Leids Universitair
Medisch Centrum

Coarctatio aortae

Utrecht, NVHVV, 10-04-2018

Derk Jan Ten Harkel

Kindercardioloog

LUMC, LEIDEN

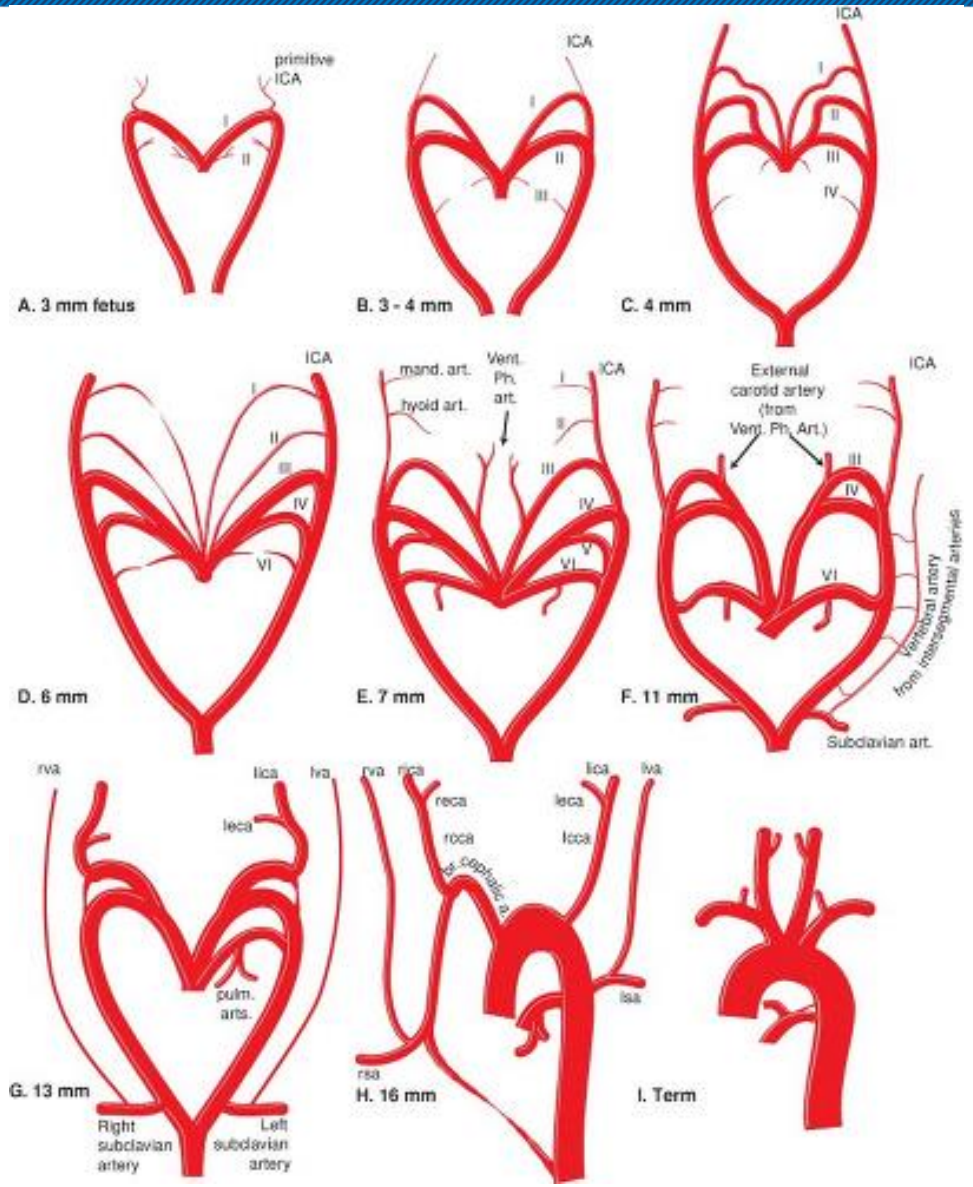
Embryologie

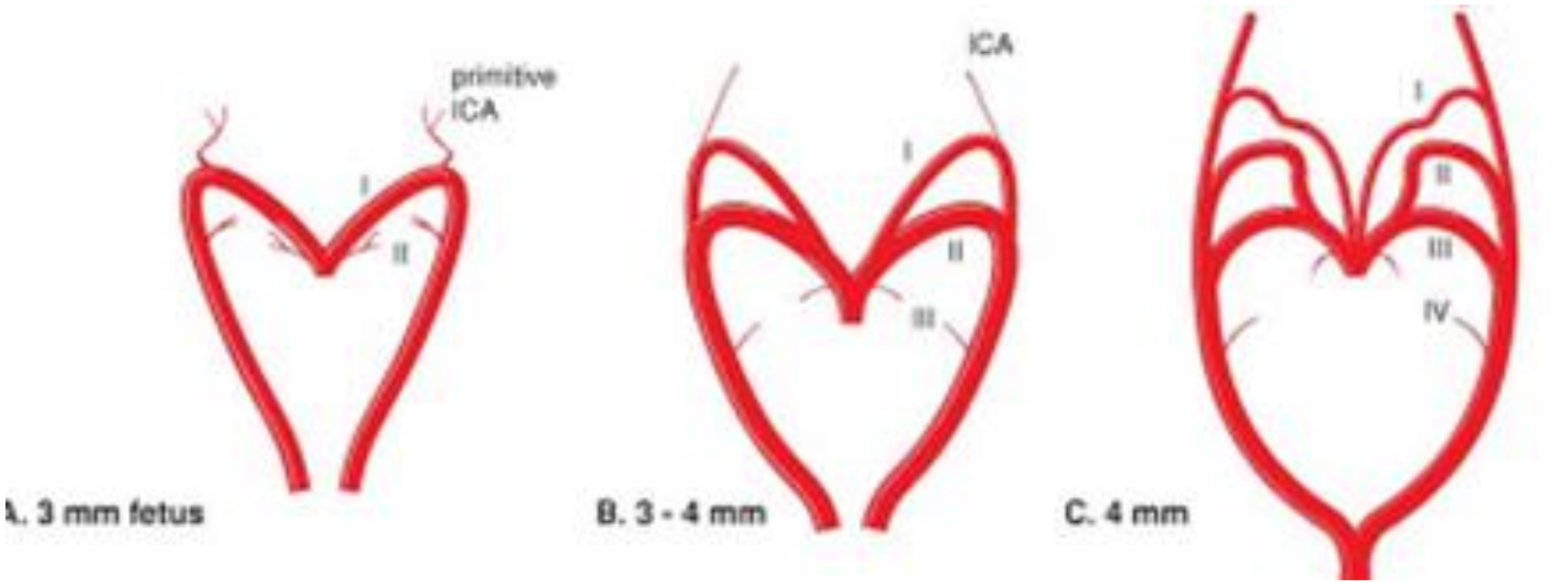
Prenatale diagnostiek

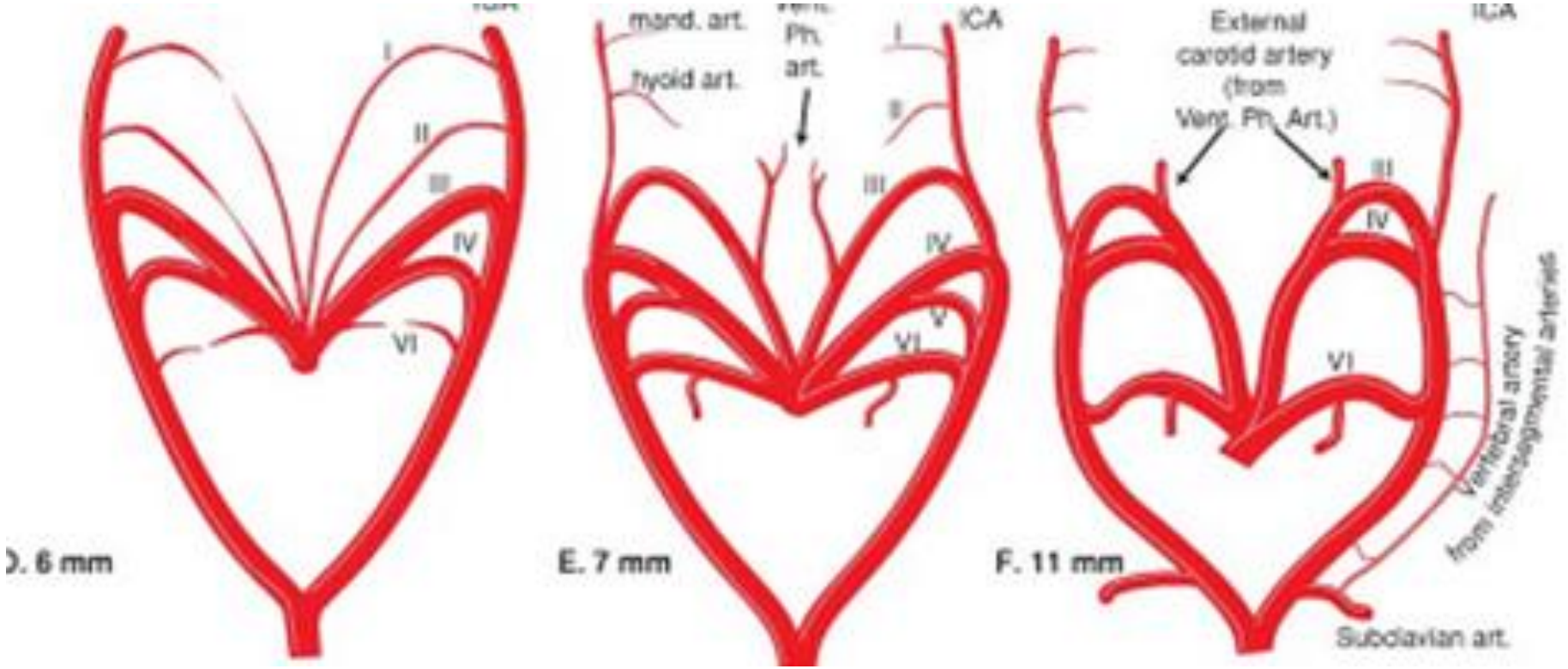
Behandeling postnataal

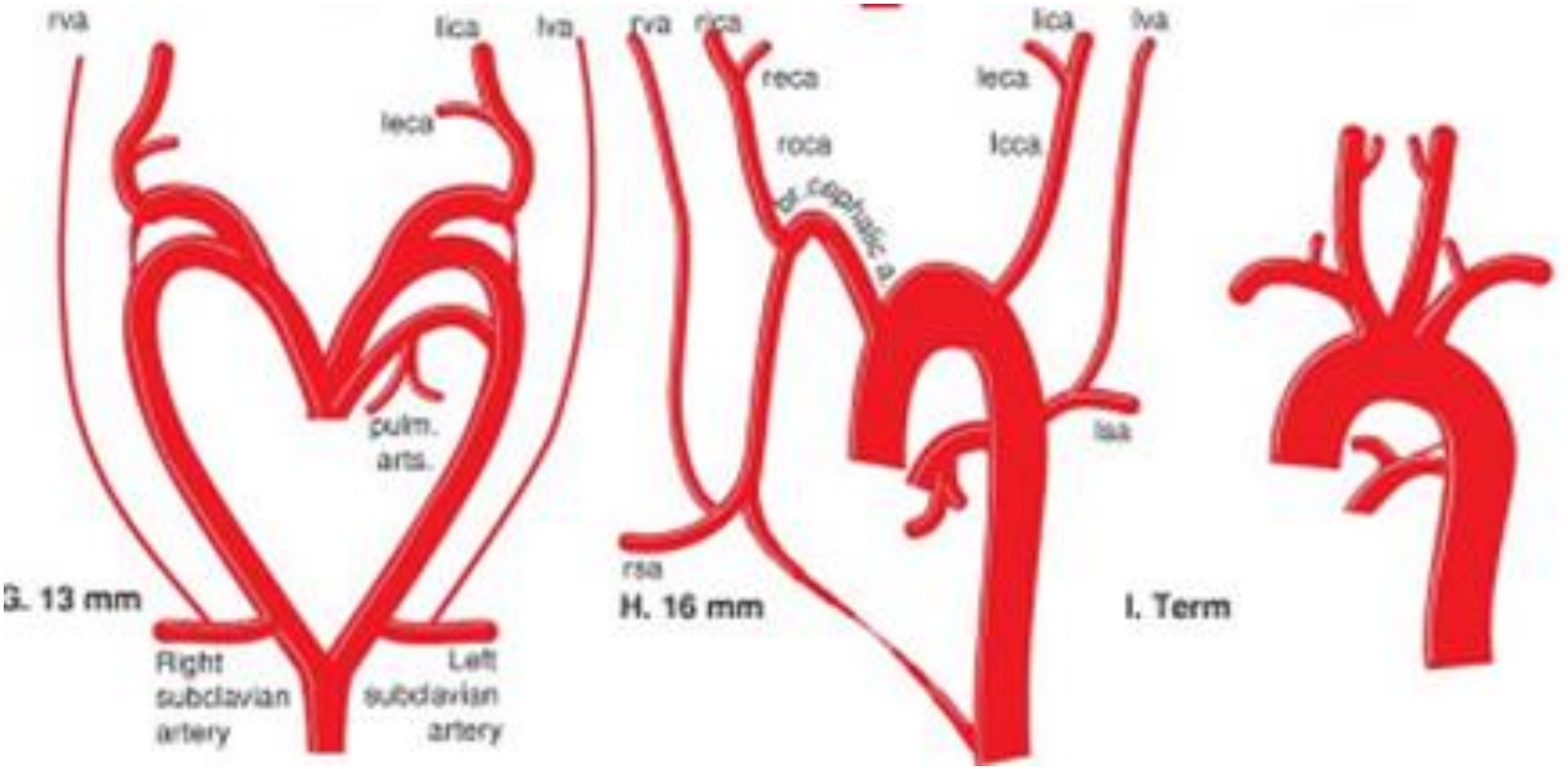
Lange termijn

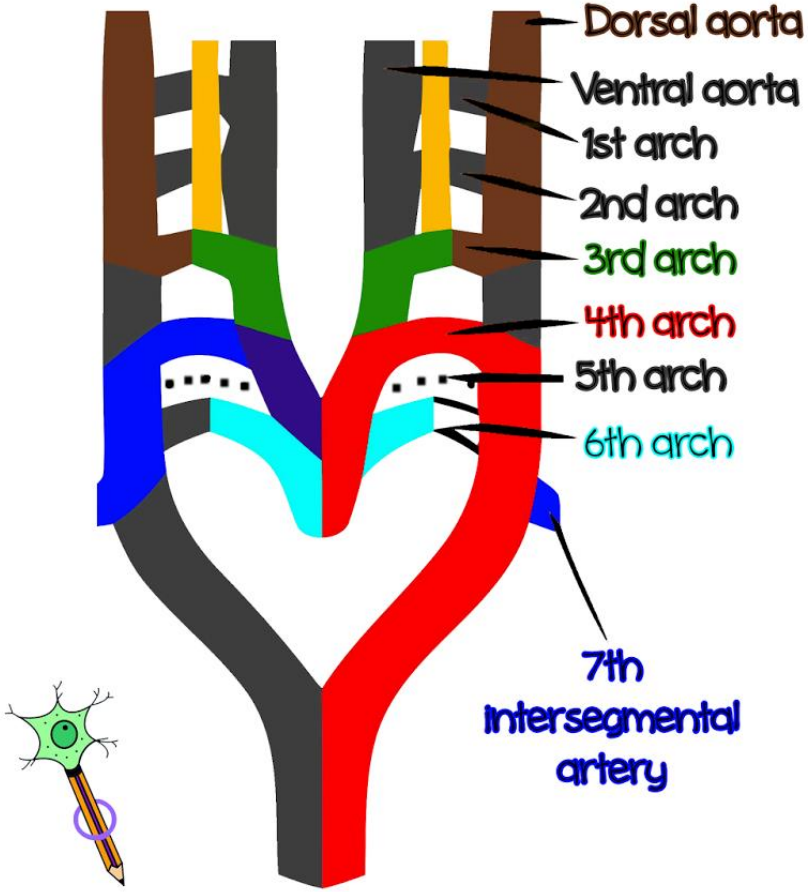
Embryologie van de aortaboog/coarctatio

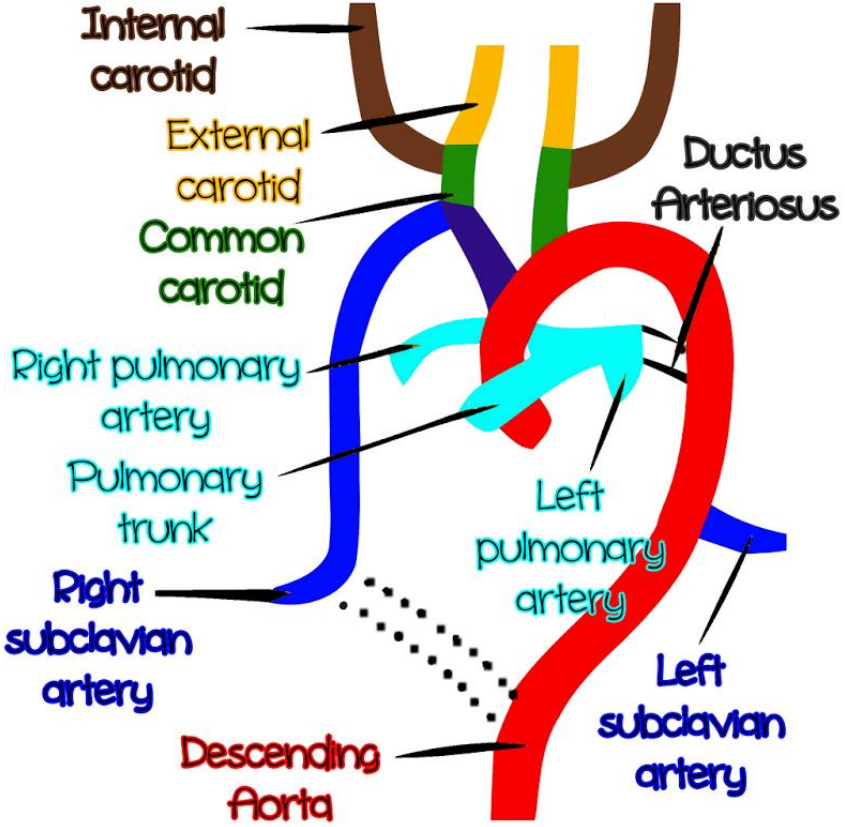

primitive ICA
A. 3 mm fetus
ICA
B. 3 - 4 mm
ICA
C. 4 mm
ICA
D. 6 mm
mand. art.
hyoid art.
Vent. Ph. art.
ICA
E. 7 mm
External carotid artery (from Vent. Ph. Art.)
ICA
Vertebral artery from intersegmental arteries
Subclavian art.
F. 11 mm
rva
lica
lva
leca
pulm. arts.
Right subclavian artery
Left subclavian artery
G. 13 mm
rva
rca
reca
roca
lica
lva
leca
lcca
br. cephalic a.
lsa
rsa
H. 16 mm
I. Term


primitive
ICA
I
II
A. 3 mm fetus
ICA
I
II
III
B. 3 - 4 mm
I
II
III
IV
C. 4 mm

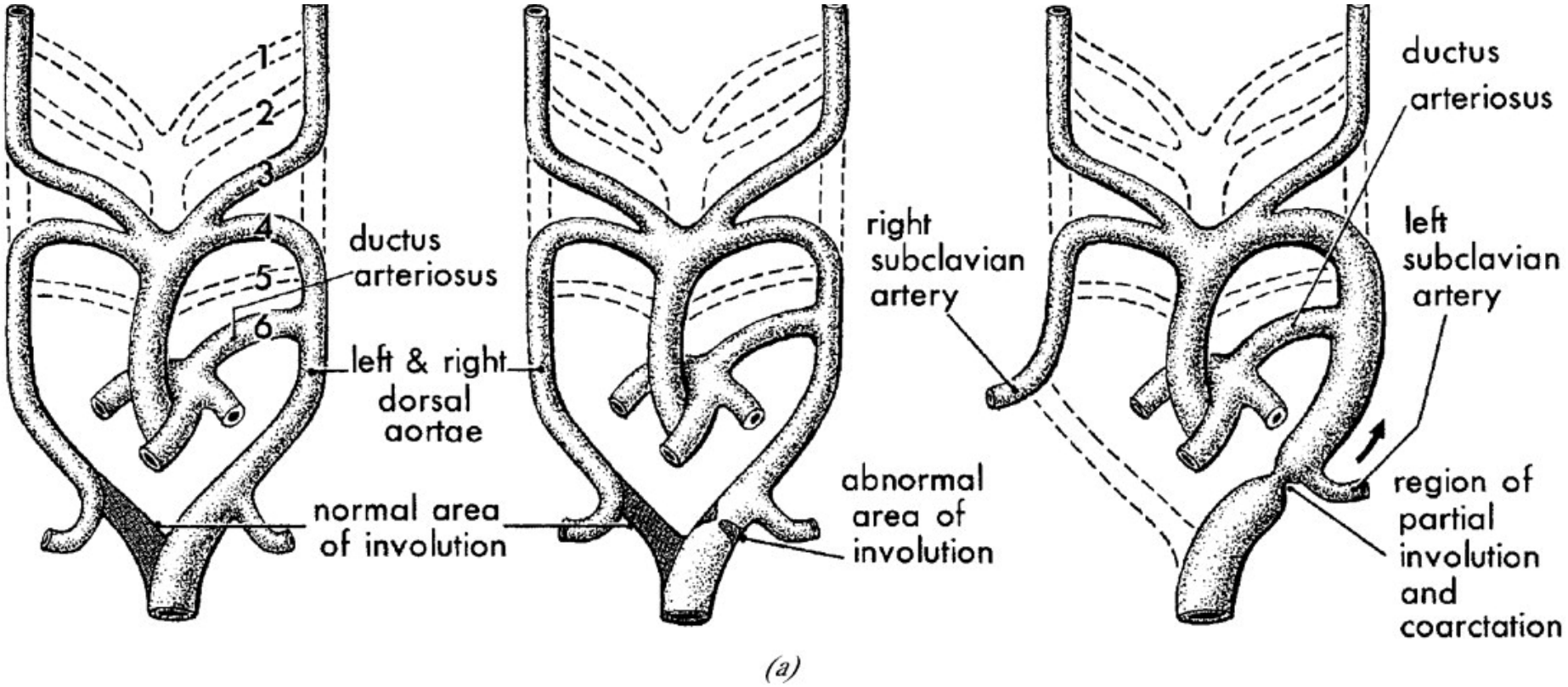

mand. art.
hyoid art.
Ph.
art.
ICA
External
carotid artery
(from
Vent. Ph. Art.)
I
II
III
IV
V
VI
E. 7 mm
F. 11 mm
Vertebral artery
from intersegmental arteries
Subclavian art.


rva
lica
lva
leca
pulm.
arts.
G. 13 mm
Right
subclavian
artery
Left
subclavian
artery
rva
rica
reca
roca
lca
leca
lca
lva
lsa
rsa
H. 16 mm
I. Term

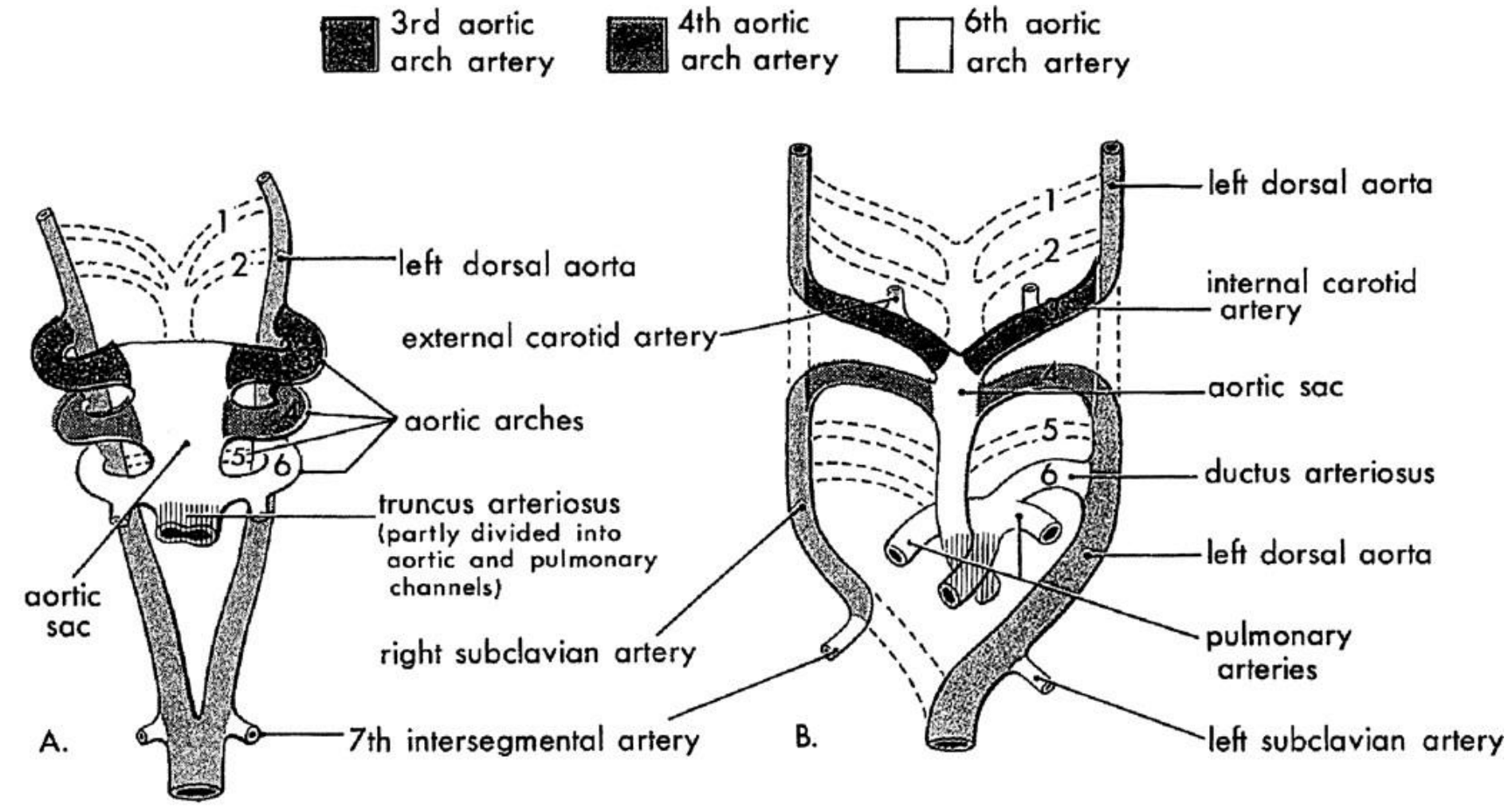


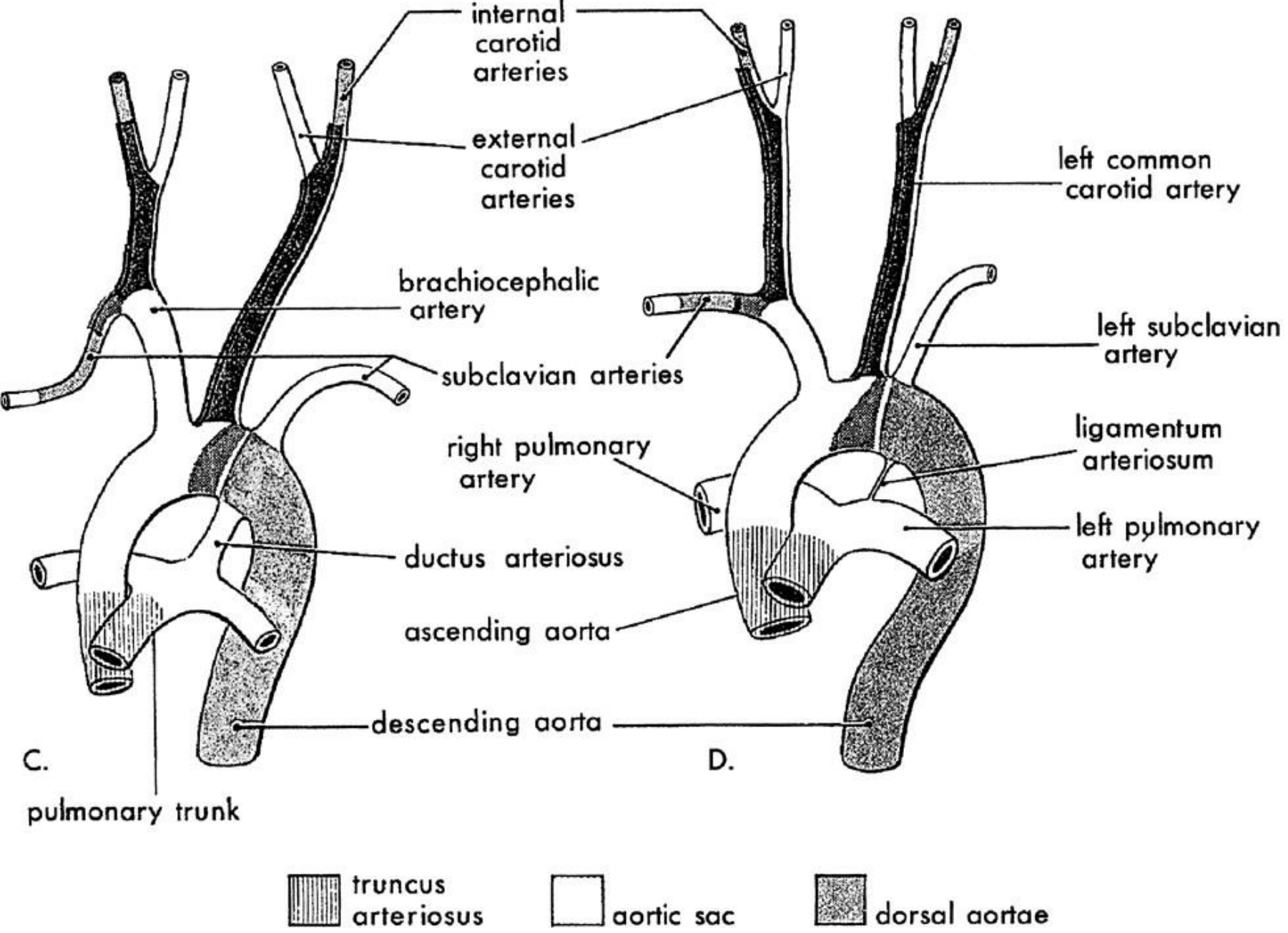


Aortic arch derivatives


1
2
3
4
5
6
ductus arteriosus
left & right dorsal aortae
normal area of involution
abnormal area of involution
right subclavian artery
ductus arteriosus
left subclavian artery
region of partial involution and coarctation

(a)


3rd aortic arch artery
4th aortic arch artery
6th aortic arch artery
1
2
left dorsal aorta
external carotid artery
aortic arches
4
5
6
truncus arteriosus (partly divided into aortic and pulmonary channels)
aortic sac
right subclavian artery
7th intersegmental artery
A.
left dorsal aorta
internal carotid artery
aortic sac
ductus arteriosus
left dorsal aorta
pulmonary arteries
left subclavian artery
B.


internal carotid arteries
external carotid arteries
brachiocephalic artery
subclavian arteries
right pulmonary artery
ductus arteriosus
ascending aorta
descending aorta
pulmonary trunk
C.
left common carotid artery
left subclavian artery
ligamentum arteriosum
left pulmonary artery
D.
truncus arteriosus
aortic sac
dorsal aortae

Prenatale diagnostiek coarctatio aortae

Table 1. Prenatal detection per category of congenital heart disease before and after introduction of the screening programme

Heart defect category	Before introduction of screening		After introduction of screening		Difference in prenatal detection (95% CI)	*P*-value
	Total (*n*)	Prenatal detection (%)	Total (*n*)	Prenatal detection (%)		
1. Septal defects	272	37.1	230	50.4	13.3 (4.7–21.9)	0.003
2. Valvular anomalies, biventricular heart	65	20.0	65	32.3	12.3 (–2.7 to 27.3)	0.110
3. Venous return anomalies	23	4.3	27	11.1	6.8 (–7.7 to 21.3)	0.380
4. Aortic arch anomalies	117	12.0	91	29.7	17.7 (6.6–28.8)	0.001
5. Conotruncal anomalies	267	26.6	229	59.8	33.2 (24.9–41.5)	<0.001
6. Hypoplastic right heart syndrome	22	50.0	18	66.7	16.7 (–13.5 to 46.9)	0.289
7. Hypoplastic left heart syndrome	85	54.1	82	97.6	43.5 (32.3–54.7)	<0.001
8. Other univentricular heart defects	83	57.8	78	94.9	37.1 (25.3–48.9)	<0.001
9. Complex defects with atrial isomerism	31	64.5	31	93.5	29.0 (10.1–47.9)	0.005
10. Miscellaneous	48	20.8	48	20.8	0	0.100
Total	1013	35.8	899	59.7	23.9 (19.5–28.3)	<0.001
Isolated CHD	619	22.8	527	44.2	21.4 (16.0–26.8)	<0.001

Van Velzen et al.; BJOG 2016

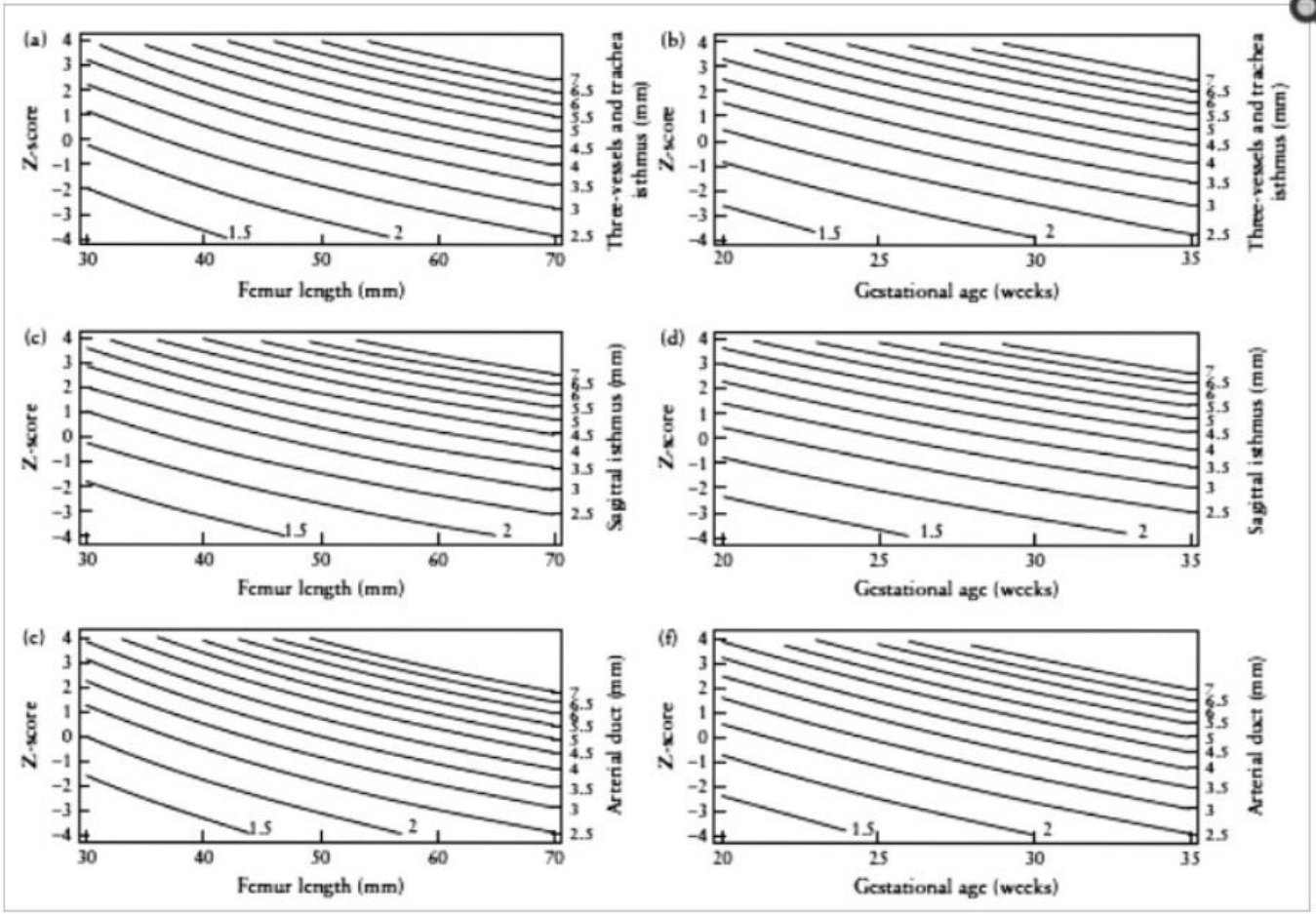
TABLE 1 Fetal echocardiographic "red flags" for coarctation of the aorta

Ventricular disproportion
RV/LV size discrepancy
Dilated RV (Z-score >2)
Hypoplastic LV (Z-score <−2)
Shone's variants (Z-score ≤−2)
Hypoplastic LV
Hypoplastic MV
Hypoplastic AV
Great vessel disproportion
Main pulmonary artery/ascending aorta ratio ≥1.7
Abnormal aortic valve
Bicuspid or dysplastic
Abnormal aortic arch dimensions and anatomy
Aortic isthmus Z-score <−2
Distal transverse arch Z-score <−2
Posterior shelf at aortic isthmus
Distal displacement of left subclavian artery
Small carotid-subclavian artery index
Abnormal isthmus/ductal relationship
IST:PDA ratio <0.7
Abnormal IST to PDA angle
Doppler abnormalities
Flow turbulence, persistence of flow into diastole, or reversal in distal transverse arch/isthmus
Abnormal growth of aortic and arch dimensions during serial evaluations
Aortic valve growth ≤0.24 mm/wk
Aortic isthmus growth ≤0.13 mm/wk

Kailin et al.; Echocardiography 2017

PMC full text: Facts Views Vis Obgyn. 2012; 4(4): 230–236.

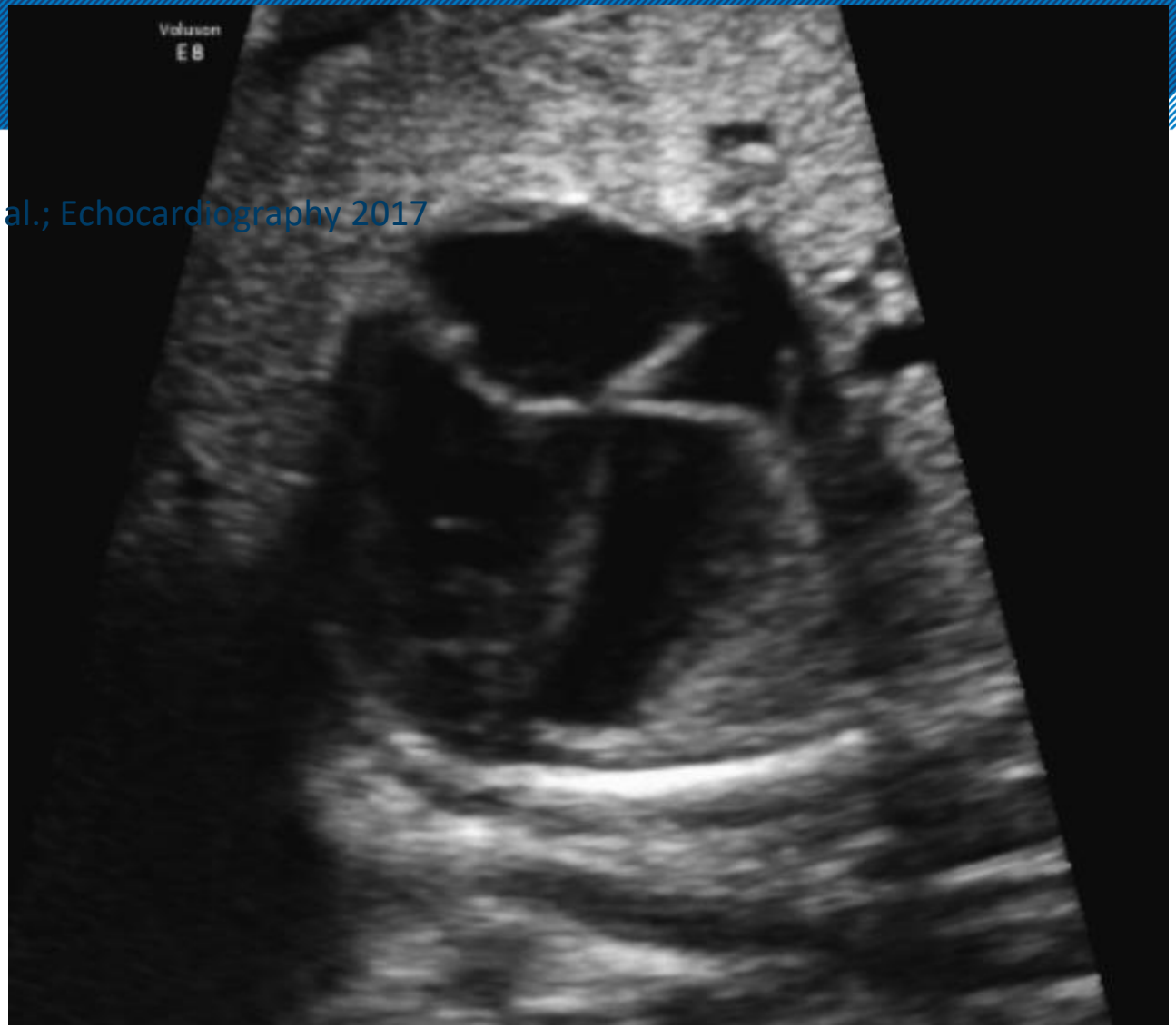
Fig. 4



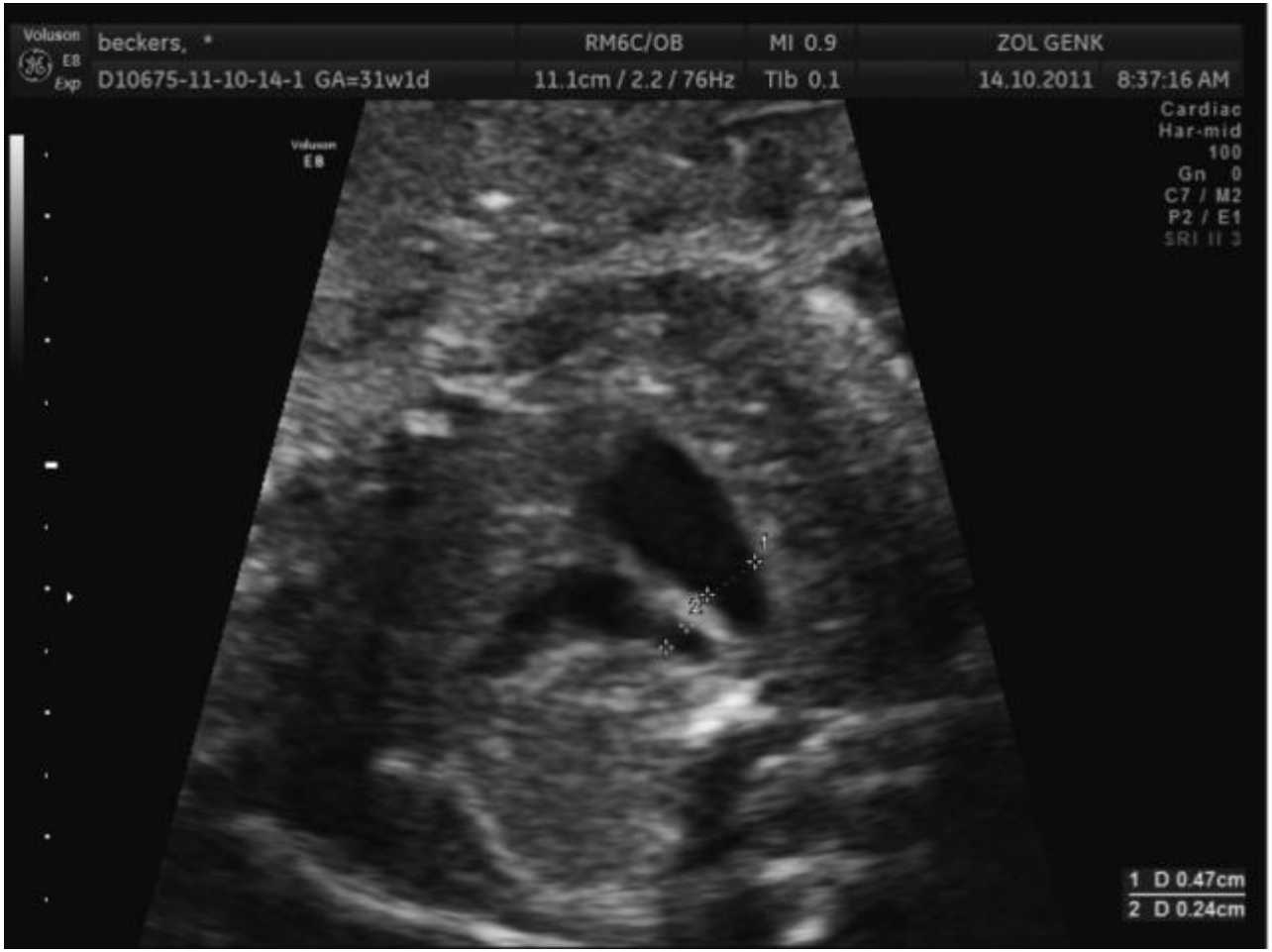
Graphical display of the z-scores for isthmal diameter in the three vessels and trachea view based on femur length (a) and on gestational age (b), for isthmal diameter in the sagittal view based on femur length (c) and on gestational age (d), and for ductal diameter in the three vessels

Kailin et al.; Echocardiography 2017

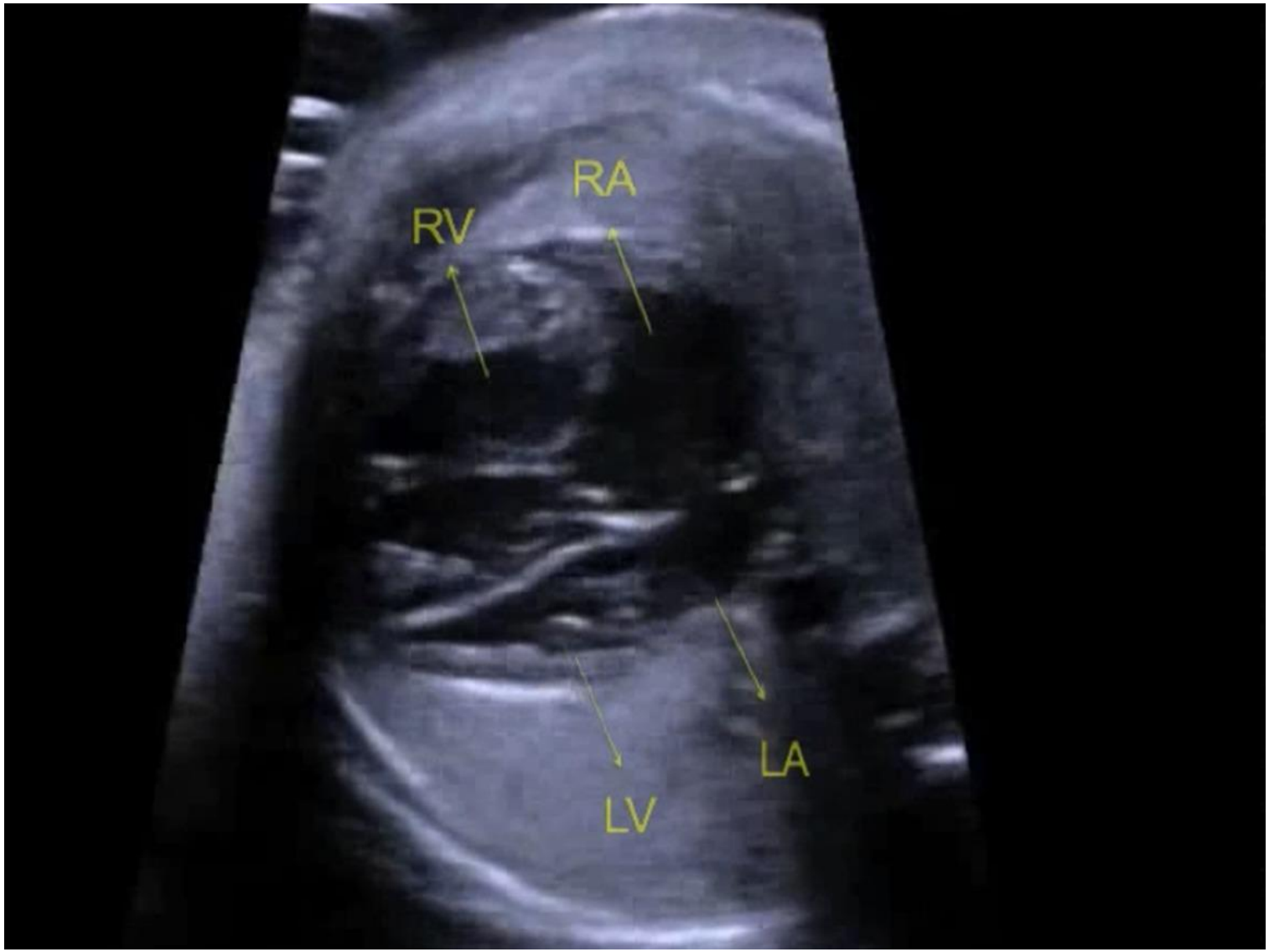
Kailin et al.; Echocardiography 2017



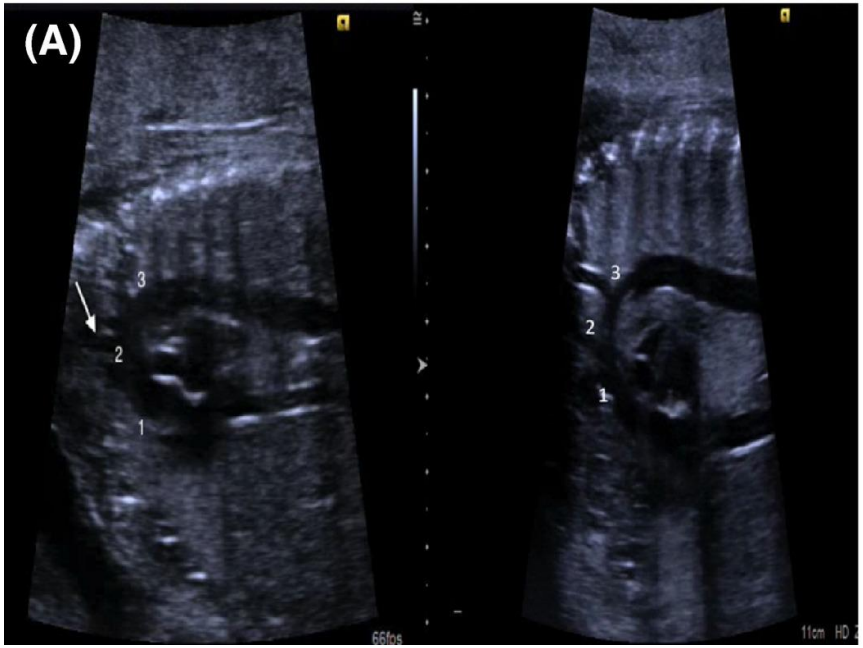
Kailin et al.; Echocardiography 2017

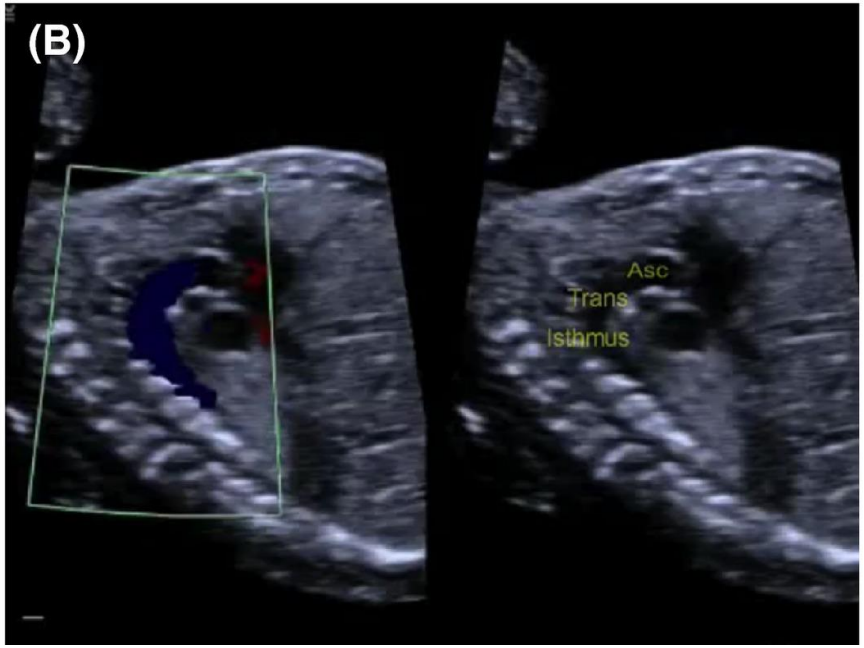


Kailin et al.; Echocardiography 2017

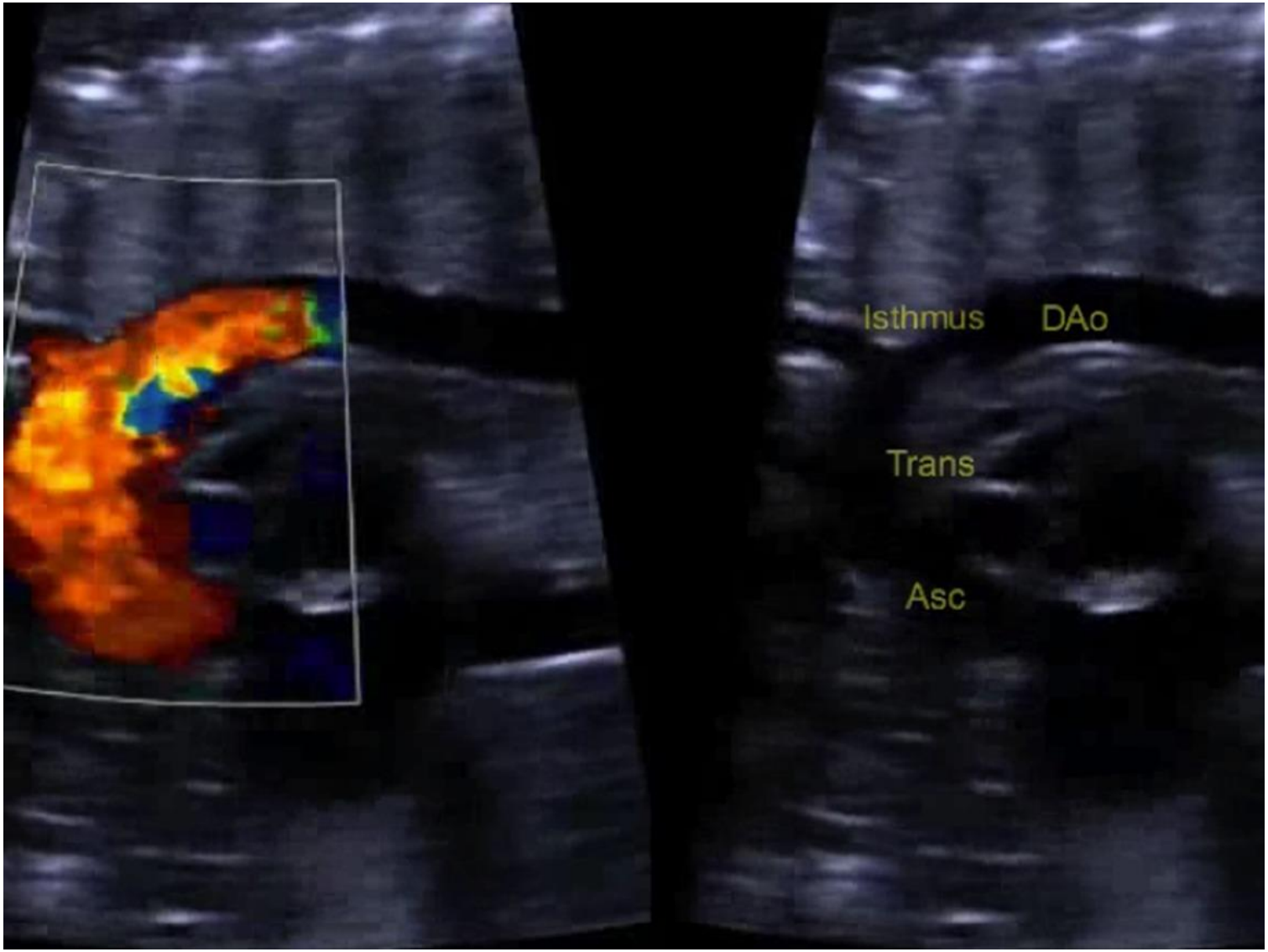


Kailin et al.; Echocardiography 2017

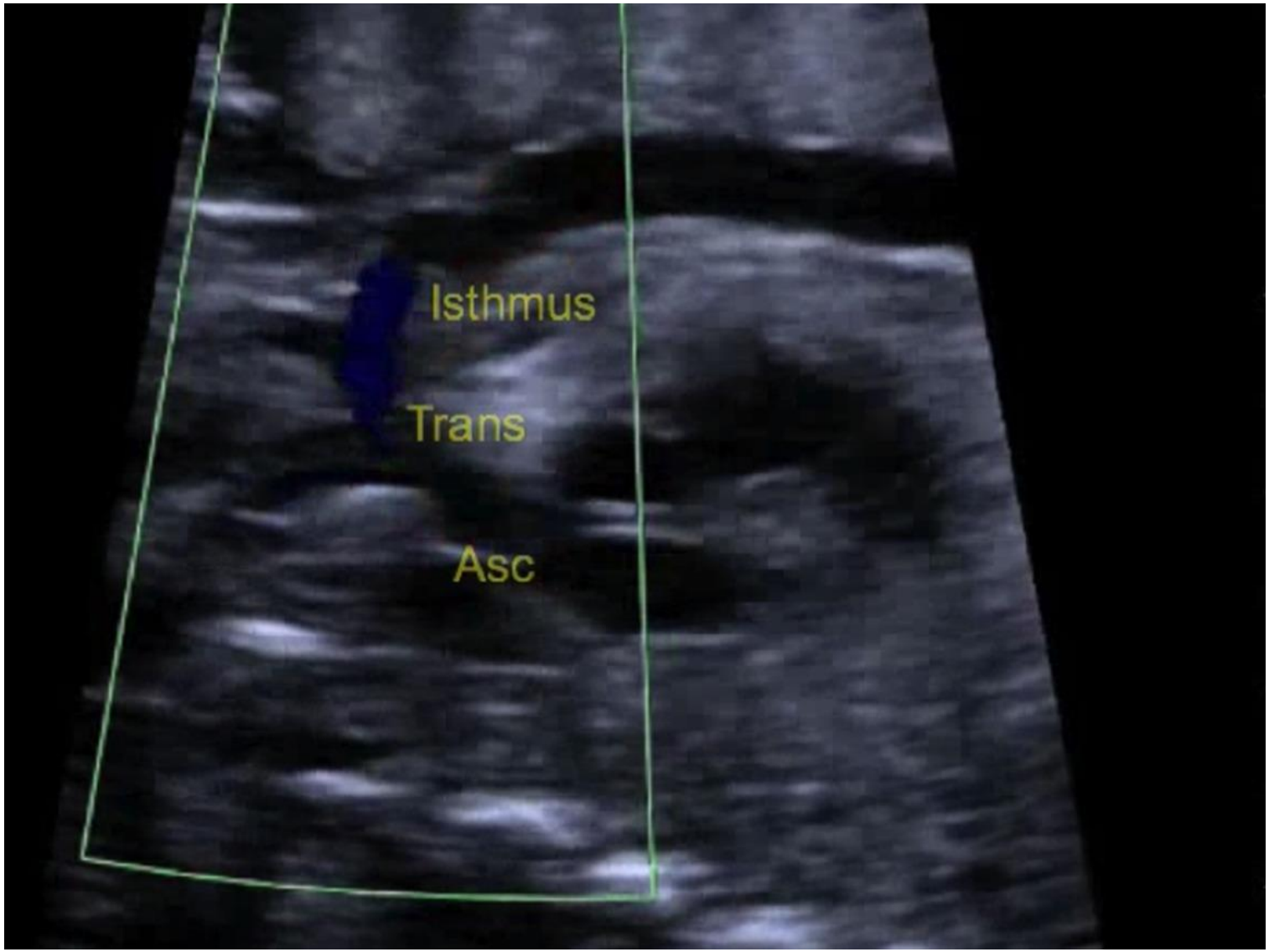




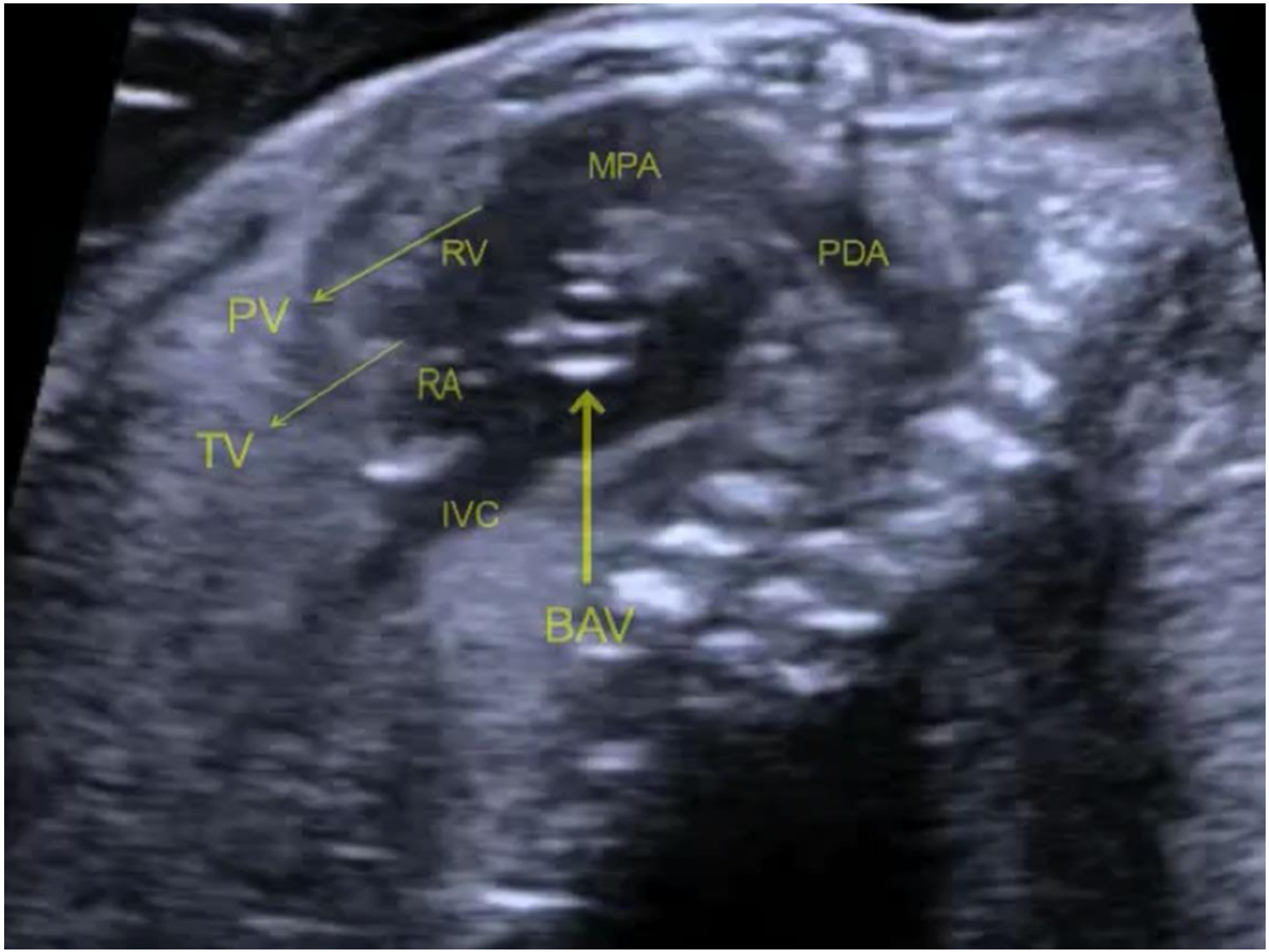
Kailin et al.; Echocardiography 2017



Kailin et al.; Echocardiography 2017

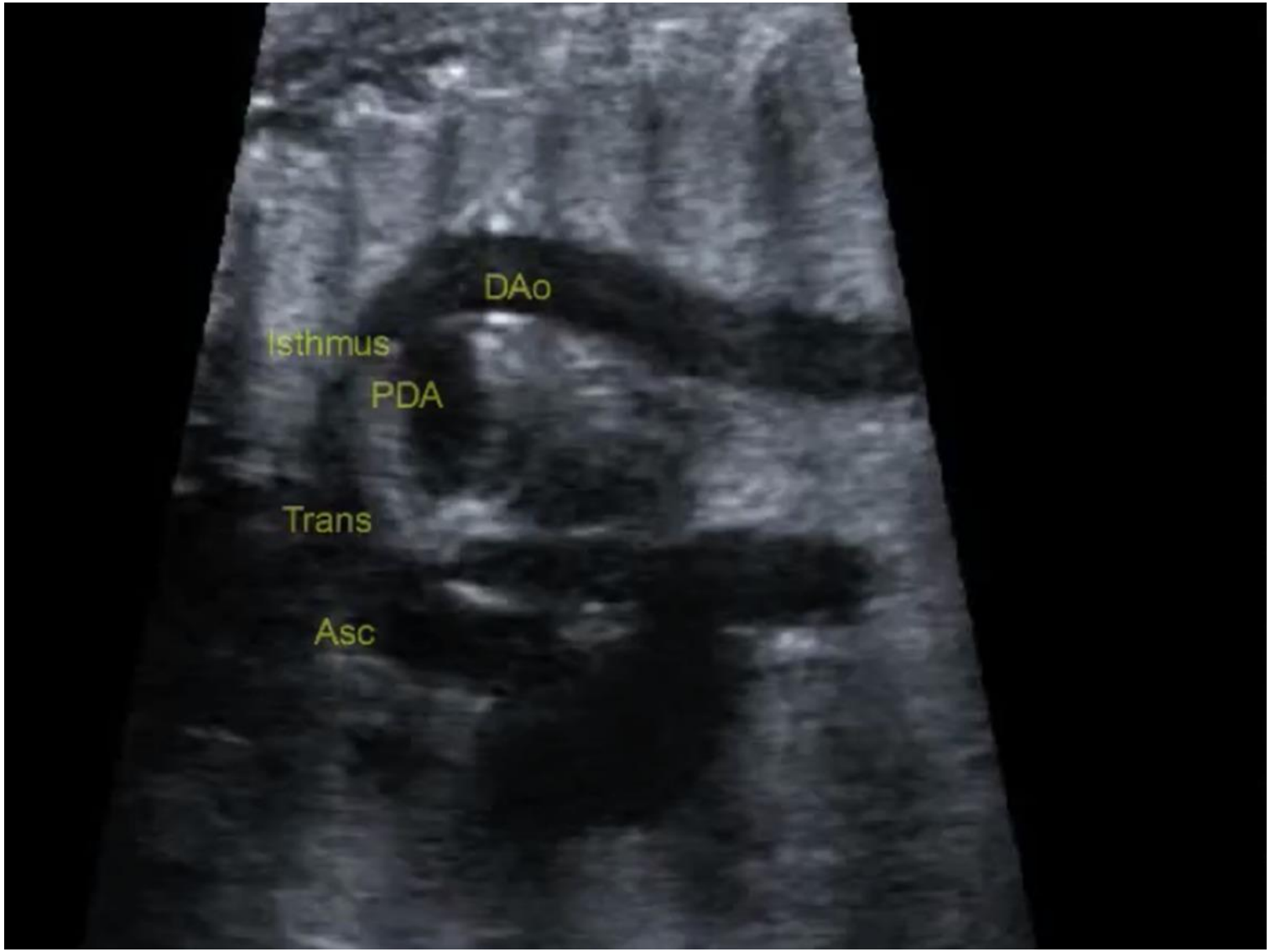


Kailin et al.; Echocardiography 2017



Kailin et al.; Echocardiography 2017

Kailin et al.; Echocardiography 2017



Kailin et al.; Echocardiography 2017

Figure 1. Case selection, inclusion and postnatal outcome

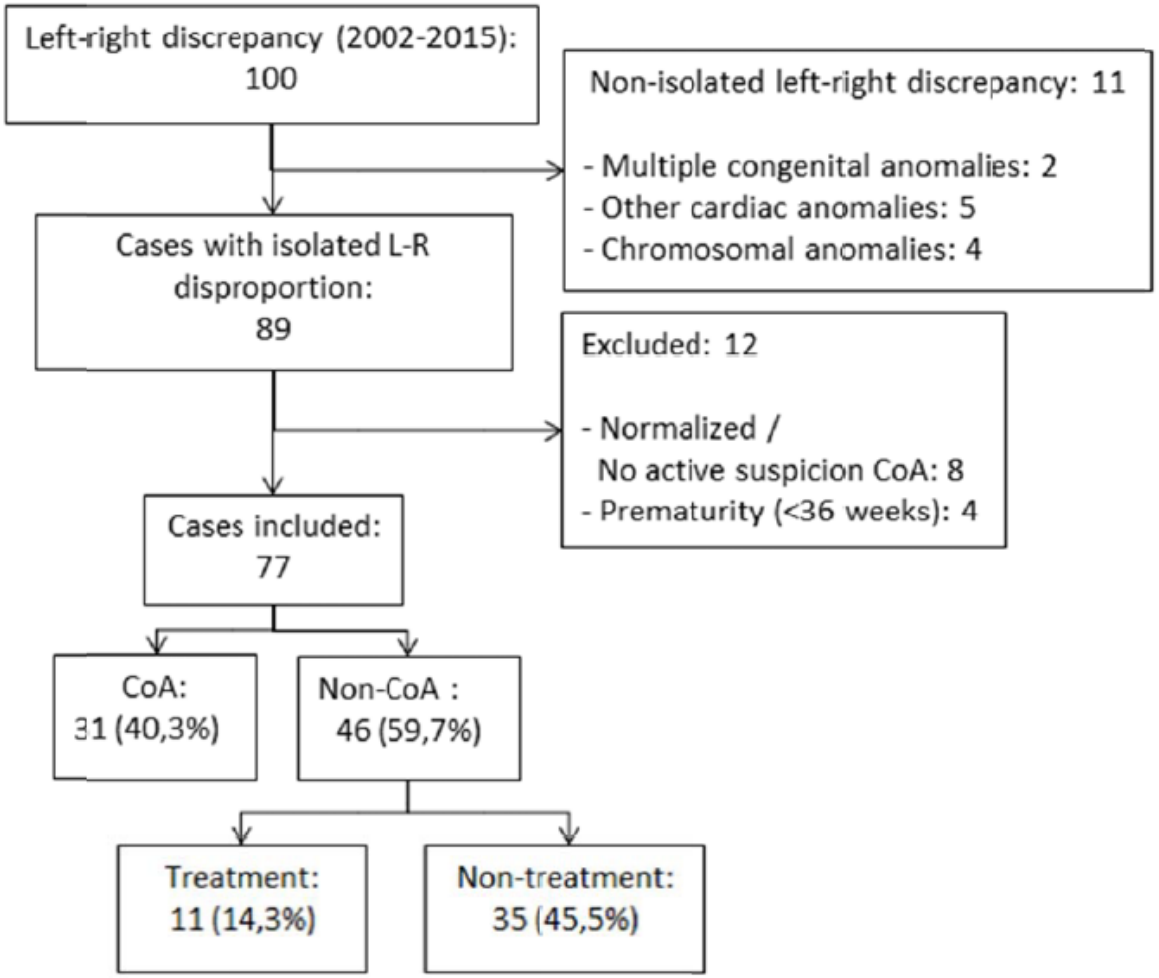
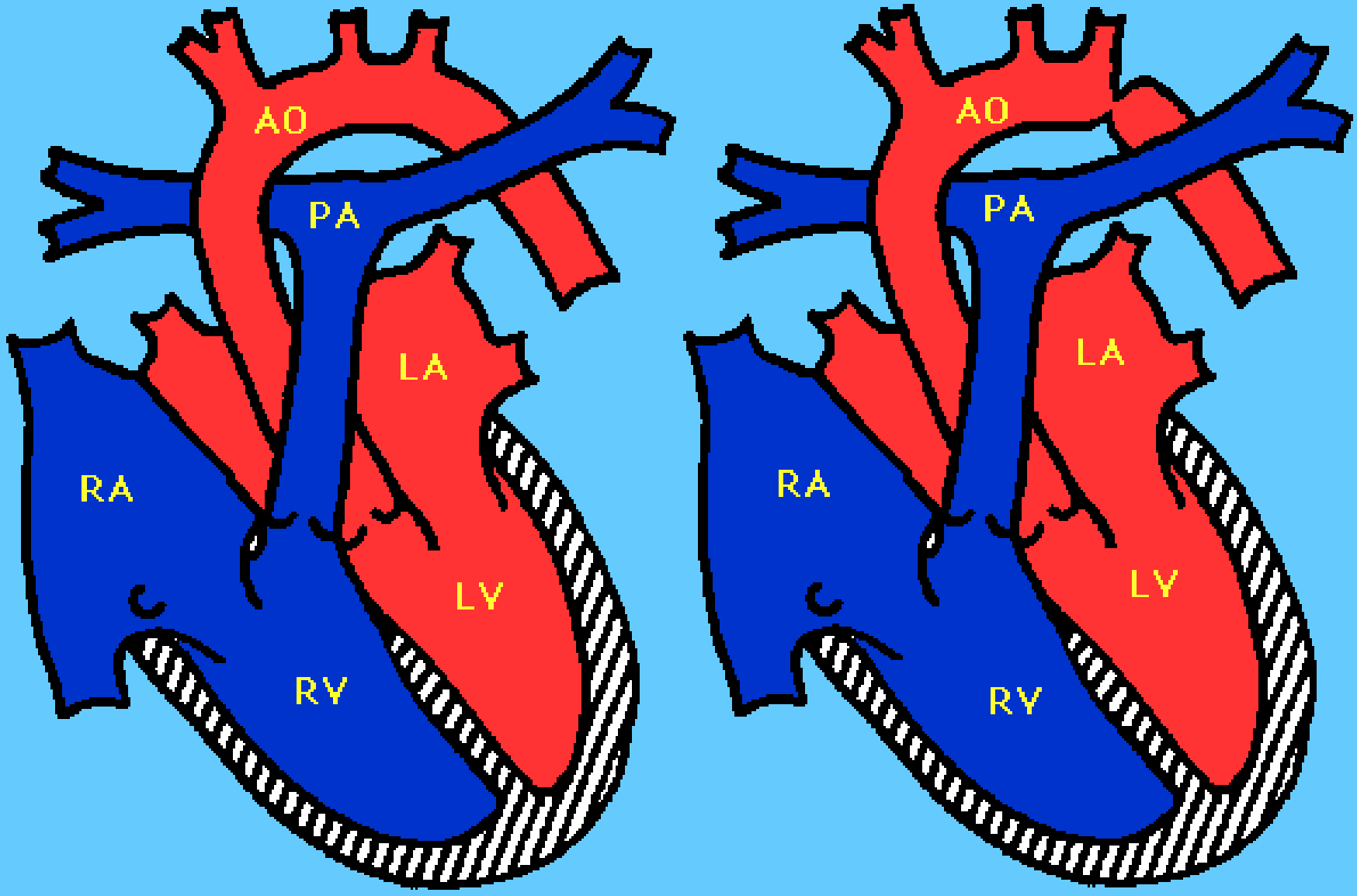


Table 1. Baseline characteristics left-right descrepancy cases				
	Non-CoA	**CoA**	***P (95% CI)***	***OR (95% CI)***
Numbers	46 (59,7)	31 (40,3)		
Sex, male	26 (56,5)	18 (58,0)	*0,89*	
Age (yrs)[#]				
Mean	4,9	4,7		
Range	[0,3-12,0]	[0,3-14,3]	*0,82* (-1,95;1,54)	
Additional prenatal findings				
Prematurely restricted or closed FO	6 (13,0)	3 (9,7)	0,65	0,74 (0,17;3,10)
PLSVC	9 (19,6)	4 (12,9)	0,44	0,61 (0,17;2,19)
Additional postnatal findings				
Bicuspid aortic valve	3 (6,5)	10 (32,3)	0,003*	6,8 (1,7;27,5)

Table 2. Clinical outcome non-CoA – No cardiovascular treatment (n=35)			
Diagnosis		**Hospital visits**	
No structural abnormalities	18	Number of admissions	43
Abnormal morphology aortic, pulmonary or tricuspid valve	6	Postnatal duration of admission (days)	
PLSVC, isolated	3	*Mean*	6,5 [2-37]
VSD + PLSVC	1	*Total*	229
ASD-II [#], isolated	1		
Genetic disorder/dysmorphic features			
Down syndrome [*]	1	Follow-up visits	
Facial dysmorphic features with thoracic scoliosis	1	*No*	25
Syndactyly with undergrowth digit IV [*]	1	**Pulmonary support**	
Humeral exostosis	1	Postnatal respiratory support	10
Other	2	PH	
		No PH	33
		Clinical PH, self-limiting	2

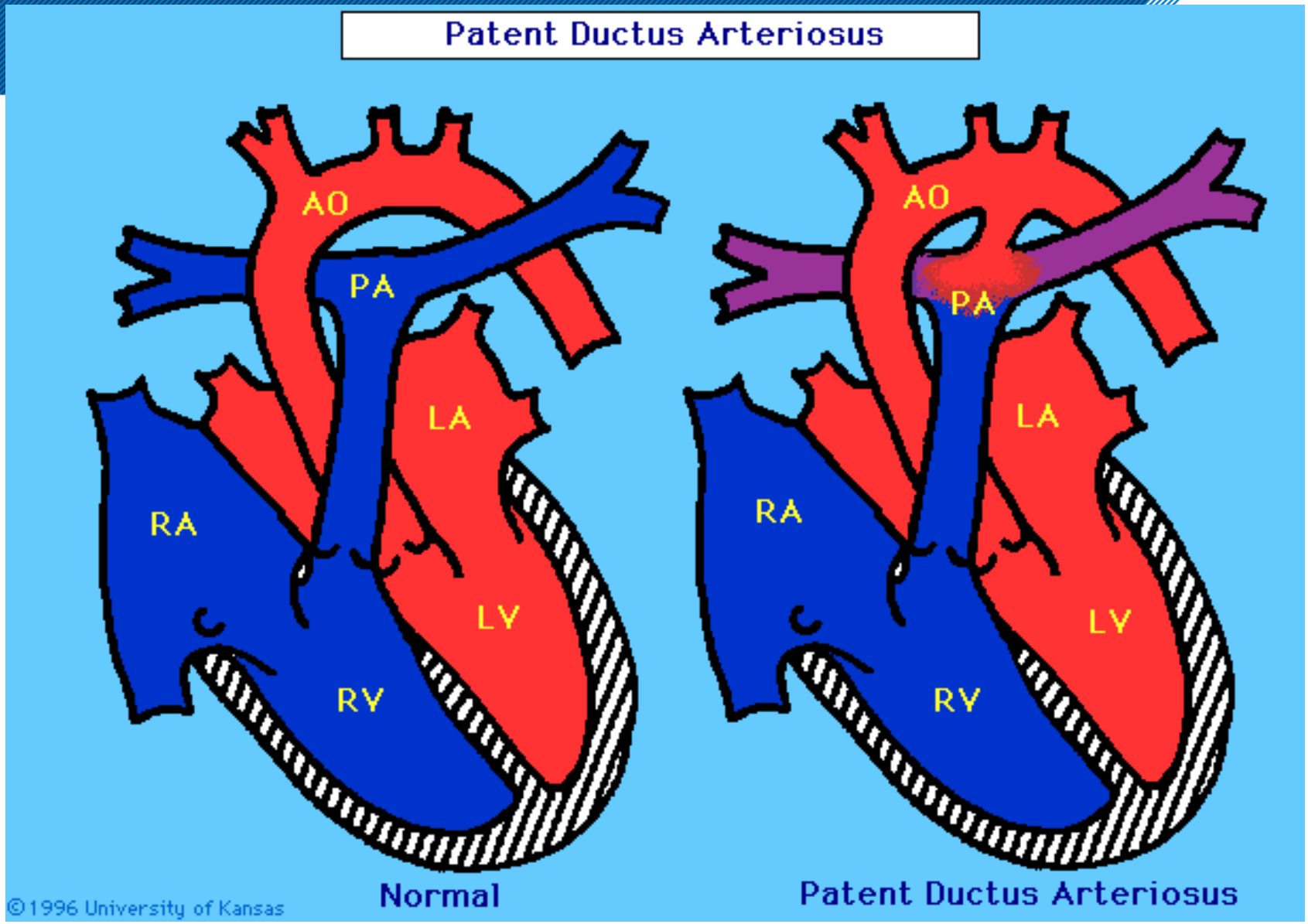
Postnatale diagnostiek coarctatio aortae

Coarctation of the Aorta



Normal

Coarctation of the Aorta


Patent Ductus Arteriosus
AO
PA
LA
RA
LV
RV
AO
PA
LA
RA
LV
RV
Normal
Patent Ductus Arteriosus
©1996 University of Kansas

Klinische presentatie

Zuigeling:

-Shock en decompensatio cordis

Oudere kinderen:

-Hypertensie bovenste lichaamshelft

-Verminderde pulsaties onderste lichaamshelft

-Cardiaal geruis

Echocardiografie bij coarctatio aortae

Exacte plaats coarctatio

Doppler signaal

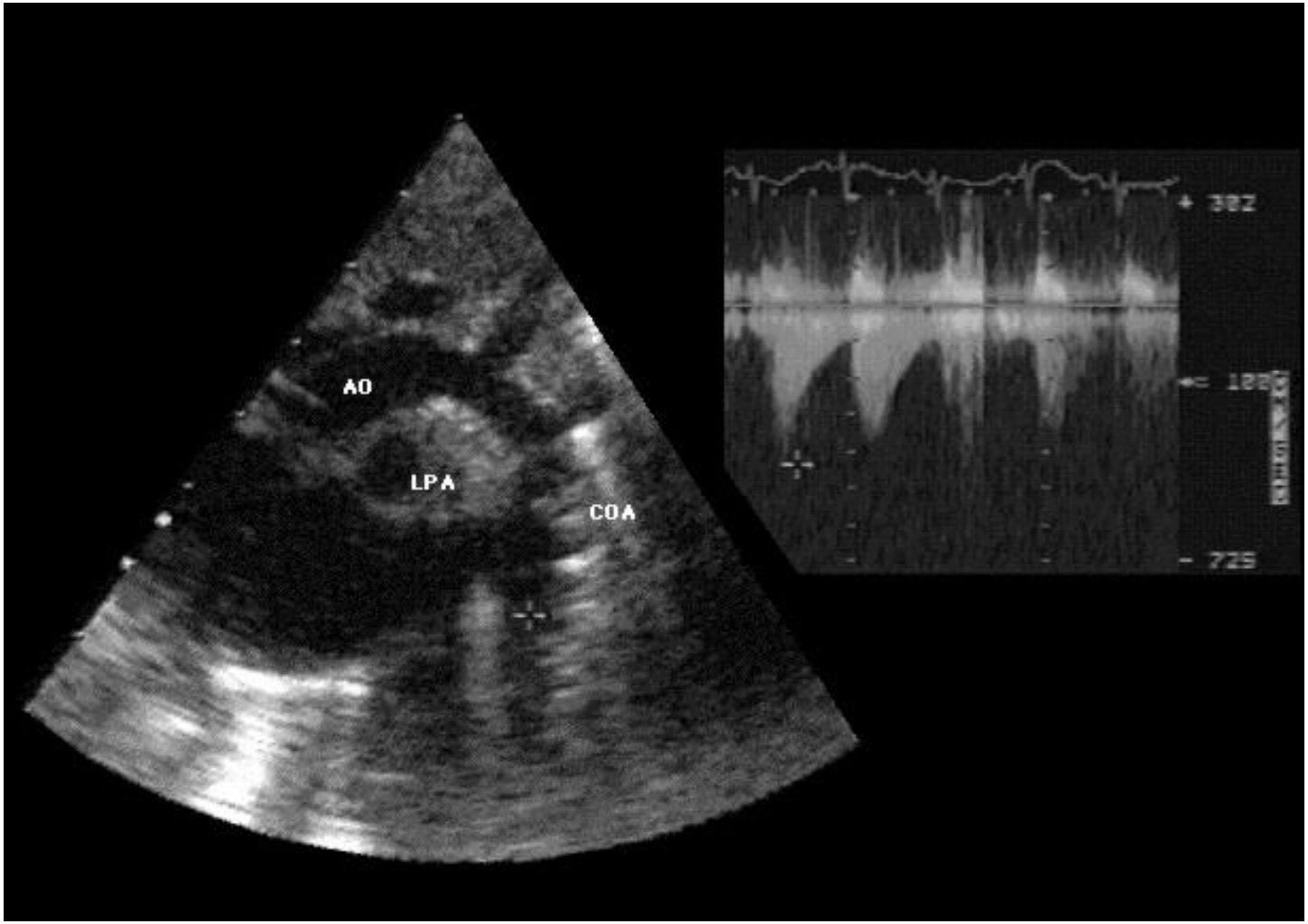
Linker ventrikelfunctie

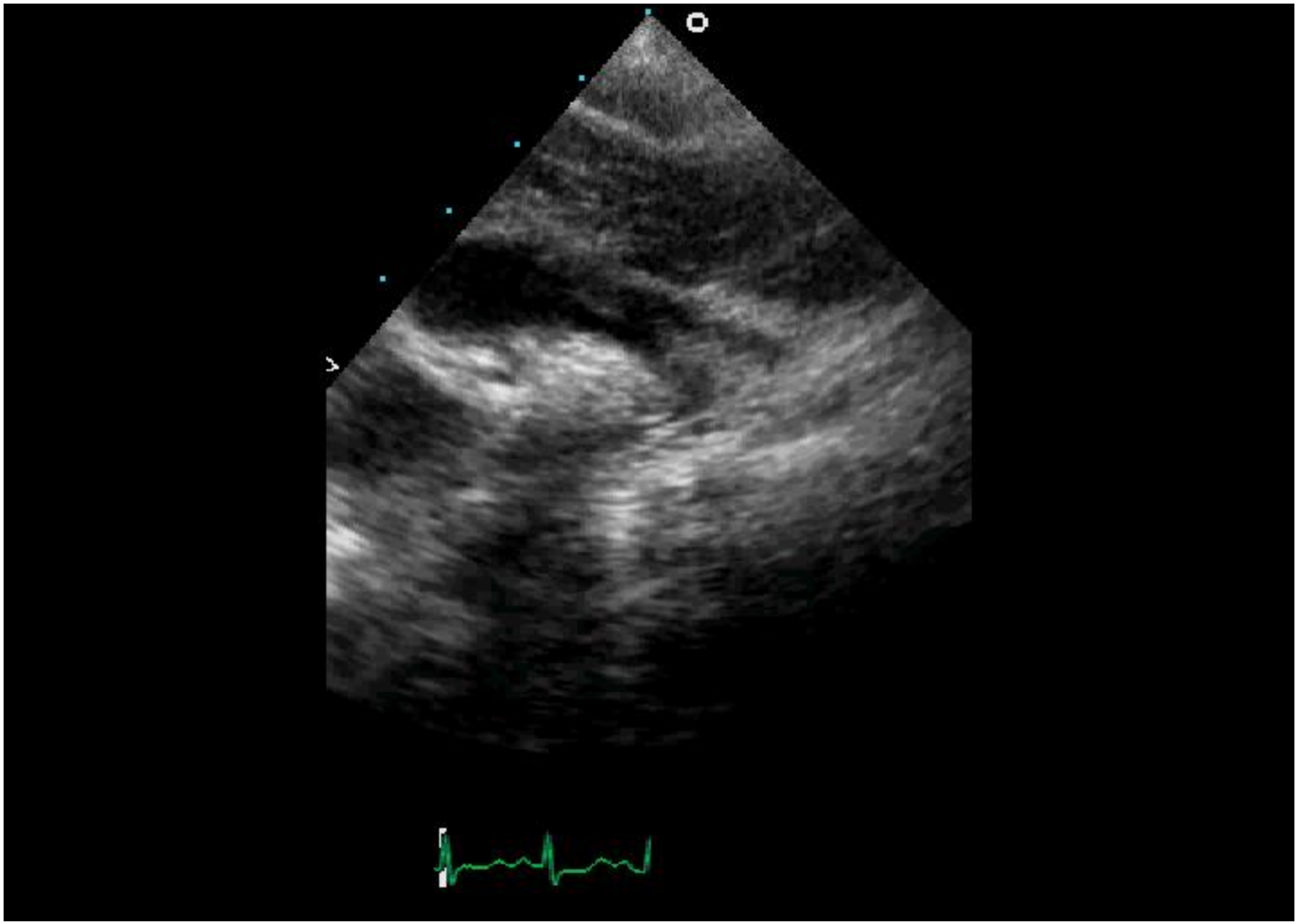
Aanwijzingen hypertrofie linker ventrikel

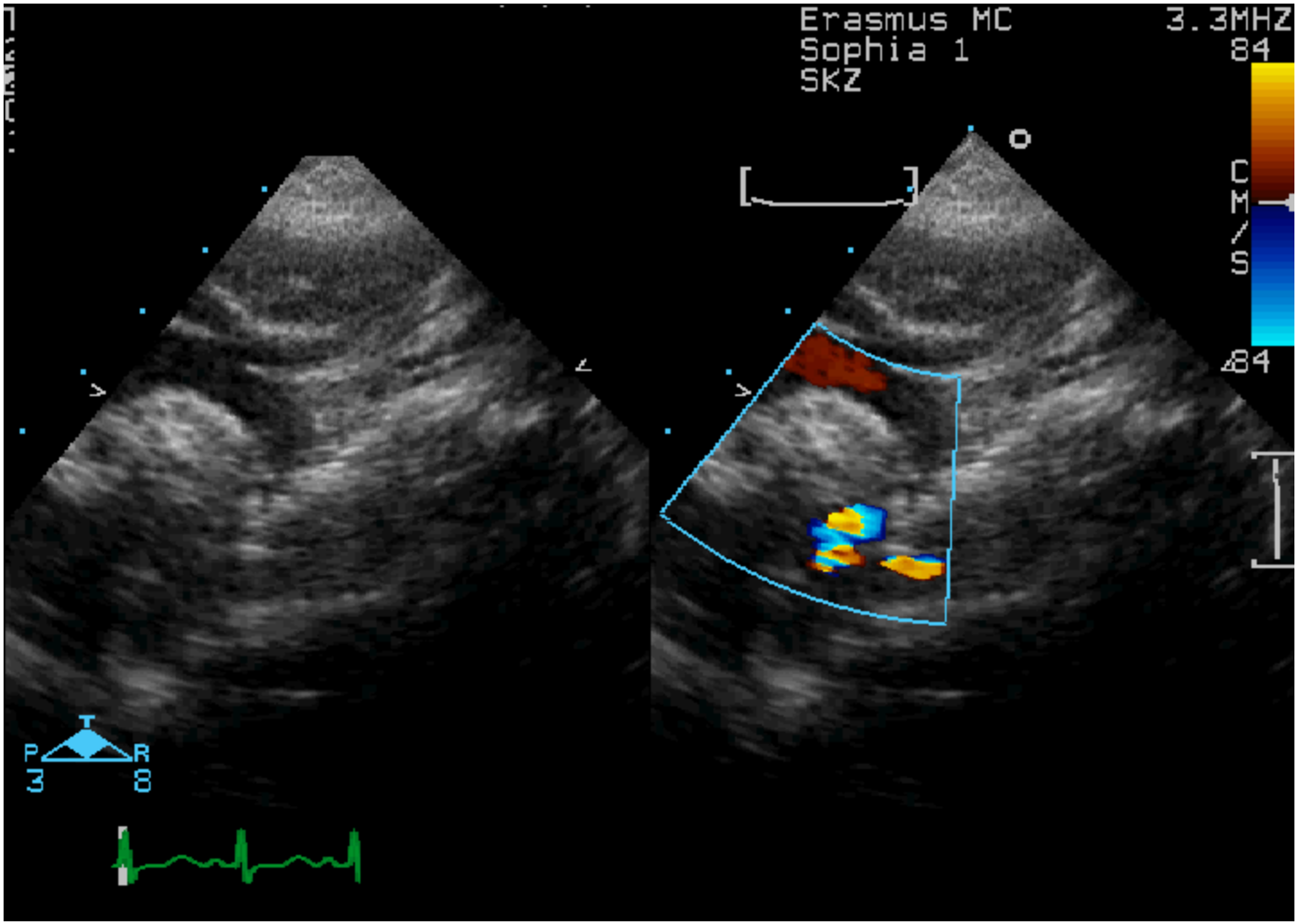
Aanwijzingen voor pulmonale hypertensie

Bijkomende afwijkingen

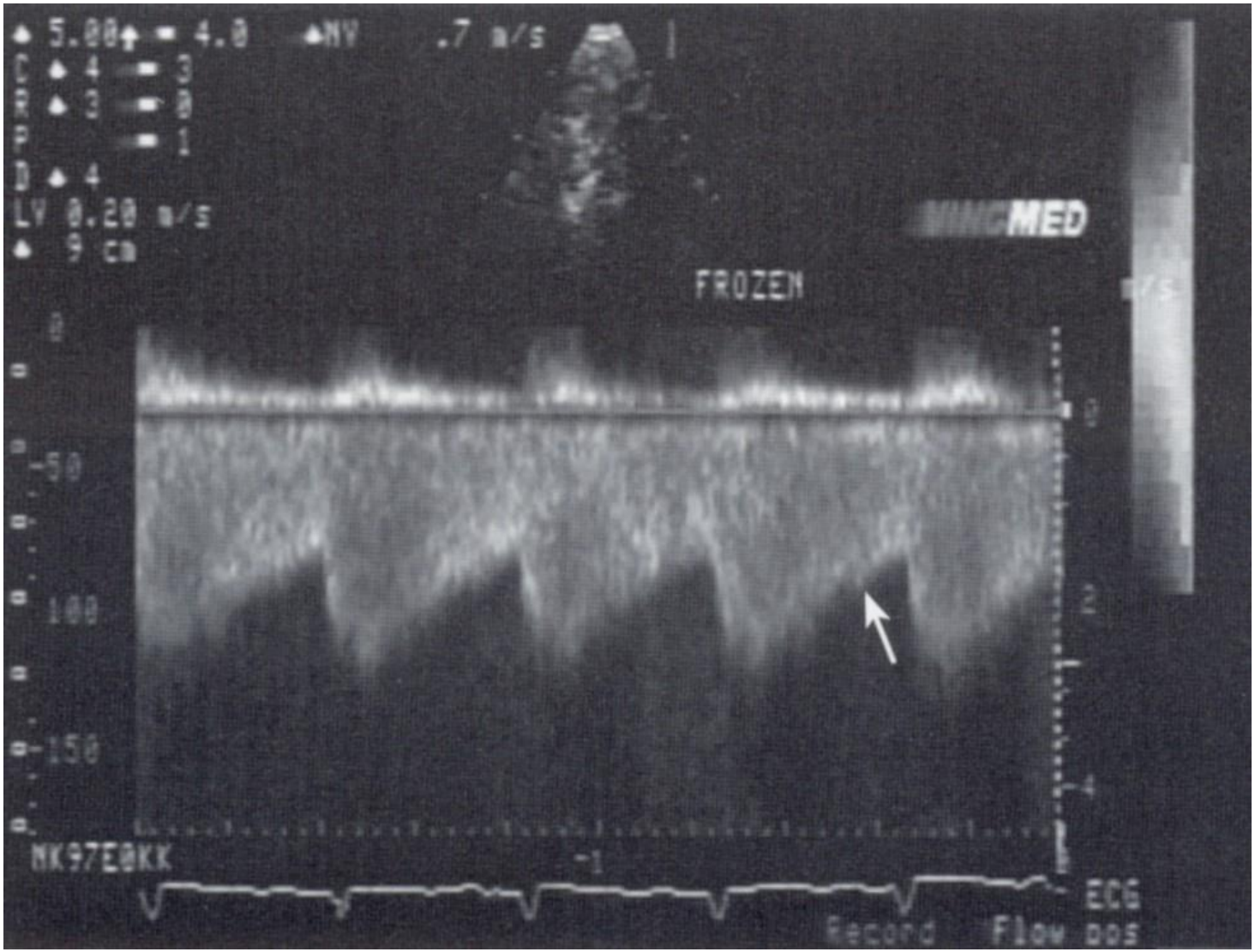
Suprasternaal



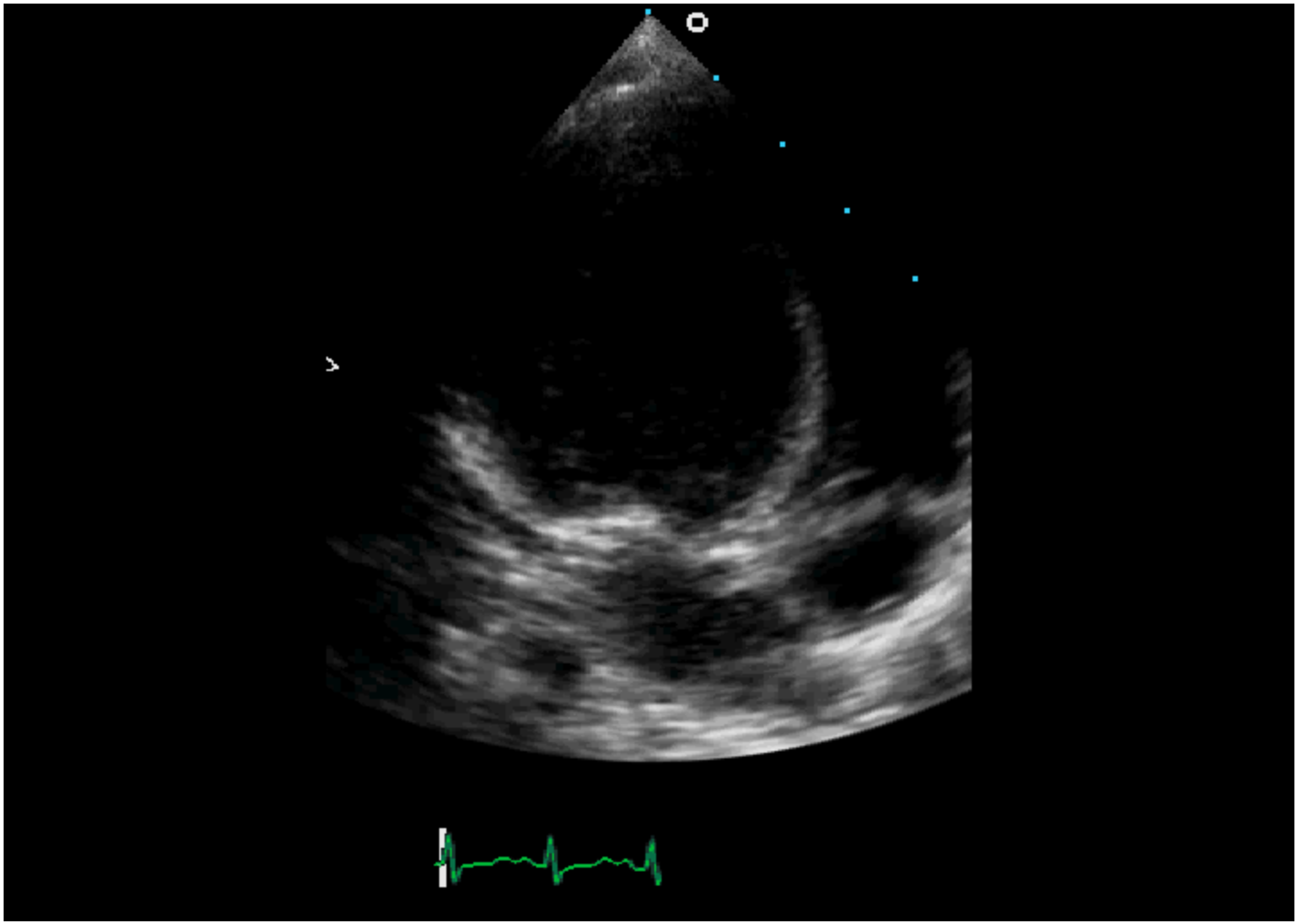



Erasmus MC
Sophia 1
SKZ
3.3MHZ
84
CM/S
84
P
T
R
3
8

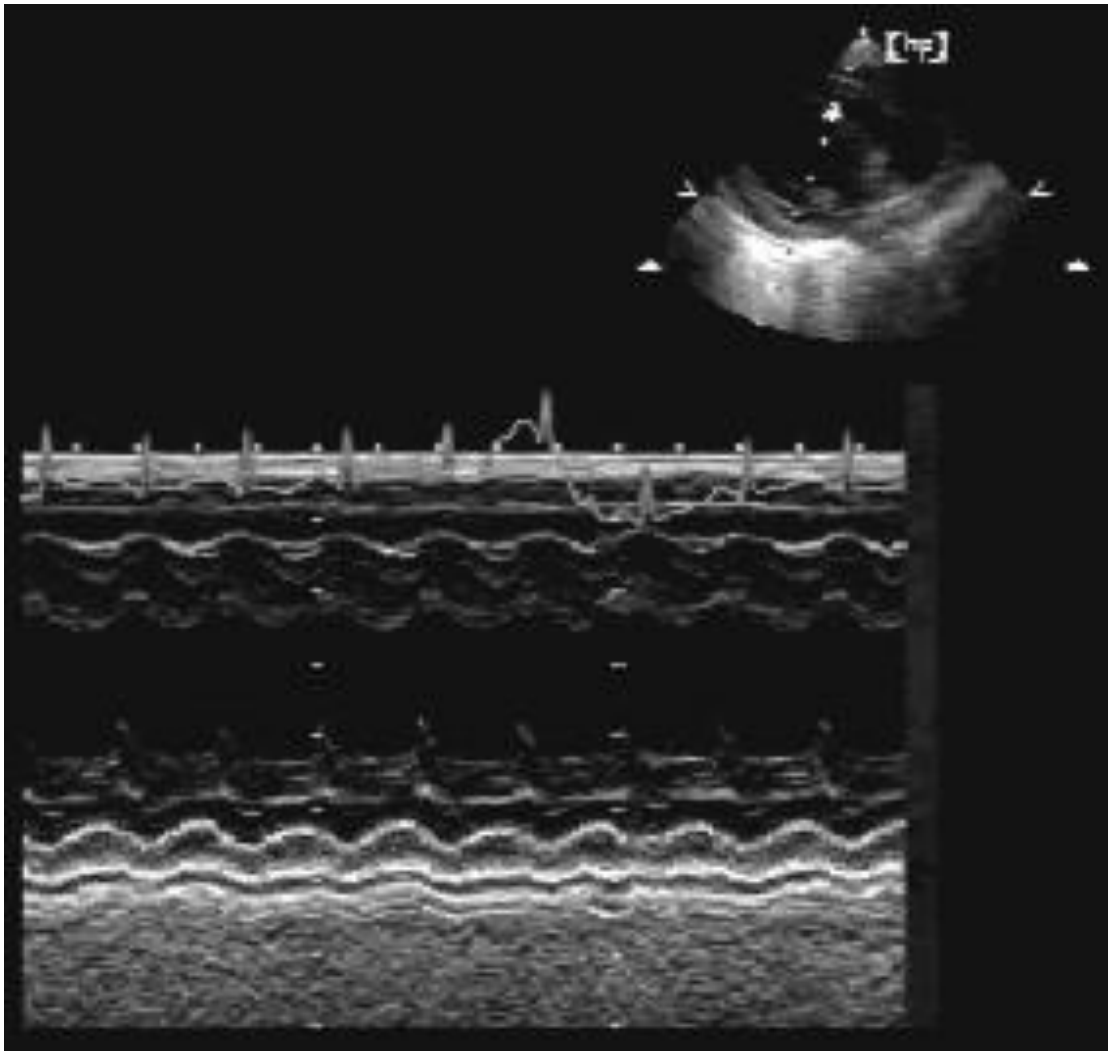
Doppler signaal



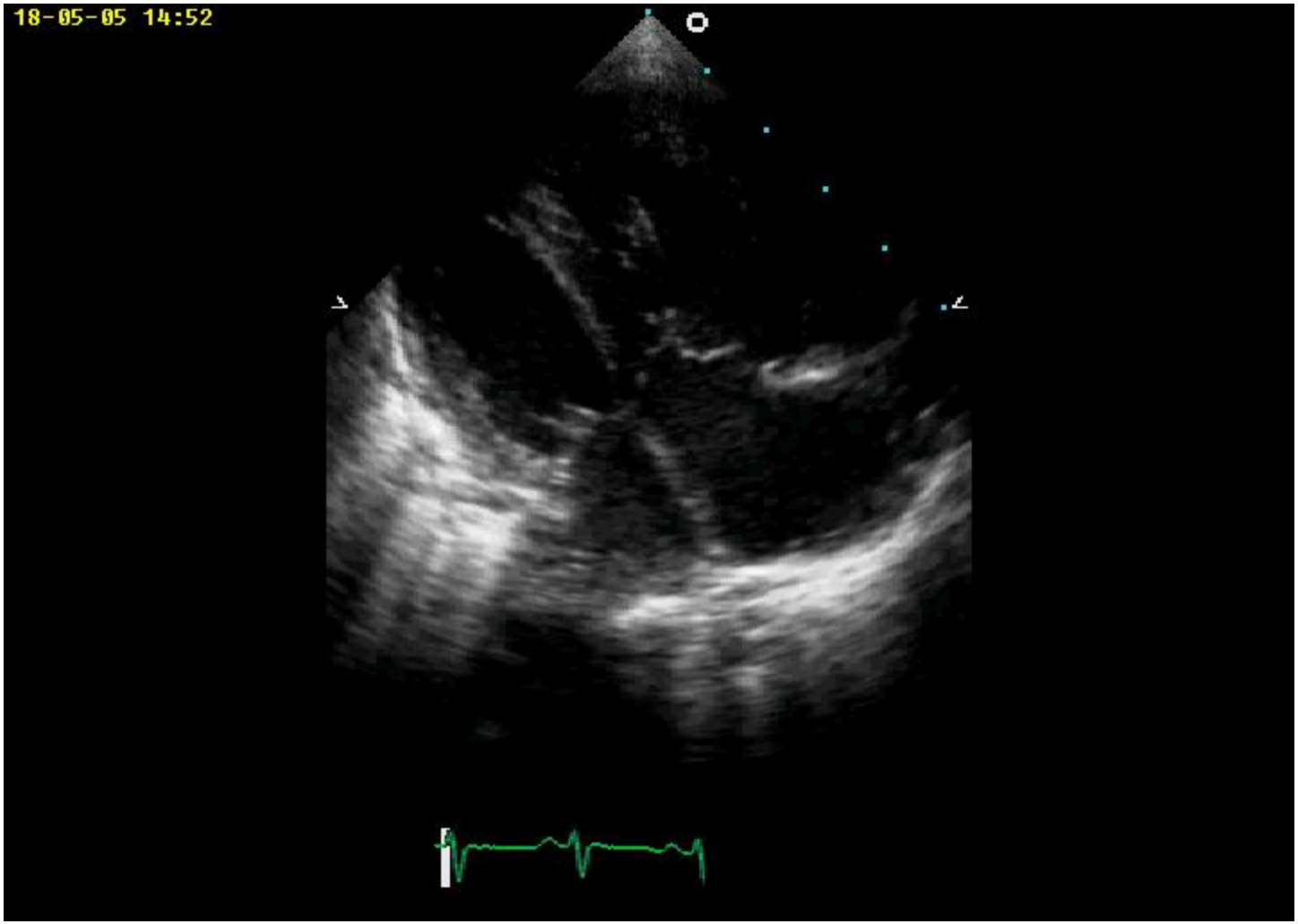
Vierkamerbeeld

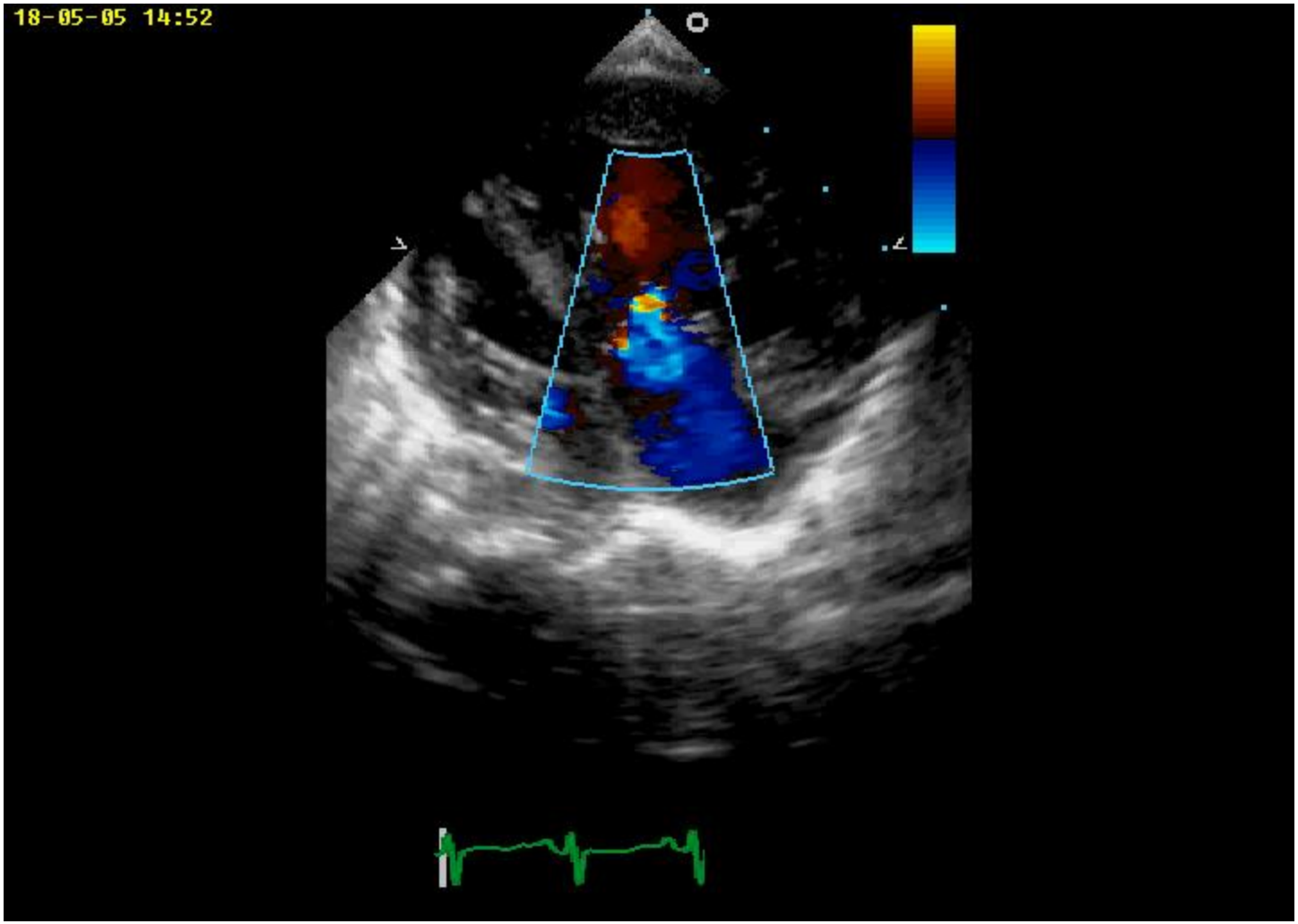


M-mode



Dilatatie rechts: PHT

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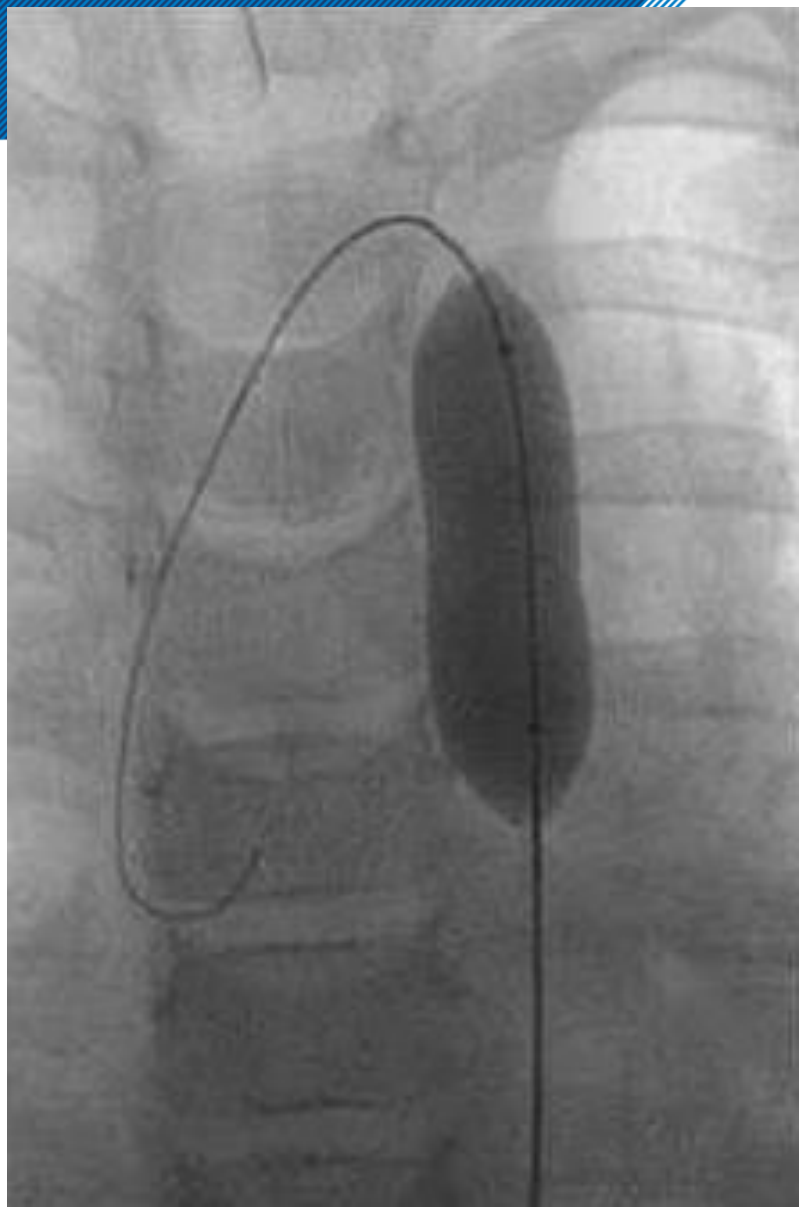
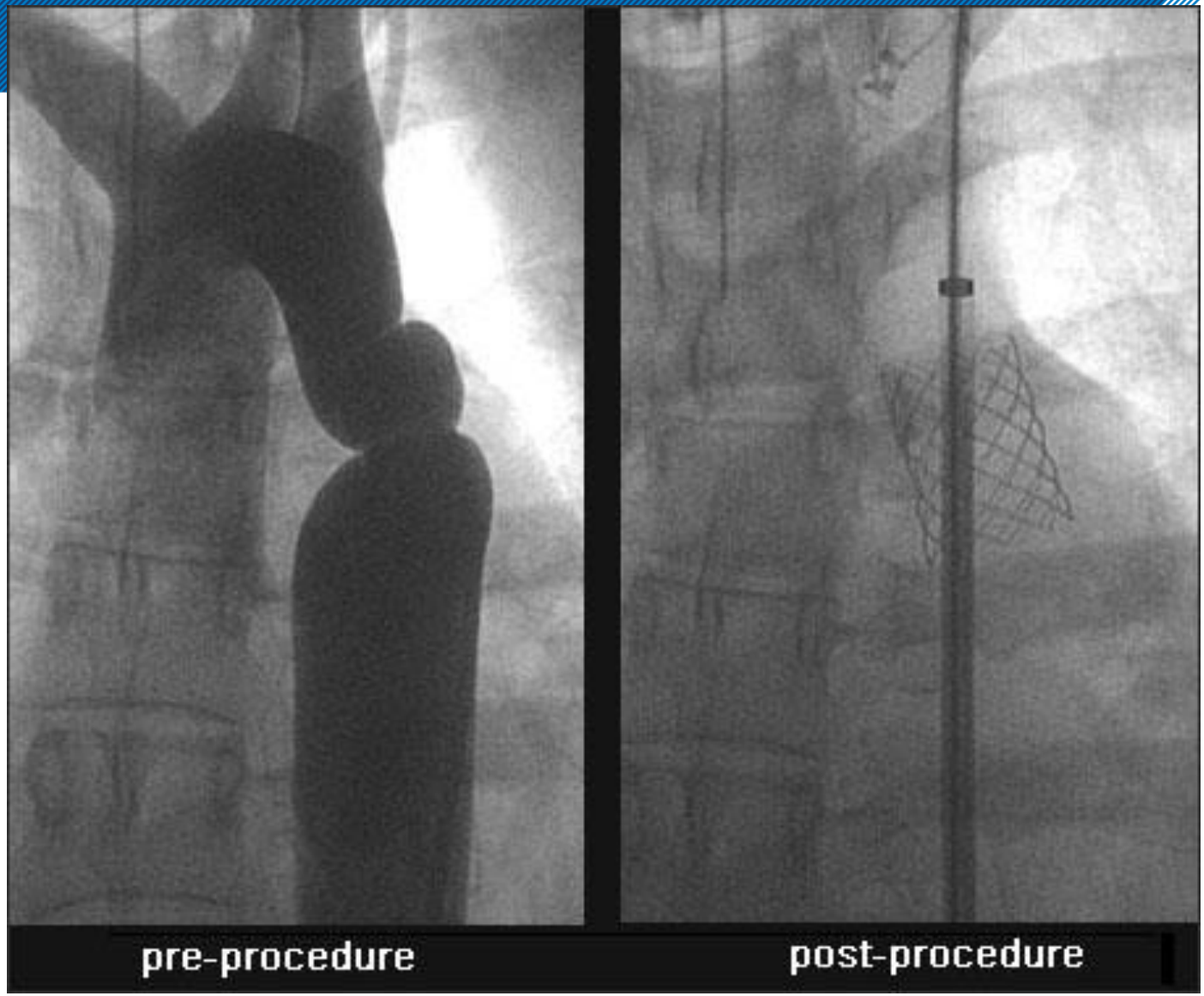



pre-procedure
post-procedure

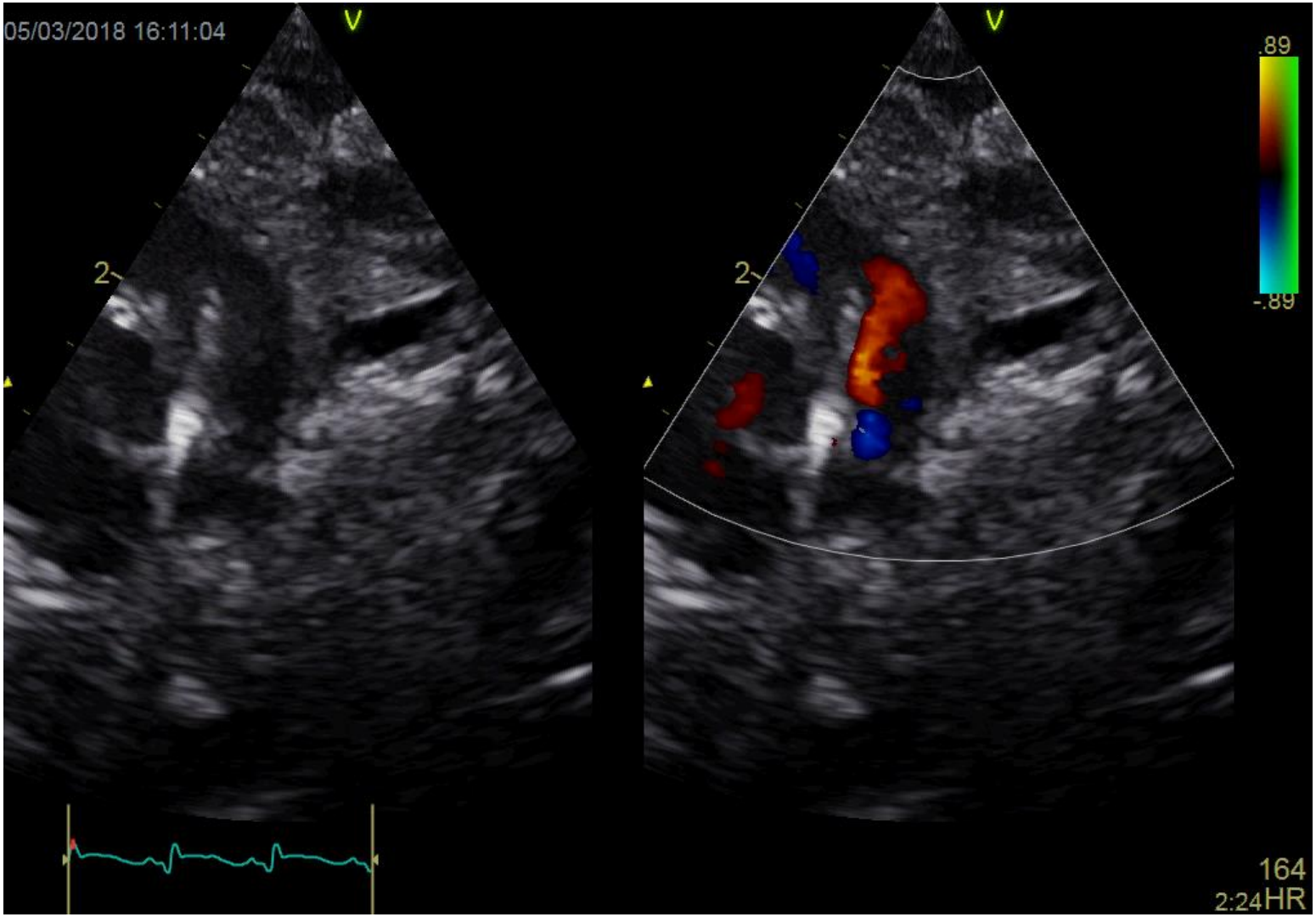
Conclusie

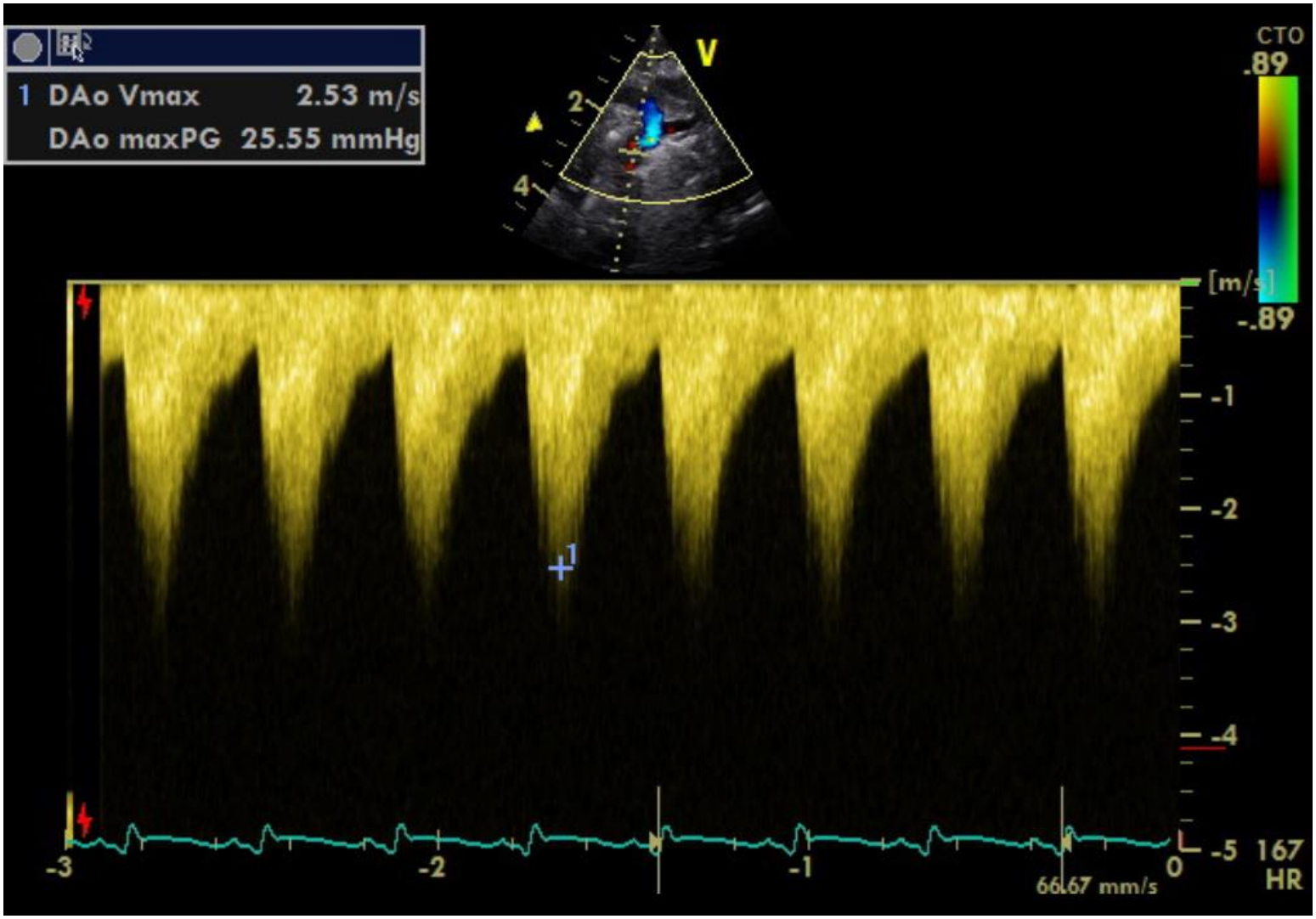
-Coarctatio aortae is een complexe afwijking

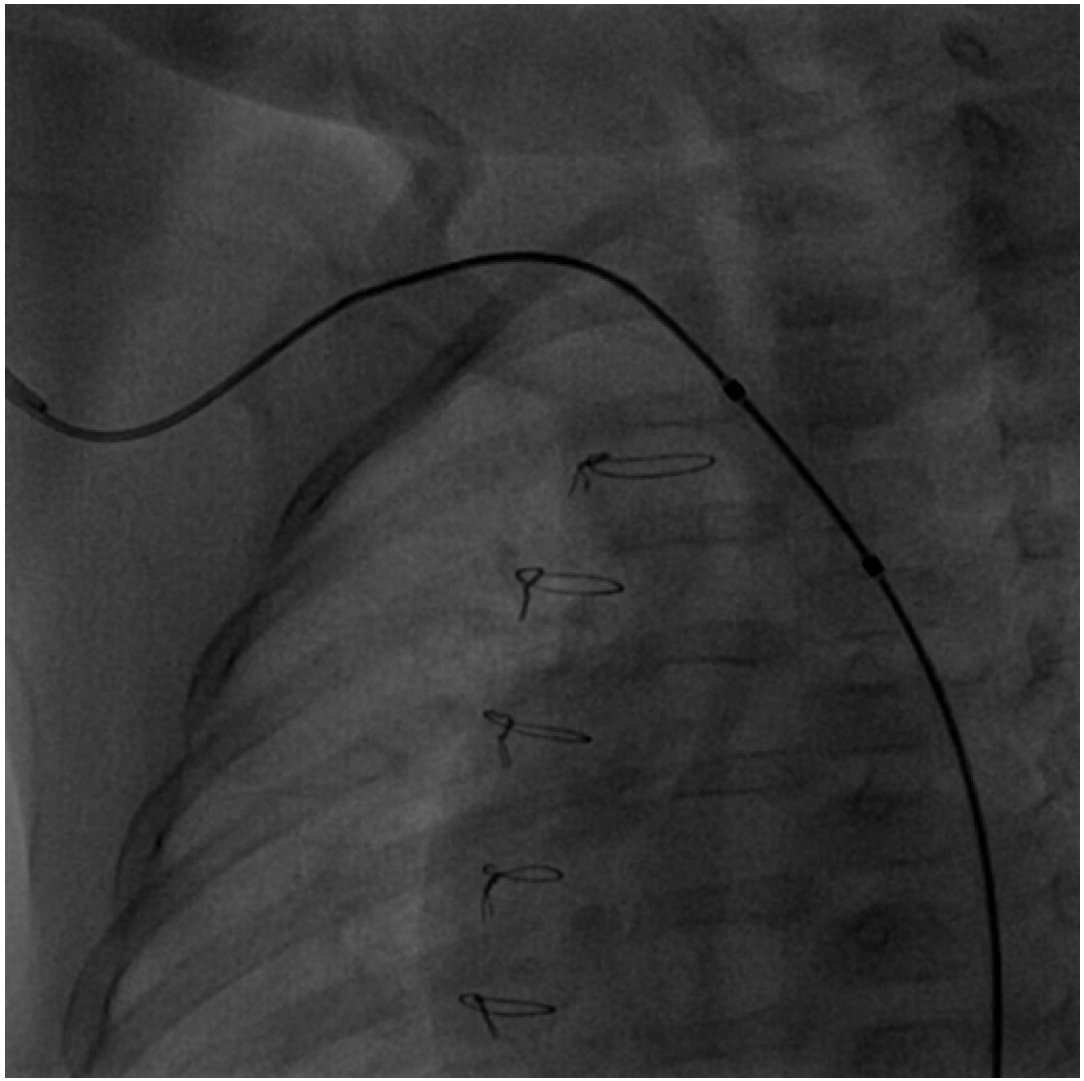
-Vaak bijkomende afwijkingen

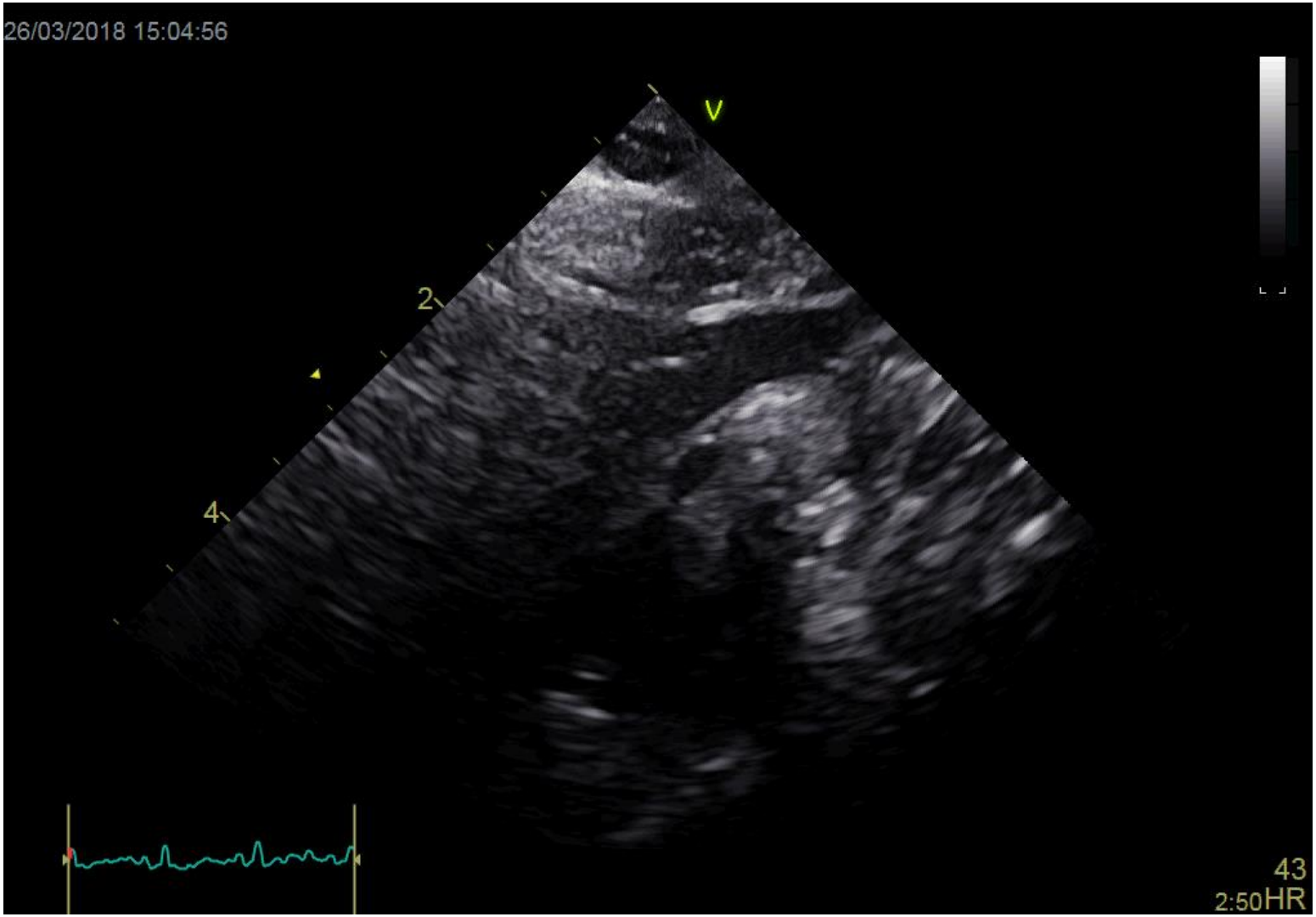
-Langdurige follow-up noodzakelijk

Patient 1


05/03/2018 16:11:04
V
2
V
2
.89
-.89
164
2:24HR


1 DAo Vmax 2.53 m/s
DAo maxPG 25.55 mmHg
V
2
4
CTO
.89
[m/s]
-.89
-1
-2
-3
-4
-5
167
HR
-3
-2
-1
0
66.67 mm/s

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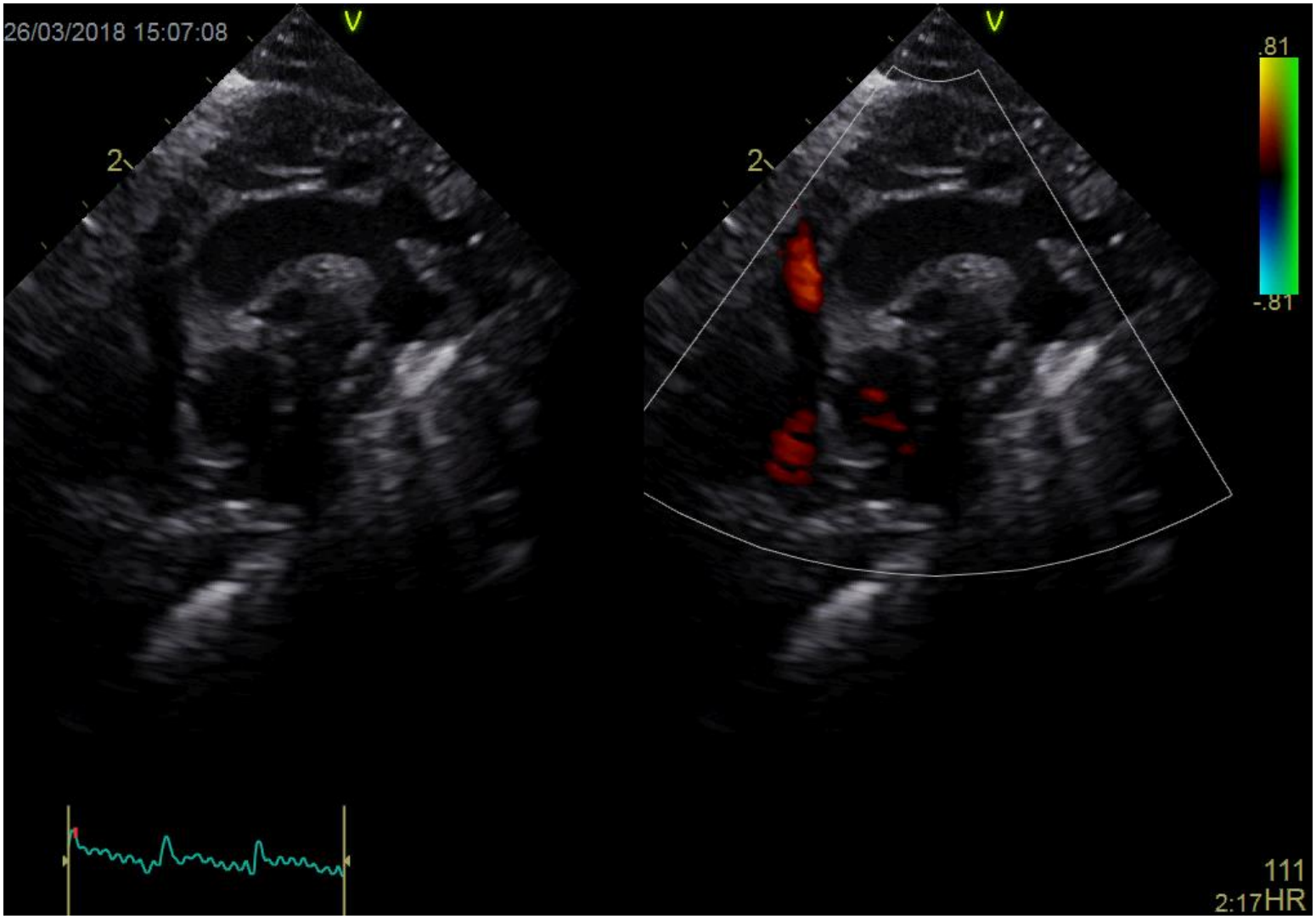
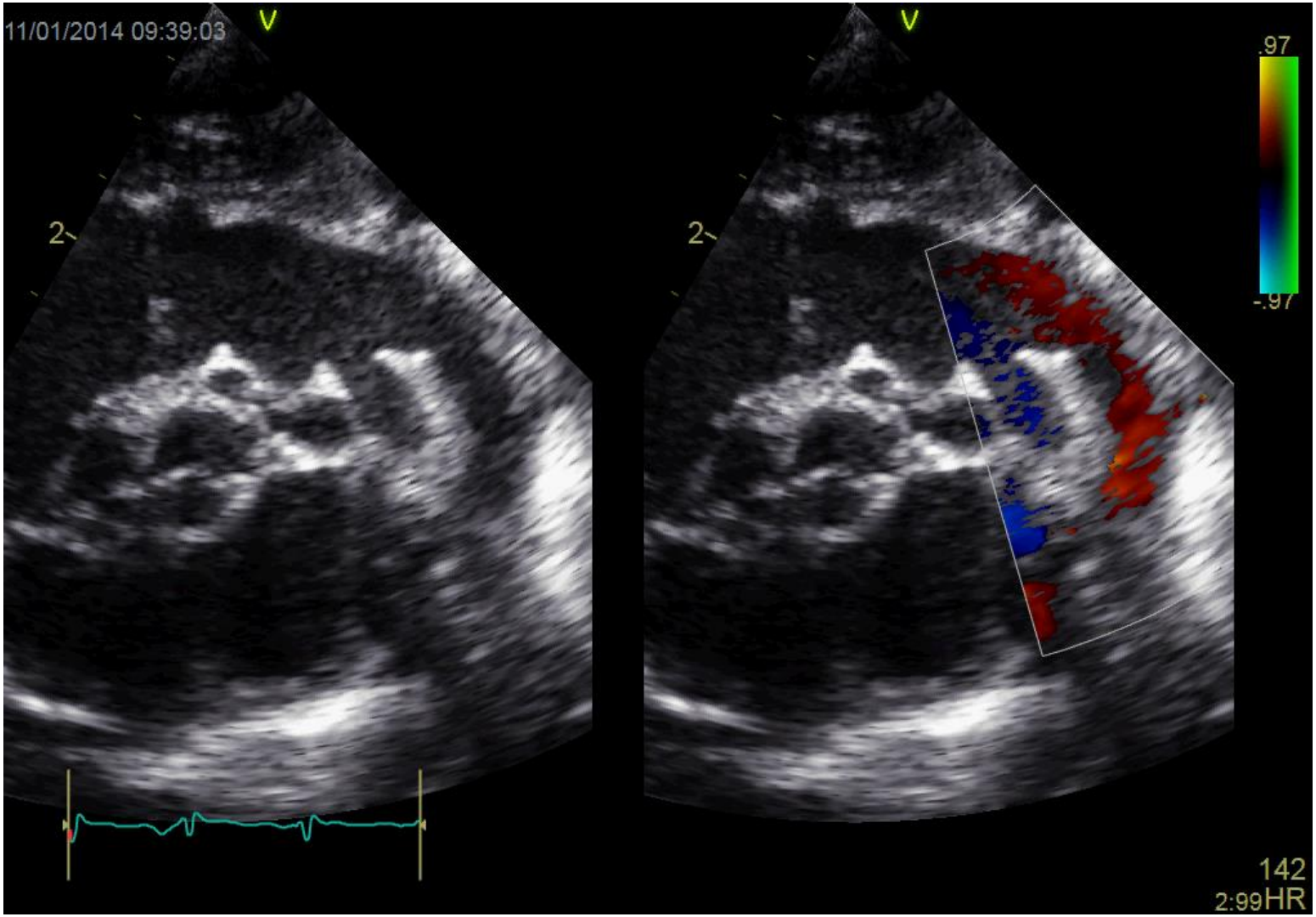
V
2
4
43
2:50HR


26/03/2018 15:07:08
V
2
V
2
.81
-.81
111
2:17HR

Patient 2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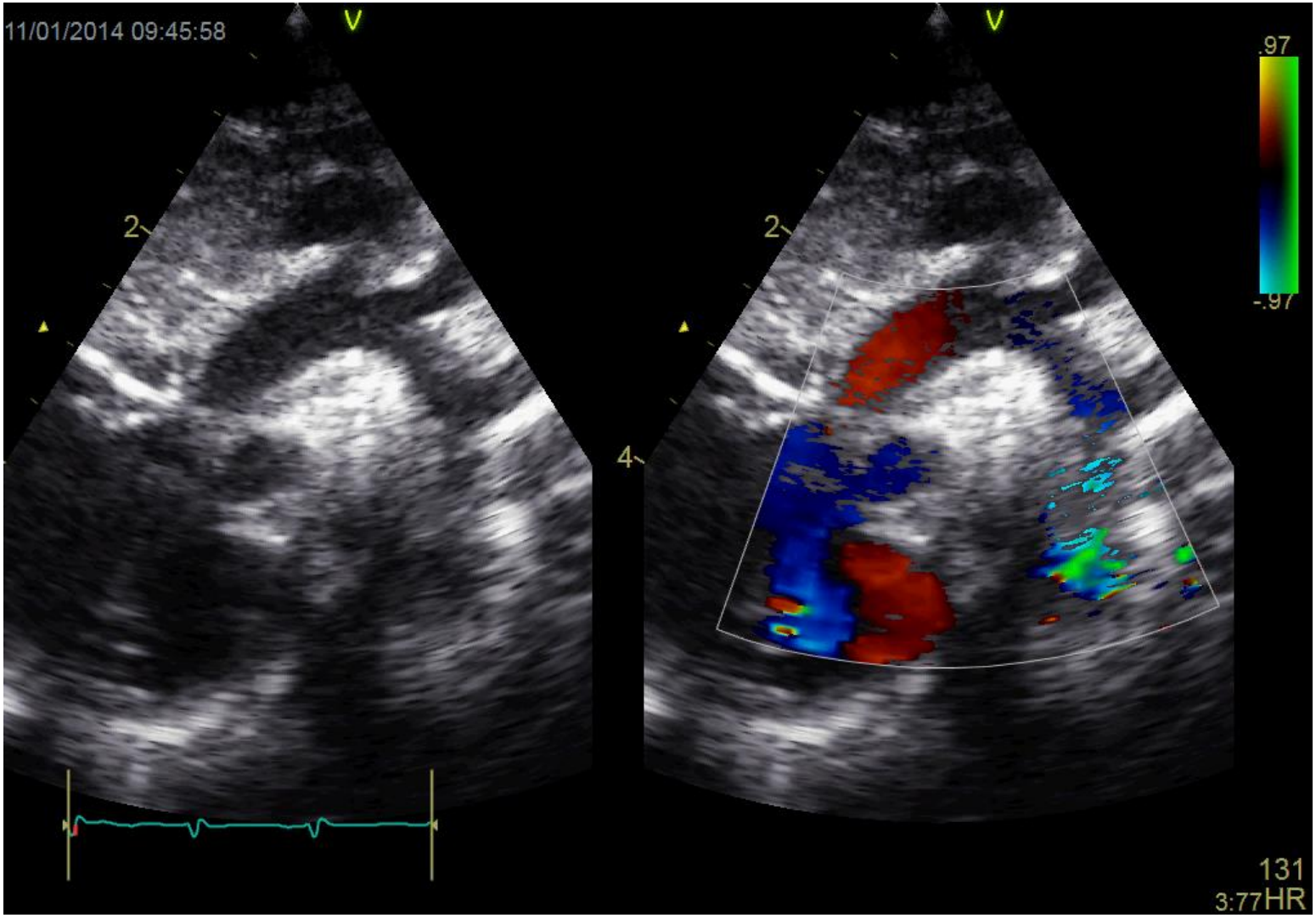
V
2
V
2
.97
-.97
142
2:99HR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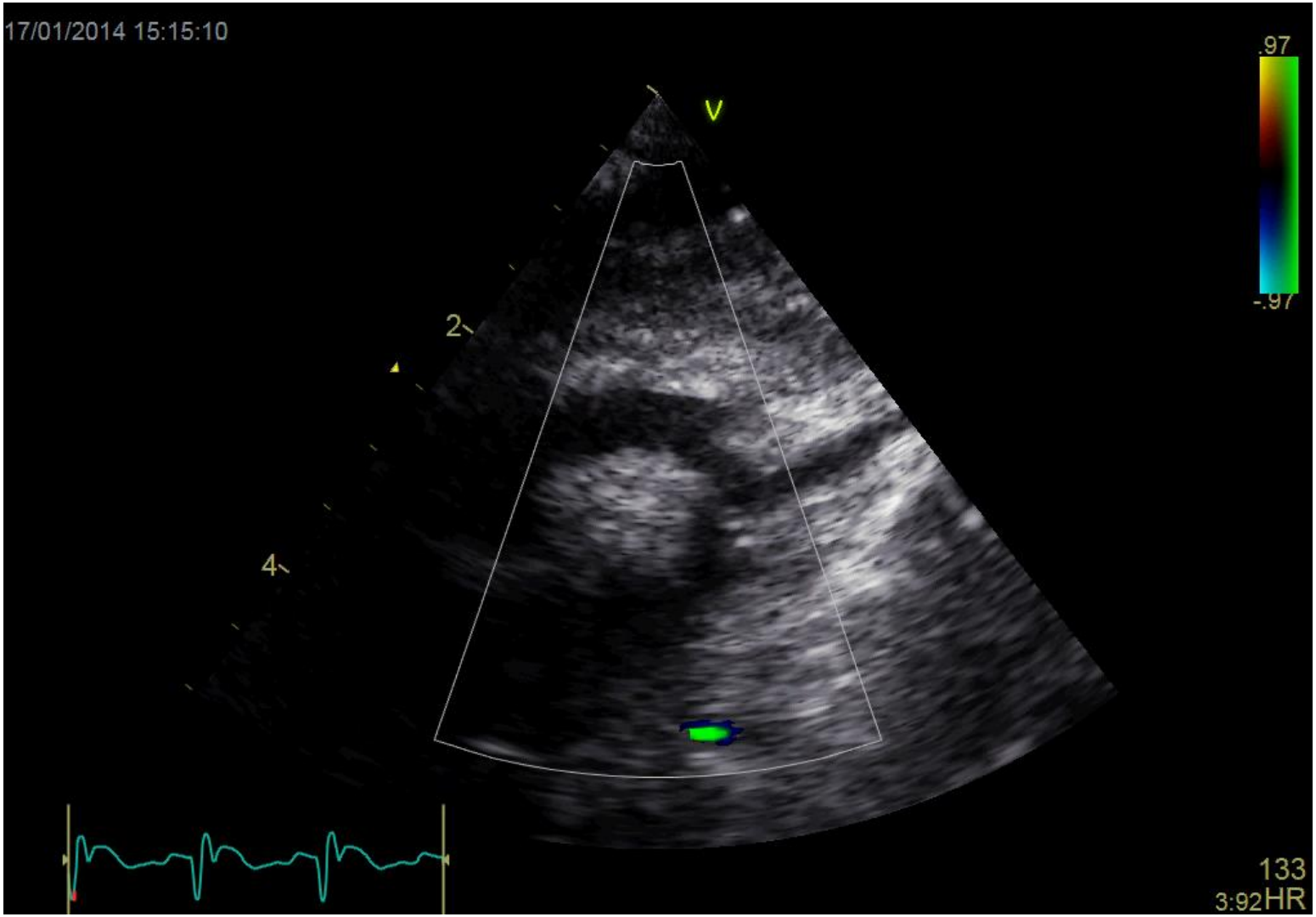
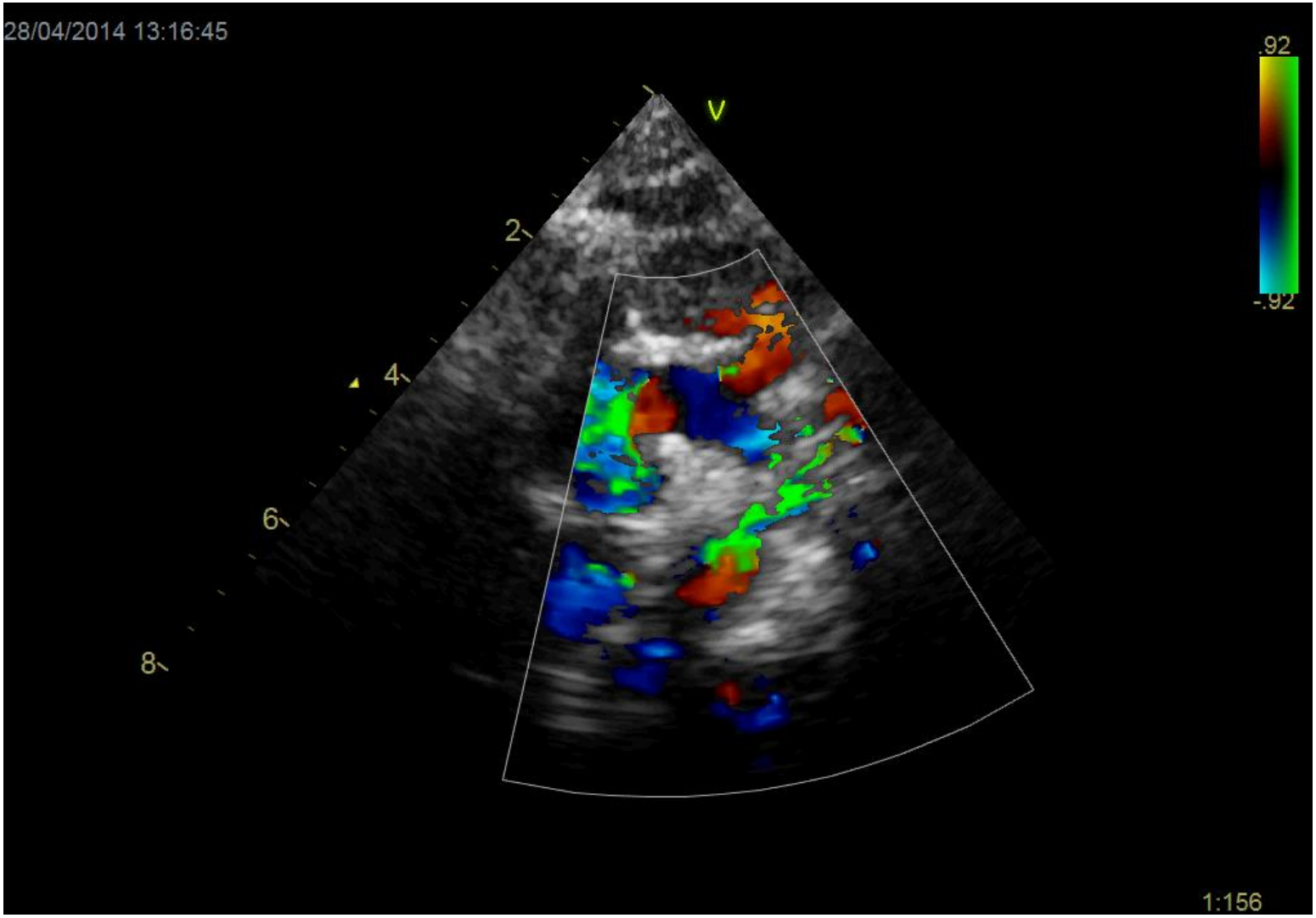
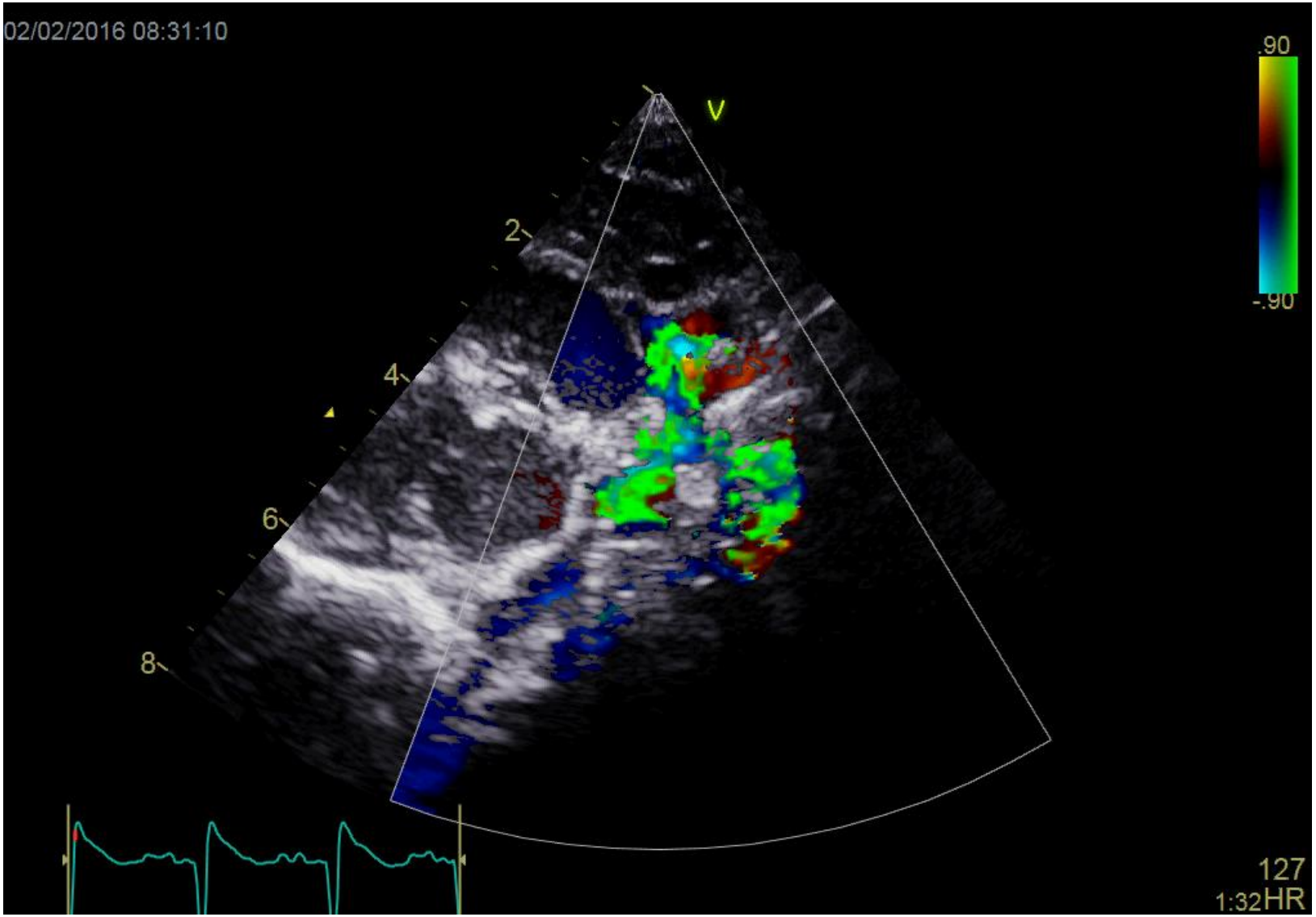
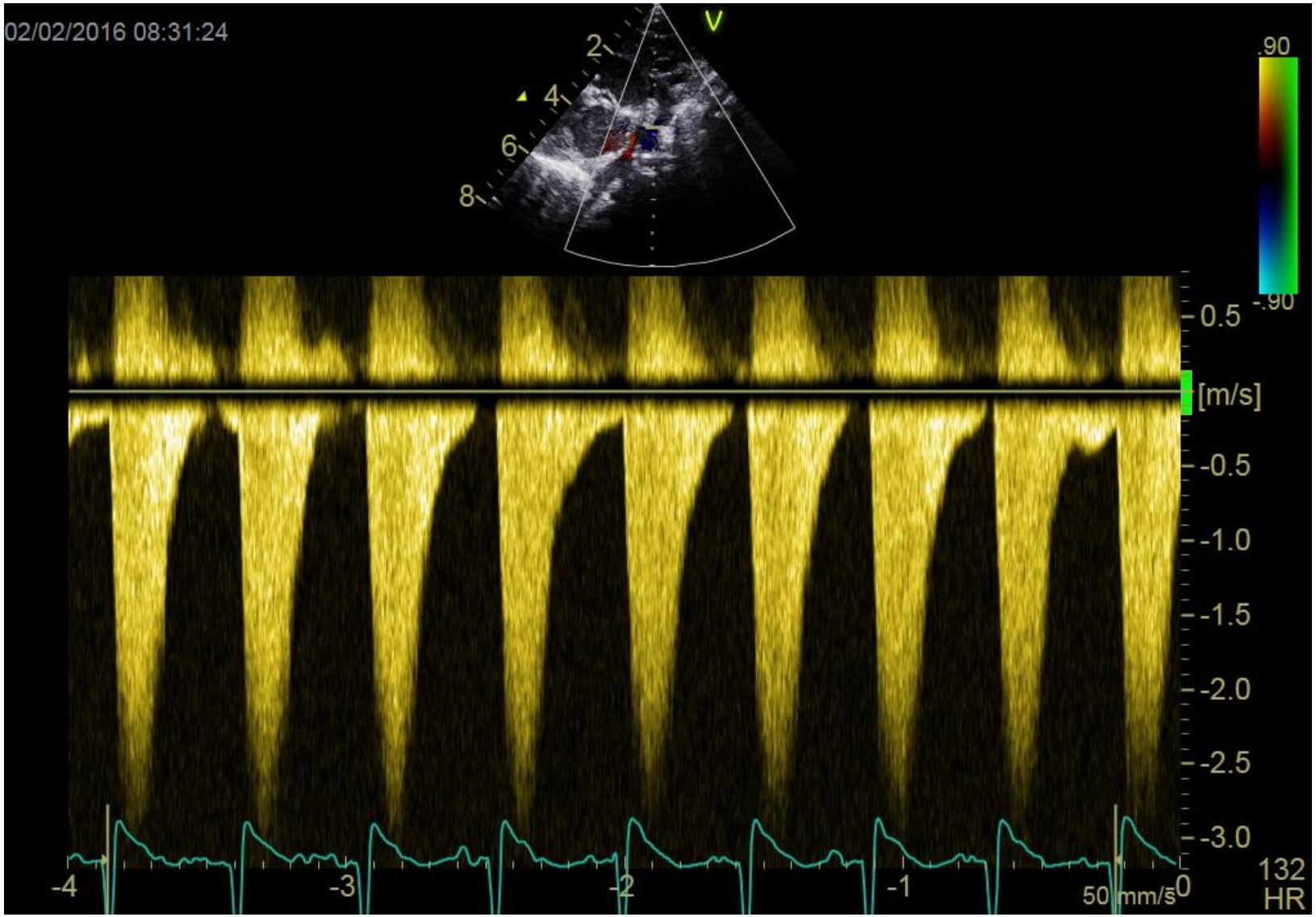
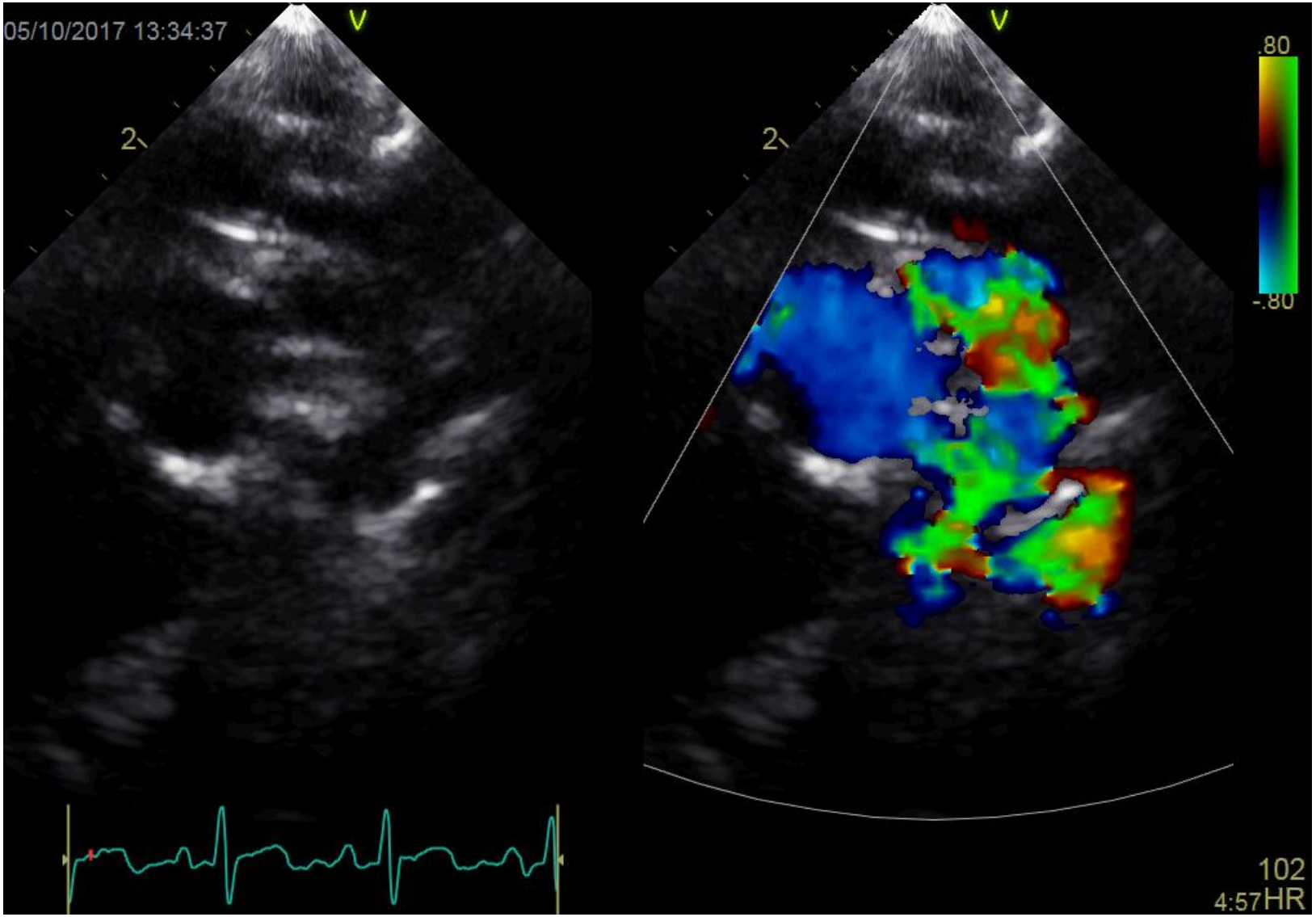
V
2
V
2
4
.97
-.97
131
3:77HR


17/01/2014 15:15:10
.97
-.97
V
2
4
133
3:92HR

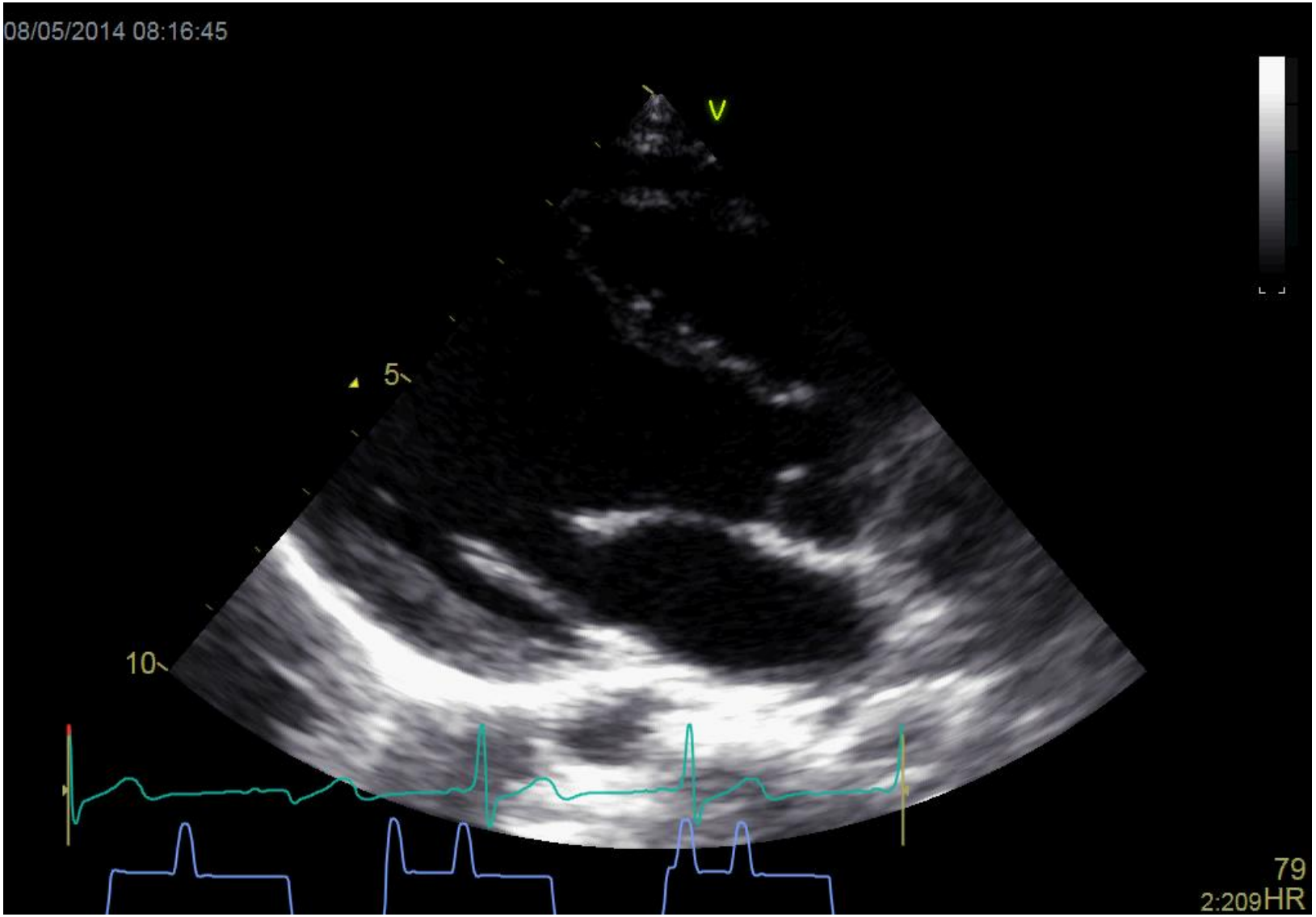

28/04/2014 13:16:45
.92
-.92
V
2
4
6
8
1:156

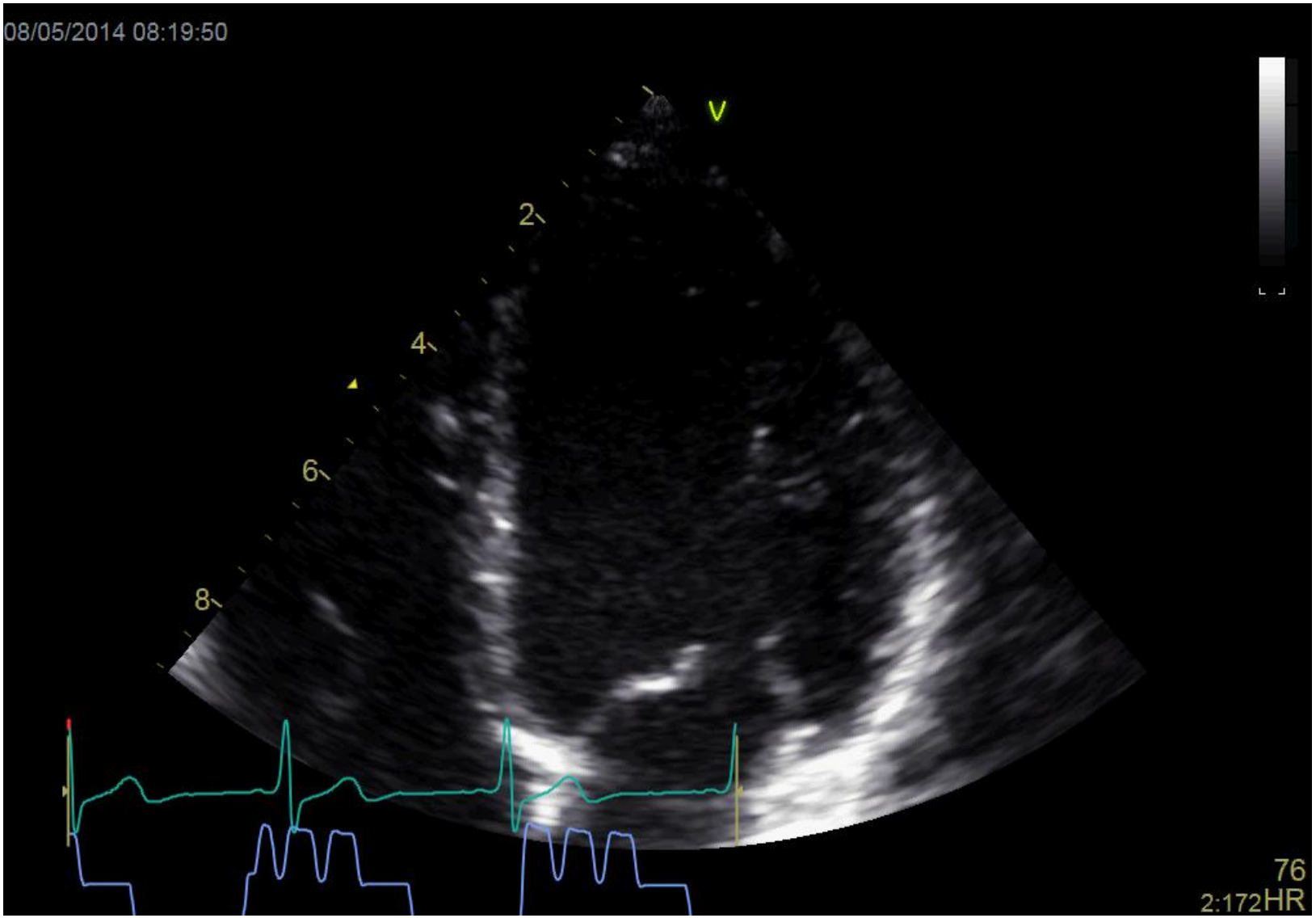

02/02/2016 08:31:10
.90
-.90
V
2
4
6
8
127
1:32HR

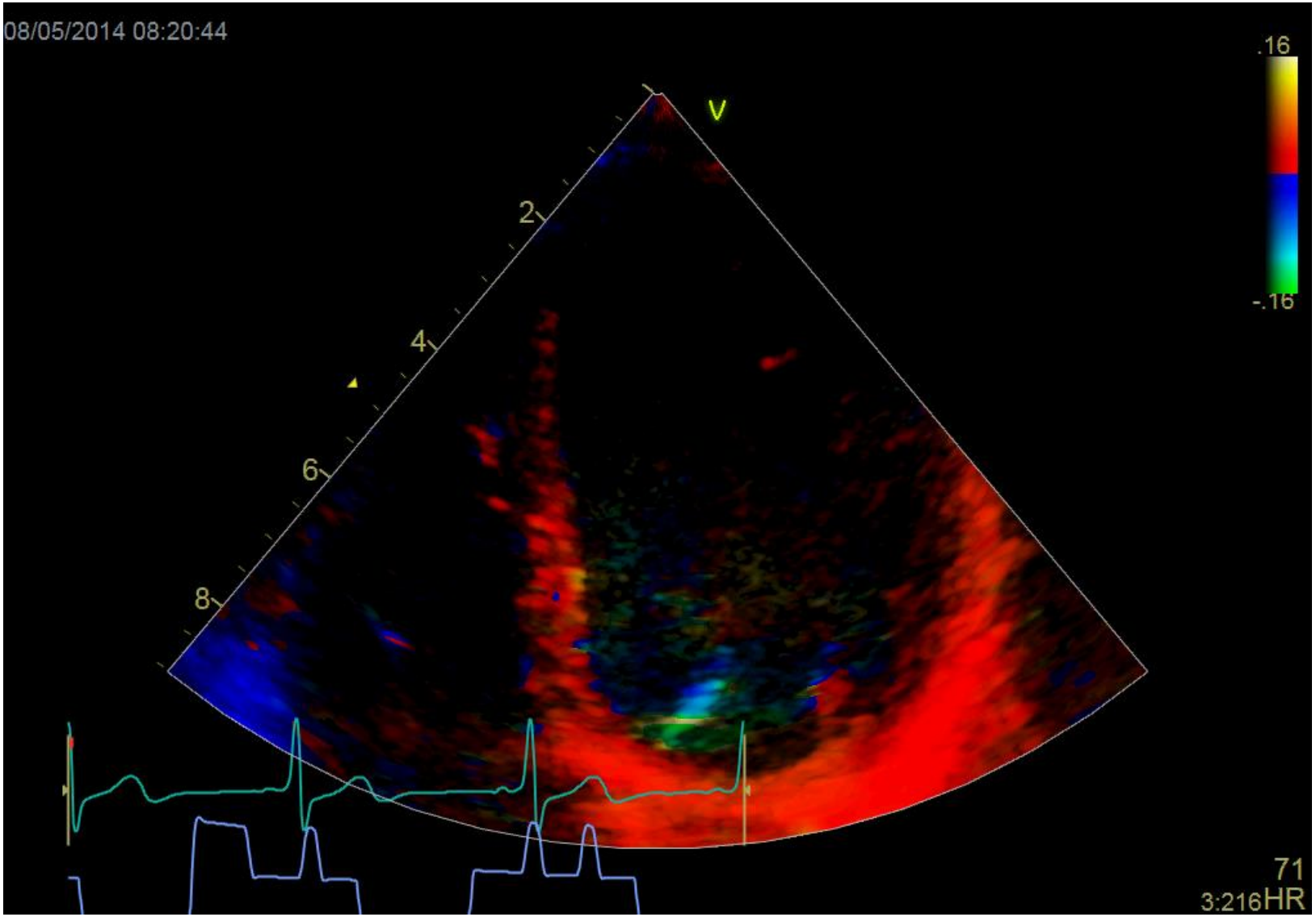

02/02/2016 08:31:24
V
2
4
6
8
.90
-.90
0.5
[m/s]
-0.5
-1.0
-1.5
-2.0
-2.5
-3.0
-4
-3
-2
-1
0
50 mm/s
132
HR

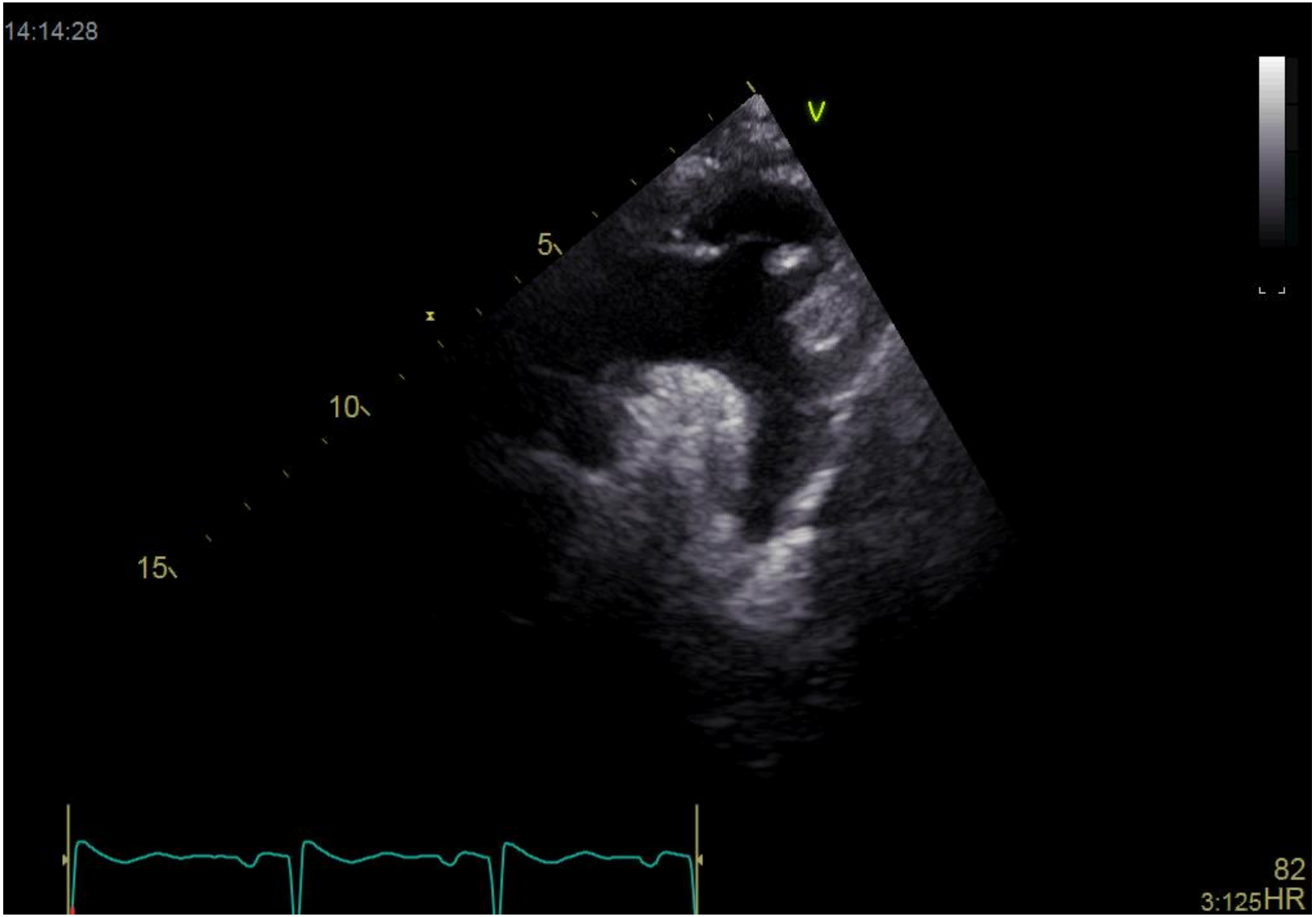

05/10/2017 13:34:37
.80
-.80
102
4:57HR

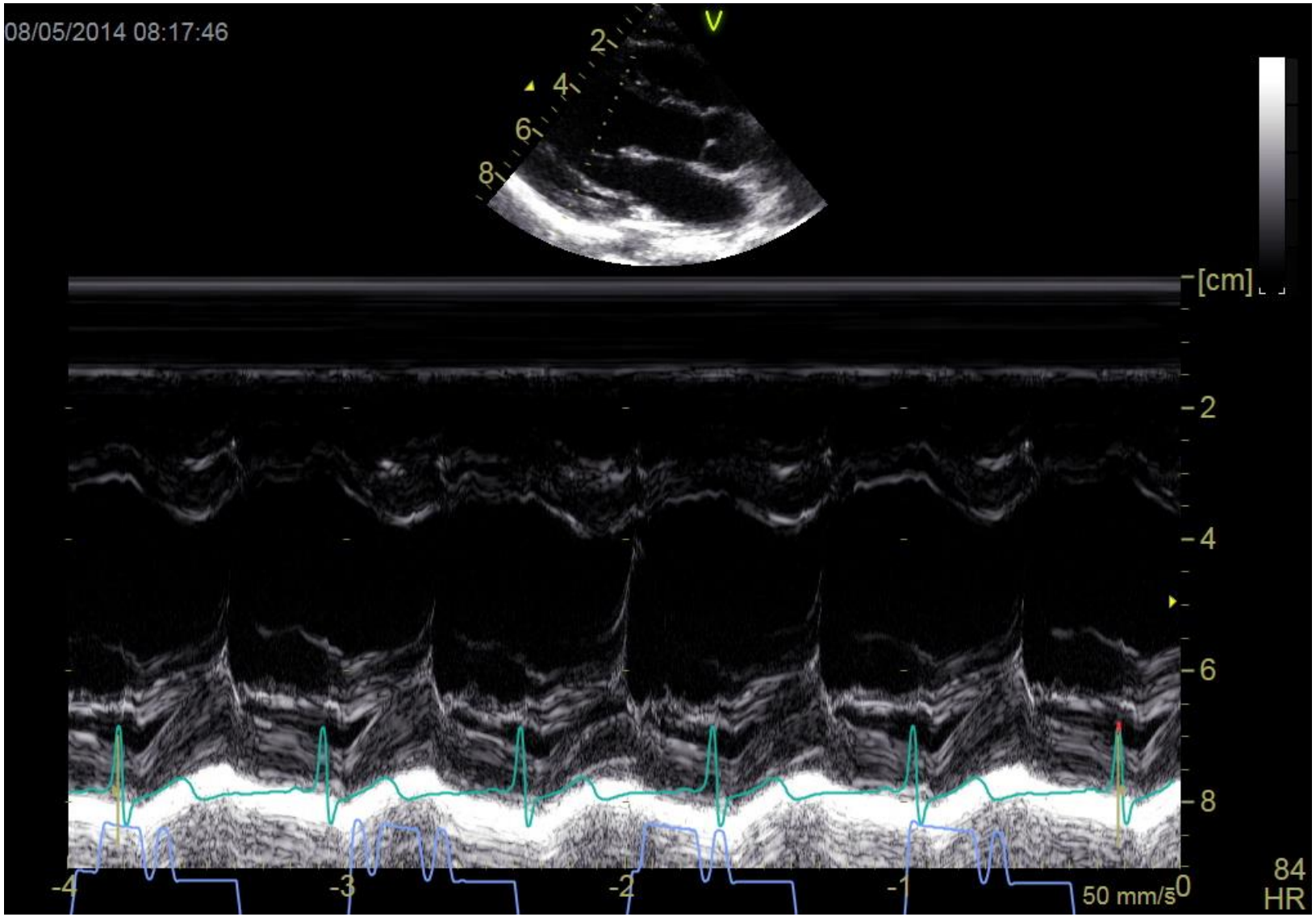
Verloop na eerste behandeling coarctatio

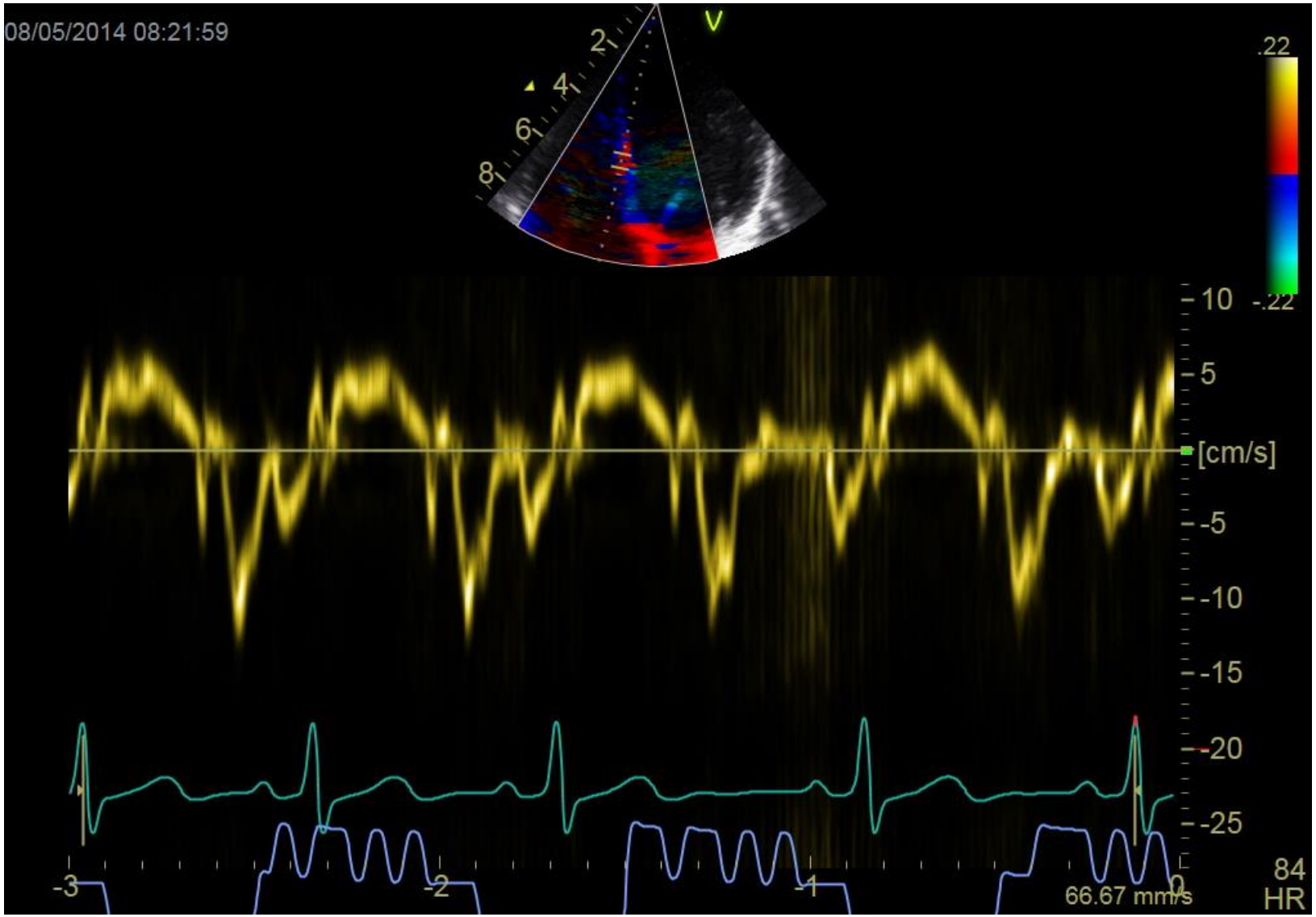

08/05/2014 08:16:45
V
5
10
79
2:209HR

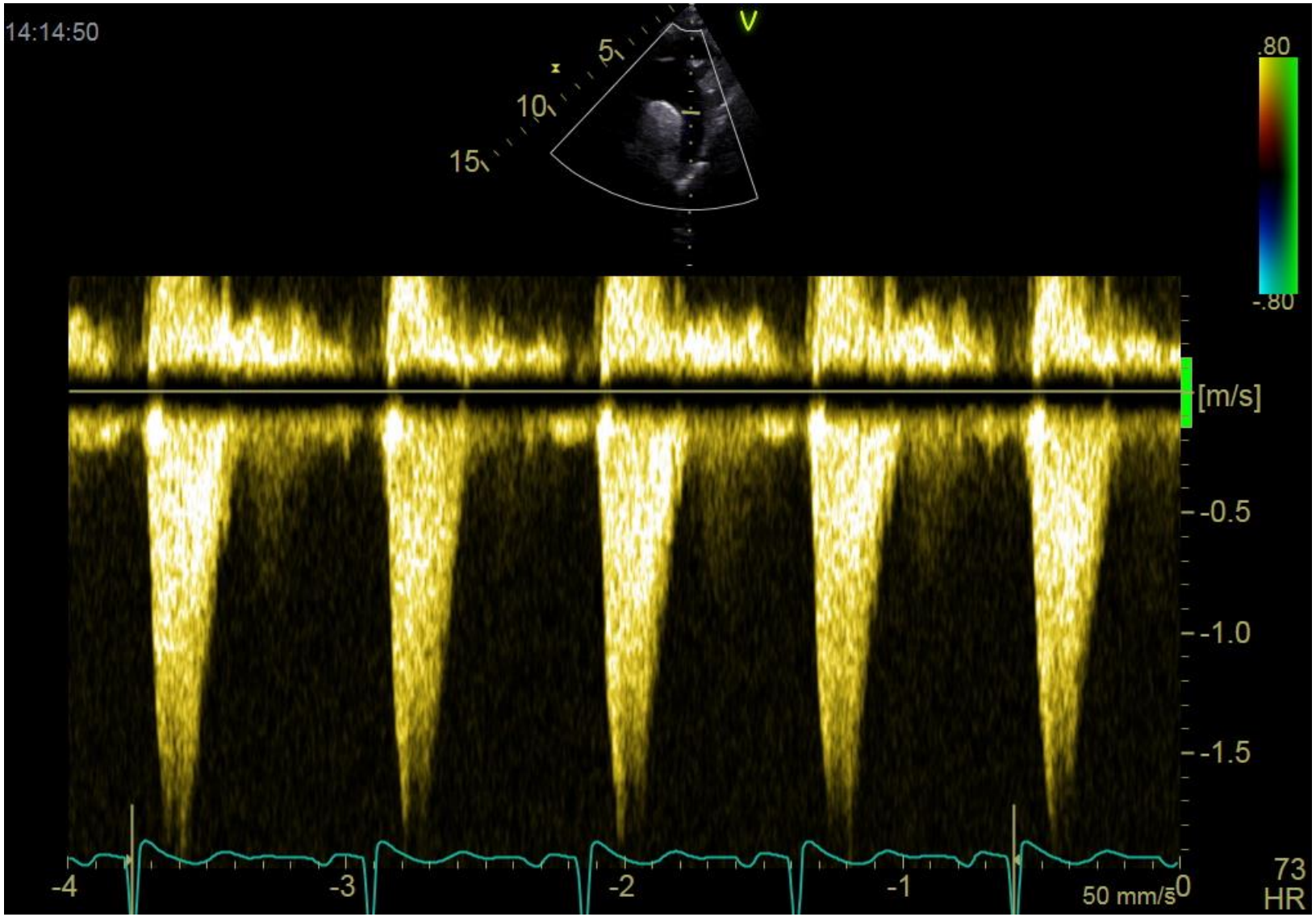

08/05/2014 08:19:50
V
2
4
6
8
76
2:172HR

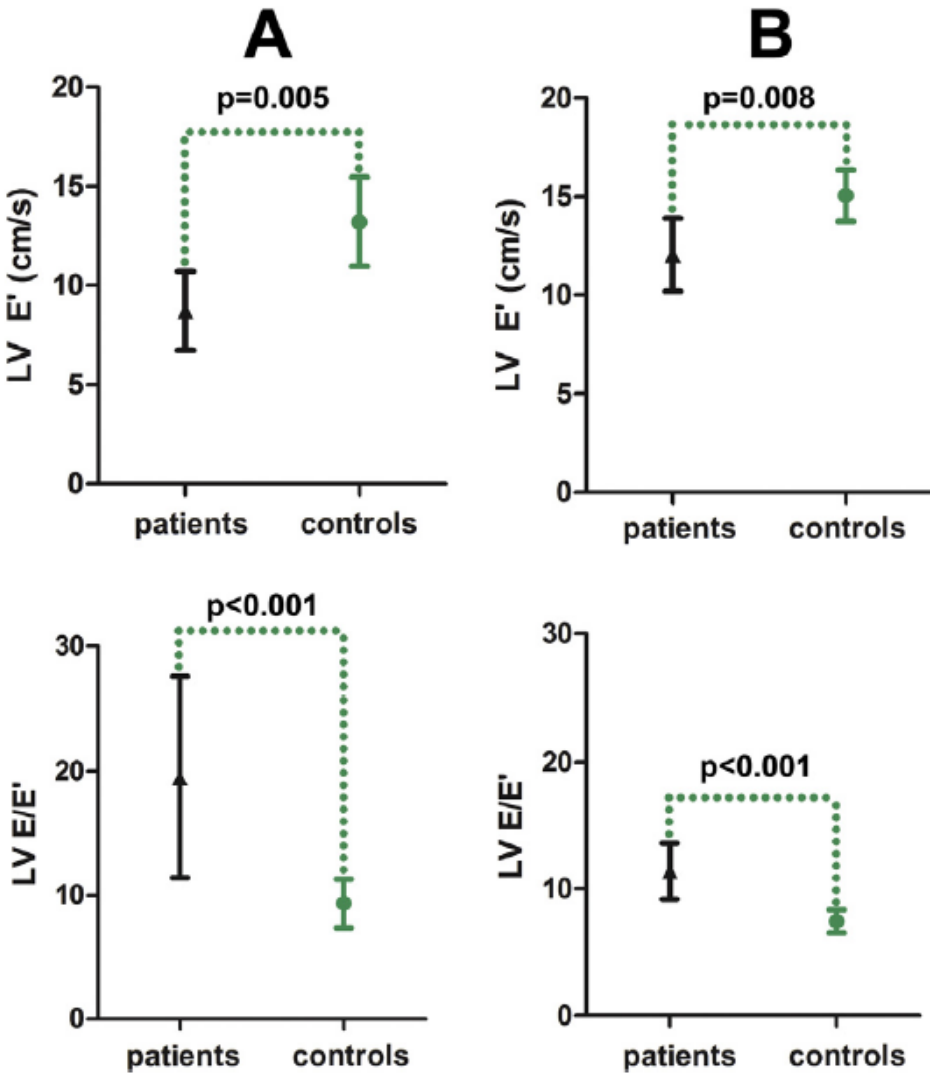

08/05/2014 08:20:44
.16
-.16
V
2
4
6
8
71
3:216HR


14:14:28
V
5
10
15
82
3:125HR

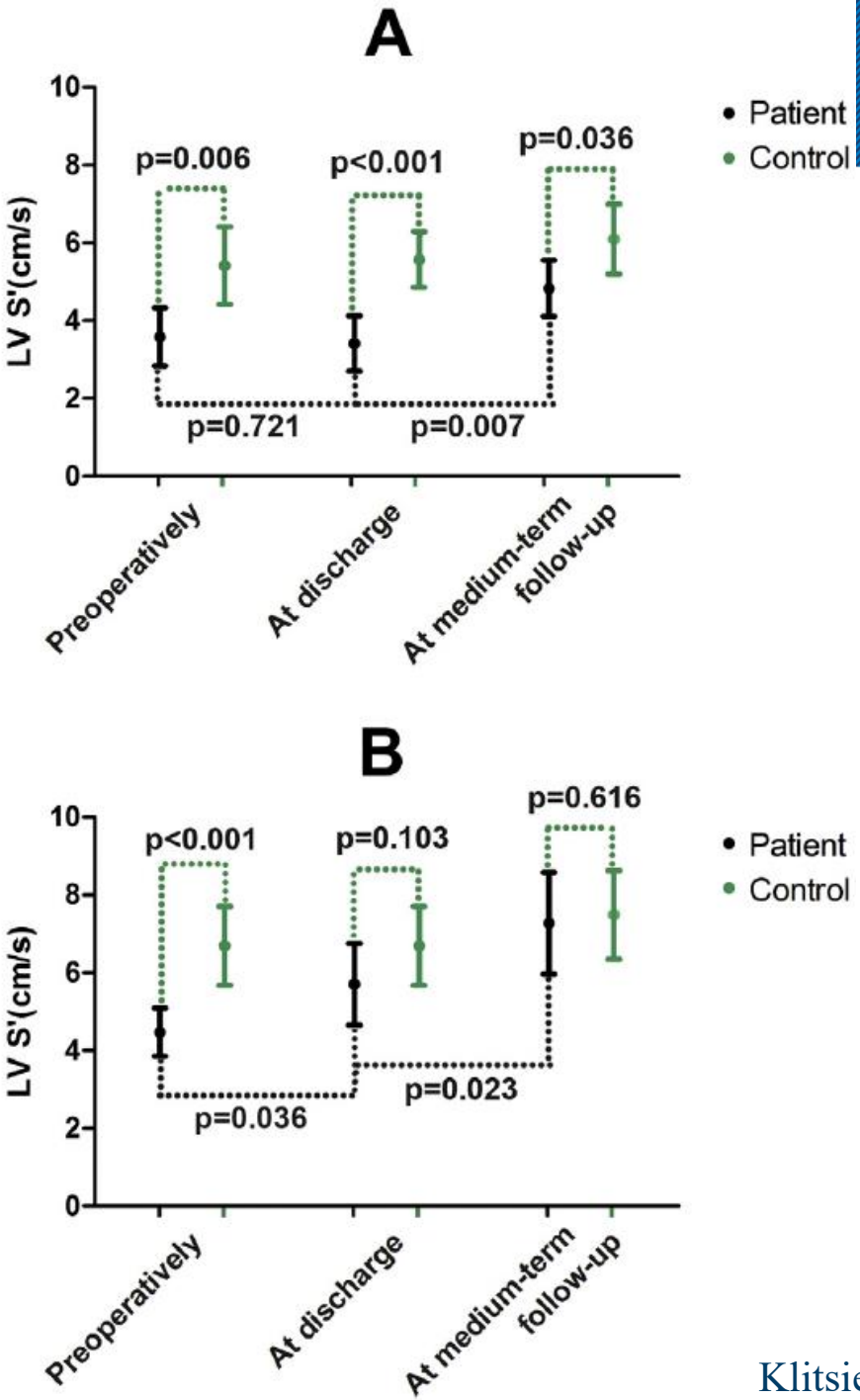

08/05/2014 08:17:46
V
2
4
6
8
[cm]
2
4
6
8
-4
-3
-2
-1
0
50 mm/s
84
HR


08/05/2014 08:21:59
V
2
4
6
8
.22
-.22
10
5
[cm/s]
-5
-10
-15
-20
-25
-3
-2
-1
0
66.67 mm/s
84
HR

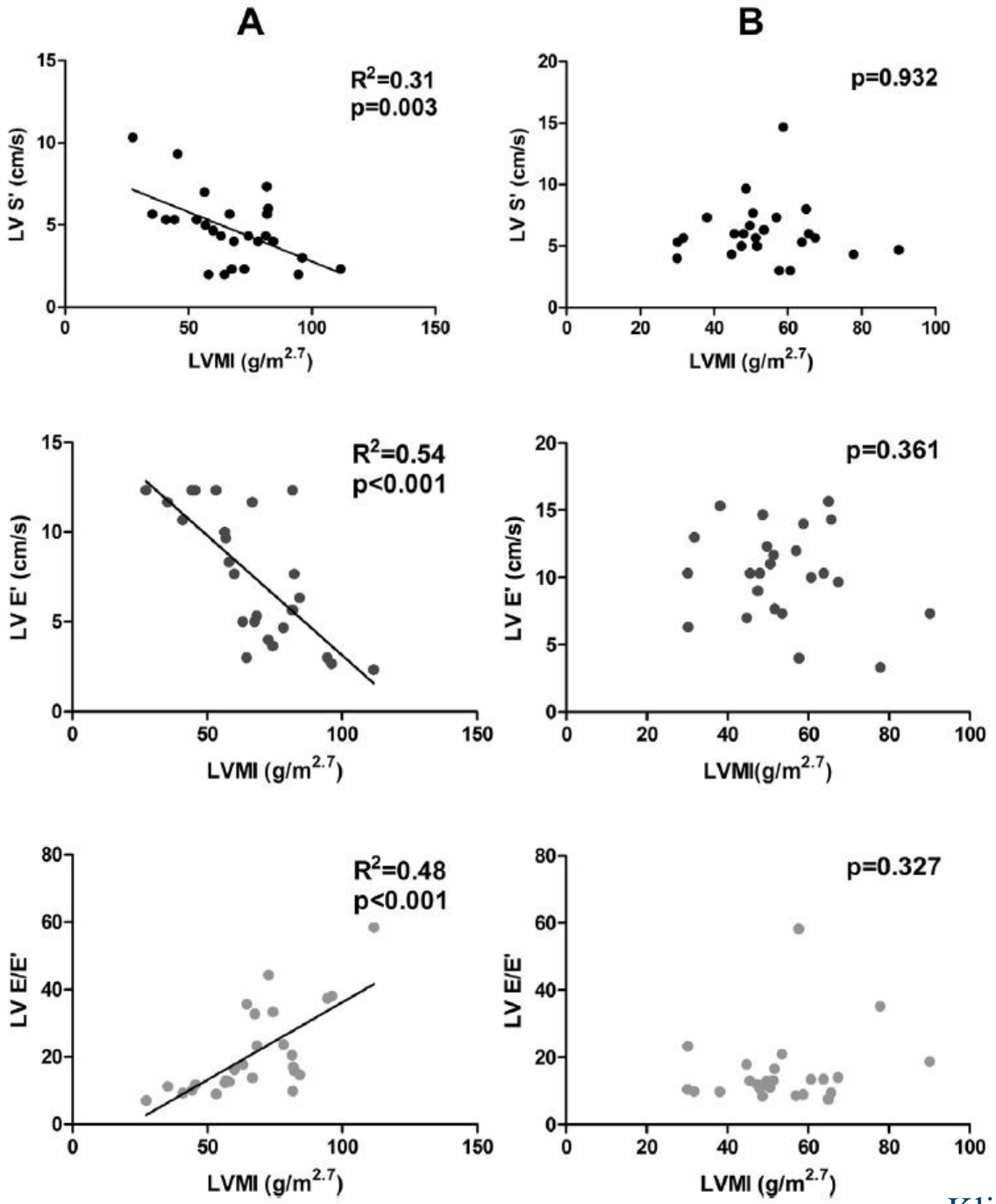

14:14:50
V
5
10
15
.80
-.80
[m/s]
-0.5
-1.0
-1.5
-4
-3
-2
-1
0
50 mm/s
73
HR



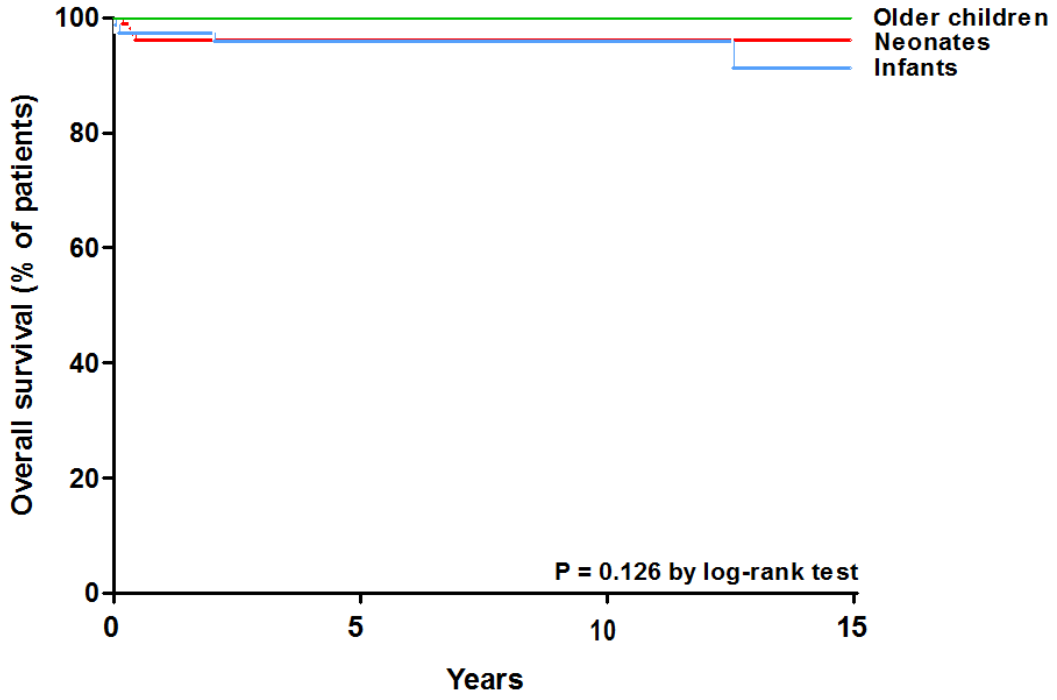
Klitsie et al.; Ann Thorac Surg 2013



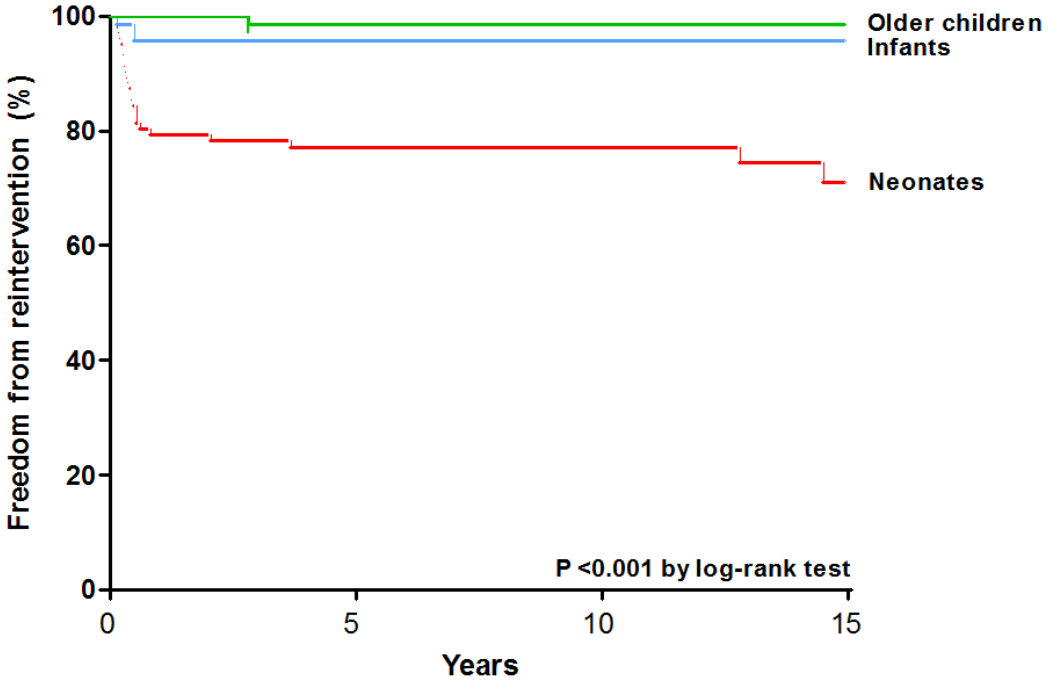
Klitsie et al.; Ann Thorac Surg 2013



Klitsie et al.; Ann Thorac Surg 2013



No. at risk				
Neonates	117	78	51	25
Infants	81	49	27	13
Older children	94	42	40	15



No. at risk				
Neonates	117	60	41	20
Infants	81	47	26	13
Older children	94	61	39	15

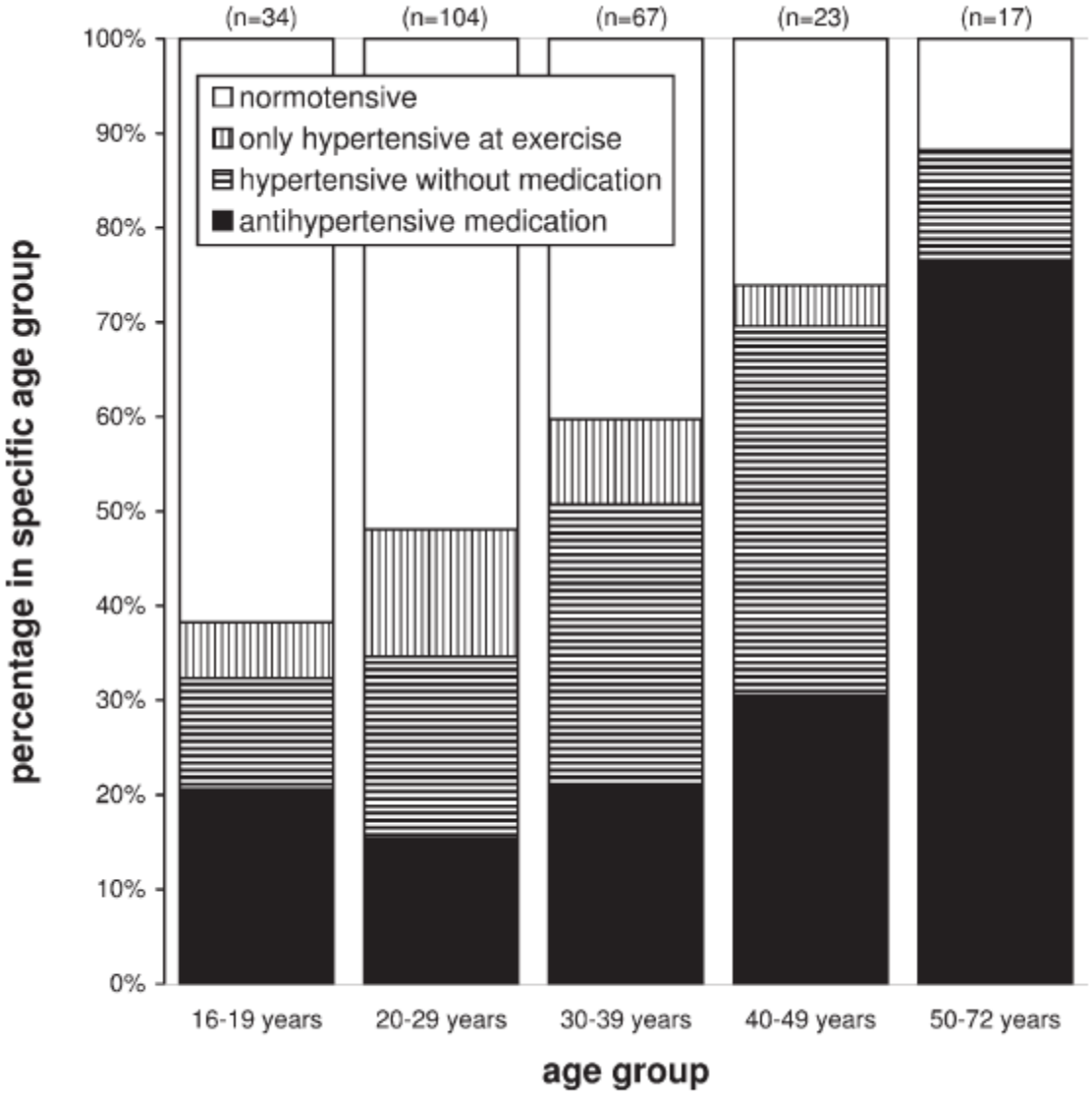


Figure 1. Prevalence of hypertension after coarctation repair.

Hager et al.; J Thorac Cardiovasc Surg 2007

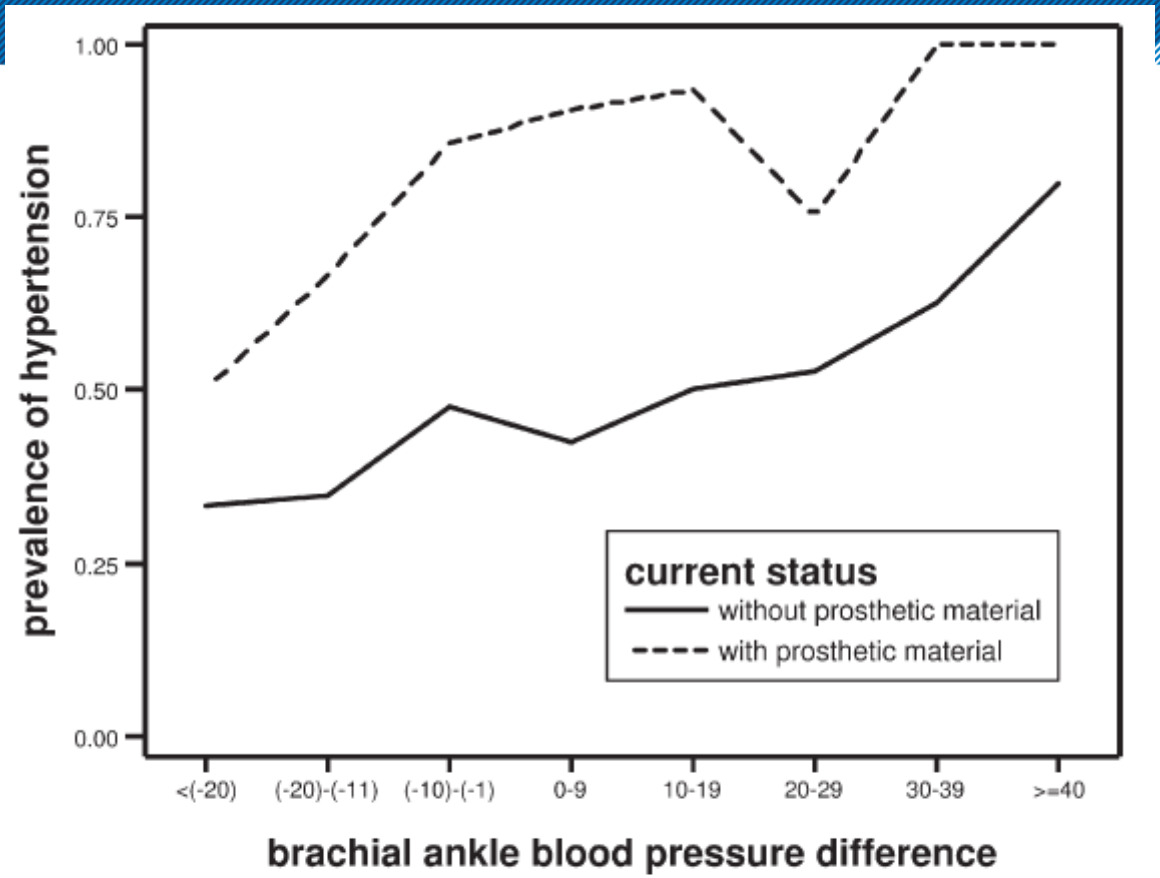
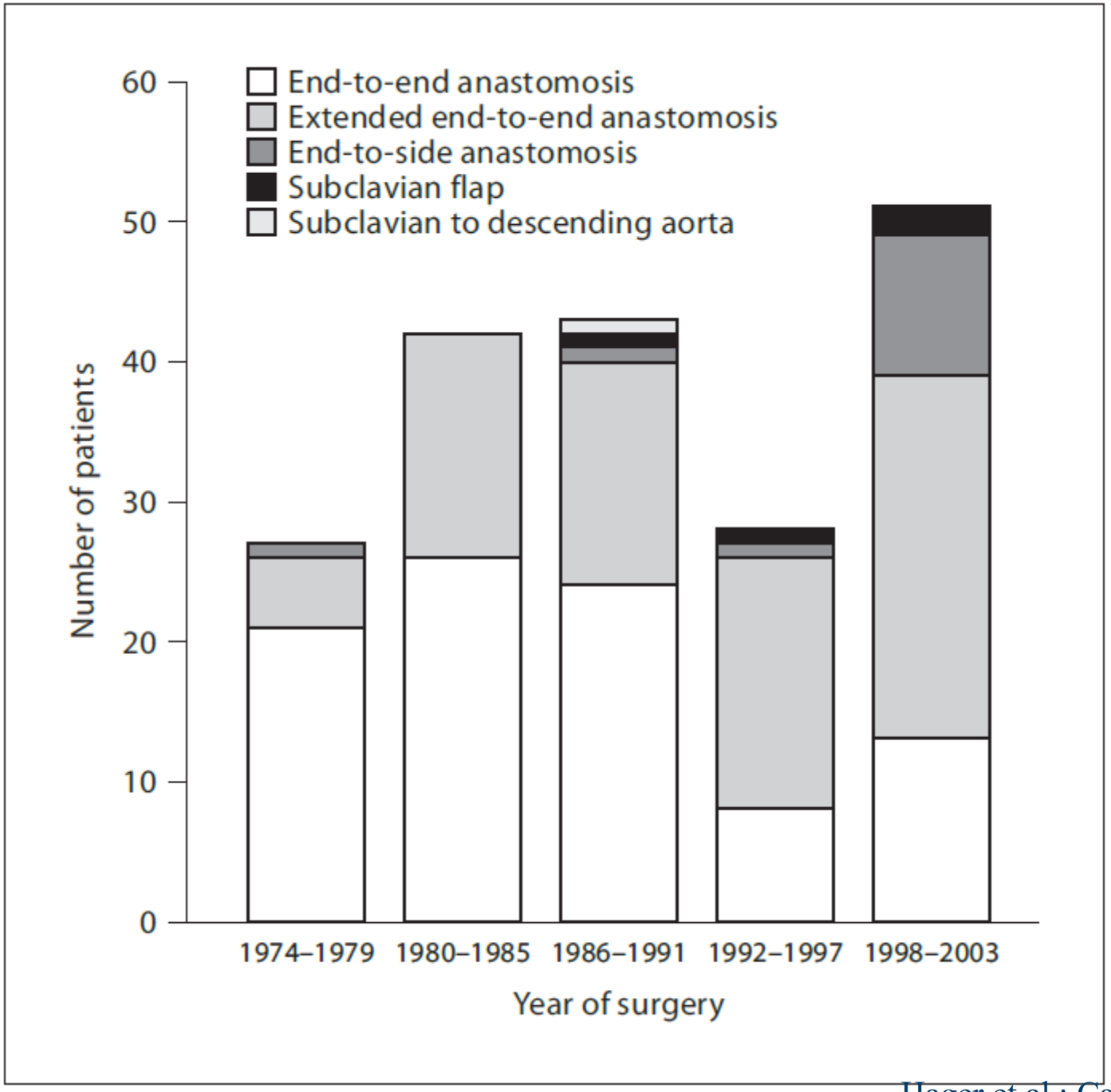
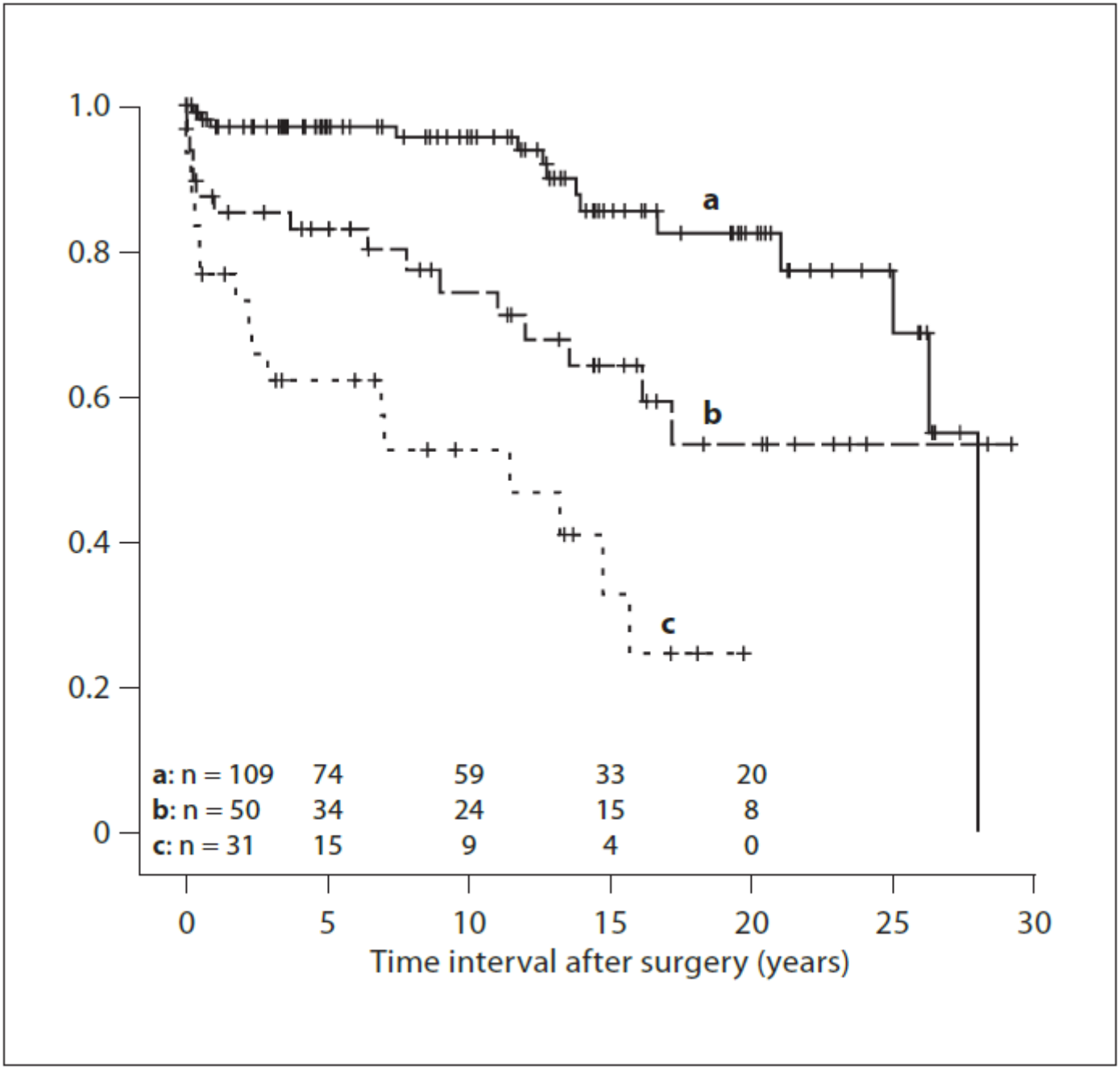


Figure 2. Prevalence of hypertension (antihypertensive drugs, hypertension at ambulatory blood pressure measurement, or hypertension at exercise) according to use of prosthetic material to repair the coarctation and according to the noninvasively measured systolic brachial–ankle blood pressure difference.

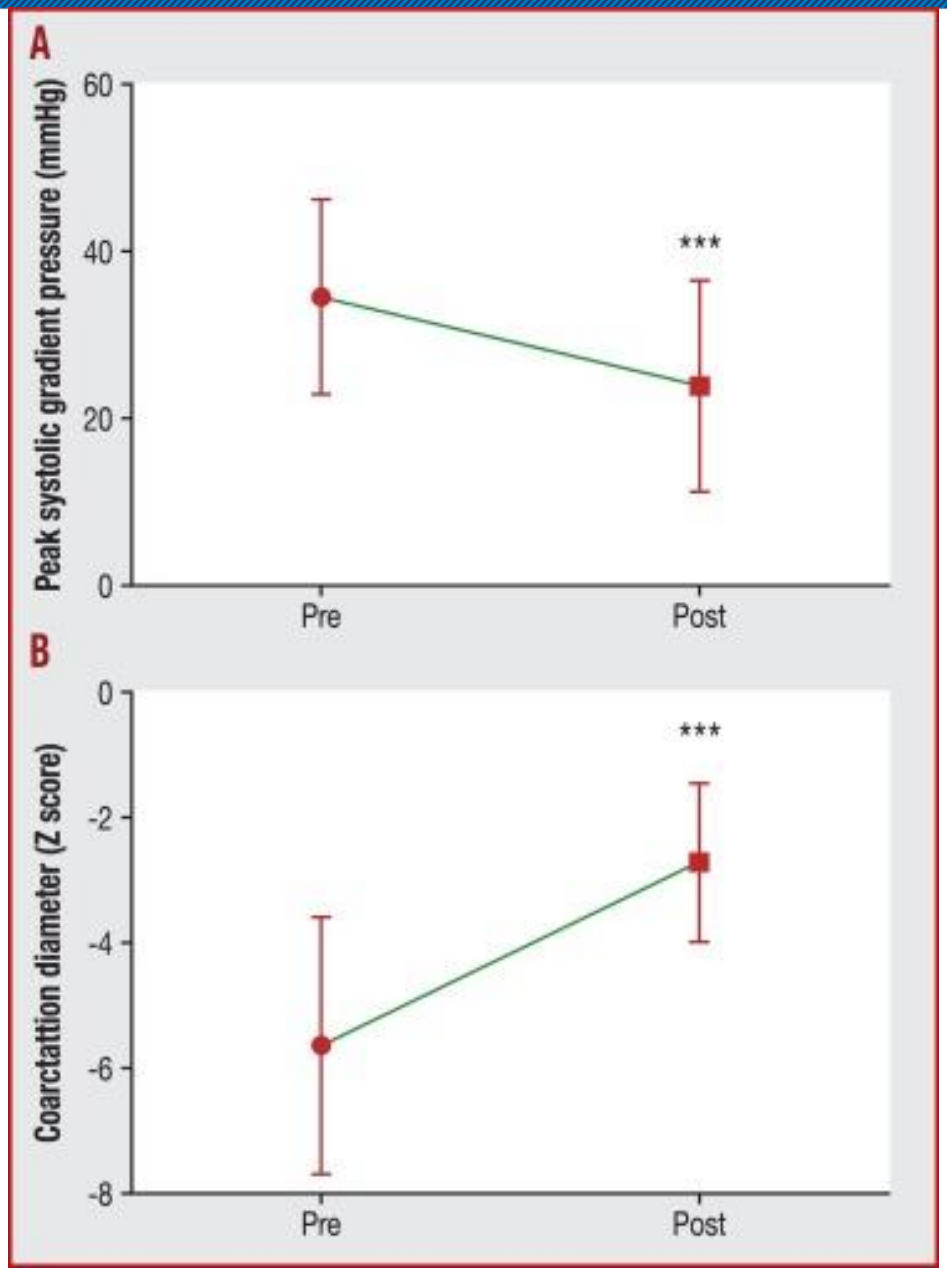
Hager et al.; J Thorac Cardiovasc Surg 2007



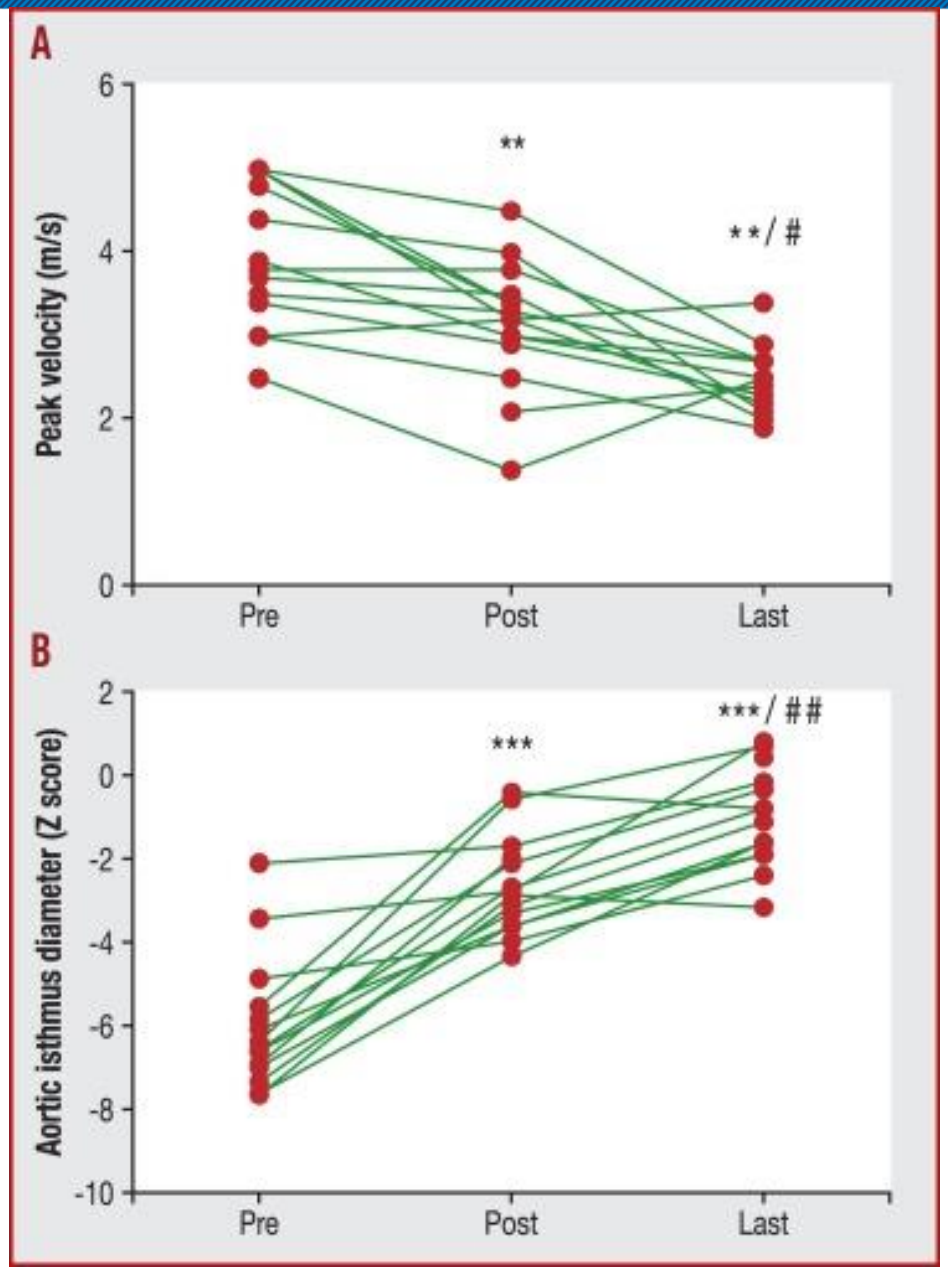
Hager et al.; Cardiology 2009



Hager et al.; Cardiology 2009



Hager et al.; Cardiology 2009



Lefort et al.; ACVD 2018

Conclusies

-Prenatale diagnostiek kan een deel van de CoA opsporen

-Behandeling meestal effectief

-Op jonge leeftijd meer kans op reinterventie

-Lange termijn effecten blijven aanwezig: verminderde LV functie en hypertensie

-Levenslange follow-up is aangewezen