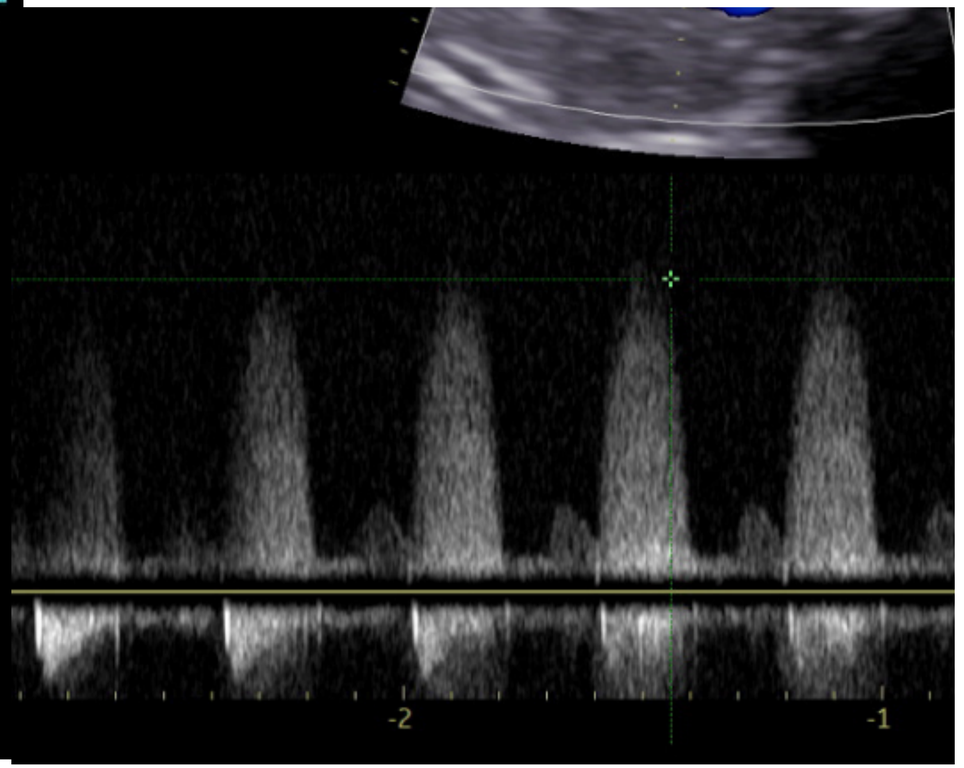




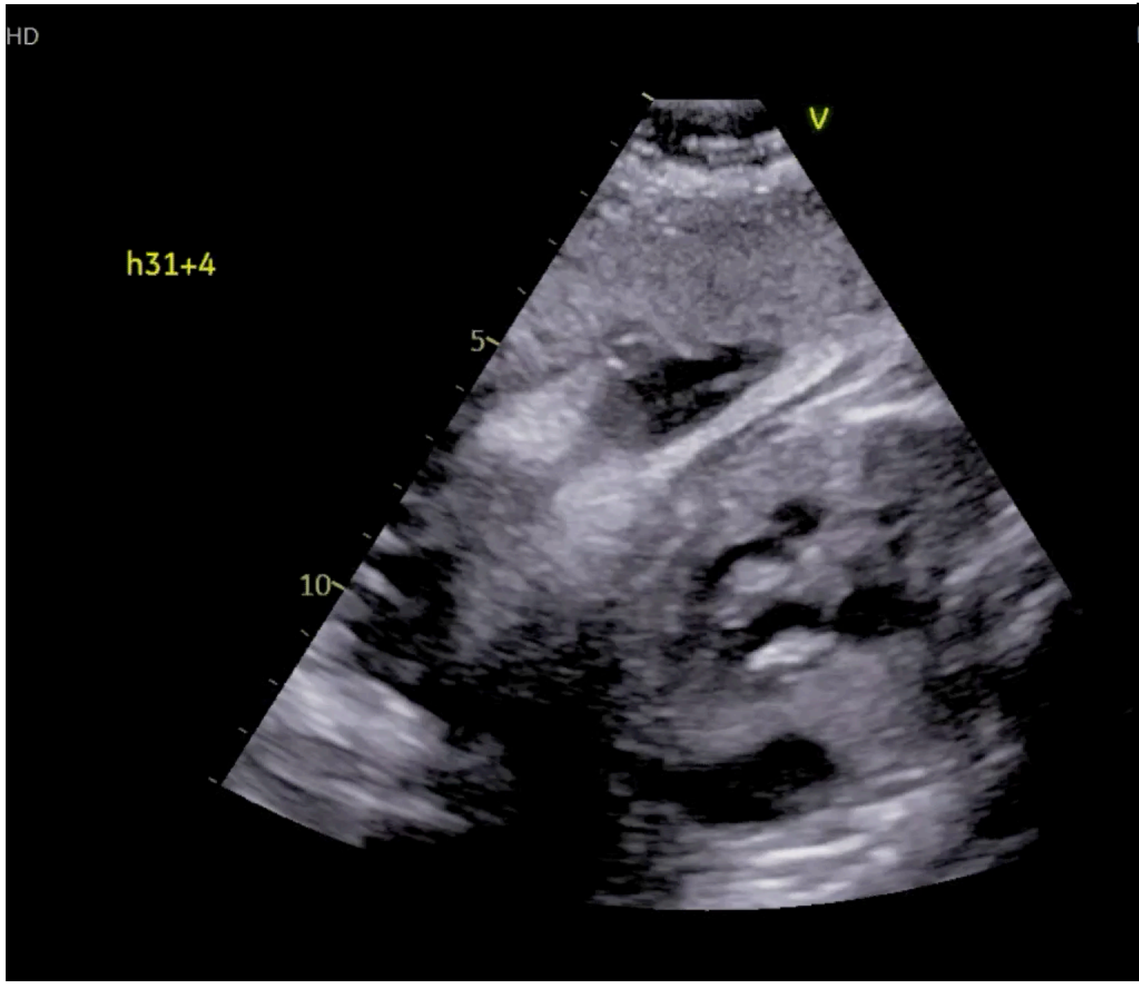
Gestational age h27+3

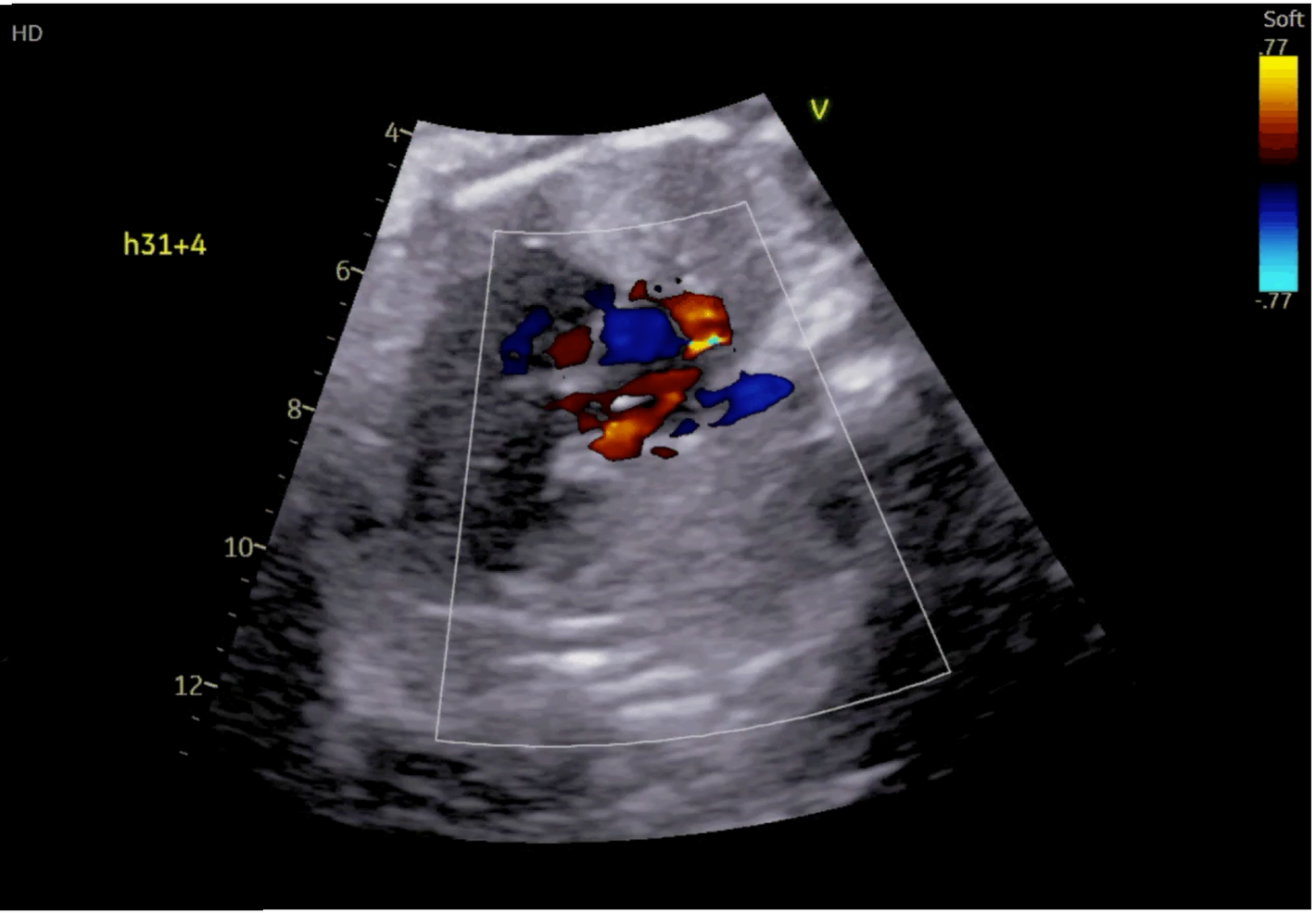


AS 80 mmHg

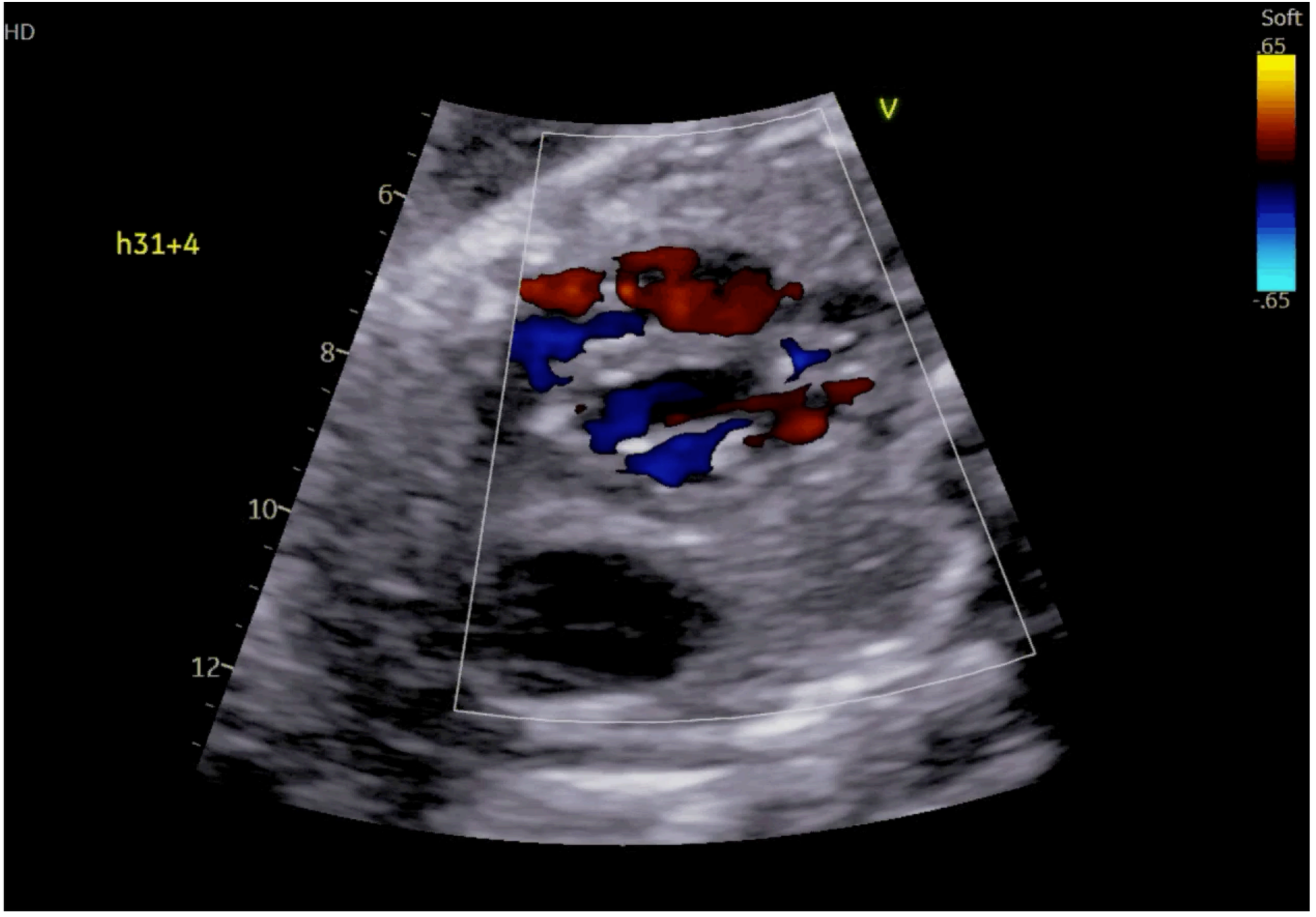


Gestational age h31+4





Gestational age h31+4



Gestational age	MR / gradient	MV-inflow	MV-annulus	AS-gradient	AoV, mm	Ao-asc, mm	LV length	LV-fxn	PFO/PVs	AoA-flow
31+4 2.0 kg	++ >70 mmHg	Monophsc	Z +1	>65 mmHg	>5.5 mm Z +0.5	>6 mm	29 mm (Z +1.3)	EFE FS 24%	L-R Mild restr	Retrogr
				Decision to treat postnatally						
34+4 2.7 kg	++ >80 mmHg	Monophsc	Z +0.2	55 mmHg No leak	> 6mm (Z +0.25)	7 mm (Z +0.5)	36 mm	EFE Reduced	L-R No restr	Retrogade
36+4	++ 90 mmHg	Monophsc	Z +1.5	49 mmHg	7	6-7	37 cm	EFE Reduced	L-R No restr	Retrogr

Decision to aim at postnatal treatment

H31+4, weight estimate 1,998 kg

- Plans for fetal intervention given up:
  - Regular visit to the perinatologist at the home town and our U Hospital at 28+4, 32+6, 33+6, 36+4 weeks of gestation.
    - the biophysical profile of the baby including growth had remained stable
  - Based on eye-balling, FS-measurements, MR and LVOTO-gradients LV function had remained stable
  - Some suspicion regarding the MV, and its role in BiV/UVH decision making
- Instead, planning of optimal timing for postnatal intervention initiated with our interventional cath-team.
- Presentation of data at the perinatal interdisciplinary (bi-monthly) case-rounds at 32+0 weeks. Glucorticoid therapy administered as planned. Postnatal therapy agreed.

Do you agree?

o Yes, I would have aimed at postnatal valvuloplasty, as well.

o No, I would have proceeded to fetal intervention at an earlier stage.

Baby boy born @ h37+5: *3.75 kg, 50.5 cm, AP 8/1/8*

First postnatal echocardiogram

- On alprostadil-infusion, spontaneous breathing (non-invasive ventilation with pressure support)
- Prior to procedure:
  - No inotropic remedy
  - Blood lactate 4.2 mmol/l, pH 7.25, HCO3 15-18 mmol/l
  - AoV-annulus 5,7 mm (Z -2), ascending Ao 7 mm (Z , distal arch 4.4 mm (Z -1.9), isthmus 5.3 mm (Z -0.2), abdominal Ao 6 mm (Z +0.1)
  - Outflow gradient 65-70 mmHg, normal coronary anatomy
  - LVIDD 20 mm, LVEF 50%, septal thickness 6.2 mm (Z +2.4), PW 5.6 mm (Z +3.4), mass 21 g (Z +3.8).
  - Haycock discriminant score Z -0.11 (LV-length 43 mm)

Baby boy born @ h37+5: *3.75 kg, 50.5 cm, AP 8/1/8*
Aortic valvuloplasty on postnatal day 1

- On alprostadil-infusion, spontaneous breathing (non-invasive ventilation with pressure support)
- Prior to procedure:
  - No inotropic remedy
  - Blood lactate 4.2 mmol/l, pH 7.25, HCO3 15-18 mmol/l
- Cardiac catheterization
  - LV 126/10 mmHg, Ao-asc 53/31 mmHg, gradient 70 mmHg
  - Tyshak-catheter 5x20 mm x3 —> Tyshak-catheter 6x20 mm x2
  - LV 83/9, gradient 18 mmHg
  - No complications
- Echocardiogram
  - MV-inflow gradient m3 mmHg
  - MR mild-to-moderate
  - No AR
  - LVOTO-gradient peak 30, m15 mmHg

Boy 1 year 3 months of age

Followed at 3 month intervals

- Active symptomless boy, no remedies
- Normal growth, 3/6 murmur at right upper sternal border, no thrill, liver size normal, femoral artery pulses well palpable
- Latest echocardiograms have been non-progressive
  - Mild central MR, inflow E 1.0 and A 1.2 m/s
  - Hypertrophic LV, papillary muscles still echogenic
  - LV 30/15 mm, EF 79%, septal thickness 6 mm, posterior wall 7 mm
  - AoV-annulus 12 mm, mild AR
  - LVOTO-gradient (slight subvalvar + valvar) mean 49 mmHg (peak flow velocity 4.8 m/s)
  - Mild retrograde flow in the distal AoA
  - No findings suggesting PH

Discussion

- Natural course of AS is not self-evident at early gestation
  - to intervene or not to intervene?
    - we decided to ask advice from professionals with vast experience - it paid off :)
    - in our case “dry run” for arranging F-AS care of future cases
  - Mitral valve raised concerns towards the end gestation
    - Did the MV chordae-dysplasia develop with EFE? Undifferentiated MV chord tendinae?
    - Prematurity < 36 wk & LBW are risk factors for HLHS / Fontan track
  - AEPC-study on “Benefits of Aortic Valvuloplasty to prevent development of HLHS”
    - focus on natural and modified history of aortic stenosis - ?to intervene or not to intervene?