



LEIDS UNIVERSITAIR MEDISCH CENTRUM

CNE Congenitale Cardiologie 20-1-2015

Congenitale hartafwijkingen

kliniek, X-thorax en echocardiografie

Arno A.W. Roest
Kindercardiologie
LUMC

Continuing Nursing Education



Congenitale hartafwijkingen

- Meest frequente aangeboren afwijking: 0.96% van geboortes
- Meeste hartafwijkingen behoeven geen interventie
- Maar als er een ingreep nodig is kan dat bijna altijd
- Dit moet dan wel in een gespecialiseerd centrum
- Overleving gestegen van 10% naar 90% op 18 jarige leeftijd

- Prenatale diagnose steeds belangrijker: Dr Rozendaal

Table 1. Incidence per Million Live Births

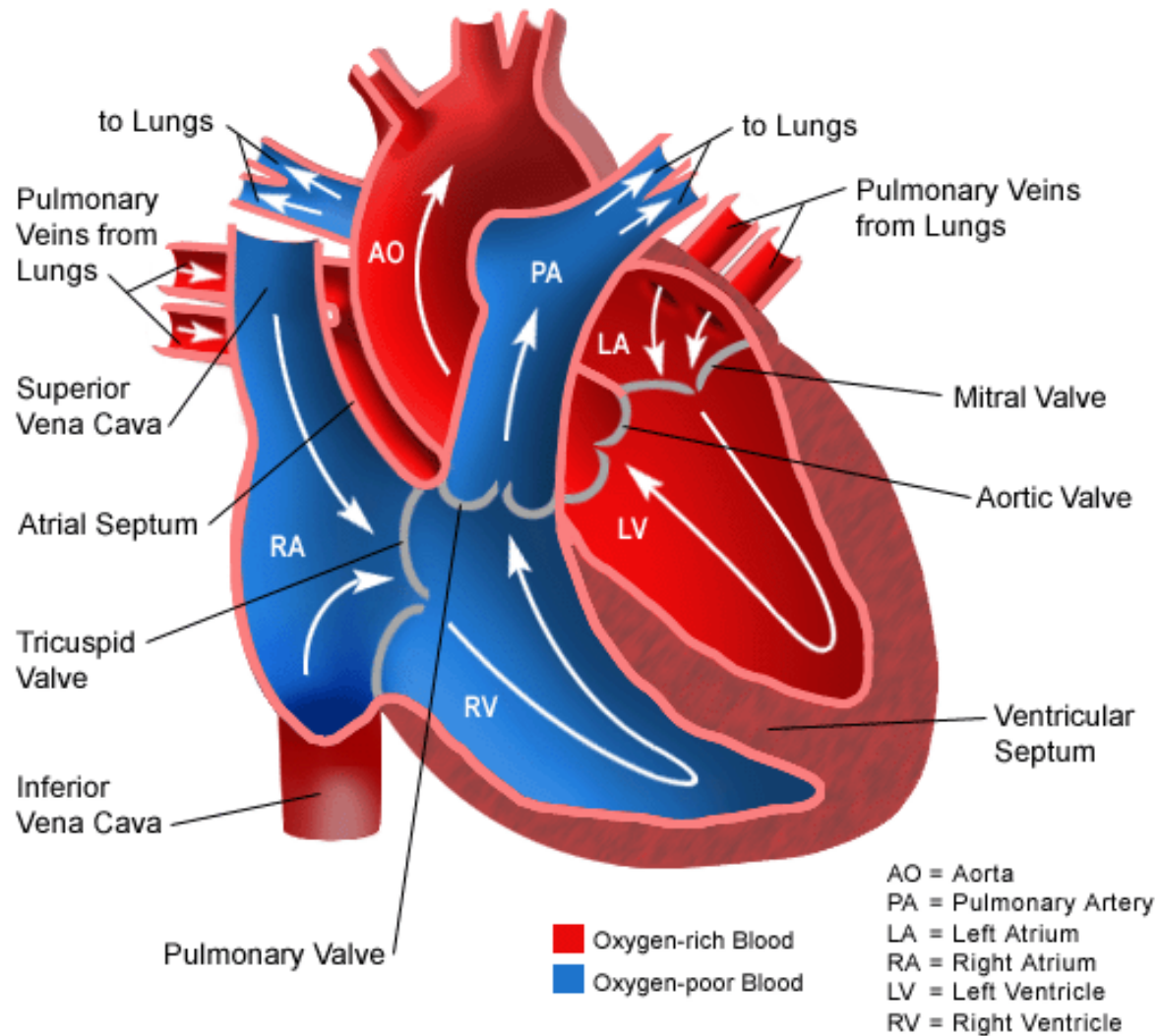
Lesion	Number of Studies	Mean	SD	Lower Quartile	Median	Upper Quartile	NERICP 1975–1977
VSD	43	3,570	2,878	1,757	2,829	4,482	345
PDA	40	799	1,399	324	567	782	135
ASD	43	941	1,043	372	564	1,059	65
AVSD	40	348	165	242	340	396	110
PS	39	729	731	355	532	836	73
AS	37	401	543	161	256	388	41
Coarc	39	409	246	289	356	492	165
Tetralogy	41	421	188	291	356	577	196
d-TGA	41	315	115	231	303	388	218
HRH	32	222	199	105	160	224	—
Tricuspid atresia	11	79	52	24	92	118	56
Ebstein's anomaly	5	114	138	38	40	161	12
Pul Atresia	11	132	123	76	83	147	69
HLH	36	266	216	154	226	279	163
Truncus	30	107	71	61	94	136	30
DORV	16	157	103	82	127	245	32
SV	23	106	70	54	85	136	54
TAPVC	25	94	46	60	91	120	58
All cyanotic	37	1,391	590	1,078	1,270	1,533	888
All CHD*	43	9,596	7,484	6,020	7,669	10,567	2,033
BAV	10	13,556	13,049	5,336	9,244	13,817	—

*Excluding bicuspid nonstenotic aortic valves, isolated partial anomalous pulmonary venous connection and silent ductus arteriosus.

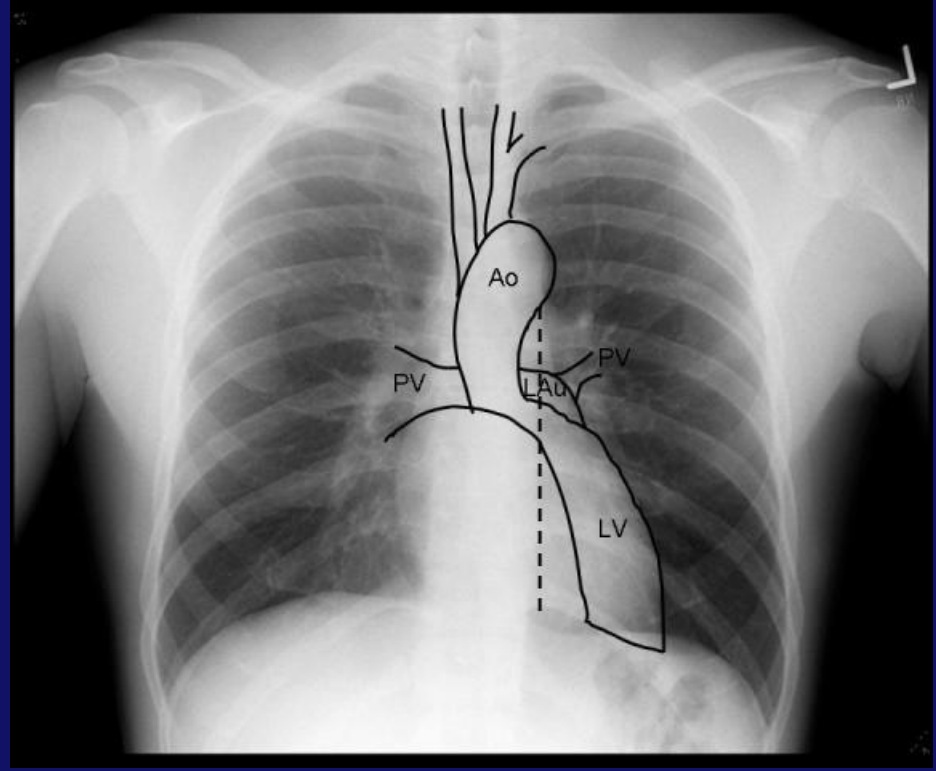
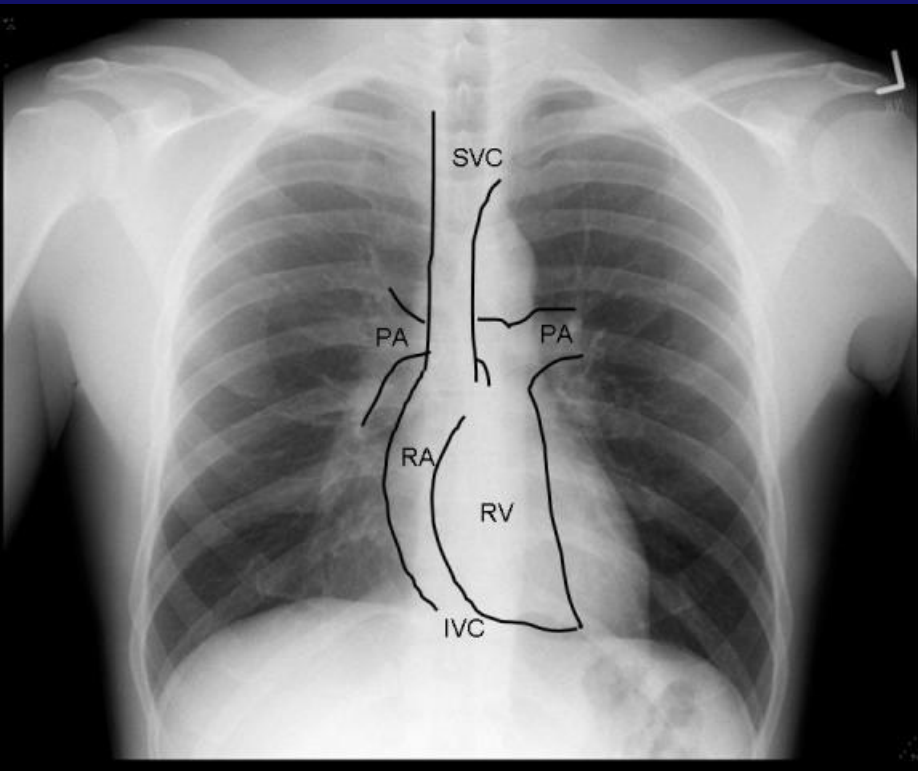
BAV = bicuspid aortic valve; CHD = congenital heart disease; Coarc = coarctation of the aorta; NERICP = New England Regional Infant Cardiac Program. Other abbreviations as in legend to Figure 5.

Normaal hart

Normal Heart

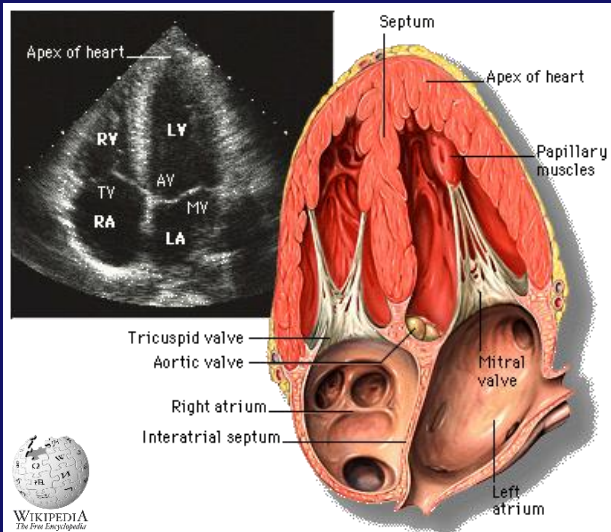


Cardiovascular structuren op de X-thorax

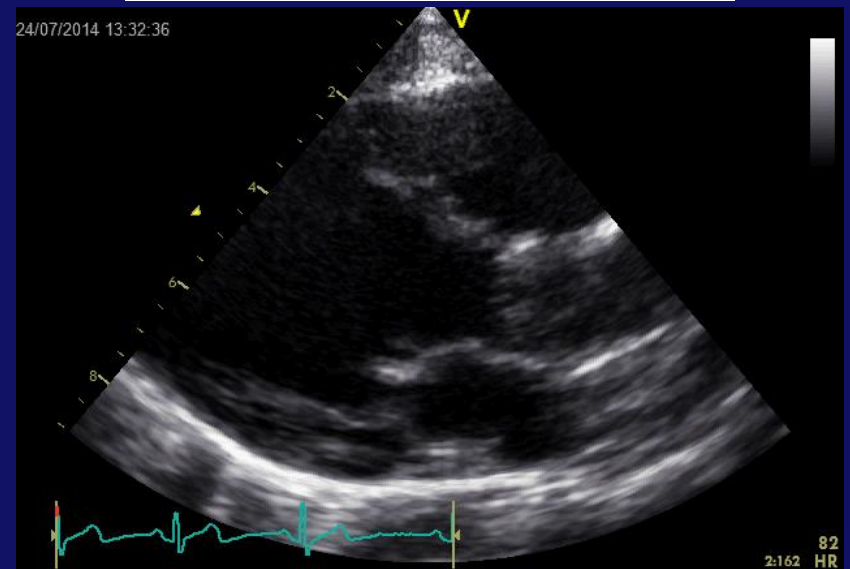
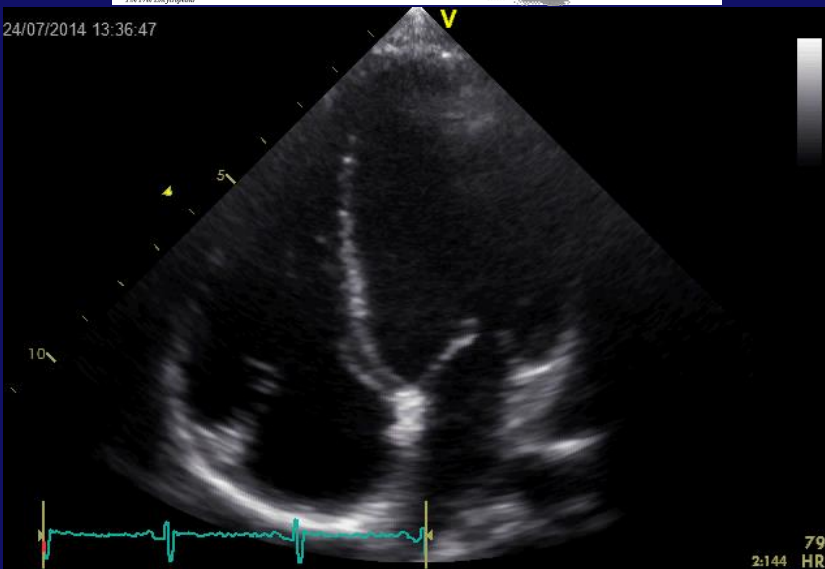
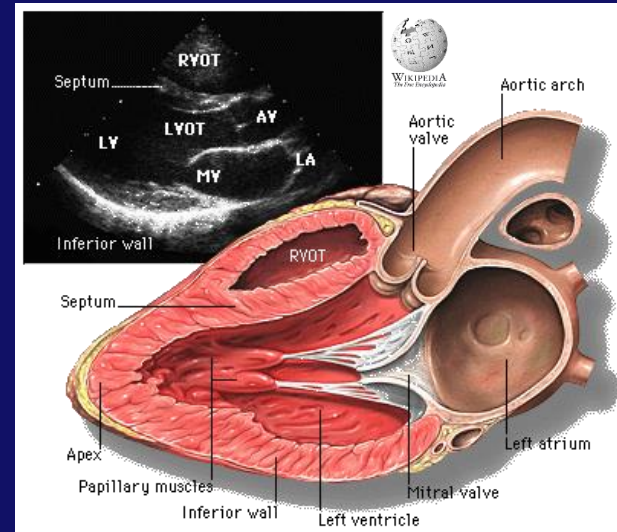


Normale echo

4 kamer beeld



Lange-as beeld

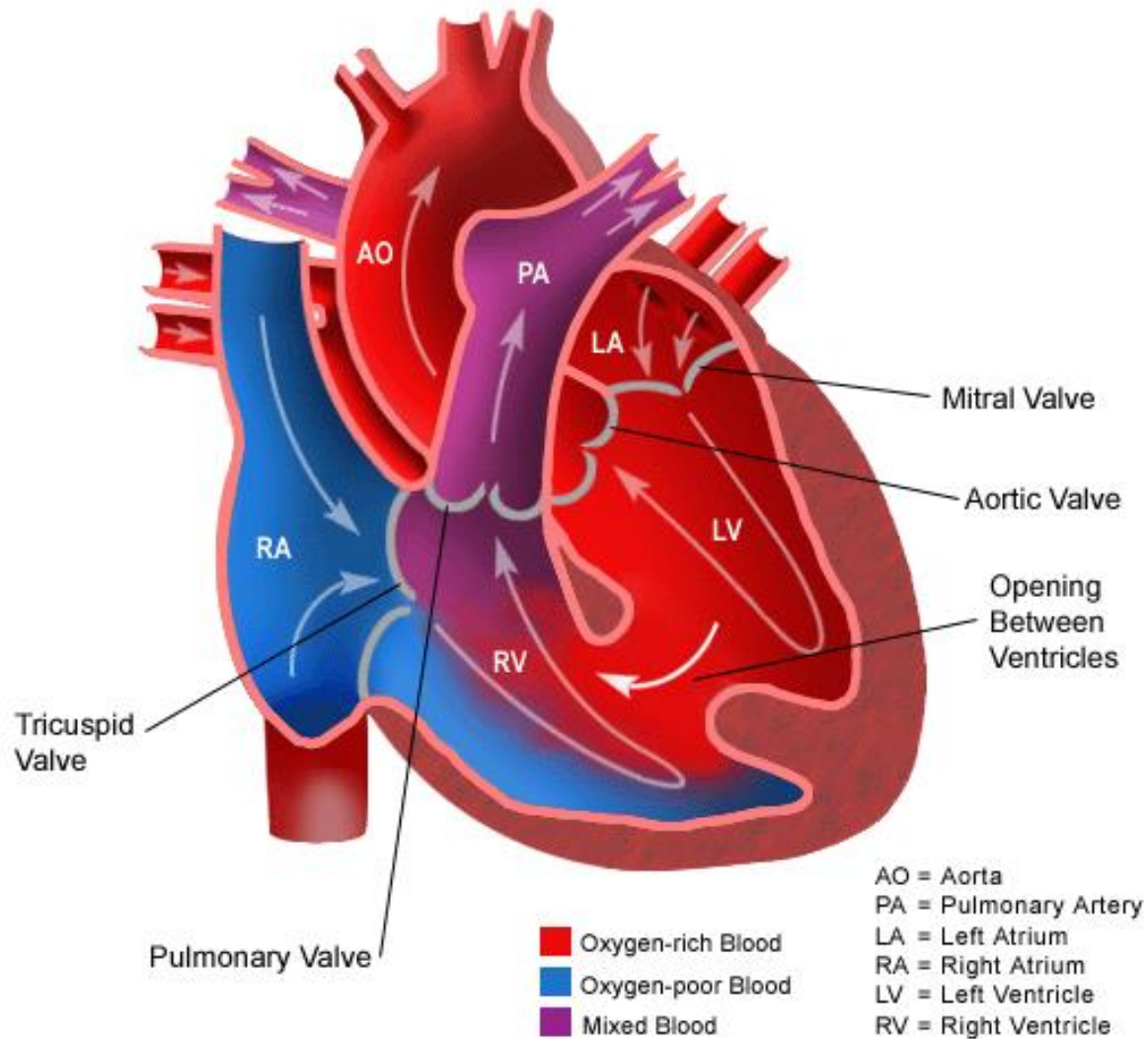


De presentatie van een aangeboren hartafwijking

- Prenatale diagnose: 20 weken echo
- Na de geboorte:
 - Cyanose
 - Tachypnoe, dyspnoe, voedings problemen
 - Hartfalen, (prenataal: hydrops foetalis)
 - Hart ruisje
 - Ritmestoornis

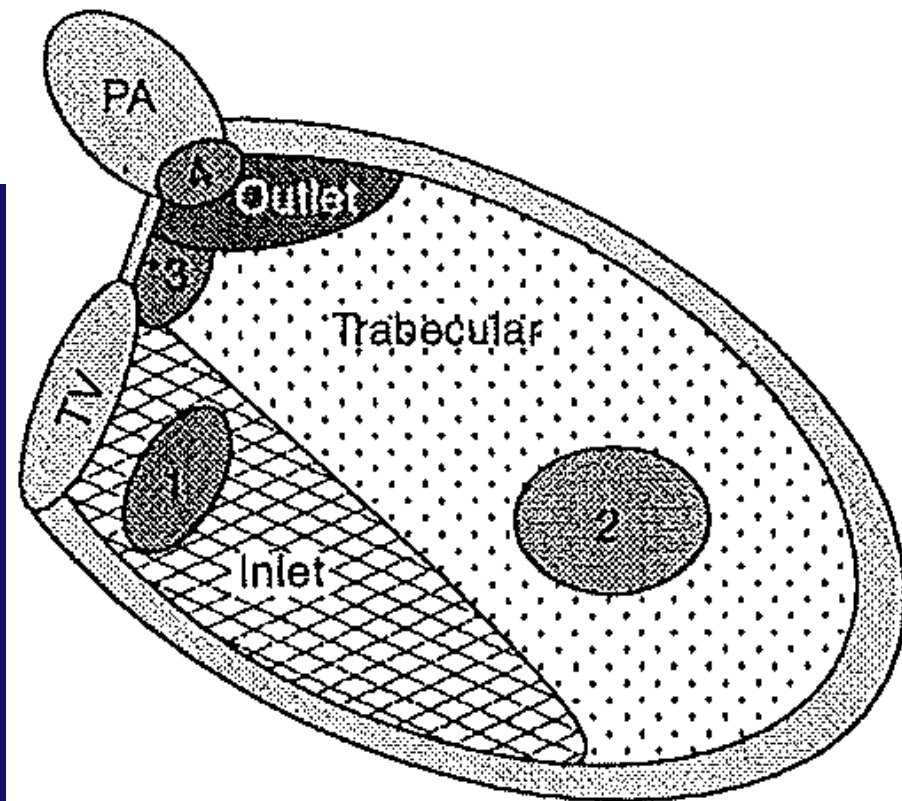
Ventrikel septum defect

Ventricular Septal Defect (VSD)

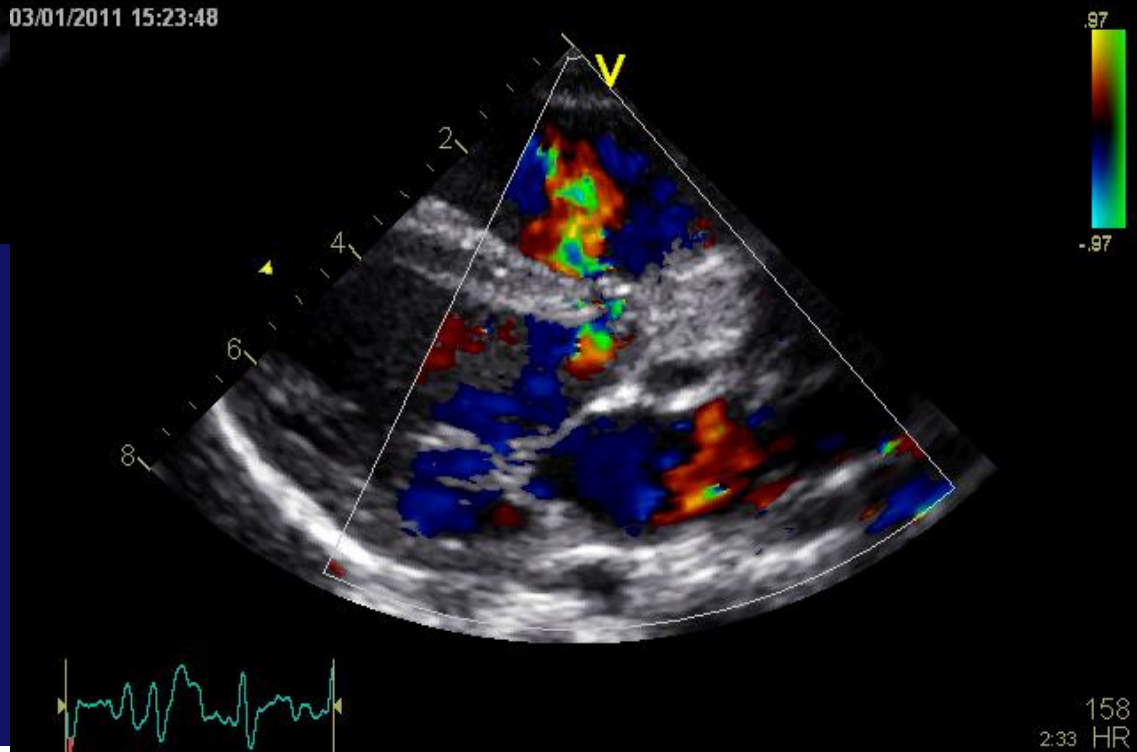
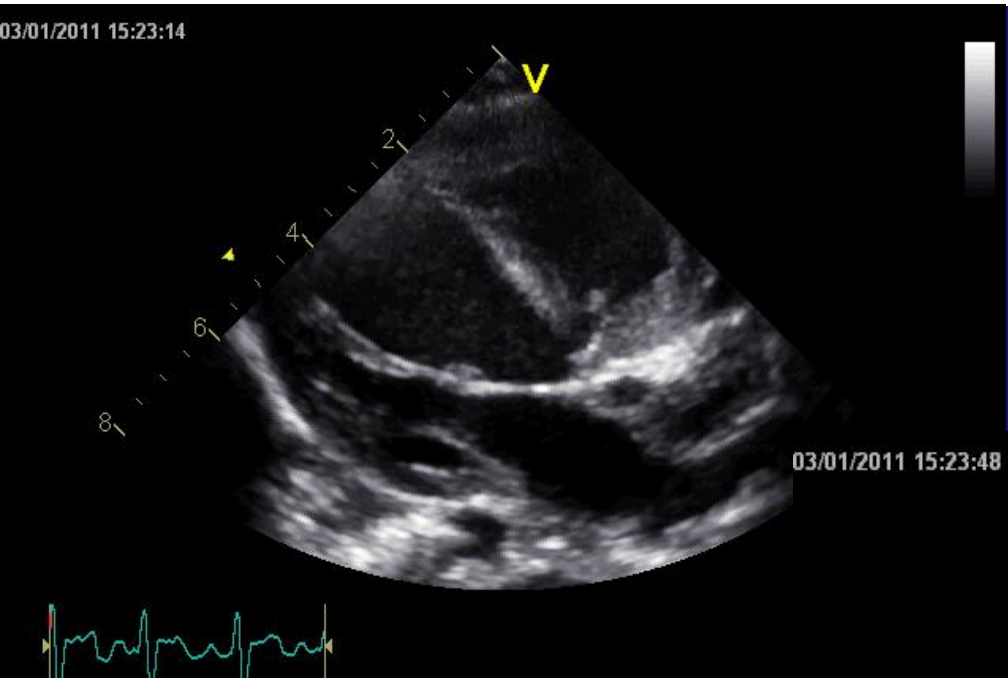


Ventricular septal defect

- Types
 - Inlet
 - Muscular/trabecular
 - Perimebraneous
 - Outlet
- Presentation
 - Small VSD: cardiac murmur
 - Large VSD: tachypnea, dyspnea, feeding difficulties
- Increased risk for endocarditis-no prophylaxis

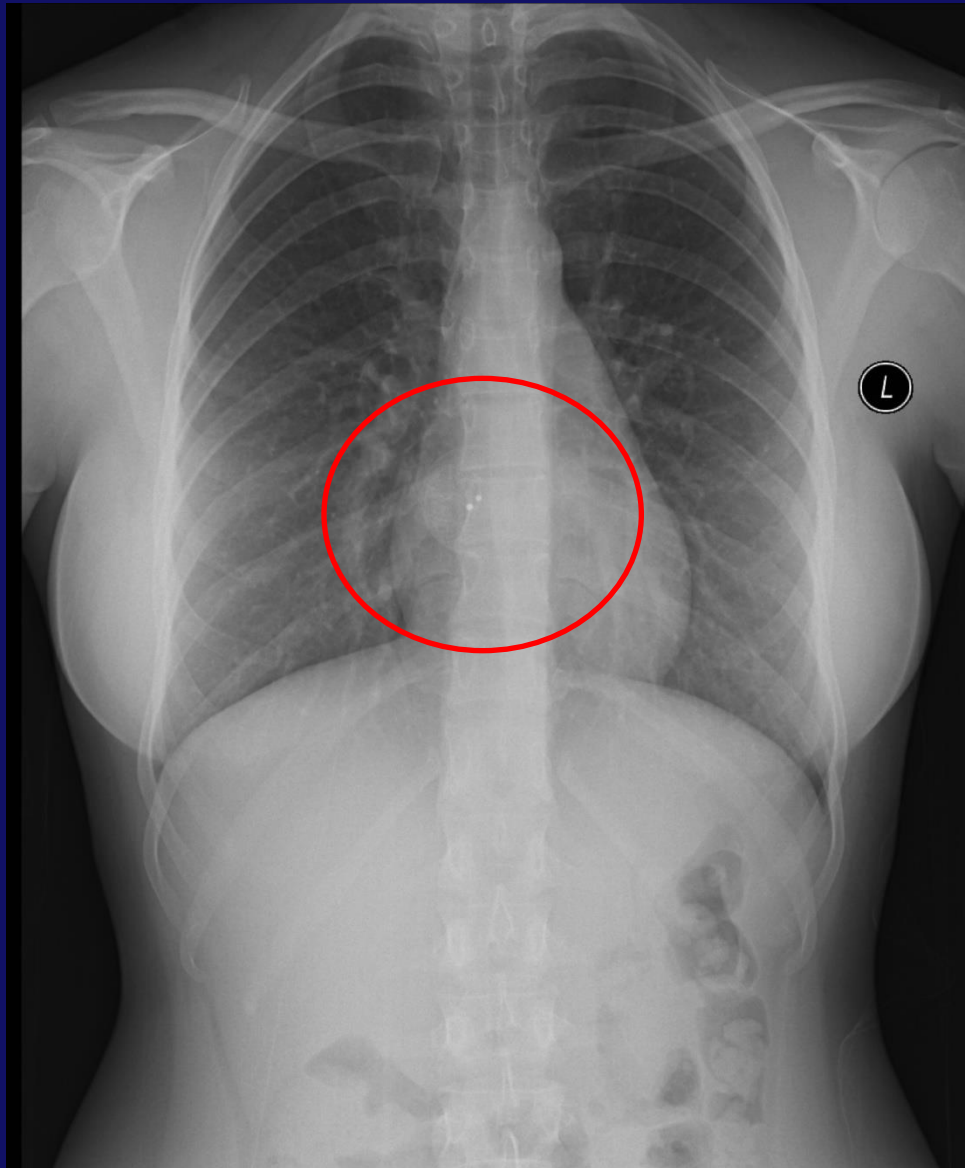


Ventricular septal defect-perimembraneous



Ventricular septal defect

- Treatment:
 - Often not needed: 70% close spontaneously
 - Signs and symptoms
 - Echocardiography
 - LV dilatation and/or dysfunction
 - Pulmonary (flow) hypertension-if untreated fixed
 - Aortic valve insufficiency in perimembraneous VSD
- Treatment options:
 - Interventional: increased risk for AV-block
 - Surgical:
 - Direct closure
 - Pulmonary artery banding if unfavorable location of VSD

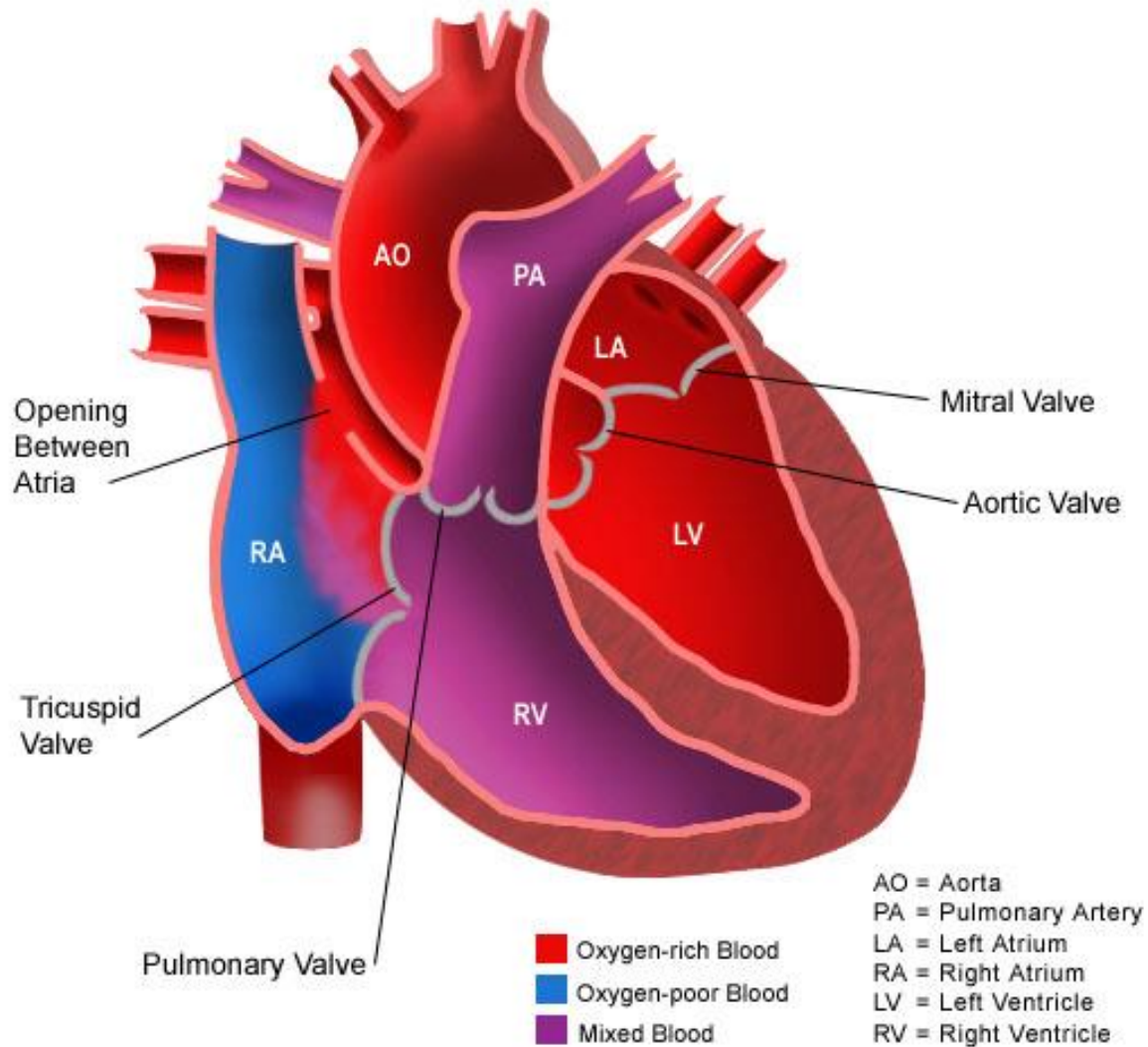


- Device om ASD te sluiten
- Device om VSD te sluiten
- Device om collateraal te sluiten
- Device om Ductus te sluiten

- Device om ASD te sluiten
- Device om VSD te sluiten
- Device om collateraal te sluiten
- Device om Ductus te sluiten

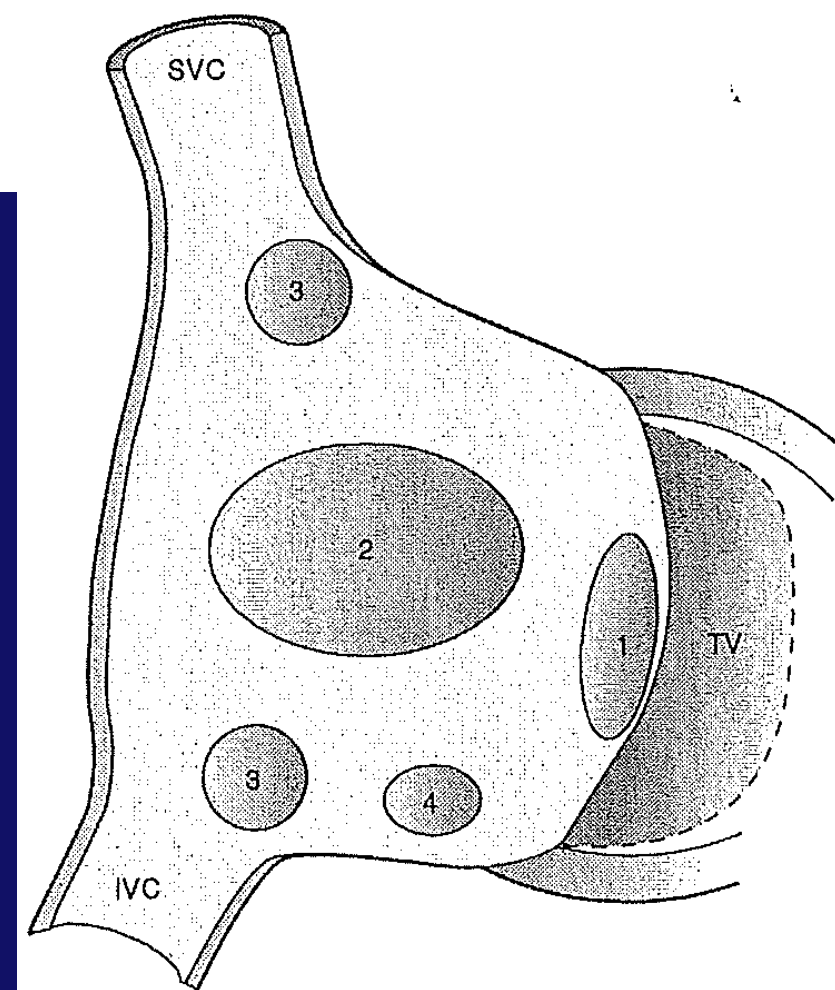
Atrium septum defect

Atrial Septal Defect (ASD)

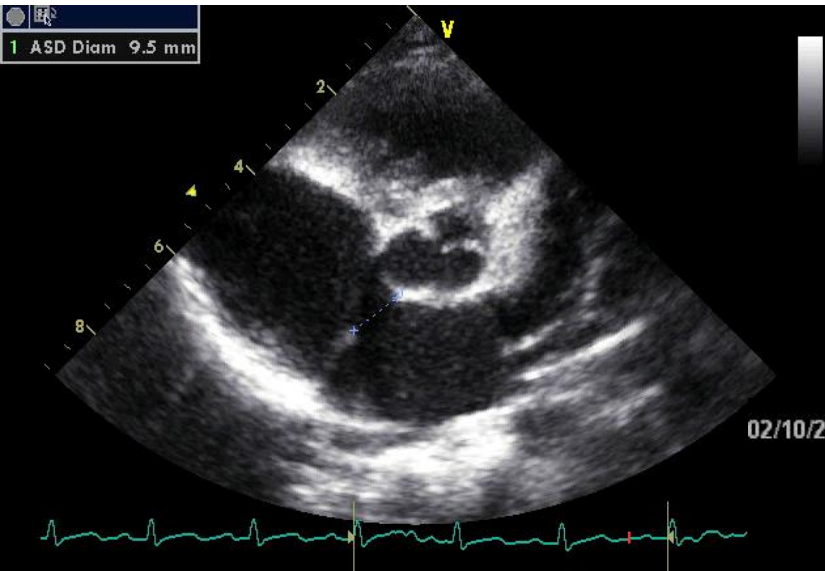


Atrial septal defect

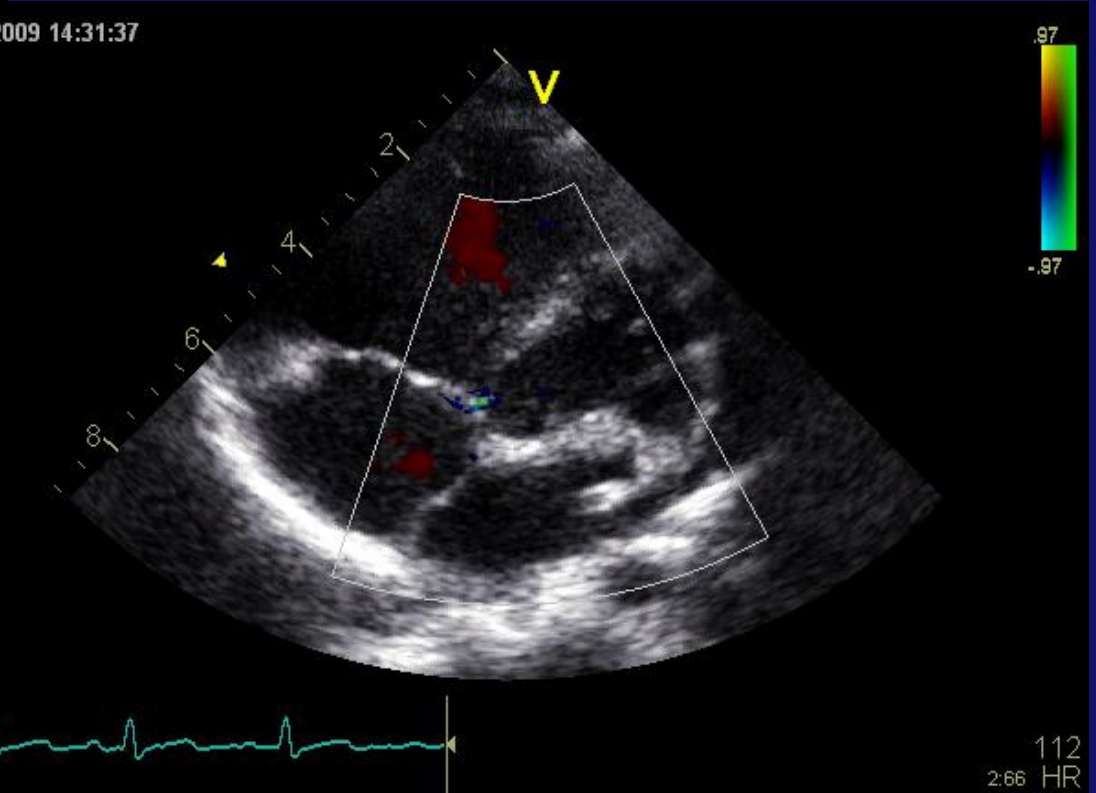
- Types;
 - ASD I-part of AVSD spectrum
 - ASD II-fossa ovalis defect
 - Sinus venosus defect
 - Sinus coronarius defect
- Presentation
 - Generally asymptomatic
 - Fixed second heart sound, PS
 - Childhood;
 - Tachypnea
 - Frequent respiratory tract infections, failure to thrive
 - Adulthood:
 - Dyspneu with exercise, fatigue
 - Arrhythmias



Atrial septal defect-secondum type



02/10/2009 14:31:37



112
2:66 HR

Atrial septal defect

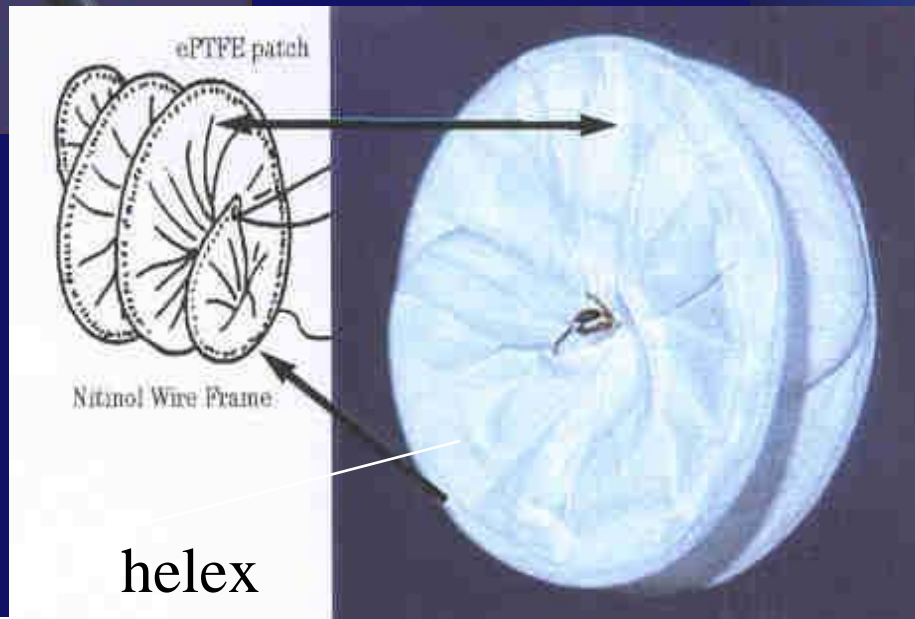
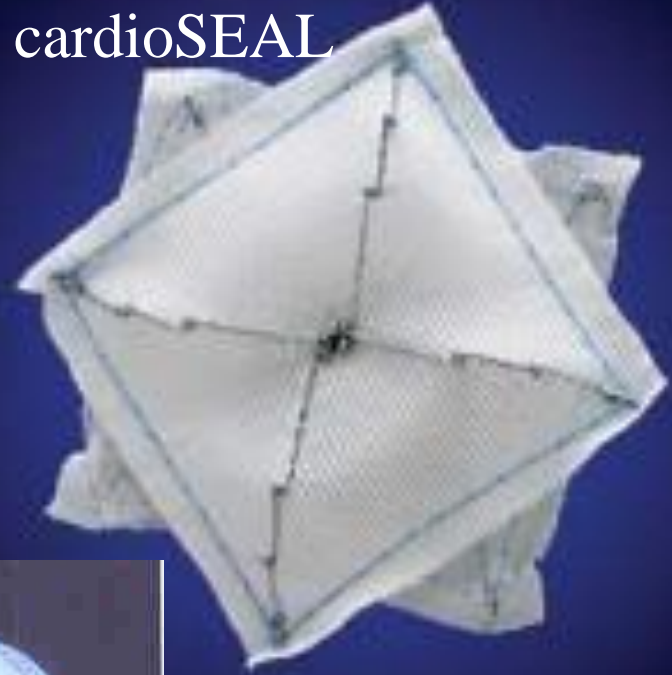
- Treatment:
 - ASD II < 3mm close spontaneously, >8mm unlikely to close
 - Signs and symptoms
 - Echocardiography
 - RA and RV dilatation
- Treatment options:
 - Interventional: ASD II with sufficient rims
 - Surgical:
 - ASD II with unfavorable anatomy
 - Other forms of ASD
- Follow-up:
 - Arrhythmias

Atrial septal occluders

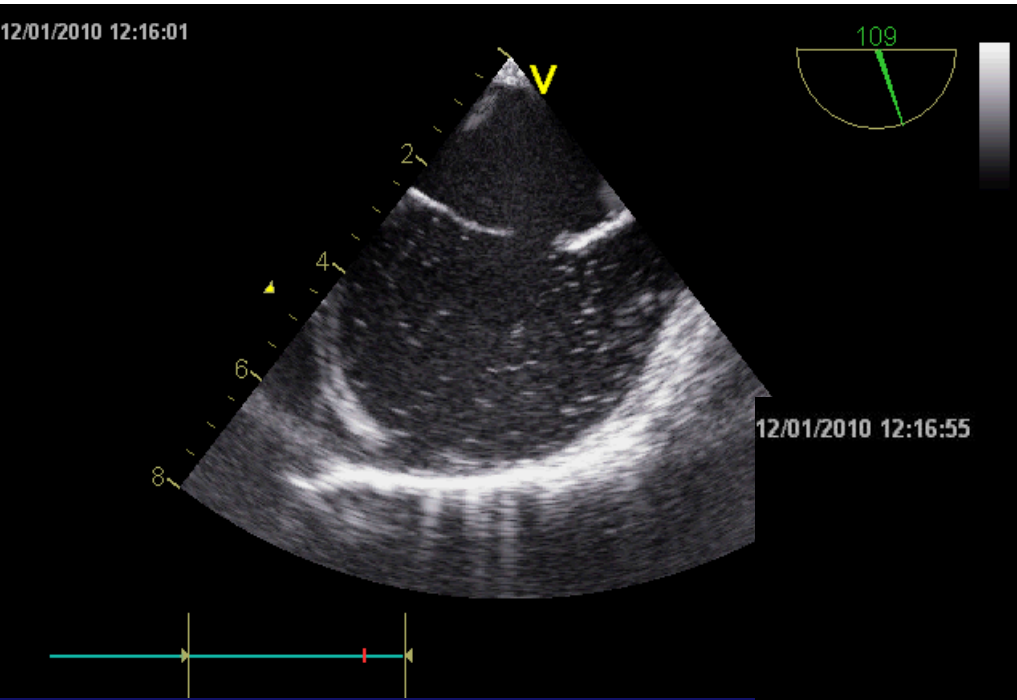
amplatzer



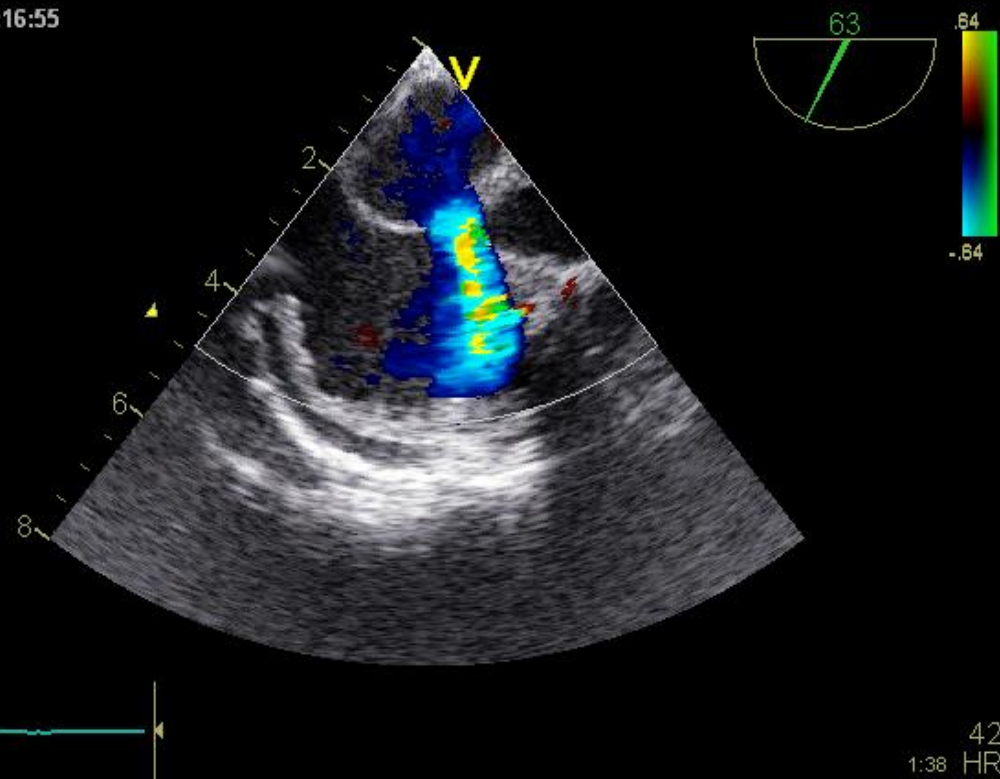
cardioSEAL



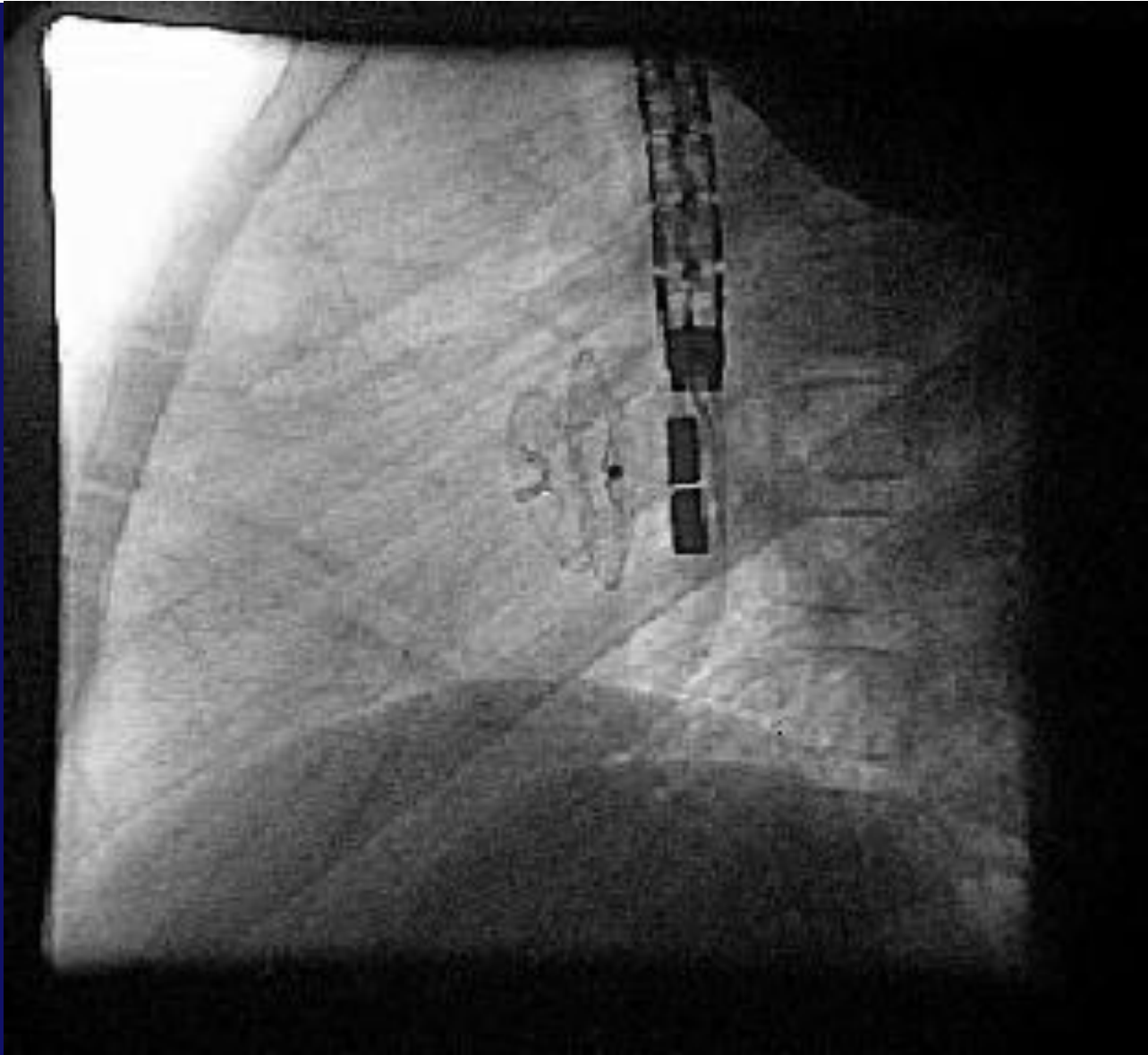
Atrial septal defect-interventional closure



12/01/2010 12:16:55

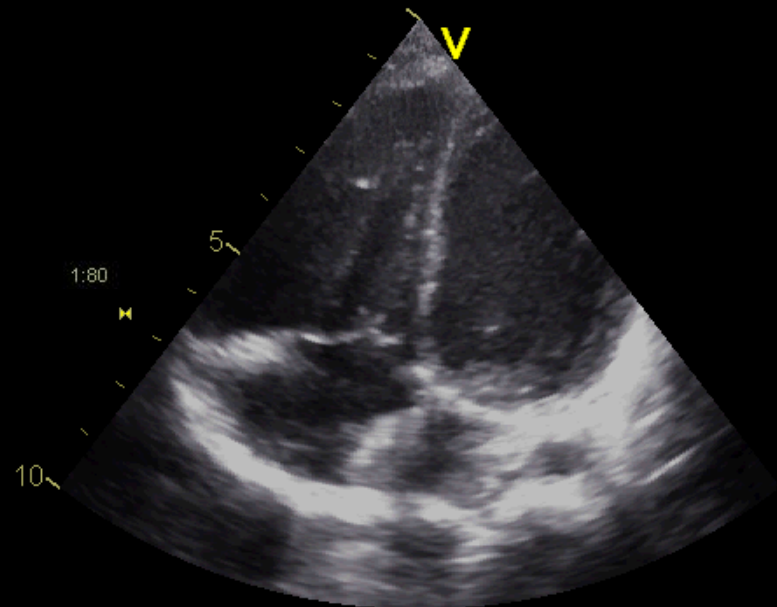
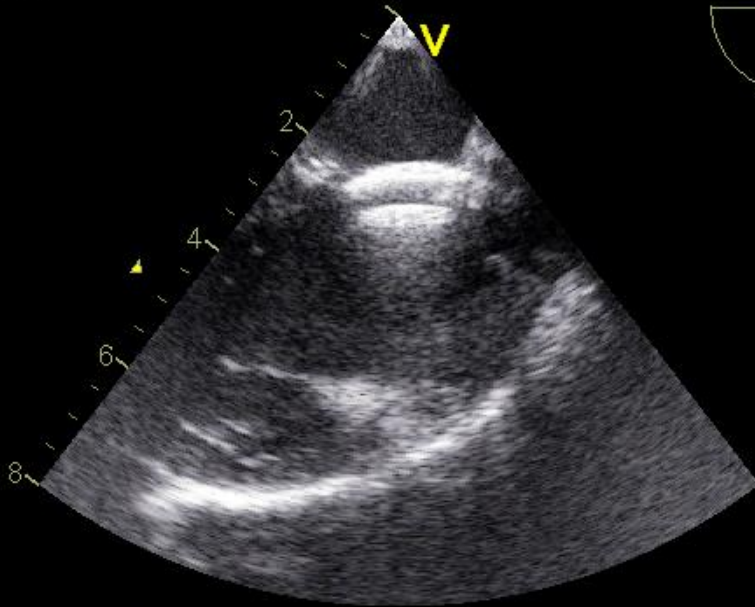


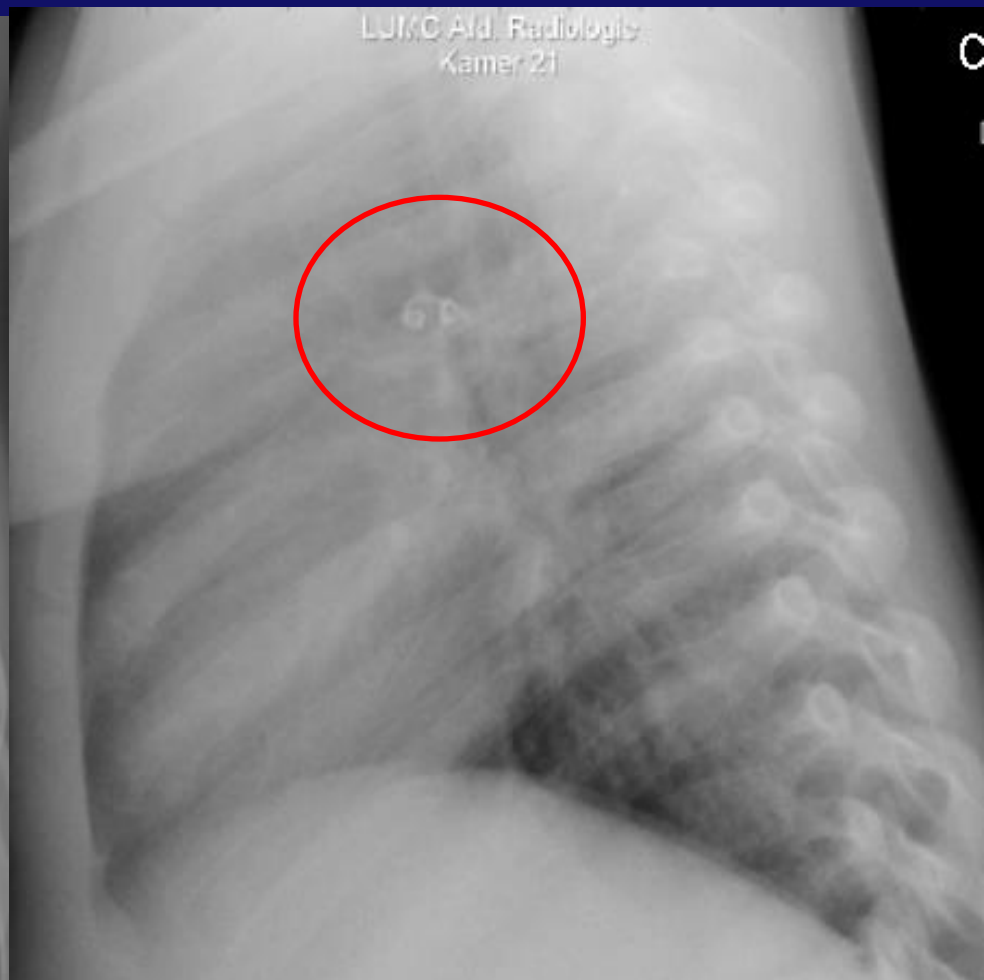
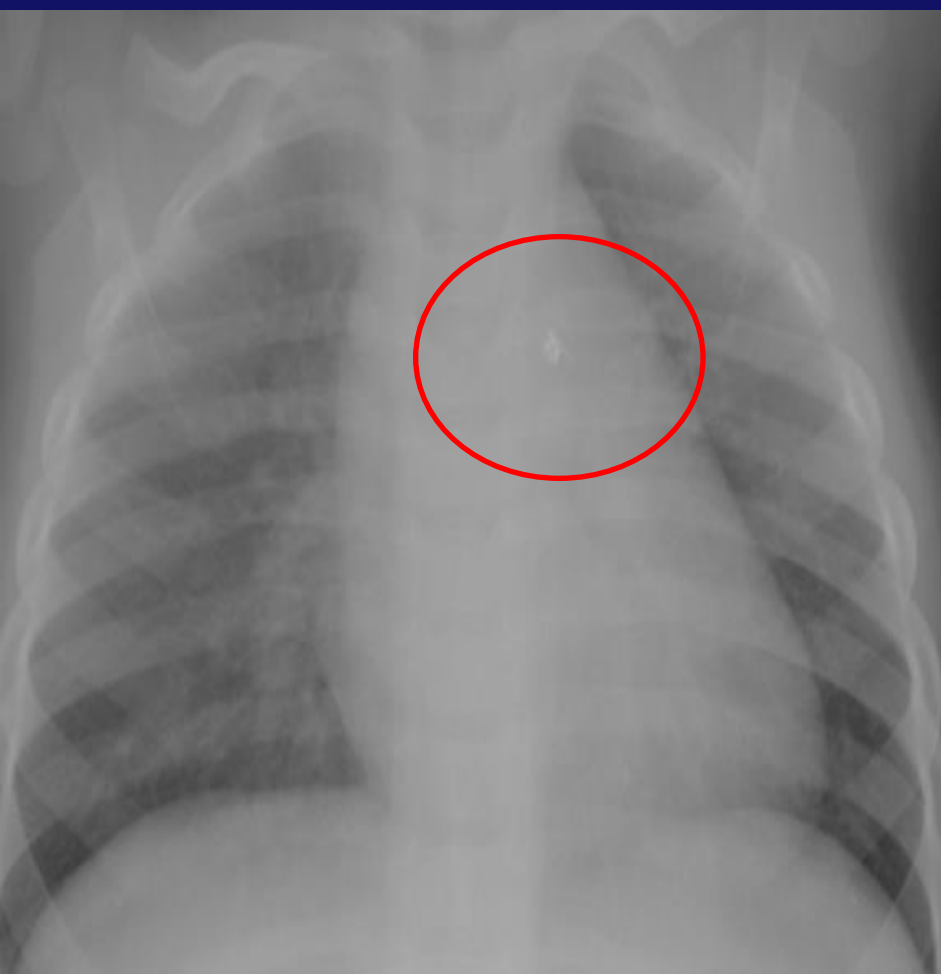
Atrial septal defect-interventional closure

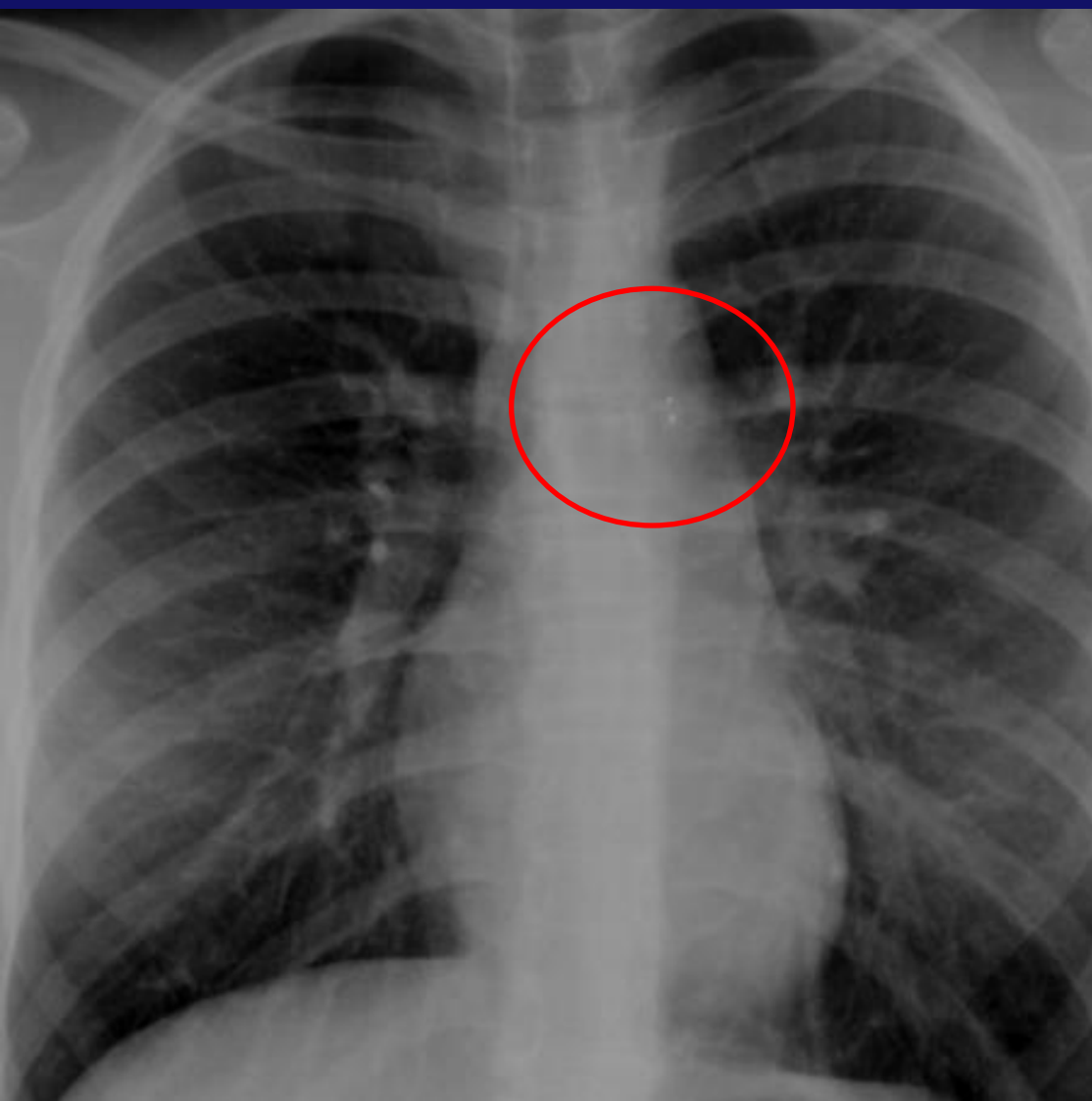


Atrial septal defect-interventional closure

12/01/2010 12:59:41



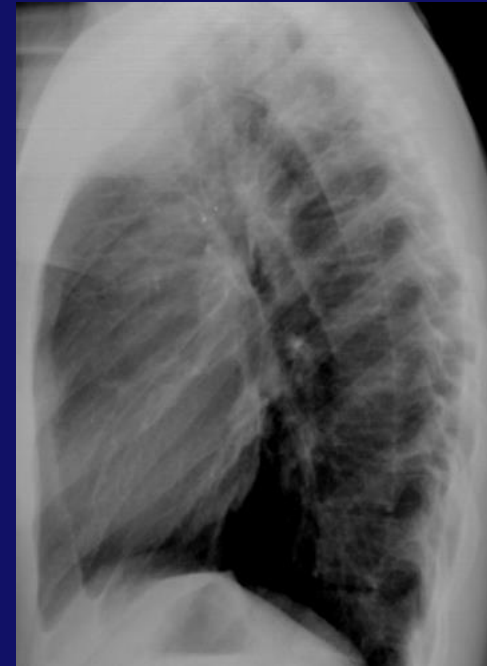




- Een stent om de ductus te sluiten
- Een coil om de ductus te sluiten
- Een plug om de ductus te sluiten
- Een chirurgische clip om de ductus te sluiten



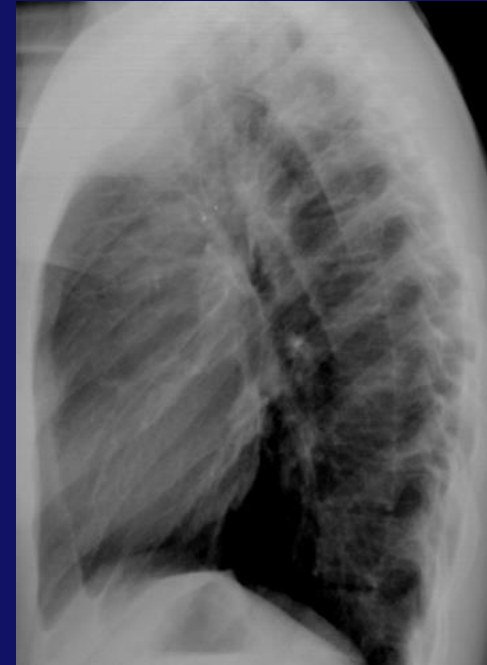
- Een stent om de ductus te sluiten
- Een coil om de ductus te sluiten
- Een plug om de ductus te sluiten
- Een chirurgische clip om de ductus te sluiten



- Een stent om de ductus te sluiten
- Een coil om de ductus te sluiten
- Een plug om de ductus te sluiten
- Een chirurgische clip om de ductus te sluiten

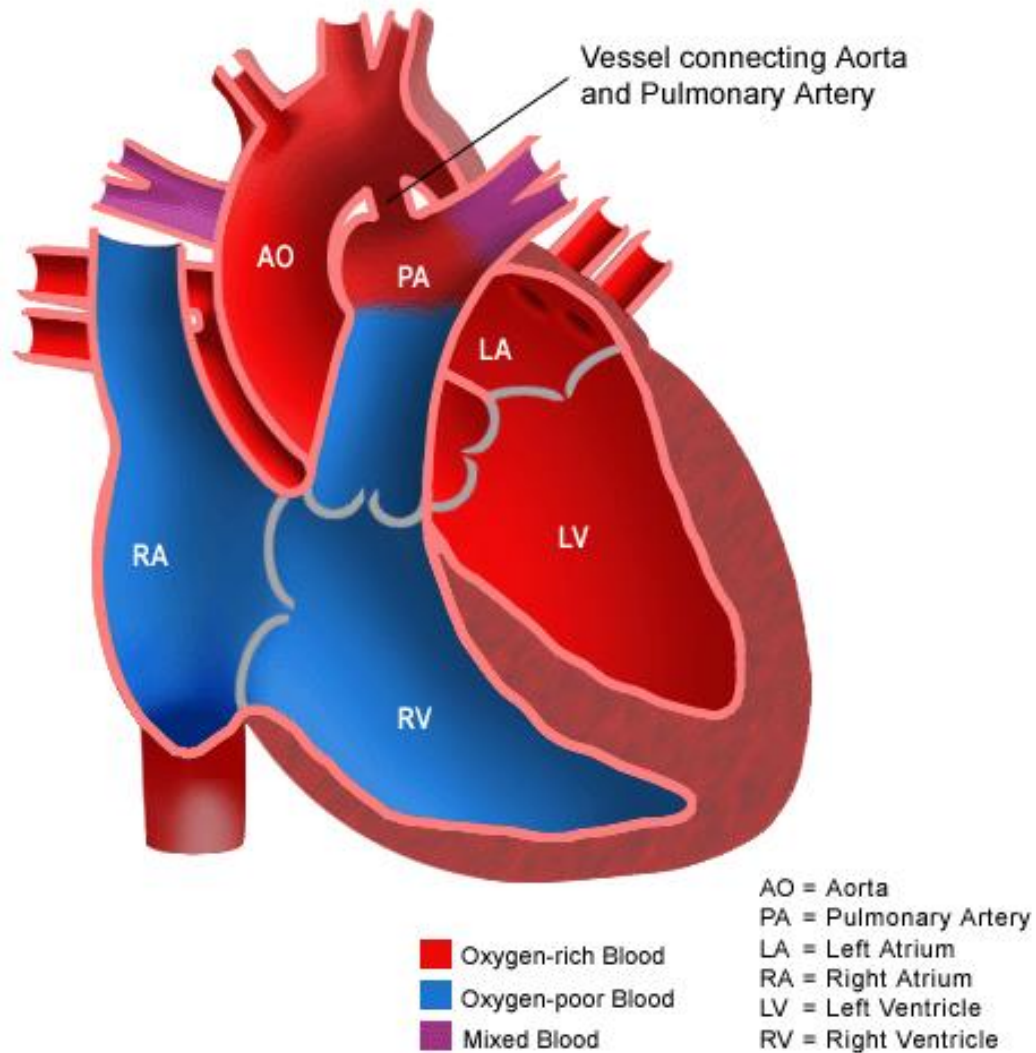


- Een stent om de ductus te sluiten
- Een coil om de ductus te sluiten
- Een plug om de ductus te sluiten
- Een chirurgische clip om de ductus te sluiten



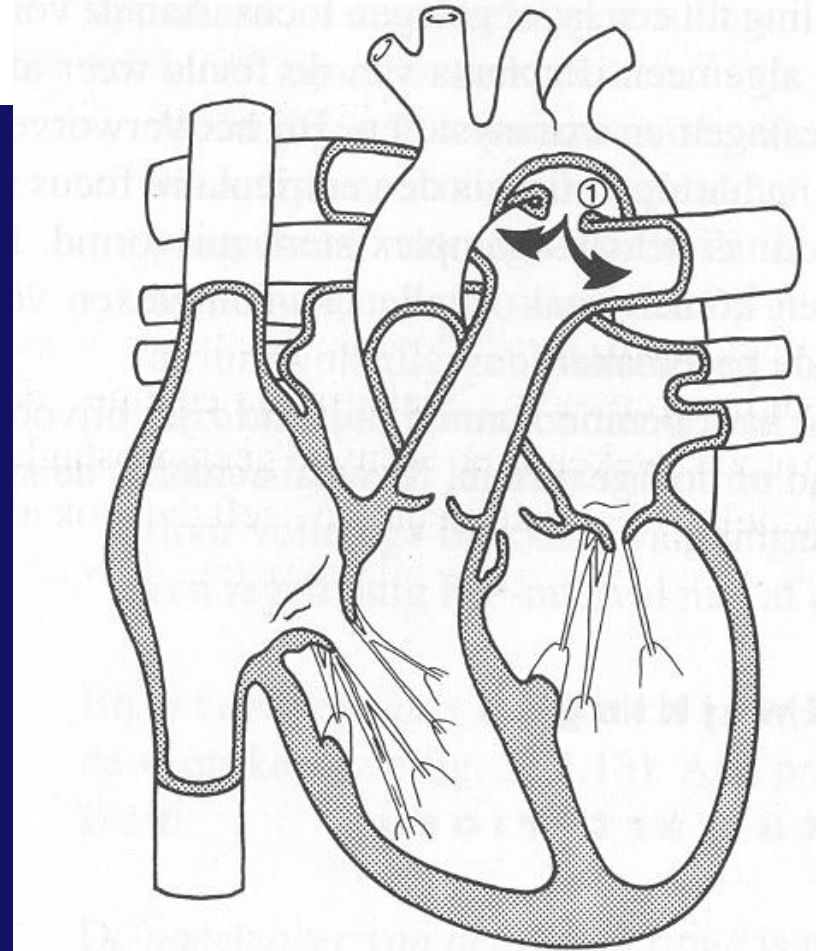
Persisterende ductus arteriosus

Patent Ductus Arteriosus (PDA)

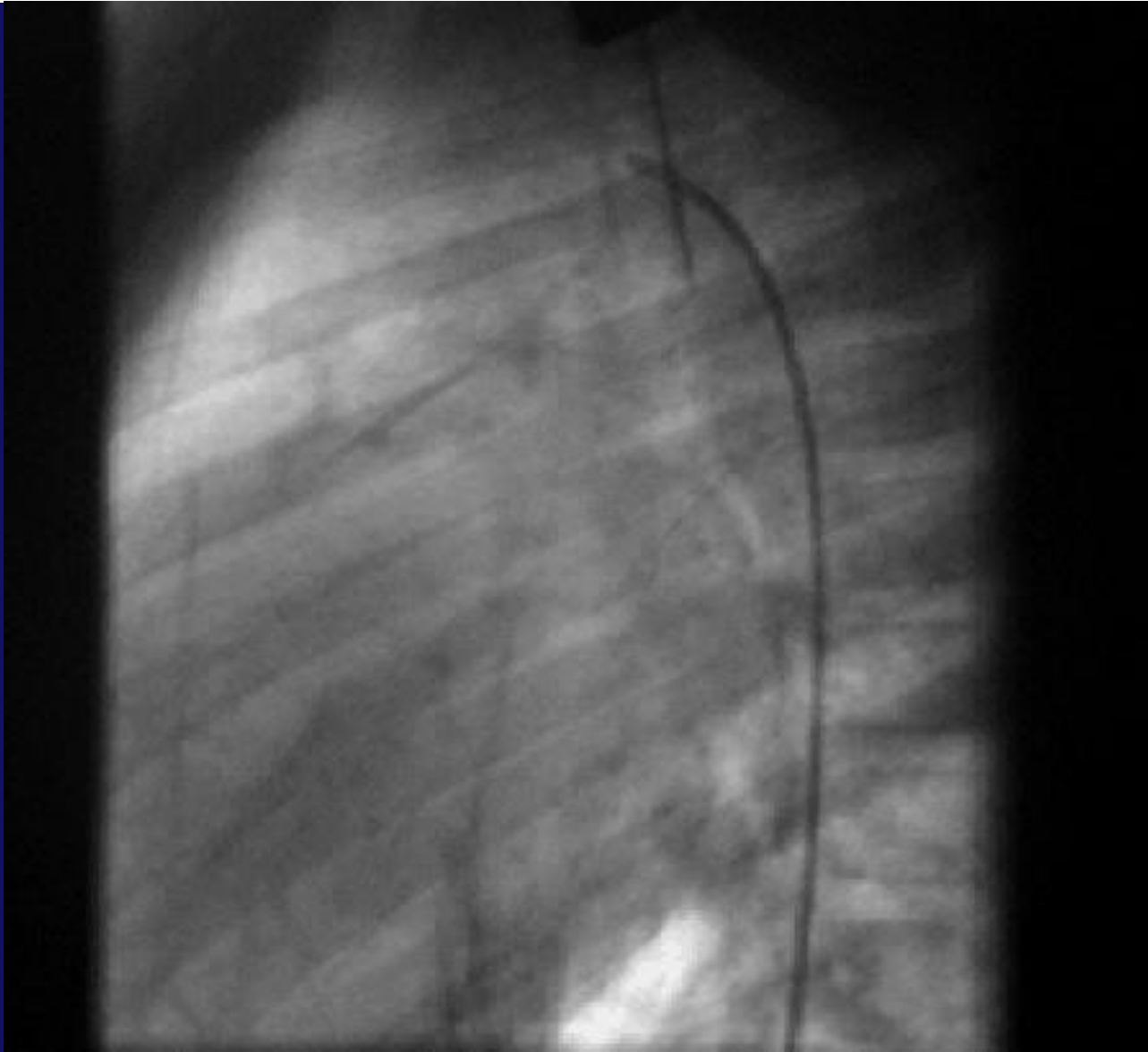


Patent ductus arteriosus

- Normal closure < first weeks
- Patients:
 - Term infant
 - Premature infant
- Term infant:
 - Large duct: cardiac failure
 - Small duct: murmur
 - Bounding groin pulses
- Preterm infant:
 - Common
 - Mostly spontaneous closure

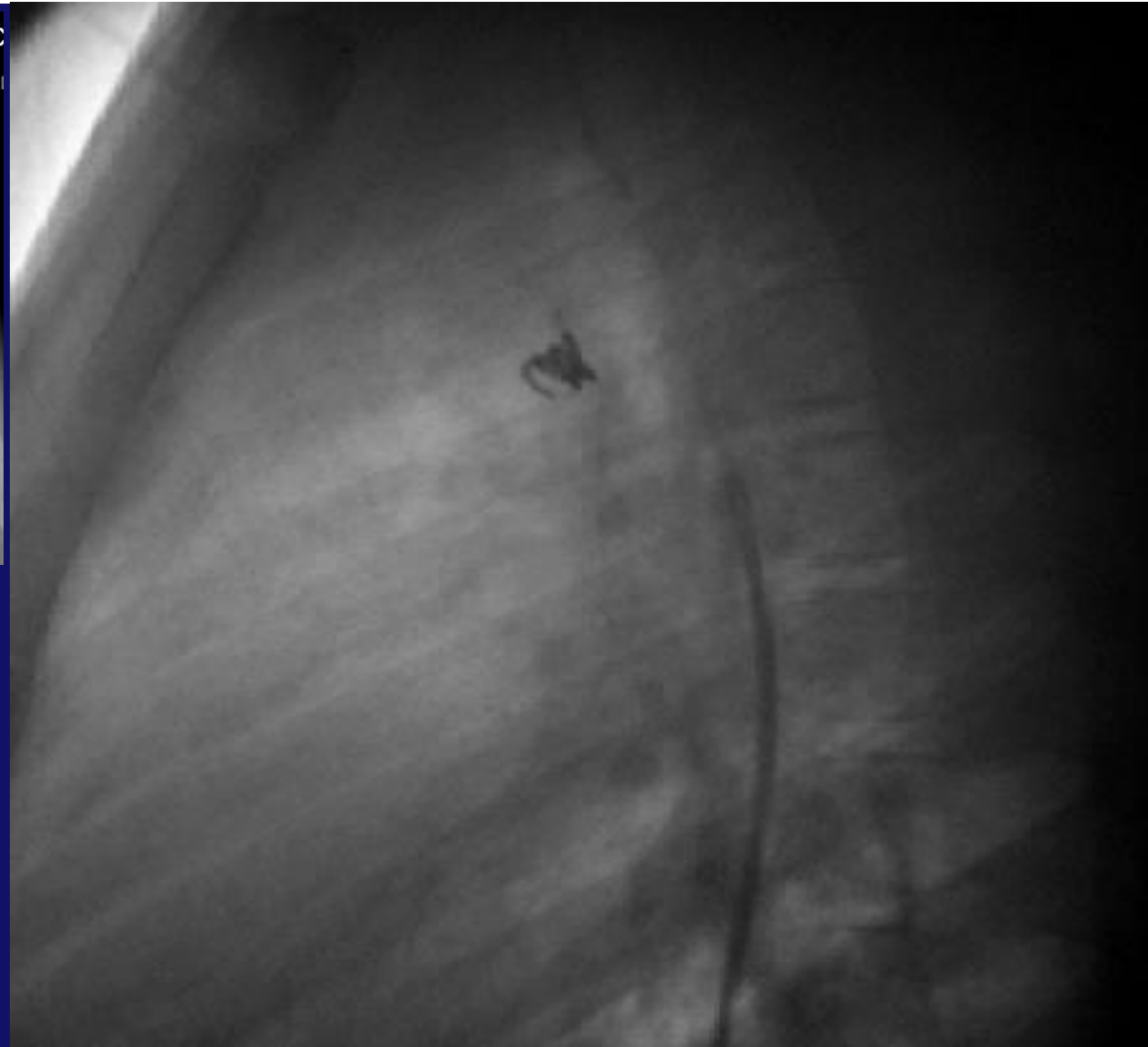


Patent ductus arteriosus

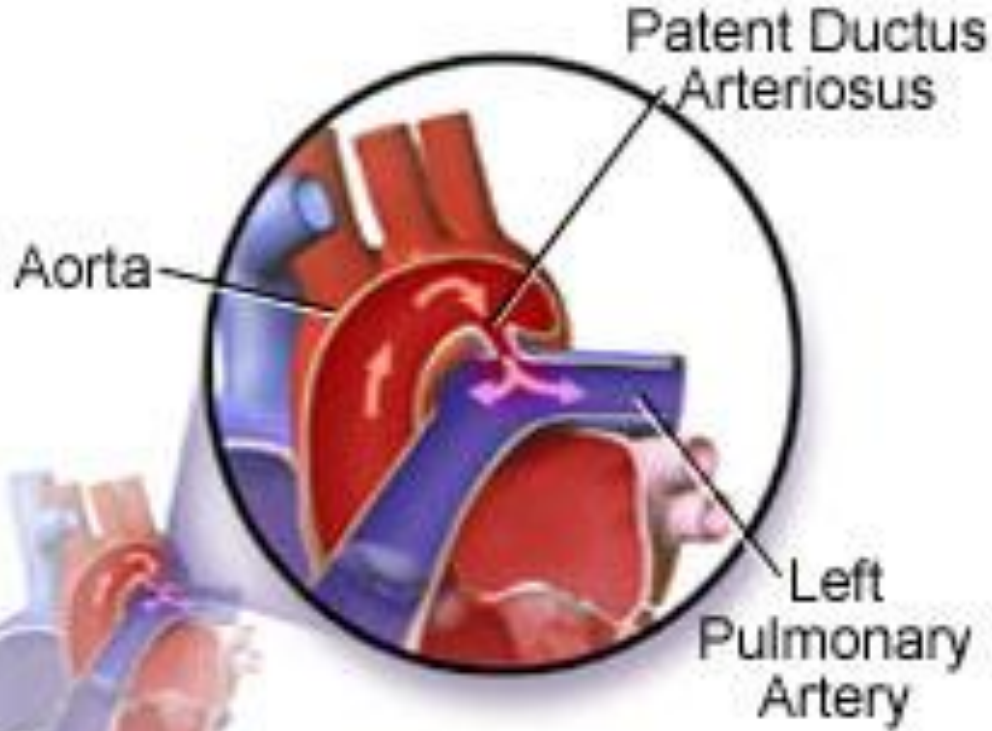


- Treatment;
 - Small duct: increased IE risk
 - Large duct: tachypnea, dyspnea, cardiac failure
 - Echocardiography:
 - LA and LV dilatation
 - Retrograde flow in abdominal aorta
- Treatment options:
 - Term infant:
 - Small duct: coil-plug
 - Large duct: ligation-plug
 - Preterm infant:
 - Pharmacological: ibuprofen, indomethacin
 - Surgical: ligation

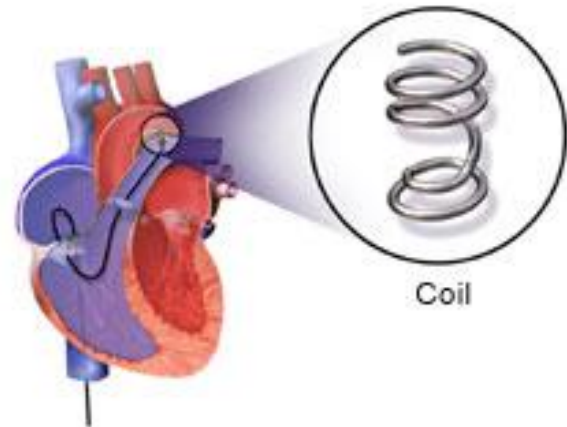
Ductus sluiting met coil



Ductus sluiting met coil

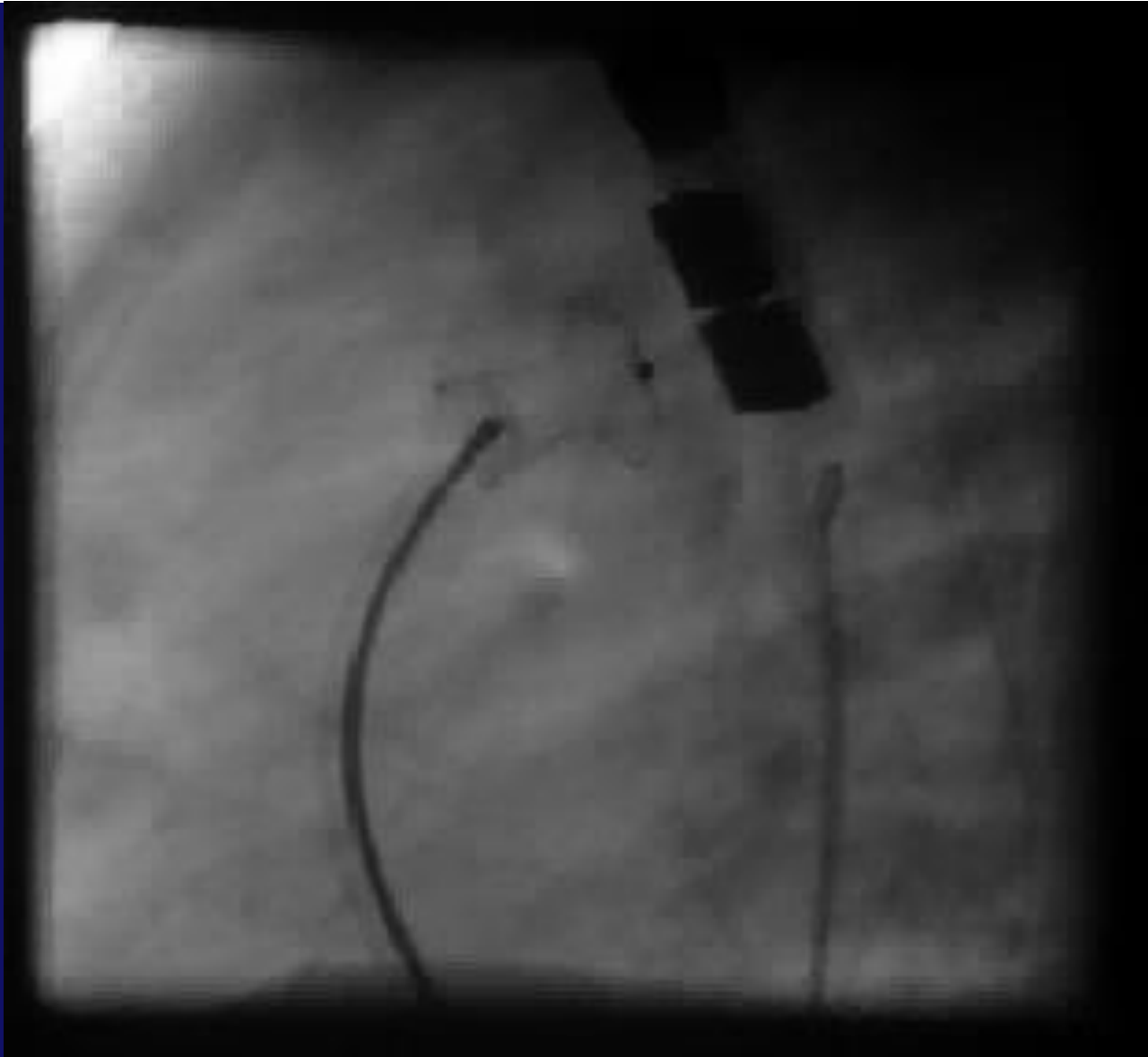


Blood Flow with Patent Ductus Arteriosus



Coil Closure of PDA





Ductus sluiting met plug

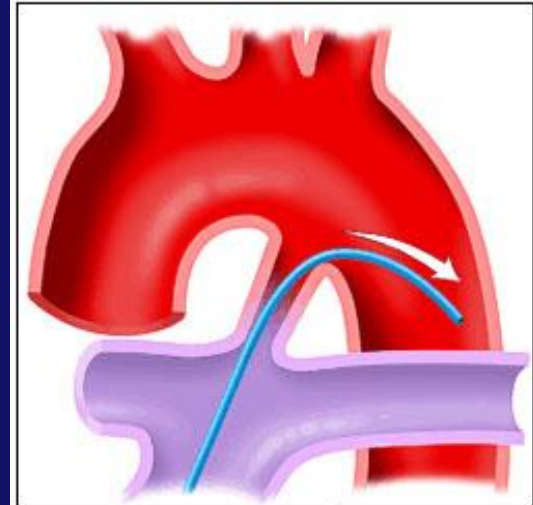
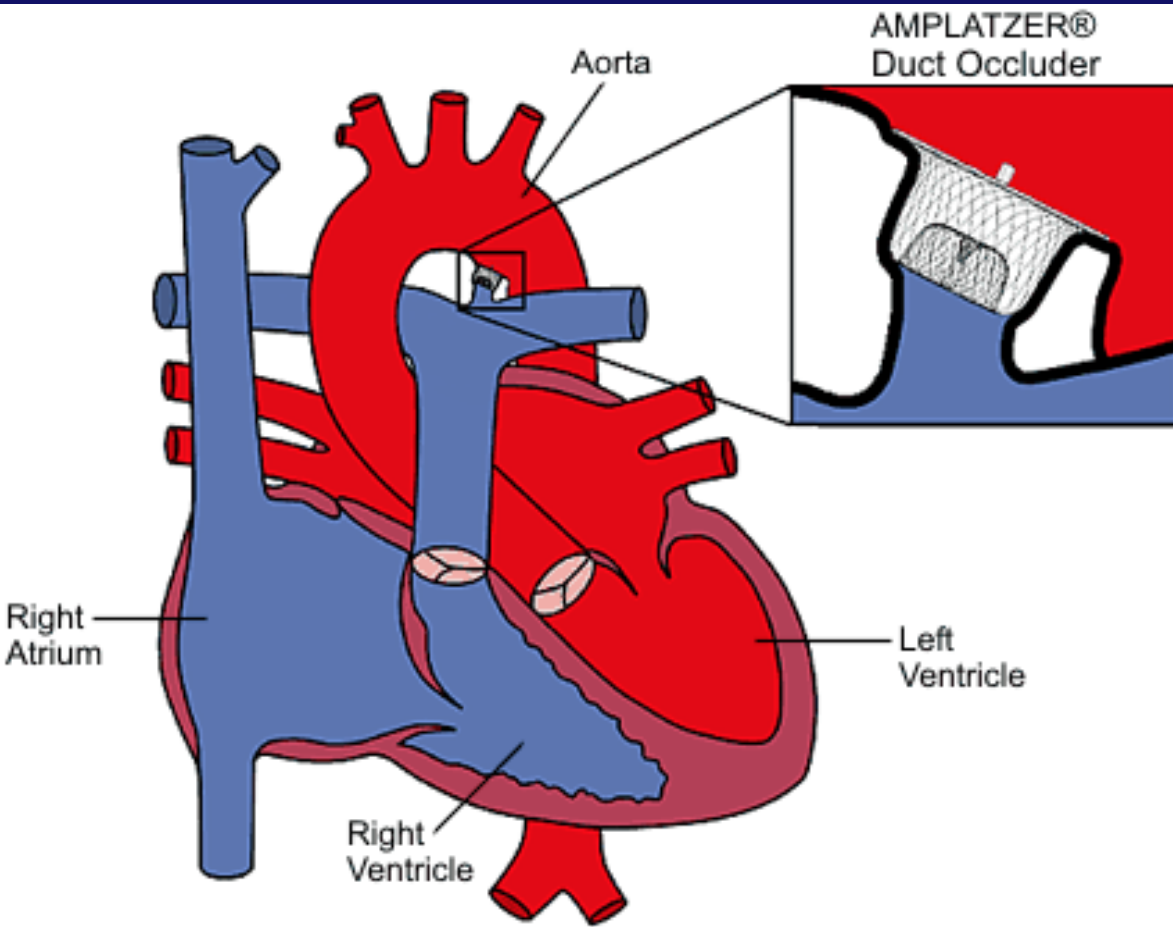
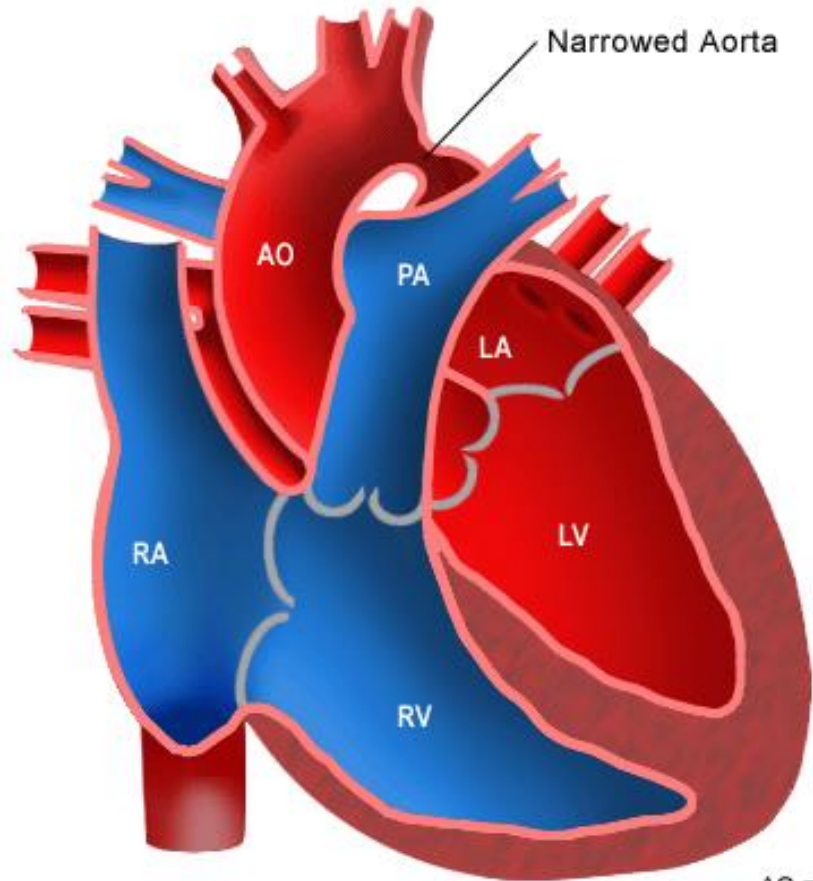


Figure 5



Coarctatie van de Aorta

Coarctation of the Aorta

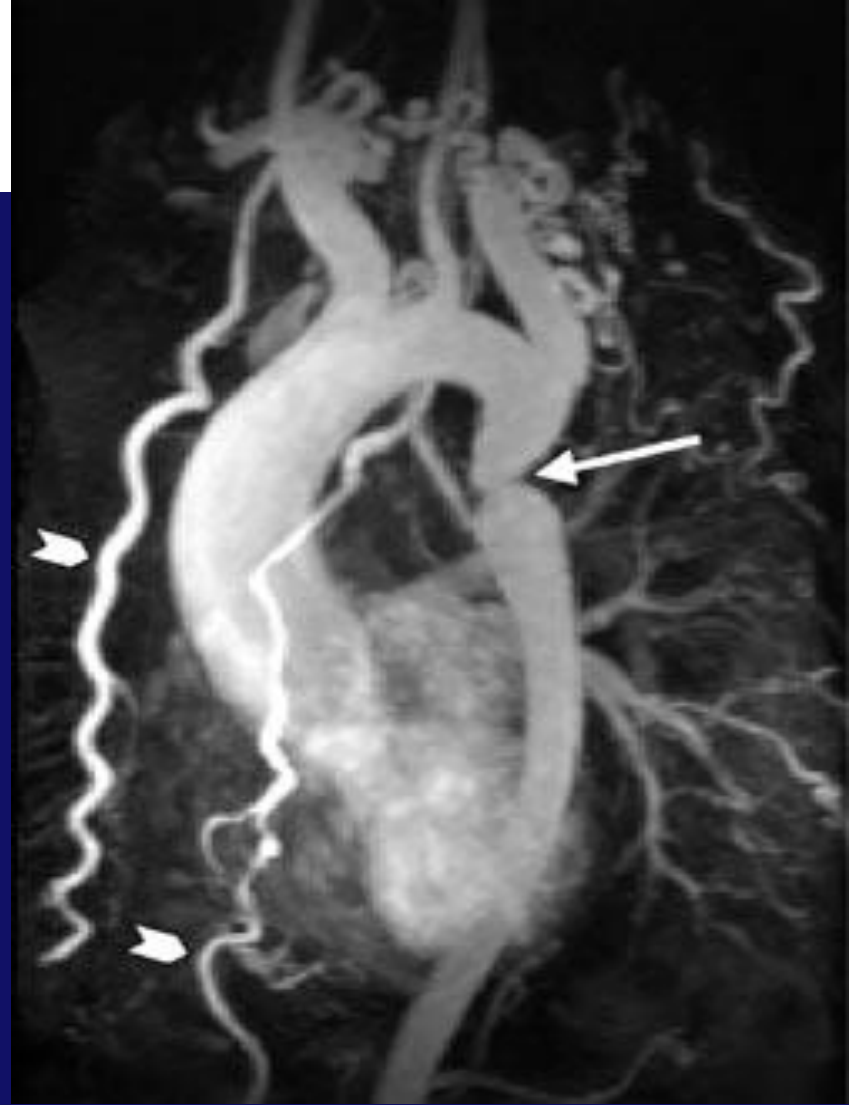


■ Oxygen-rich Blood
■ Oxygen-poor Blood

AO = Aorta
 PA = Pulmonary Artery
 LA = Left Atrium
 RA = Right Atrium
 LV = Left Ventricle
 RV = Right Ventricle

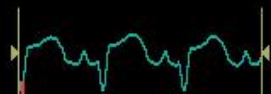
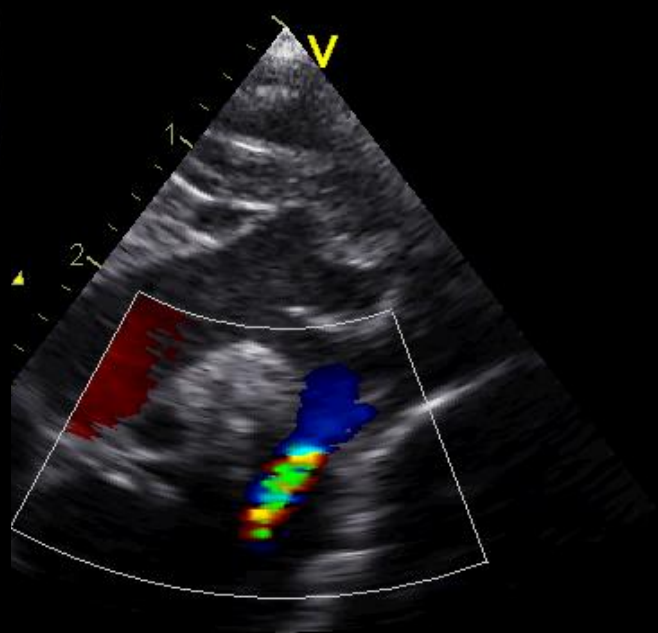
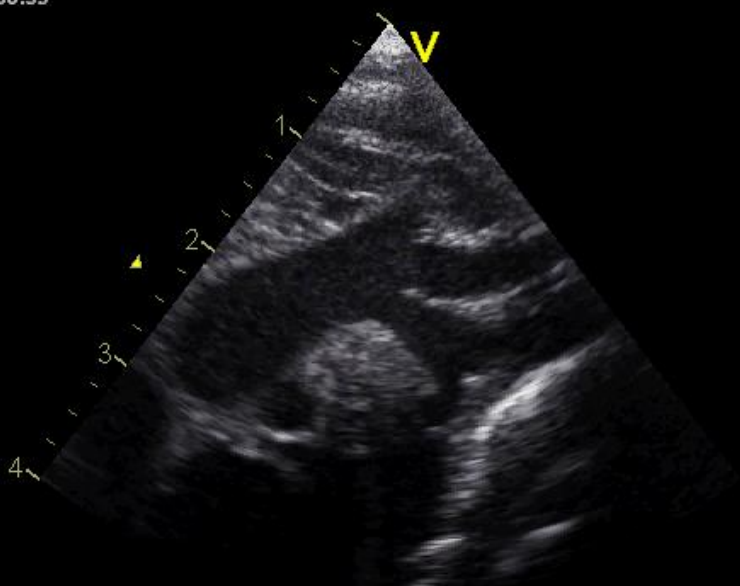
Coarctation of the Aorta

- Types:
 - From discrete narrowing
 - To interrupted aortic arch
- Presentation:
 - Asymptomatic
 - Hypertension
 - Severe coarctation:
 - Tachypnea, dyspnea
 - LV failure, shock, acidosis
- High incidence of bicuspid aortic valve

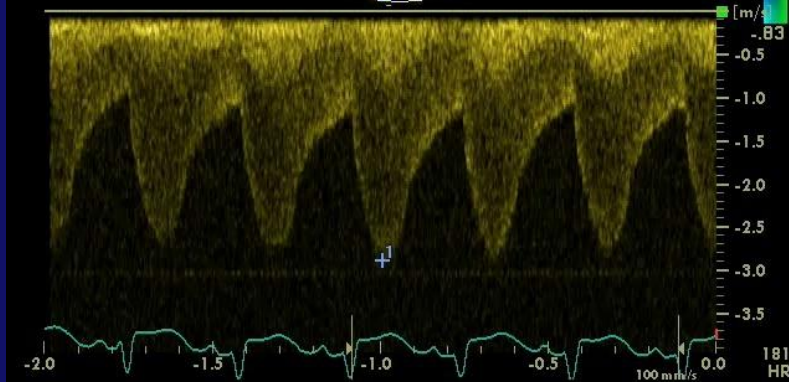


Coarctation of the Aorta

22/11/2010 14:08:39



1	DAo Vmax	2.90 m/s
	DAo maxPG	33.65 mmHg

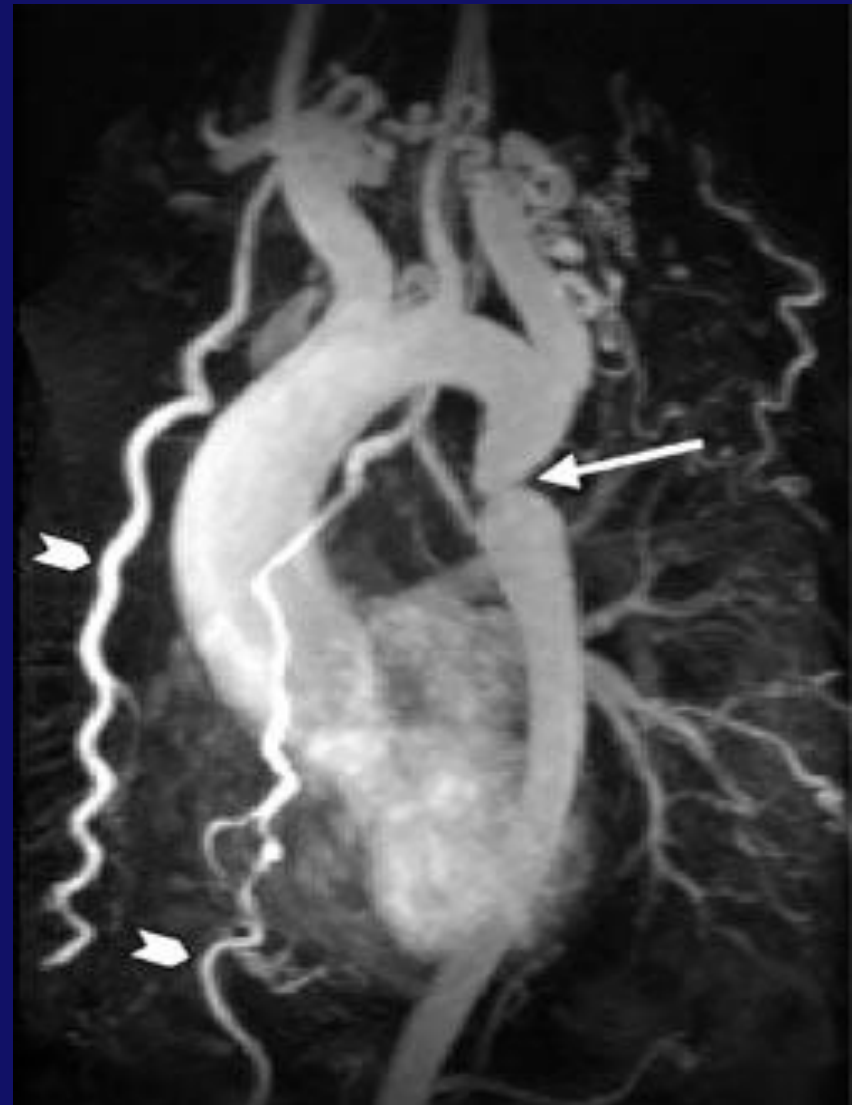
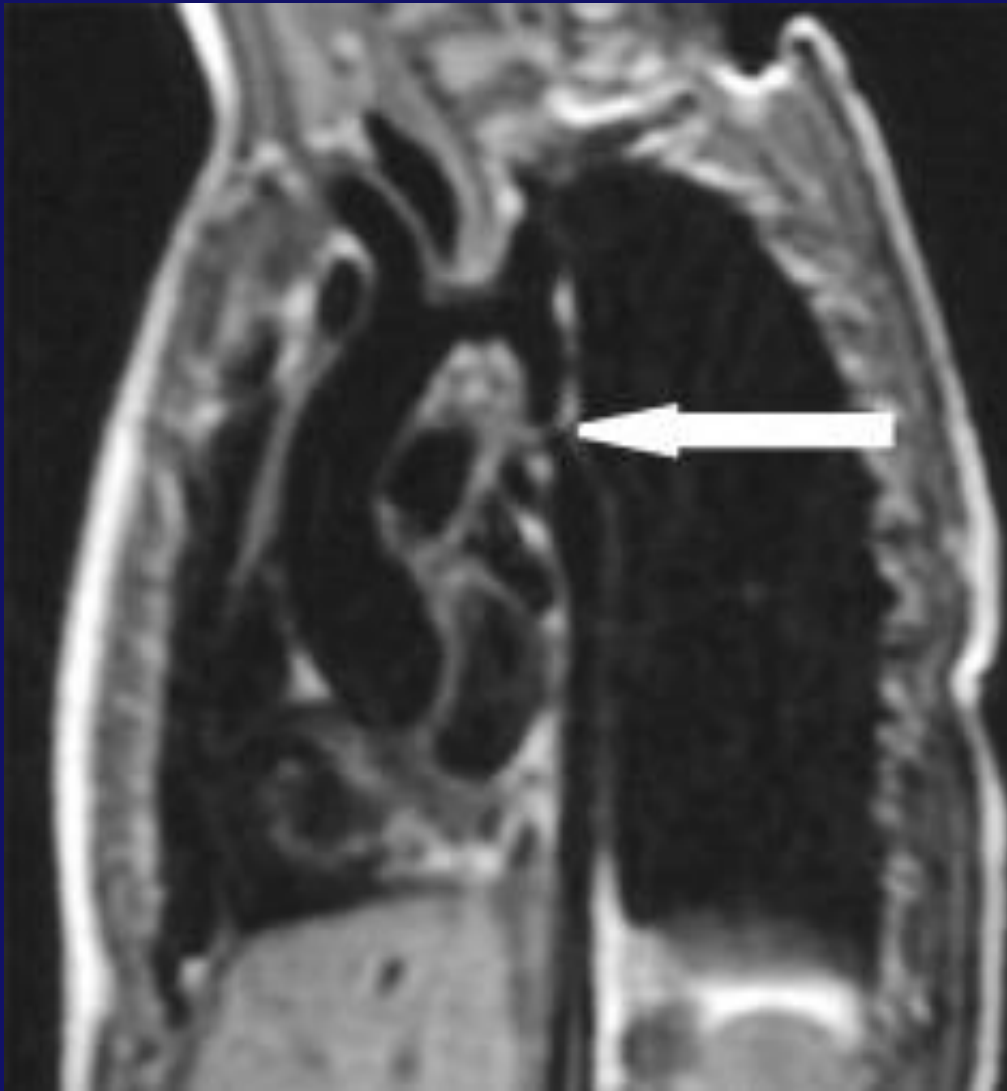


171
2:34 HR

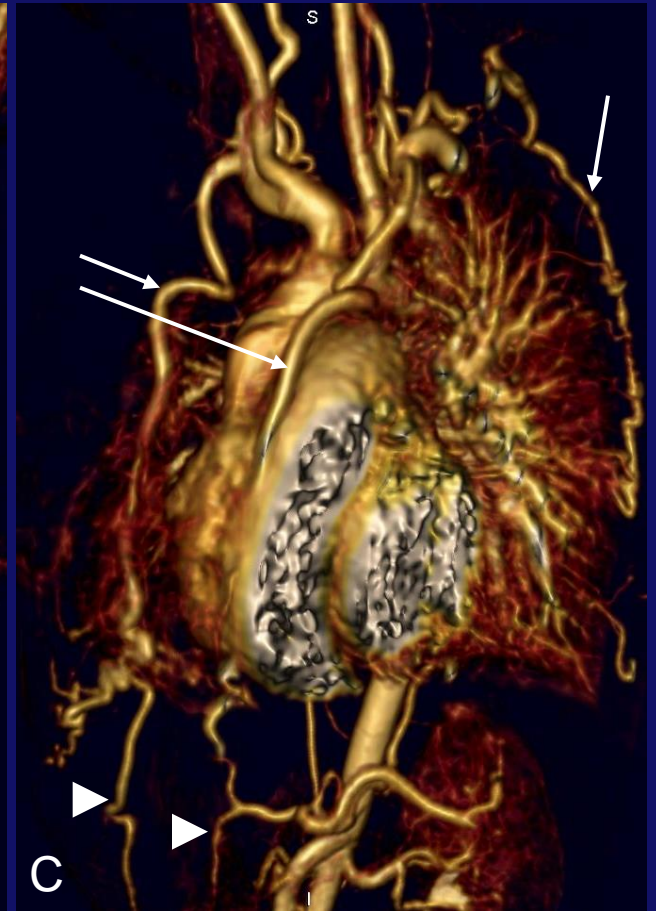
Coarctation of the aorta

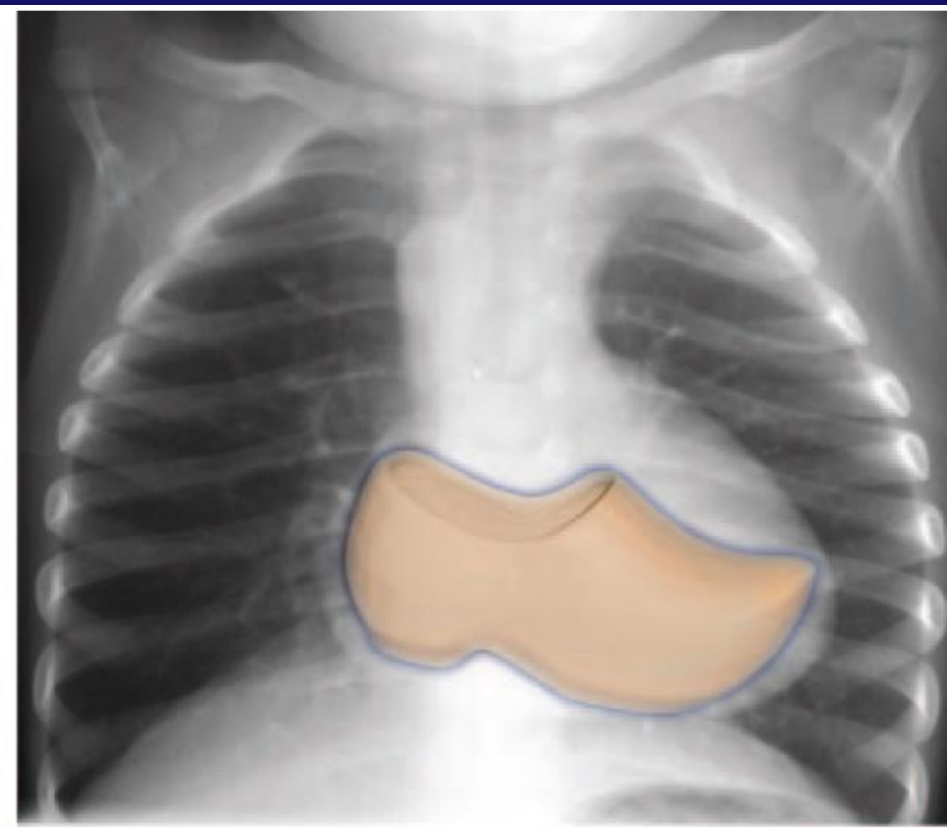
- Treatment:
 - LV failure due to severe stenosis
 - Hypertension
 - Echocardiography:
 - Narrowing desc aorta
 - Specific flow pattern: diastolic flow continuation
- Treatment:
 - Surgical: resection and end-to-end anastomosis
 - Interventional: re-coarct: balloon dilatation or stent
- Long-term complications
 - re-coarctation
 - aneurysm
 - Hypertension

MRI in Coarctatio Aortae



MRI in Coarctatio Aortae

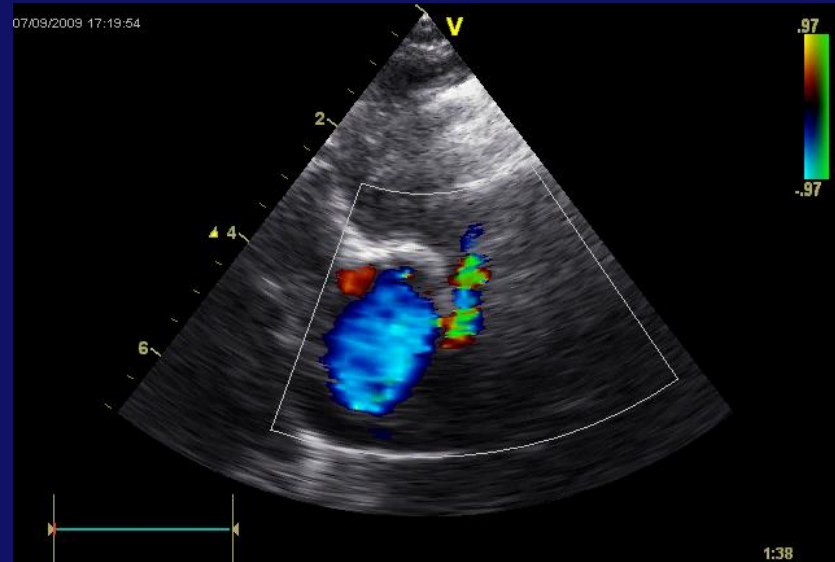
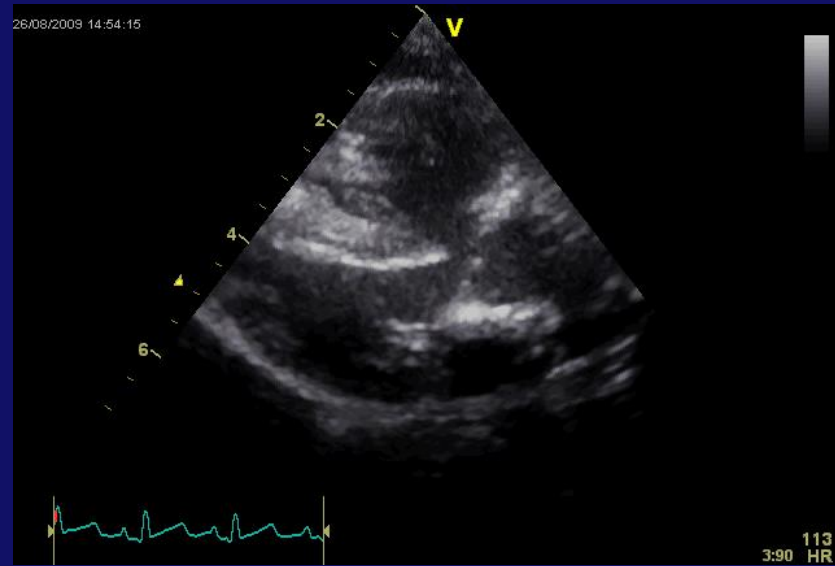
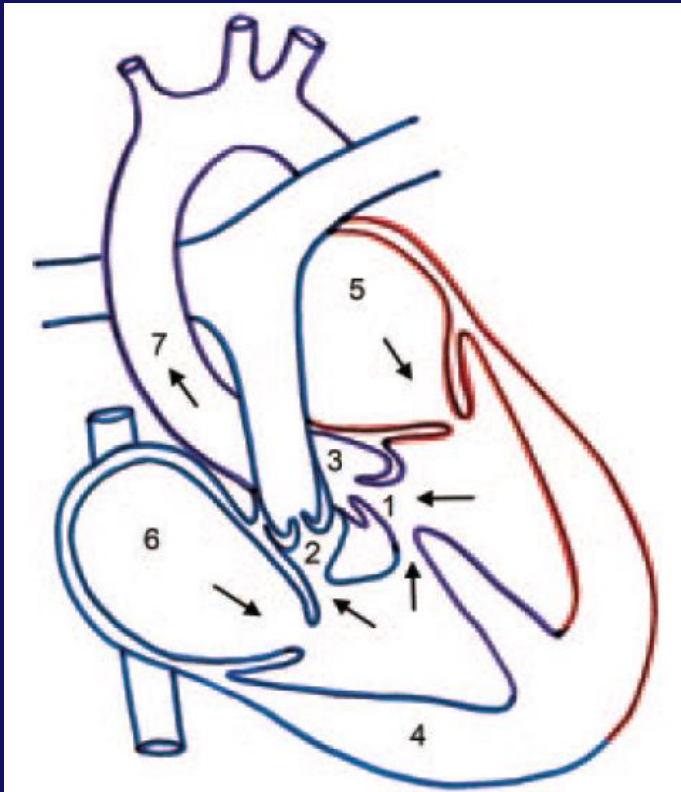
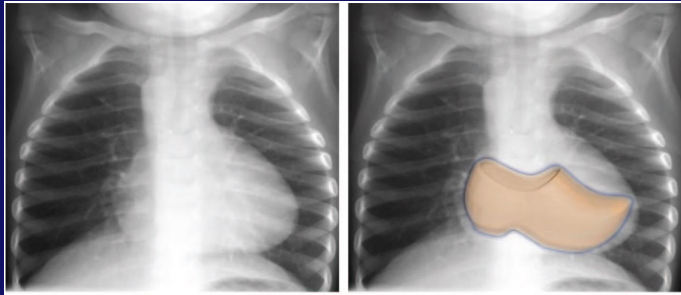




Welke hartafwijking

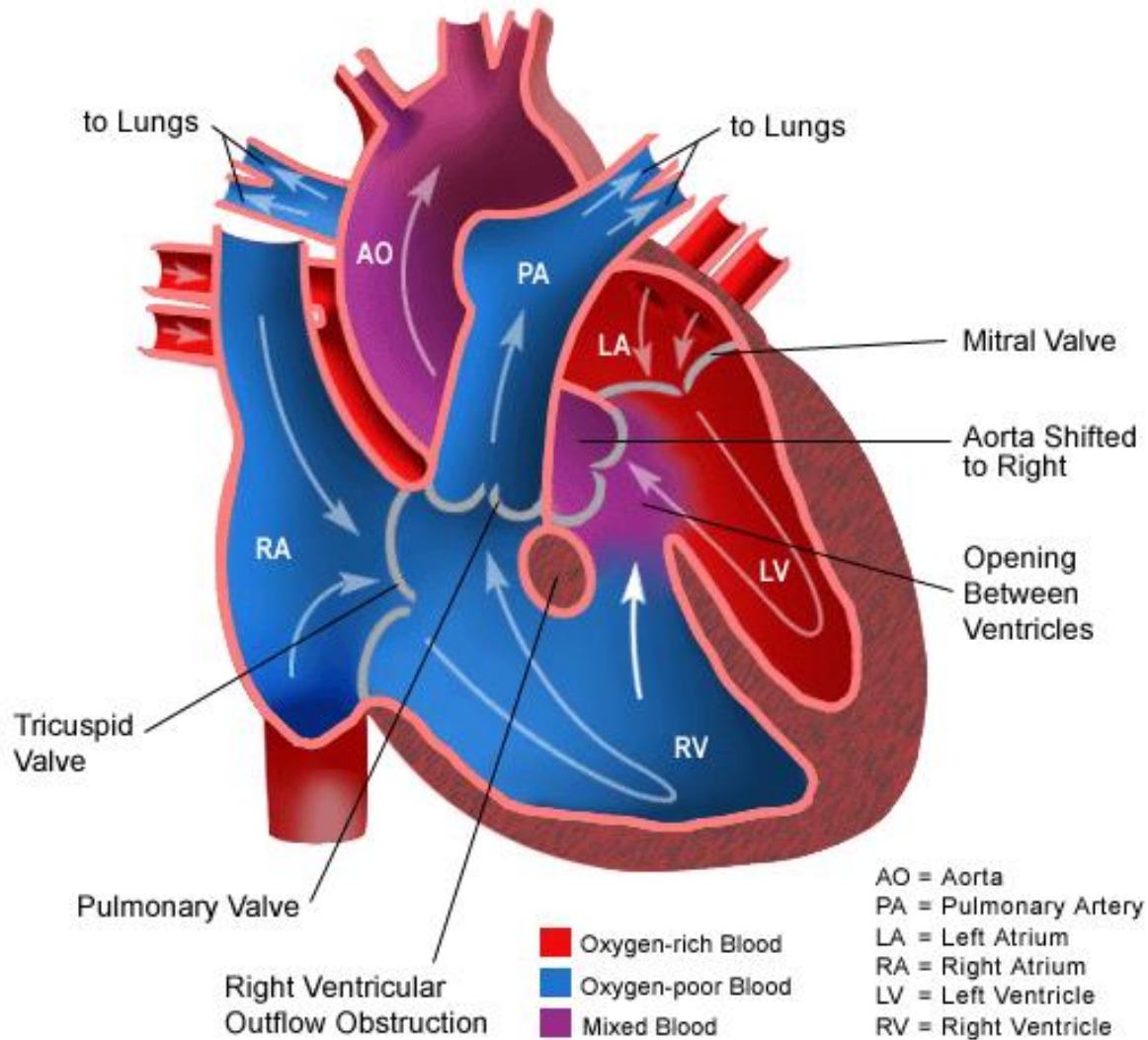
- Tetralogie van Fallot
- Coarctatio Aortae
- Morbus Ebstein
- Hypoplastisch Linker Hart Syndroom

- Tetralogie van Fallot
- Coarctatio Aortae
- Morbus Ebstein
- Hypoplastisch Linker Hart Syndroom



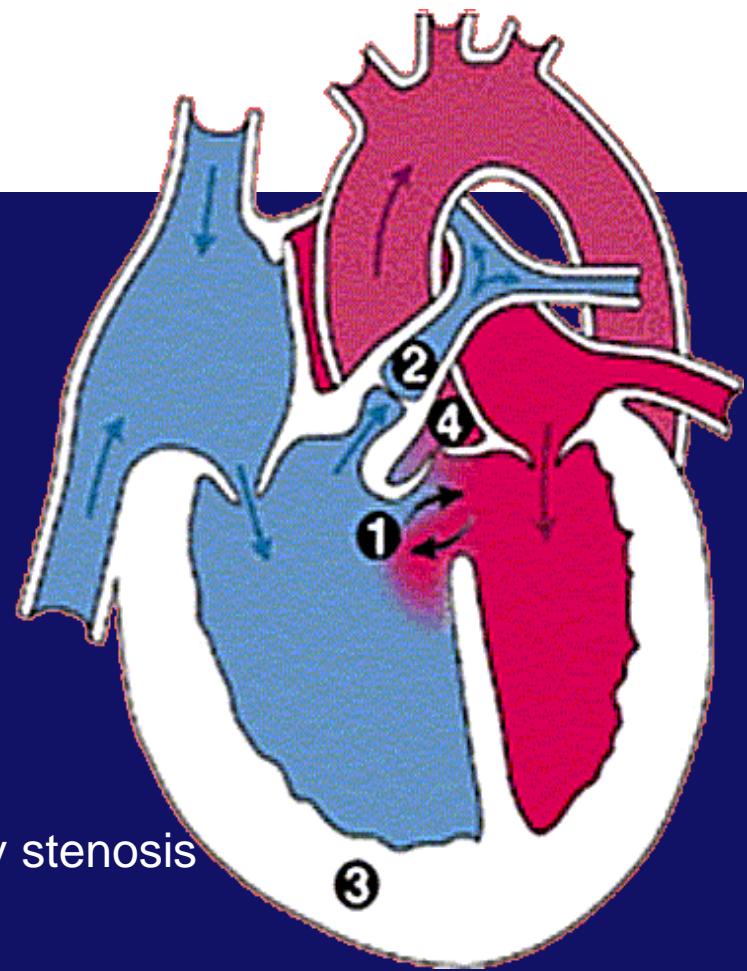
Tetralogie van Fallot

Tetralogy of Fallot (TOF or "Tet")

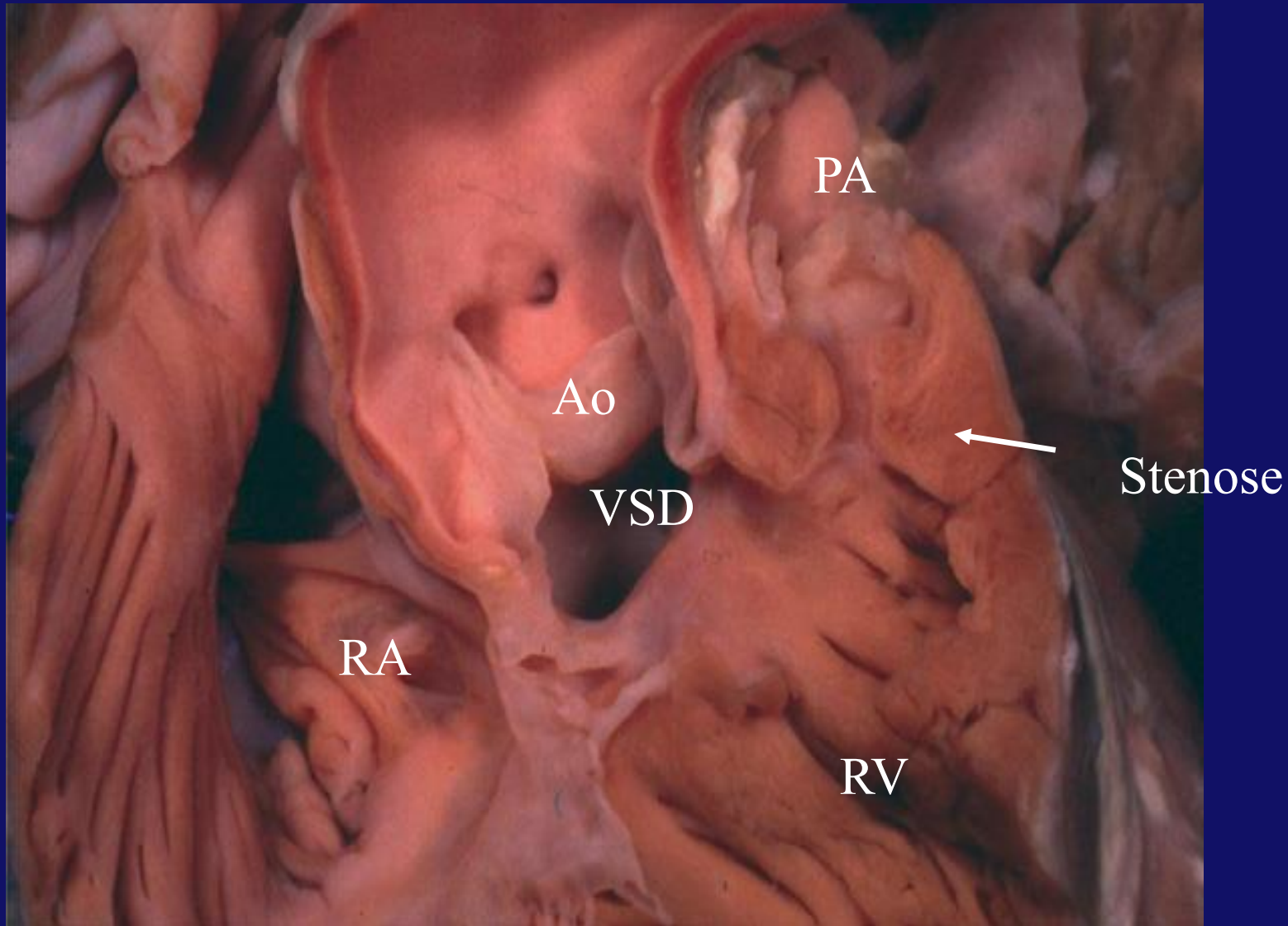


Tetralogy of Fallot

- Anterior displacement outlet septum:
 - Ventricular septal defect
 - Pulmonary stenosis
 - Hypertrophy of right ventricle
 - Overriding aorta
- Types:
 - Pink Fallot: no cyanosis,
 - Blue fallot: cyanosis: severe pulmonary stenosis
 - Absent pulmonary valve
 - Pulmonary atresia
- Genetics:
 - 22q11 deletion, Trisomy 21
 - Often de novo

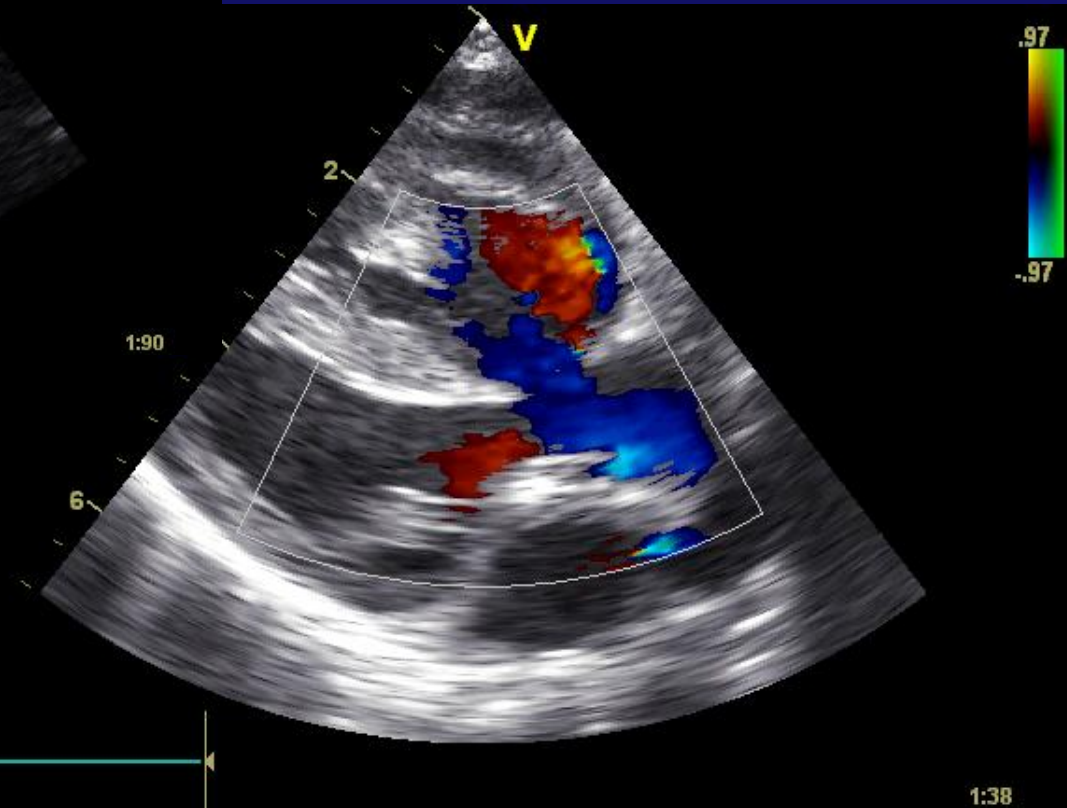
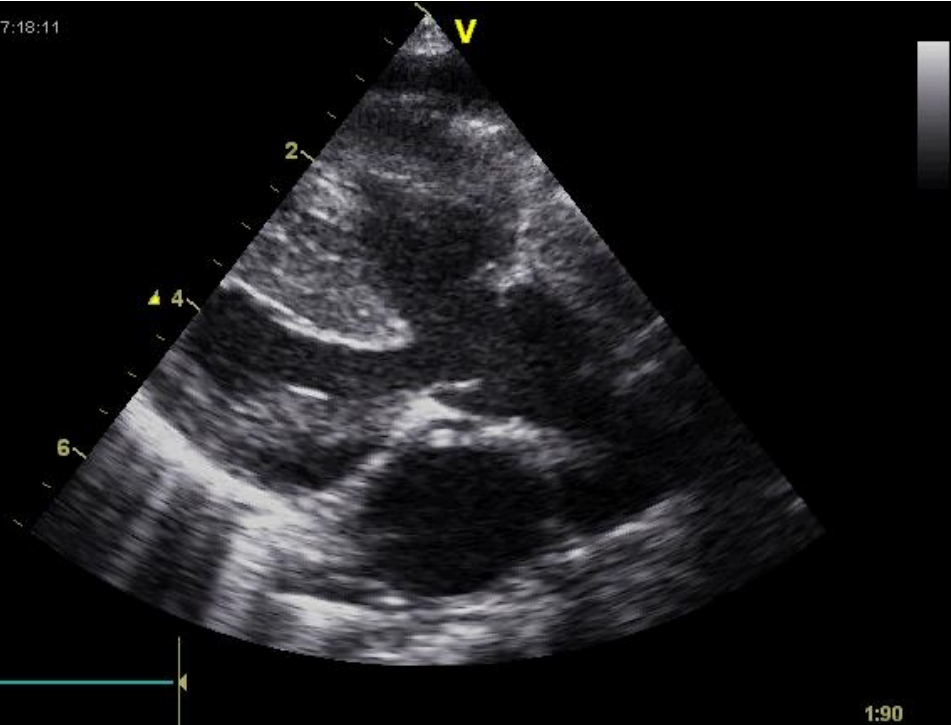


Tetralogy of Fallot



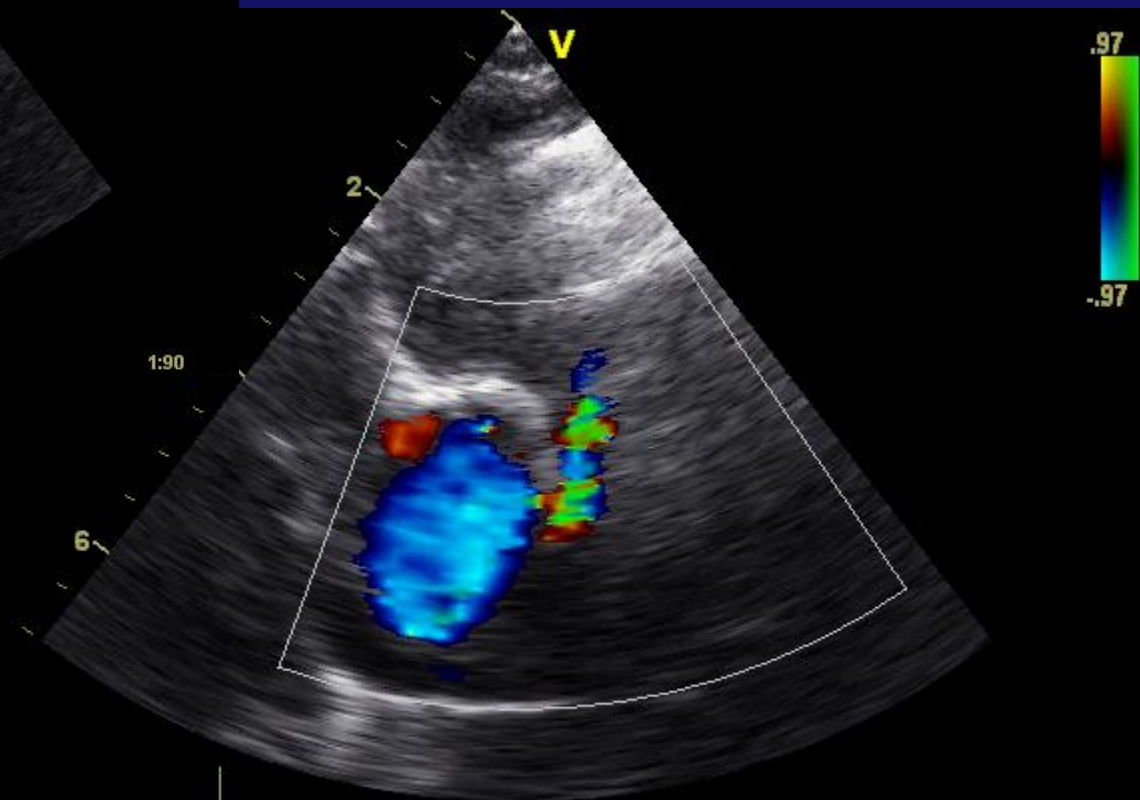
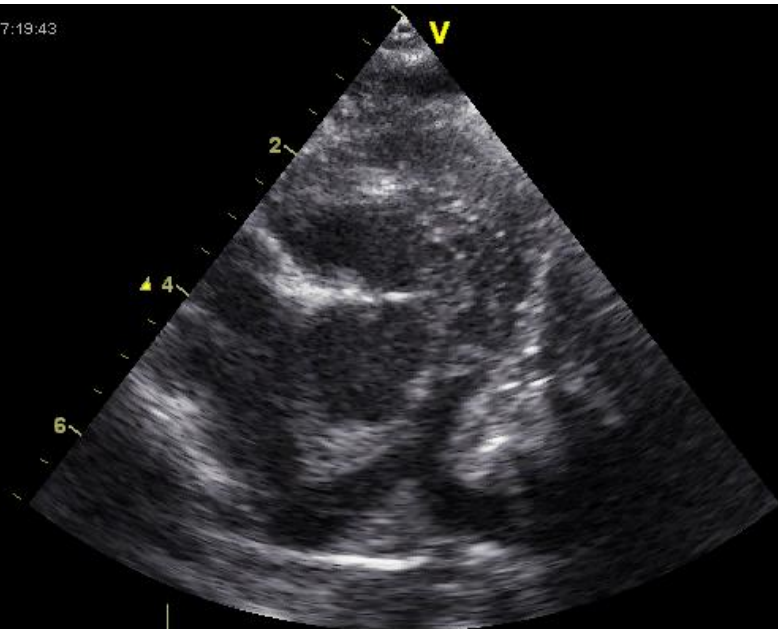
Tetralogy of Fallot-VSD/overriding Ao

07/09/2009 17:18:11



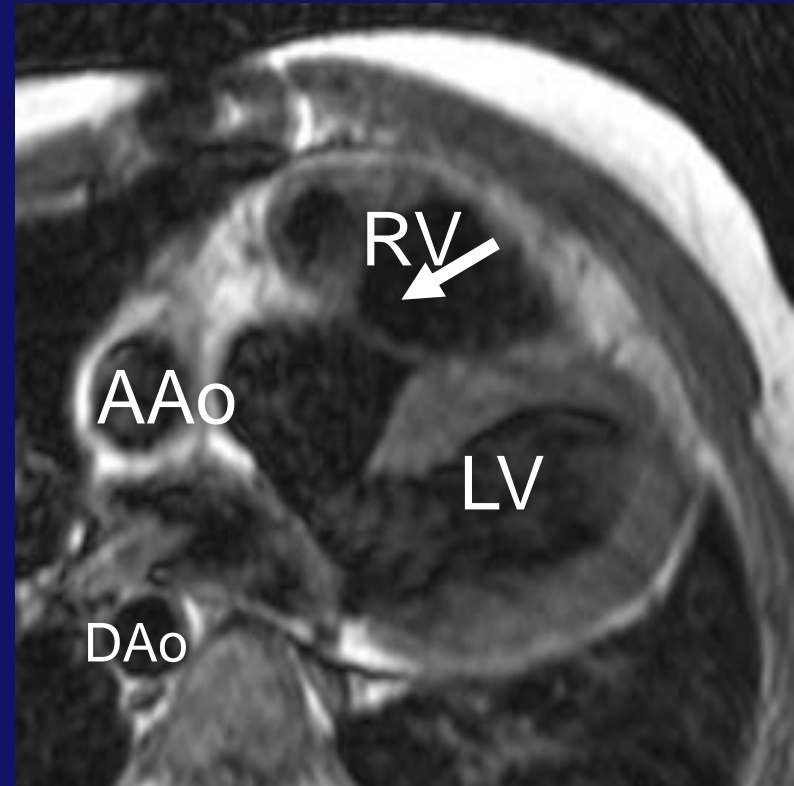
Tetralogy of Fallot-pulmonary stenosis

07/09/2009 17:19:43

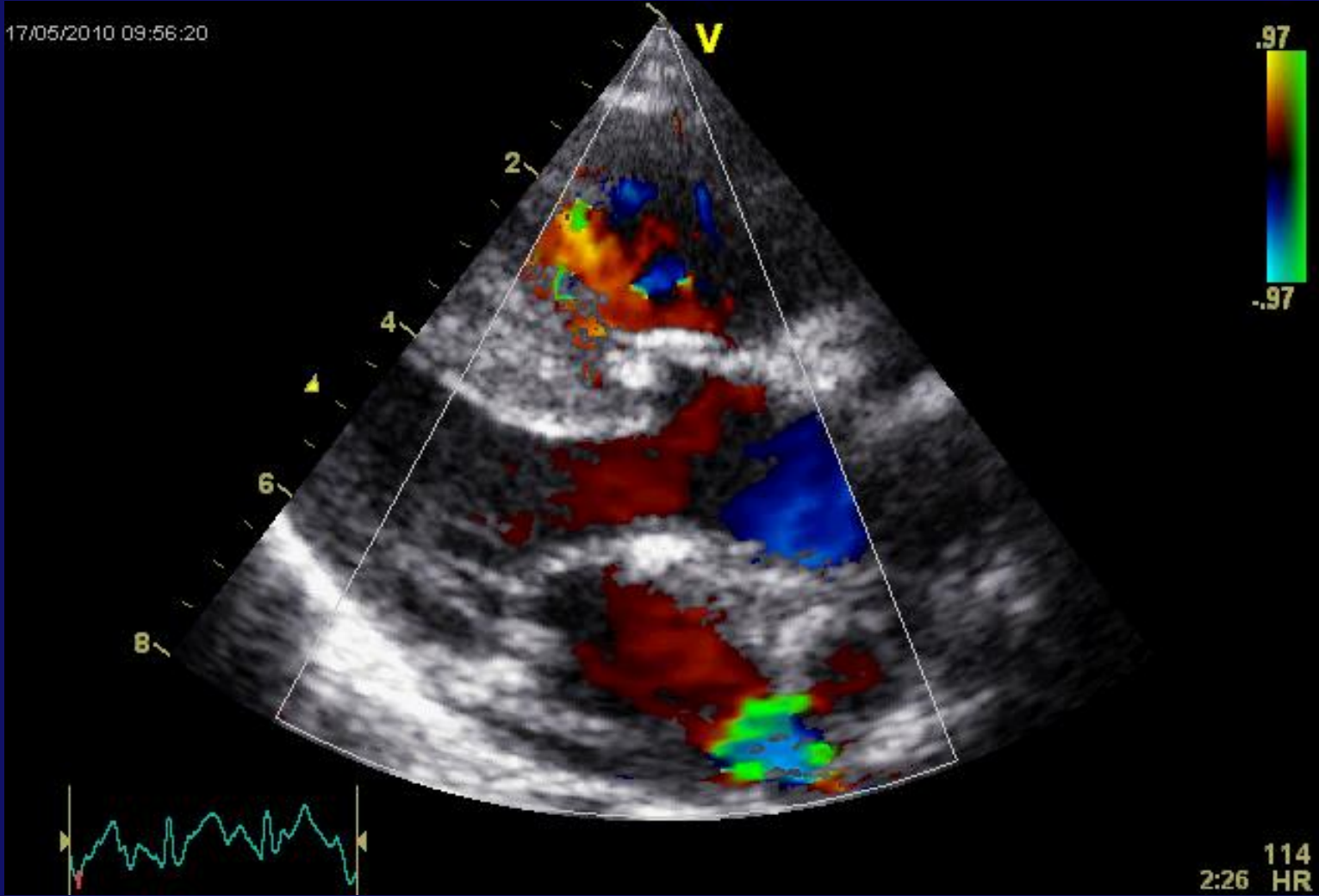


Tetralogy of Fallot

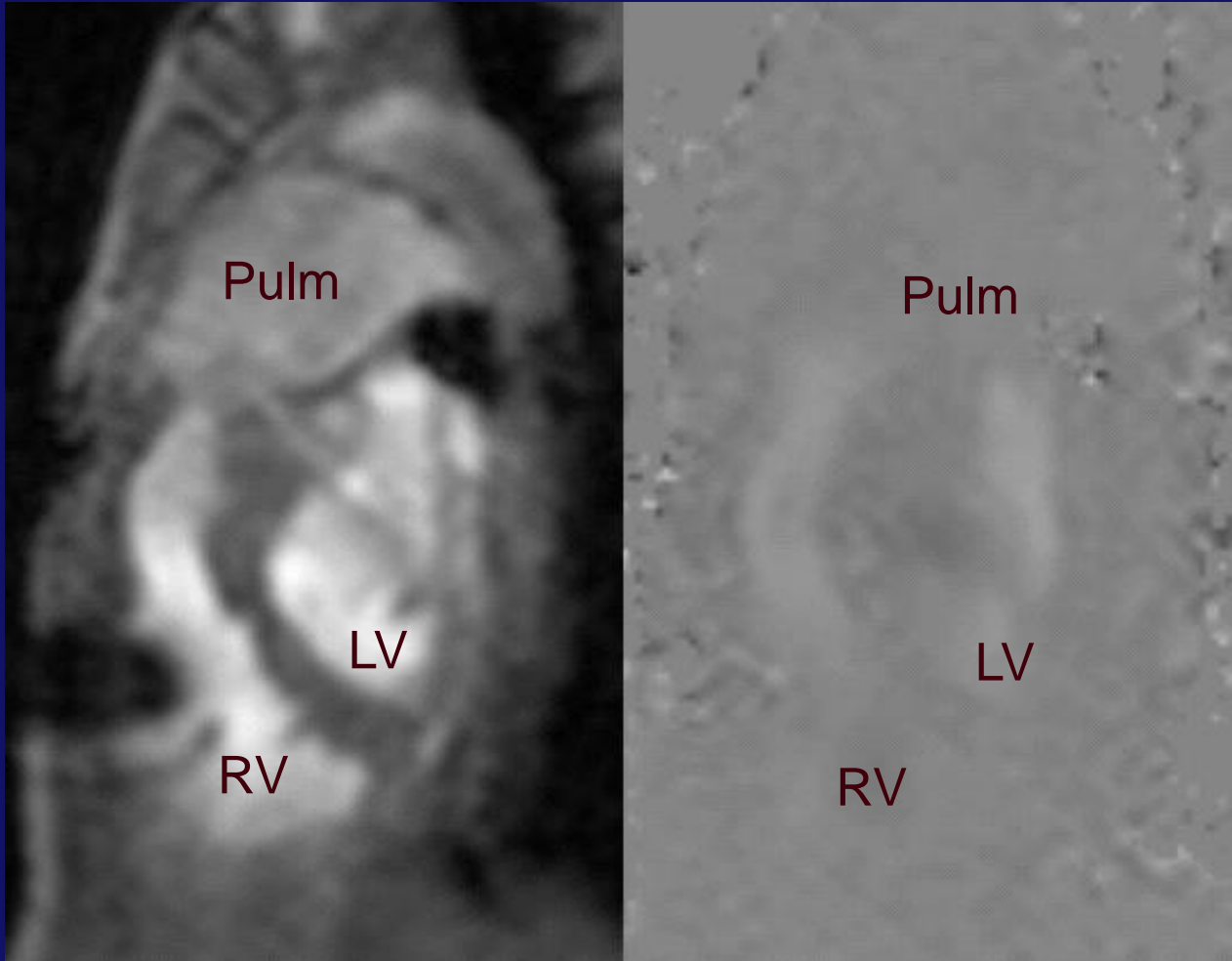
- Treatment:
 - Within first year of life
 - Sometimes palliative shunt is needed: modified BT shunt
 - Surgical correction:
 - Closure VSD
 - Relief pulmonary stenosis
- Follow-up:
 - Pulmonary regurgitation
 - Pulmonary stenosis
 - RV dilatation
 - Impairment of RV and LV function
 - Complications of palliative shunts



Tetralogy of Fallot-VSD closure



Sagittal cine MRI and flow map of pulmonary regurgitation & stenosis



RVOT dilatation after Fallot repair

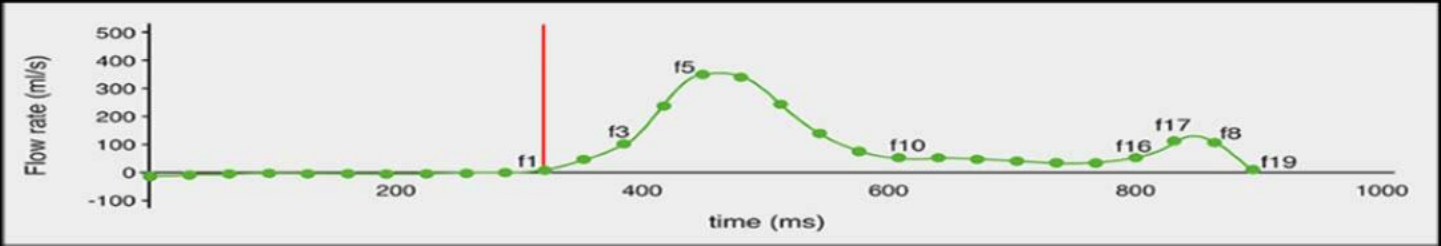
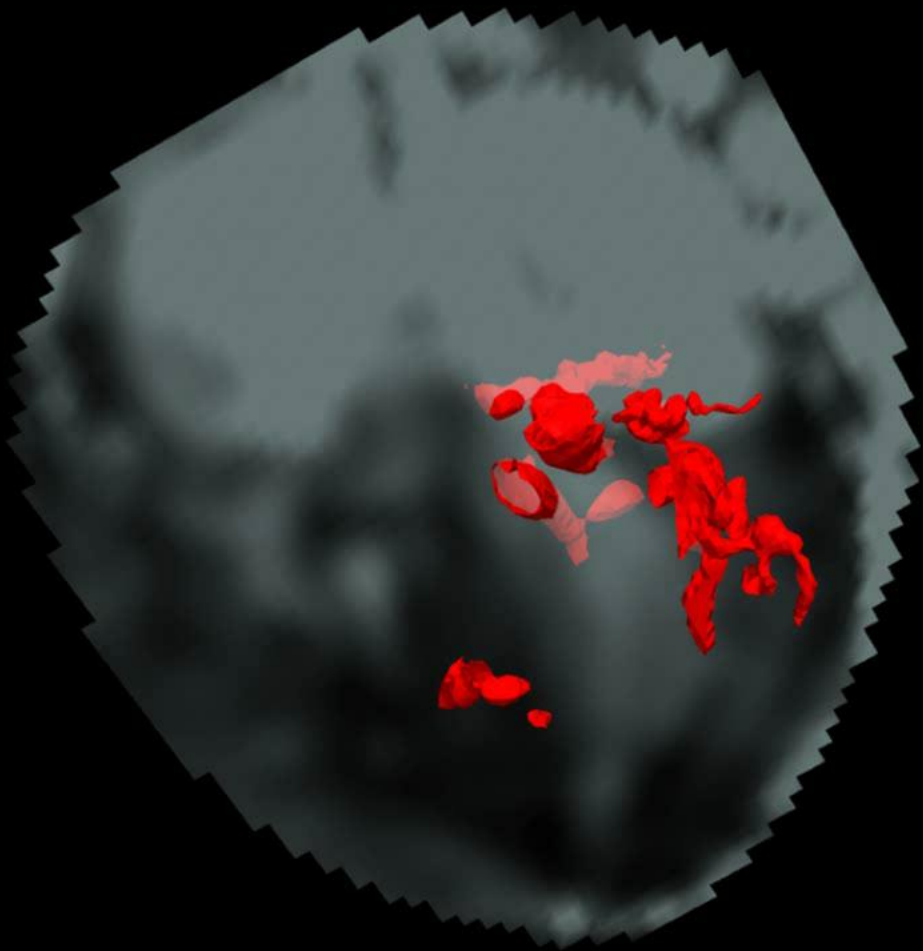


...and now something completely different

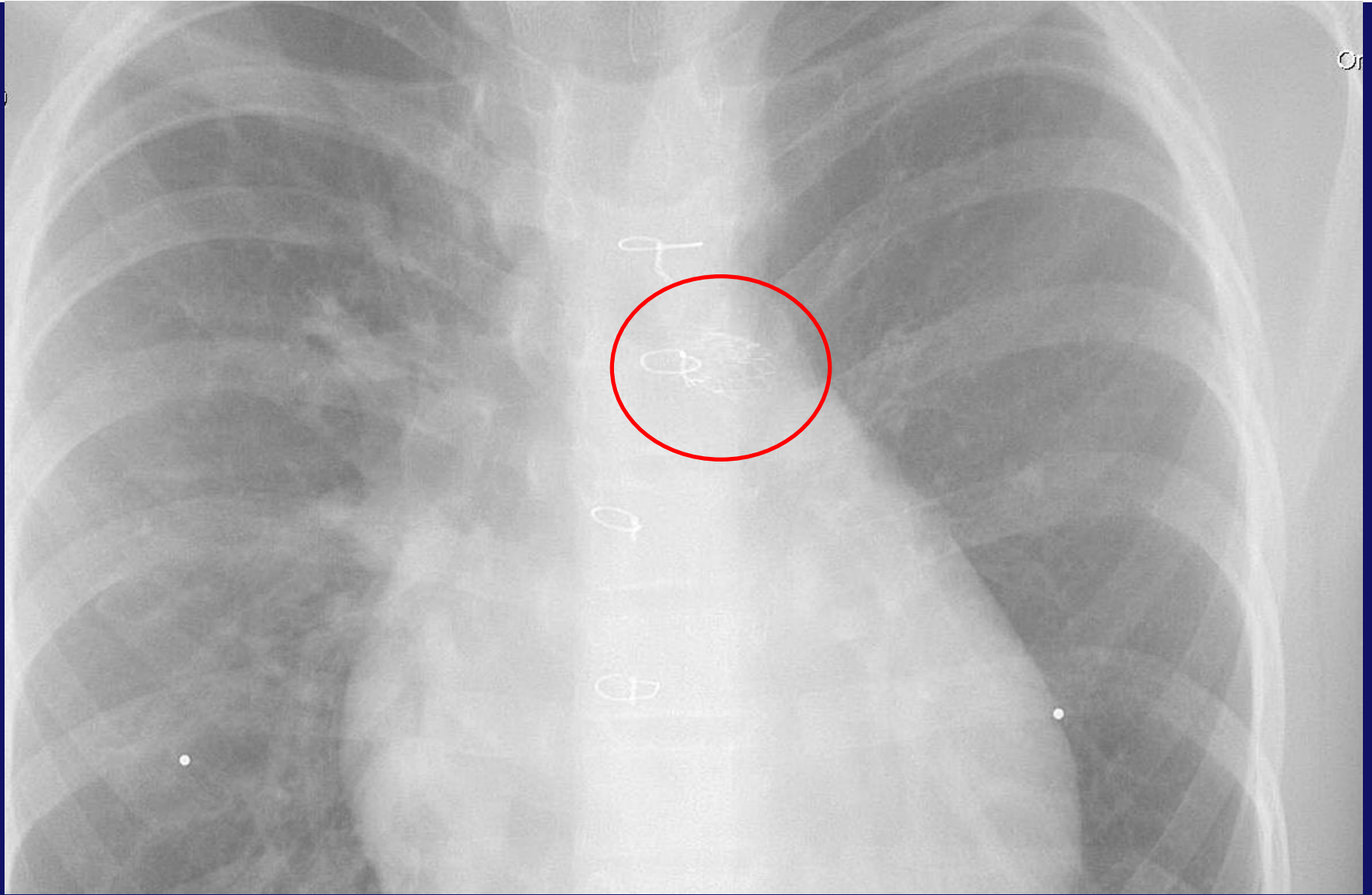


Vortex





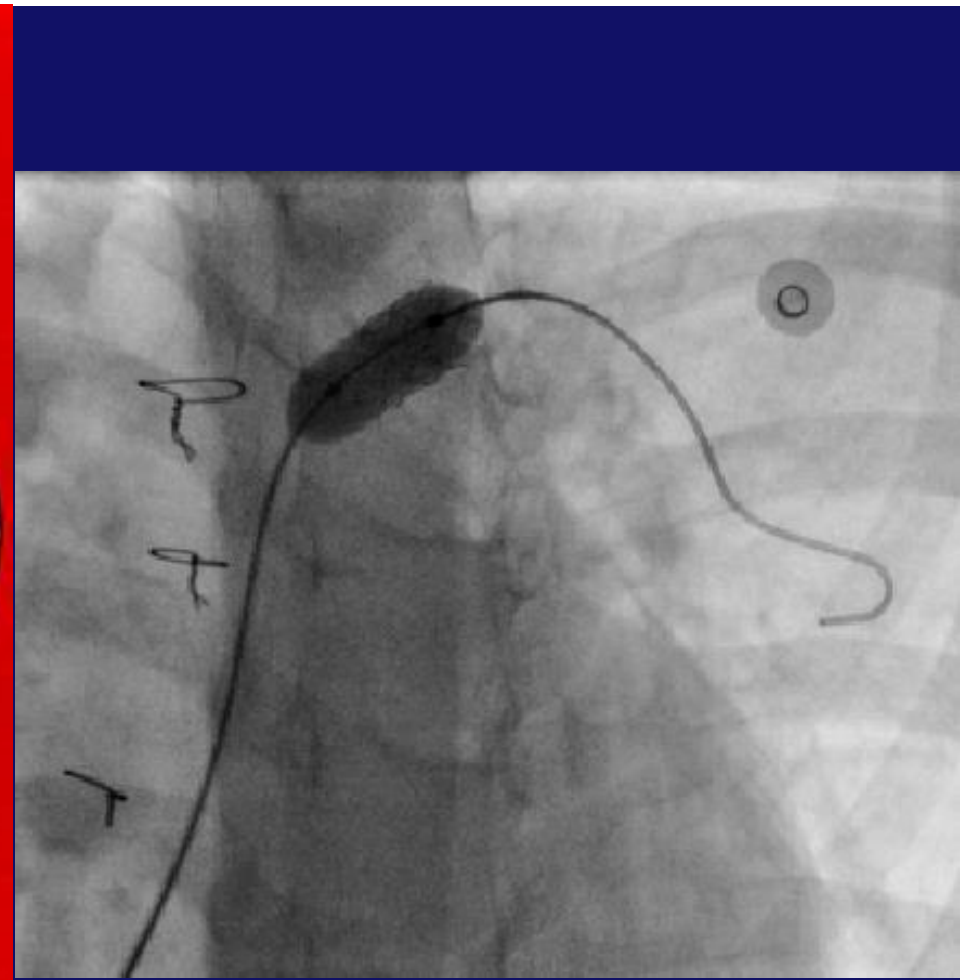
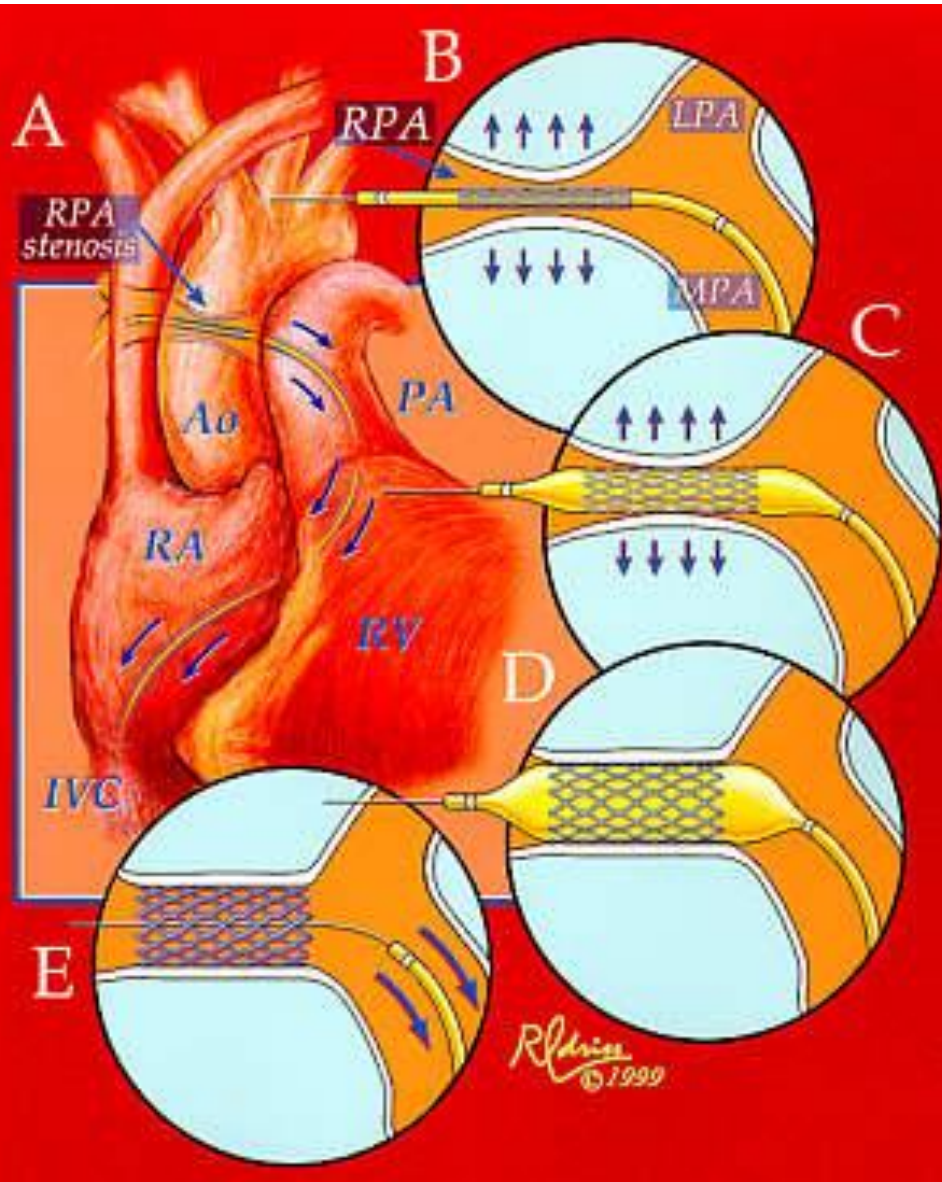
Sp correctie tetralogie van Fallot

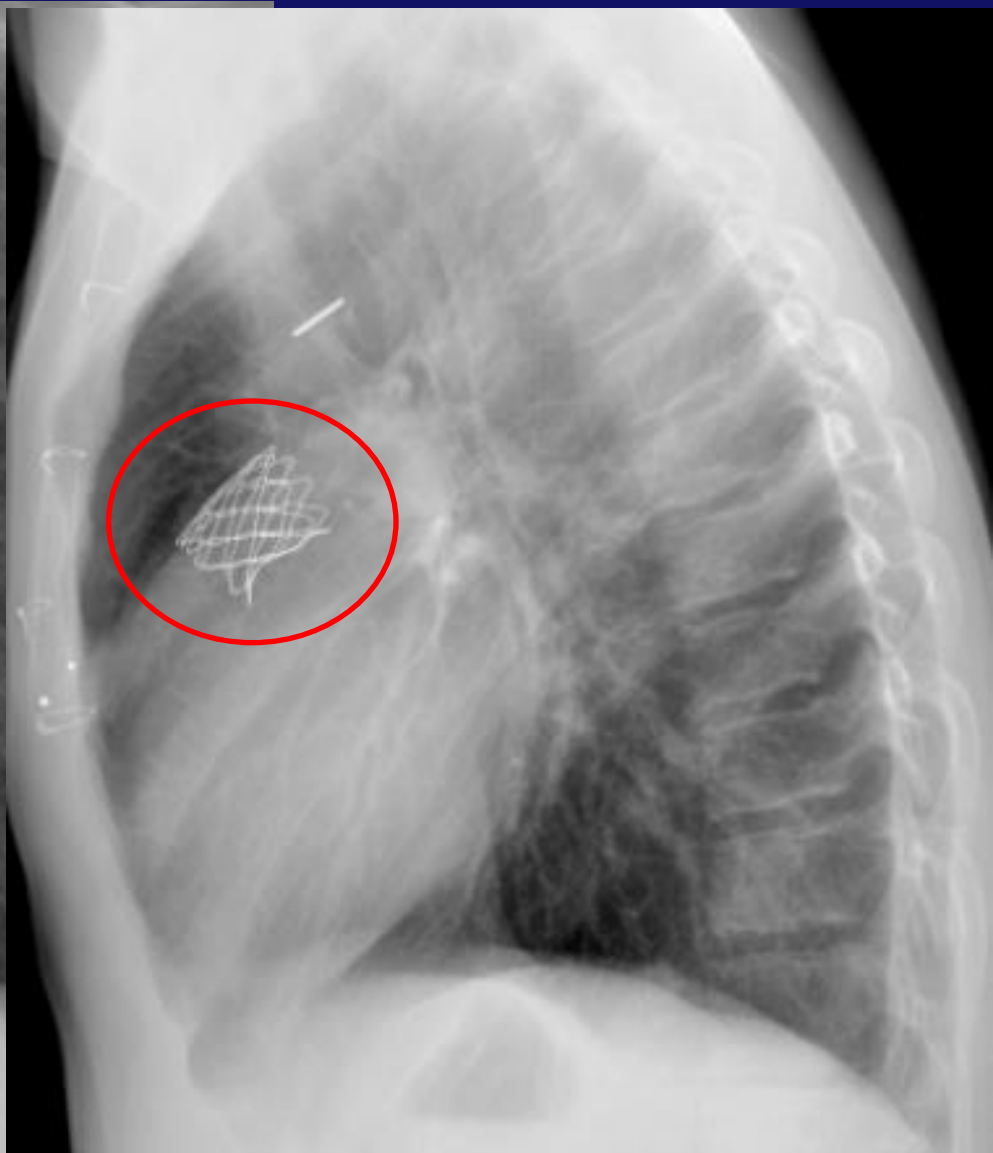


- Stent in de ductus
- Stent in de linker longslagader
- Stent in de rechter longslagader
- Stent in de hoofdbronchus

- Stent in de ductus
- Stent in de linker longslagader
- Stent in de rechter longslagader
- Stent in de hoofdbronchus

Stenting pulmonaal arteriën

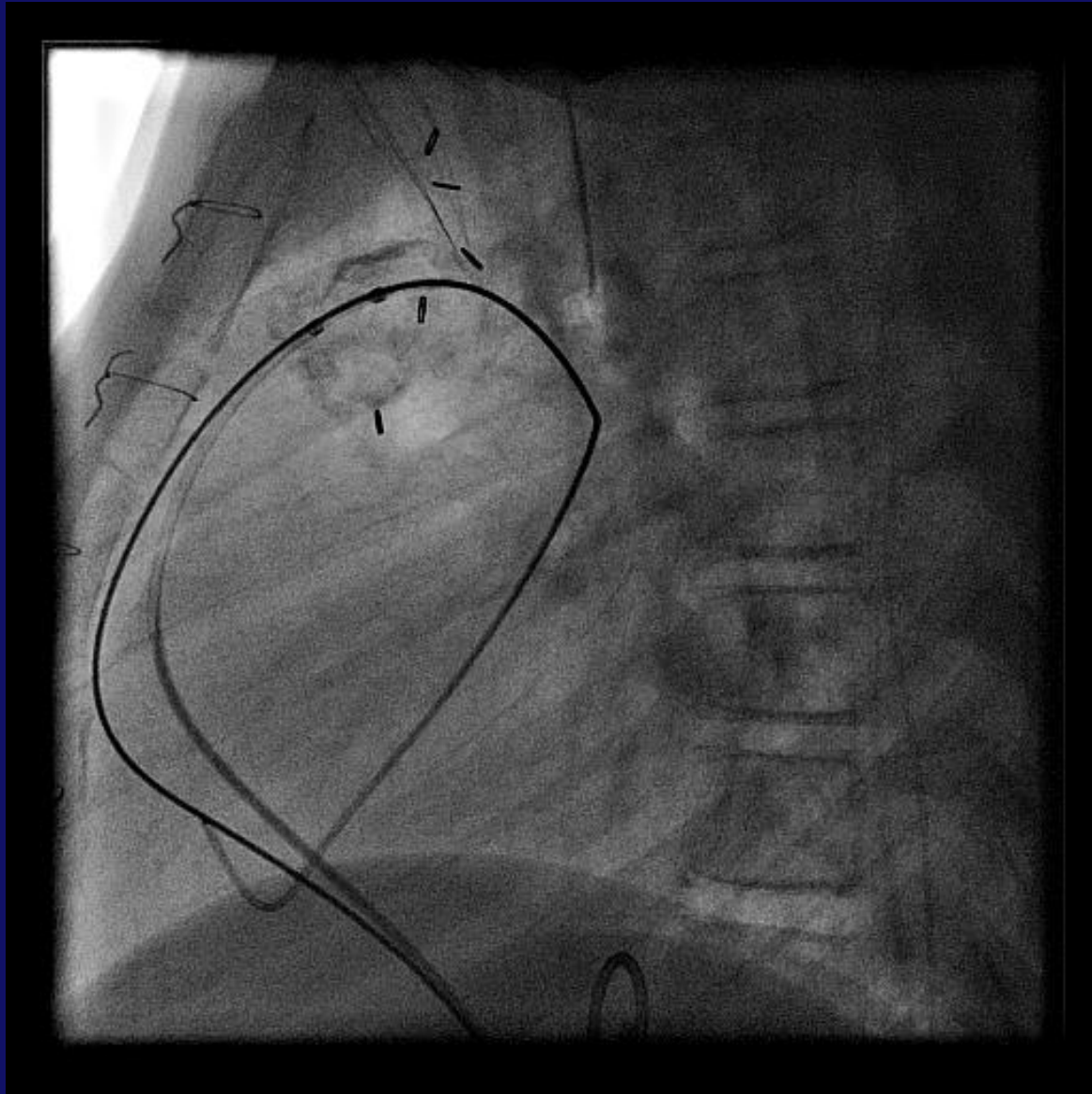


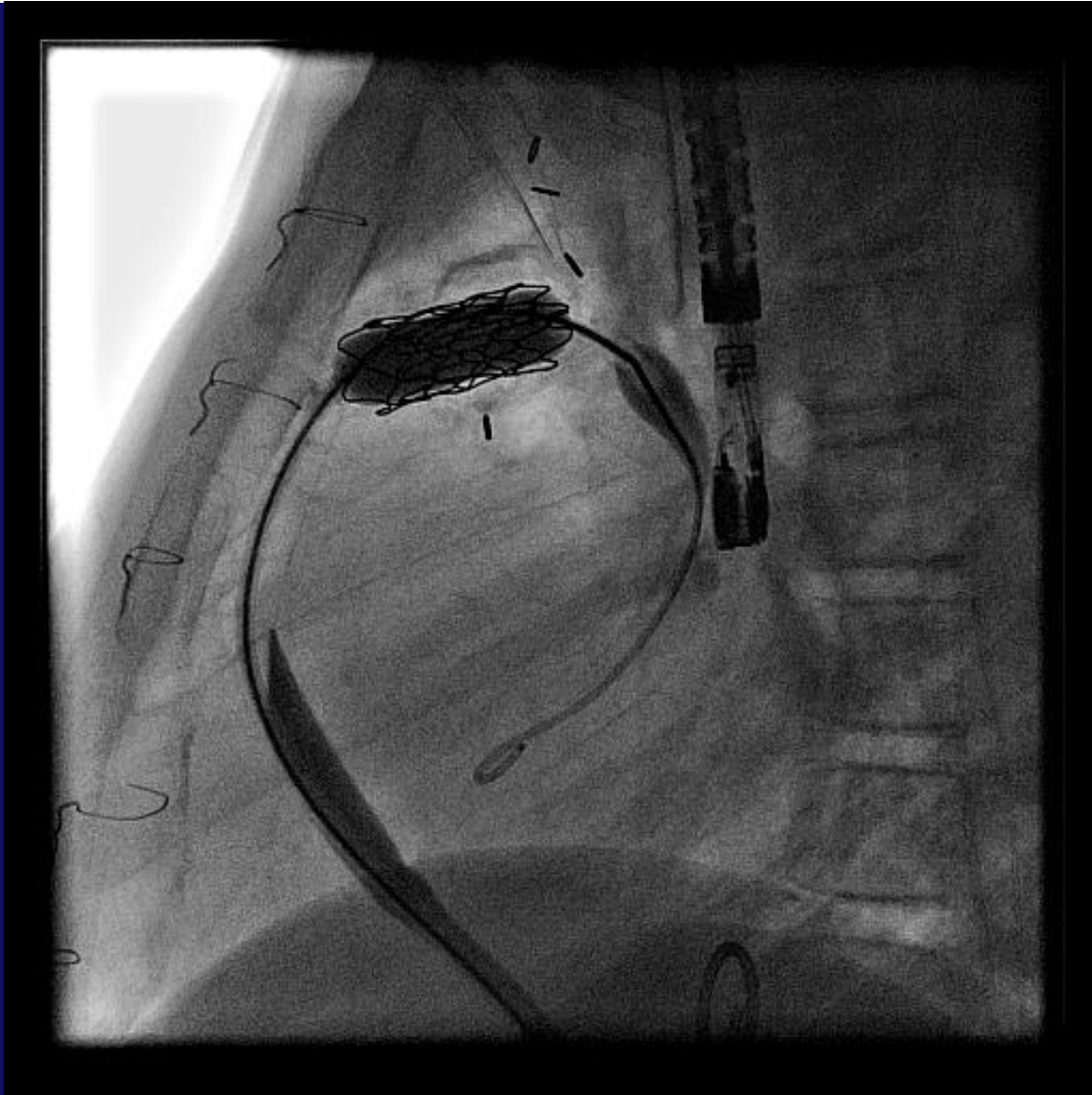


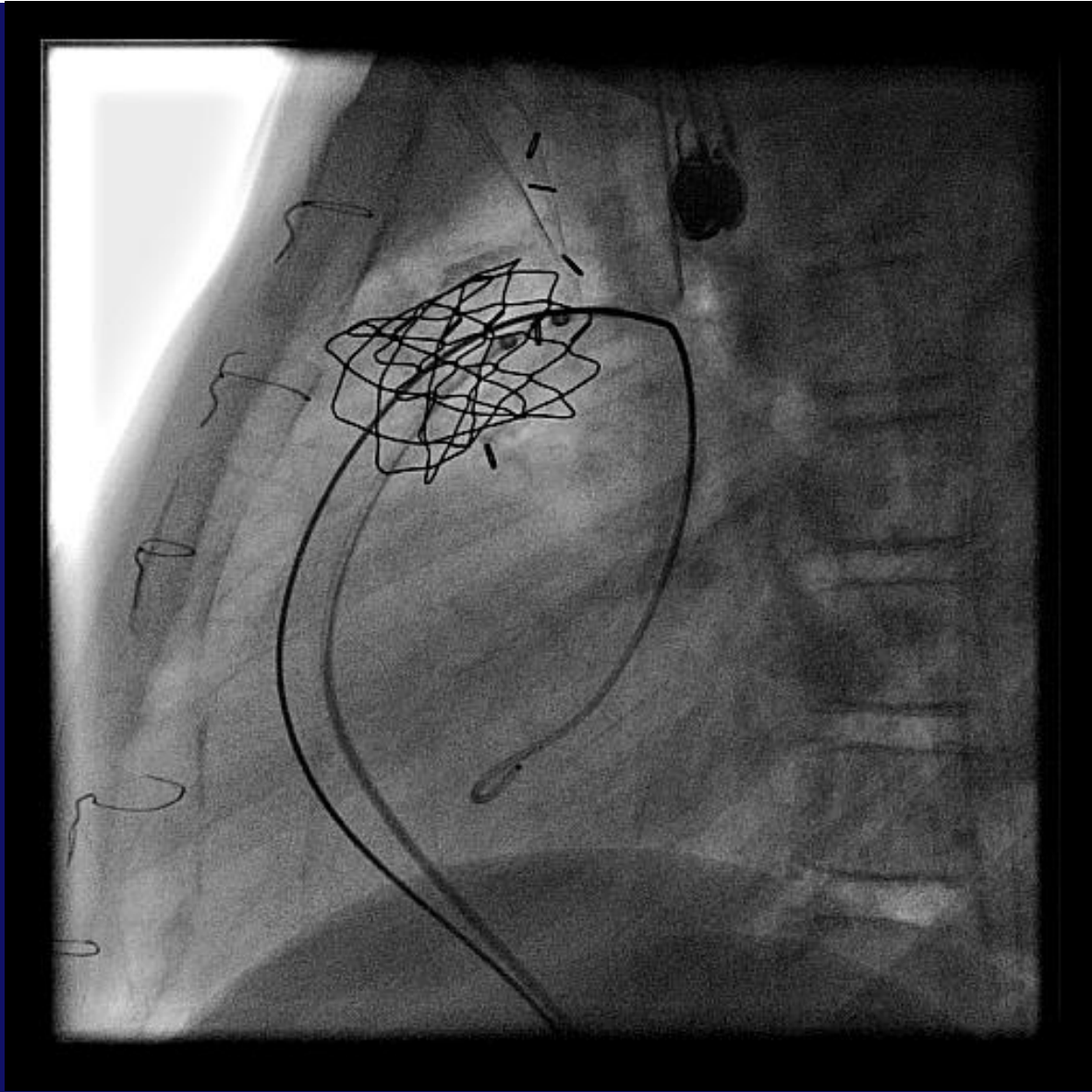
- Een stent in de aorta
- Een stent in de truncus pulmonalis
- Een stent in de ductus
- Een stent in de vena cava superior

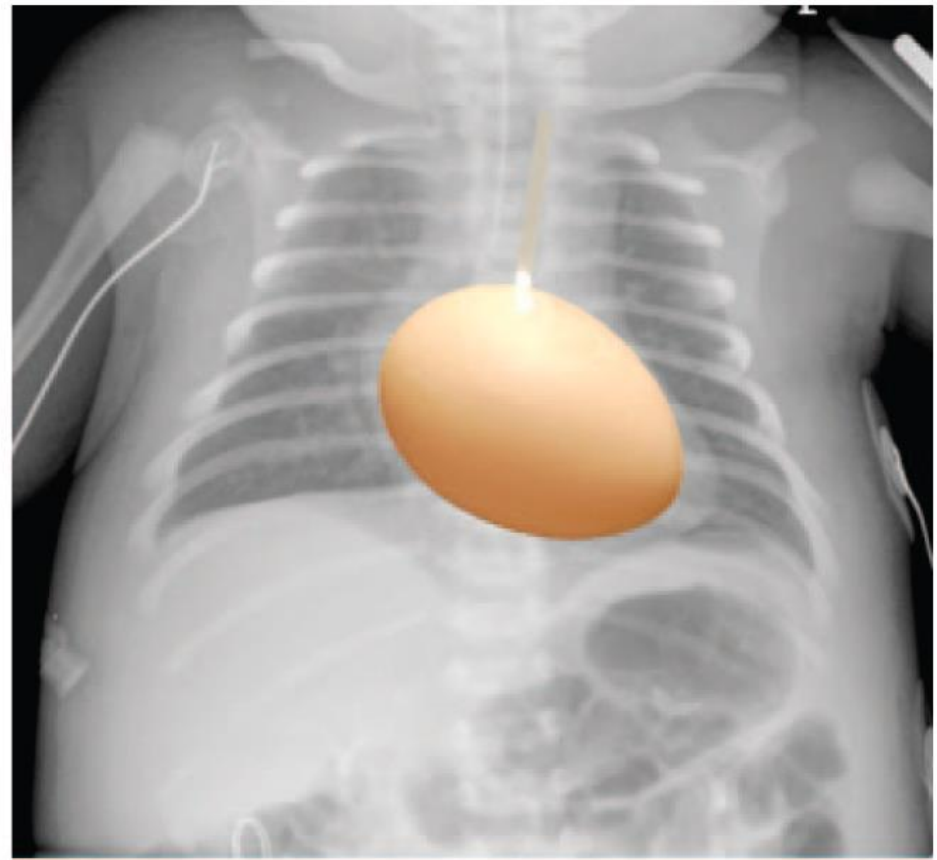
- Een stent in de aorta
- Een stent in de truncus pulmonalis
- Een stent in de ductus
- Een stent in de vena cava superior









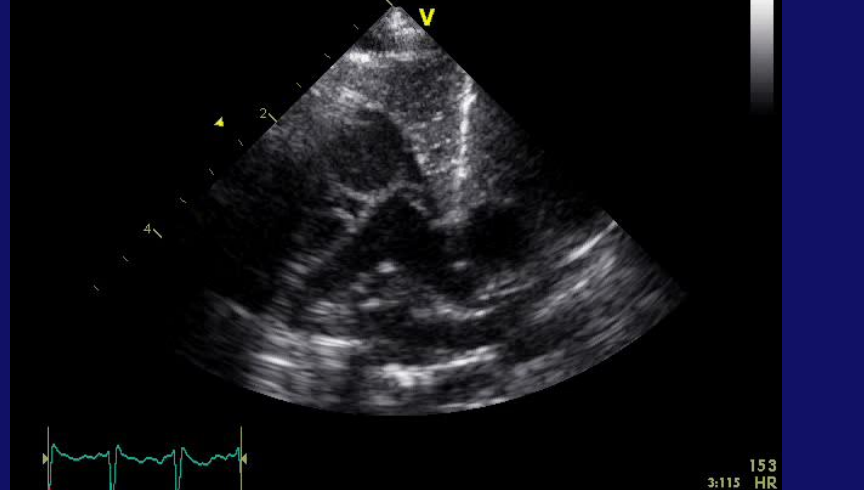
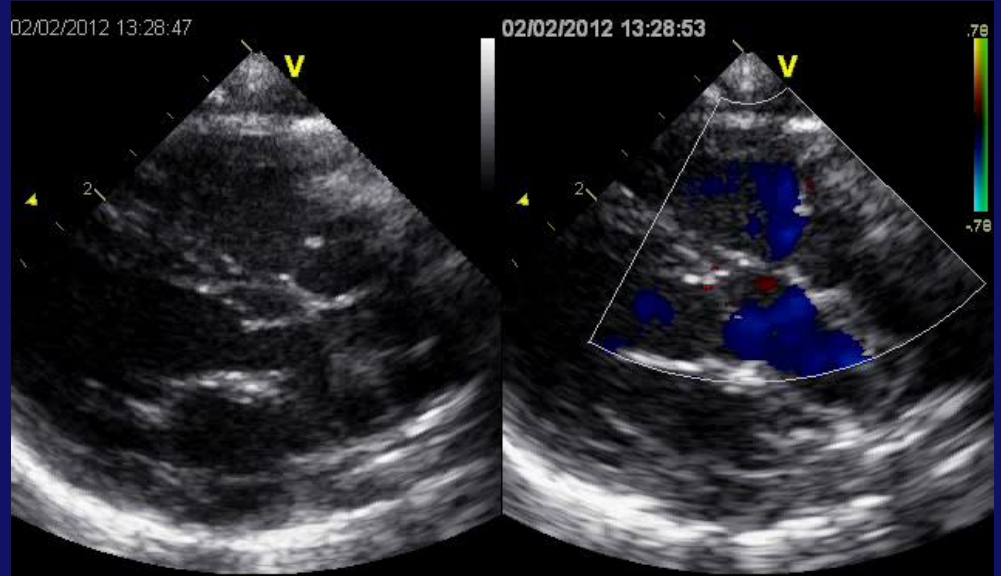
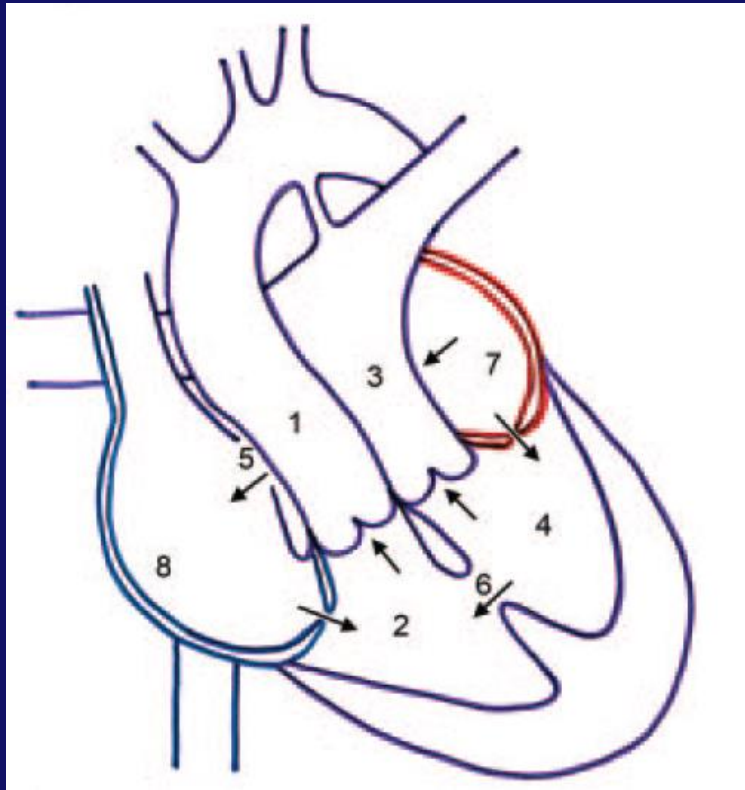
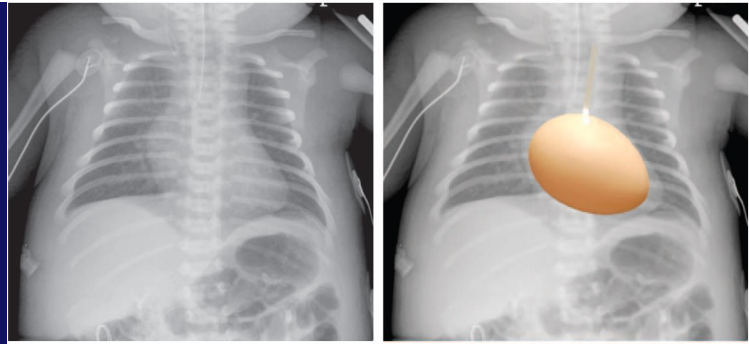


Welke hartafwijking

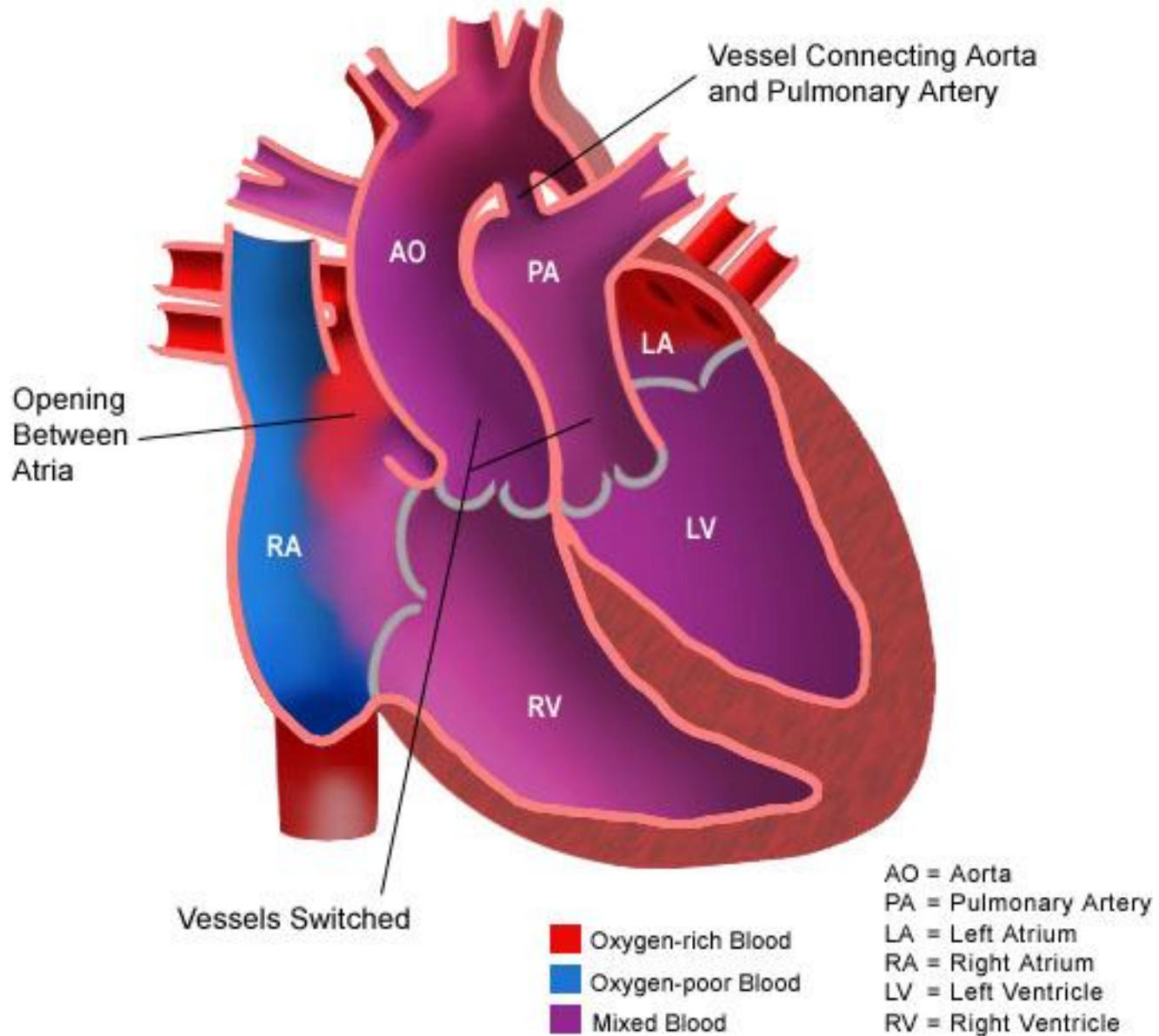
- Scimitar syndroom
- Transpositie van de grote vaten
- Pulmonalis stenose
- Morbus Ebstein

Welke hartafwijking

- Scimitar syndroom
- **Transpositie van de grote vaten**
- Pulmonalis stenose
- Morbus Ebstein

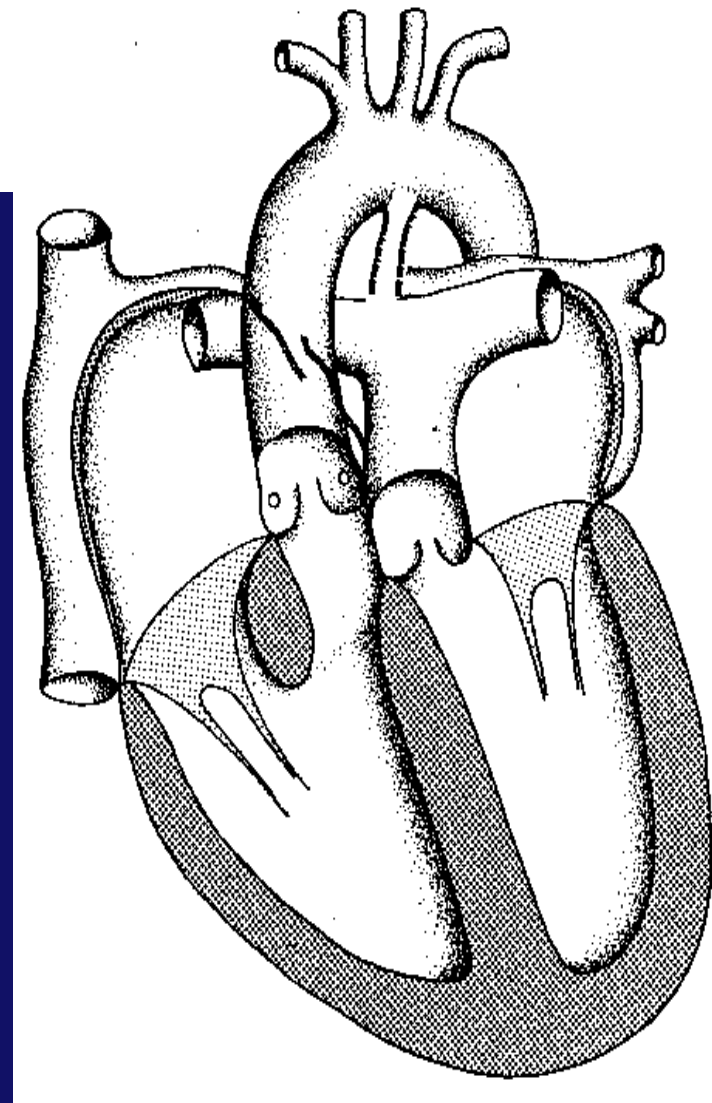


Transposition of Great Arteries

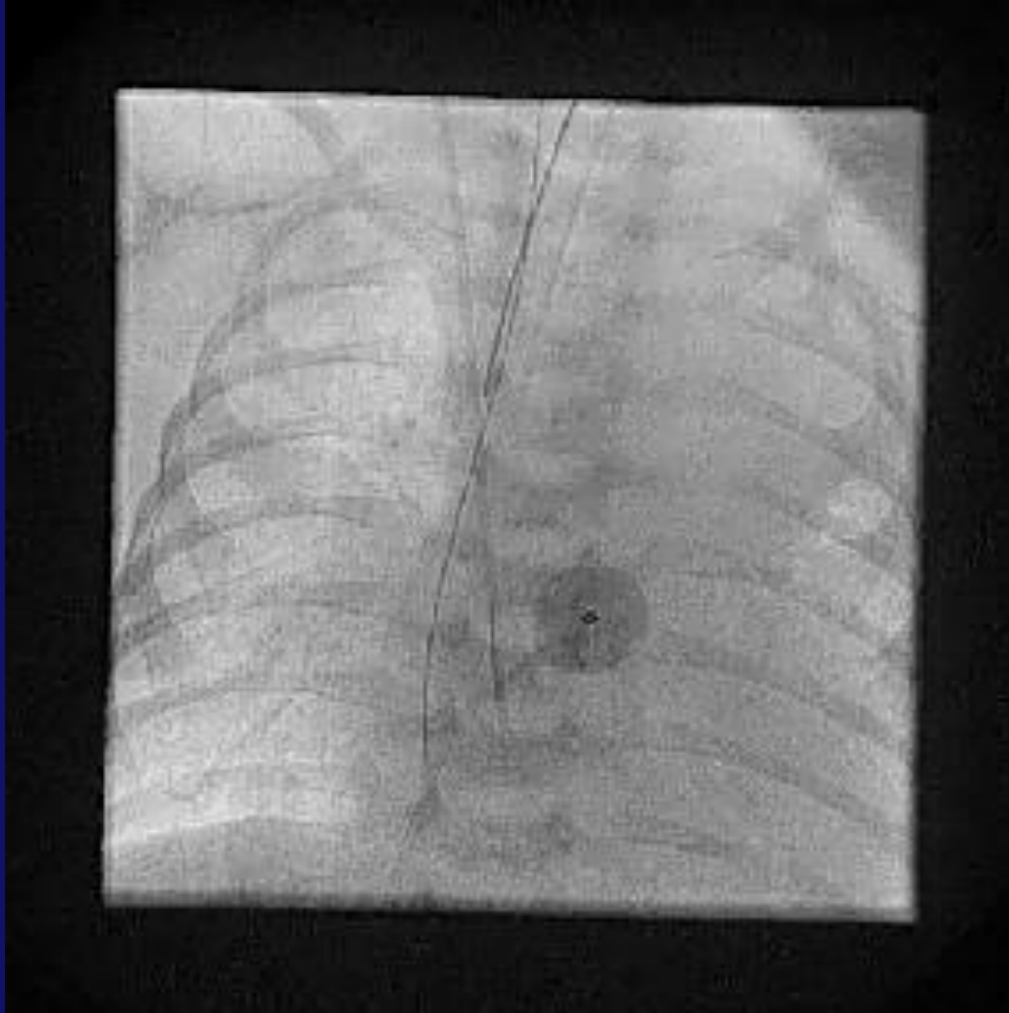


Transposition of Great Arteries

- Types:
 - Simple TGA: small ASD/VSD
 - Complex TGA: PS, DORV
- Presentation:
 - Cyanosis
 - No respiratory distress
- Survival depends on:
 - Early recognition
 - Atrial communication
 - Lesser extent: ductal patency
- If atrial communication insufficient: Rashkind septostomy



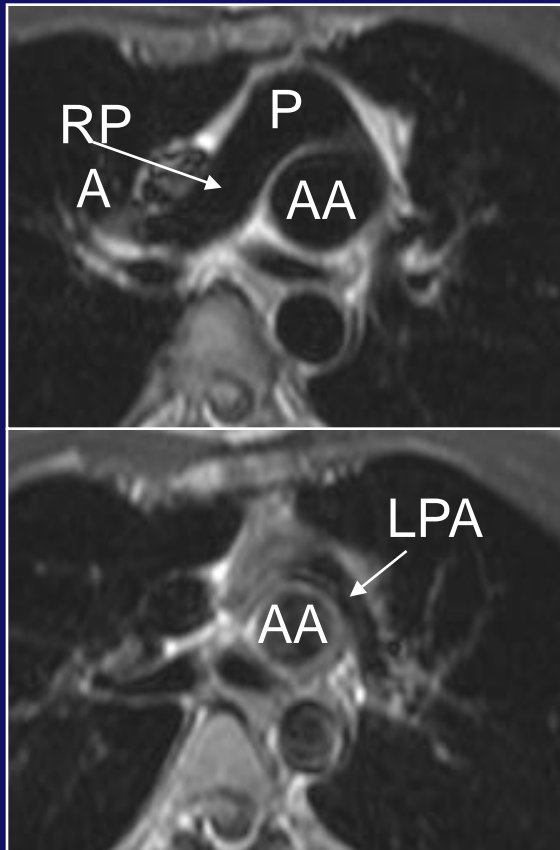
Rashkind for Transposition of Great Arteries



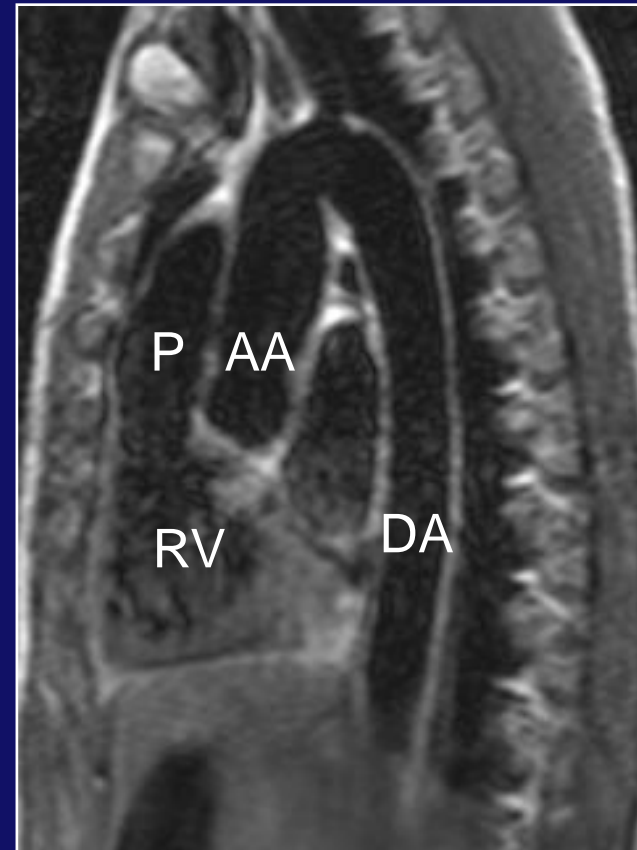
Transposition of the Great Arteries

- Treatment:
 - Preferably within first 2 weeks of life
 - Currently: arterial switch operation (ASO)
 - Until early eighties: atrial redirection: Mustard/Senning
- Follow-up:
 - ASO:
 - Aortic insufficiency
 - Narrowing RV outflow tract and pulmonary stenosis
 - (Neo) aortic root dilatation
 - Mustard/Senning
 - Baffle obstruction or leakage
 - RV dysfunction due to systemic pressure
 - Atrial arrhythmias due to atrial enlargement

Transposition of the Great Arteries after ASO



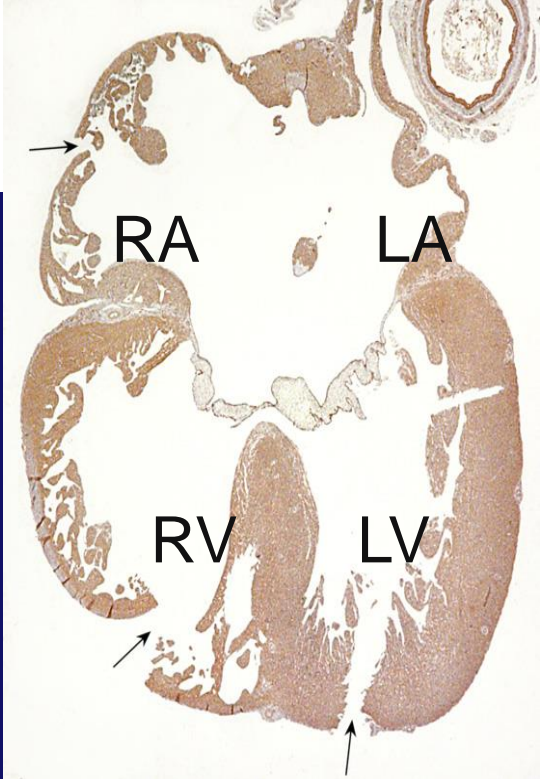
transverse sections



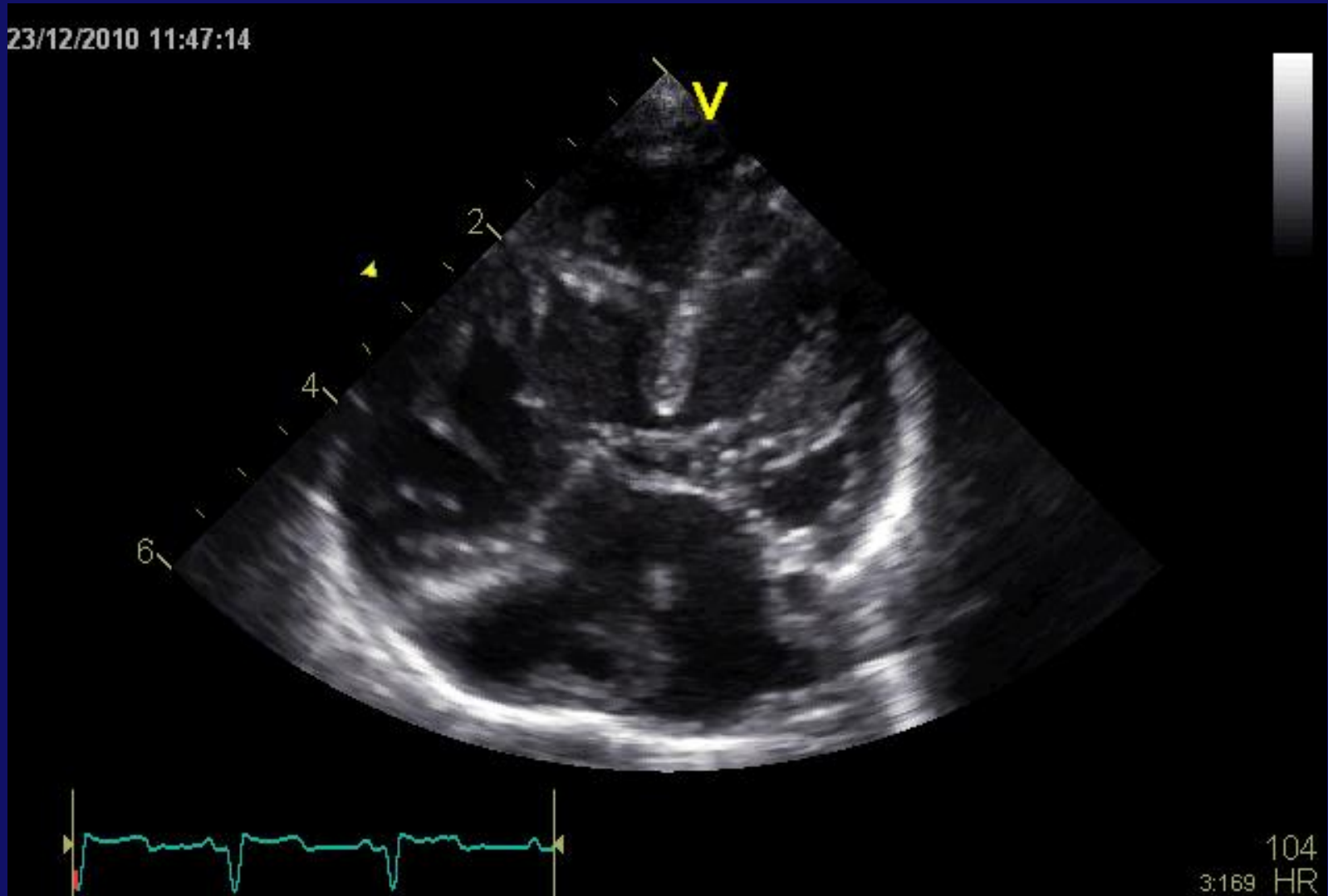
sagittal section

Atrioventricular septal defect

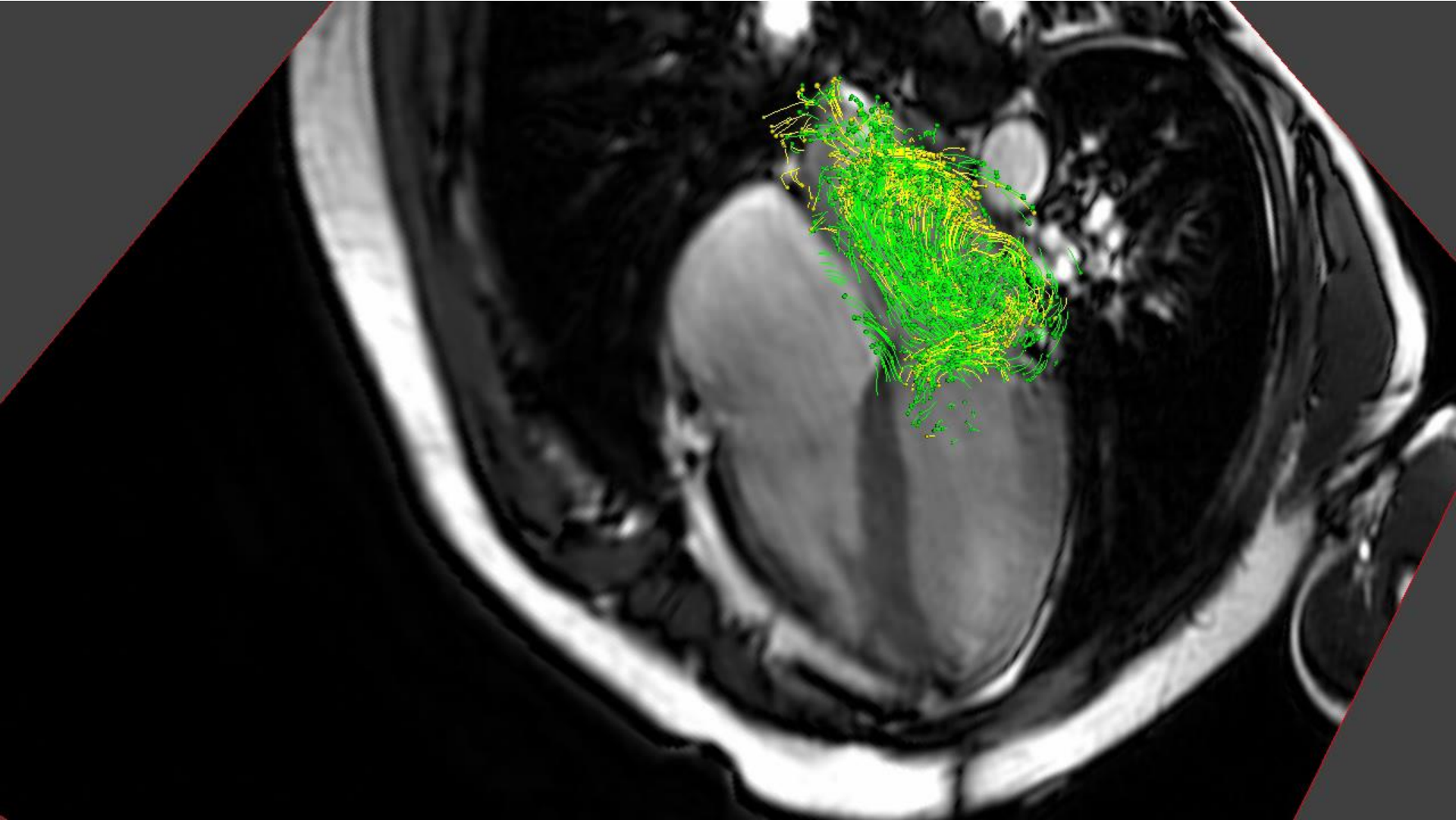
- Types:
 - Partial: ASD I
 - Intermediate: ASD I, small VSD
 - Complete: common AV-valve
 - Unbalanced AVSD
- Genetics:
 - Trisomy 21
- Presentation:
 - Partial-interm: asymptomatic in children
 - Complete:
 - Increased pulmonary flow
 - Failure to thrive
 - Pulm hypertension



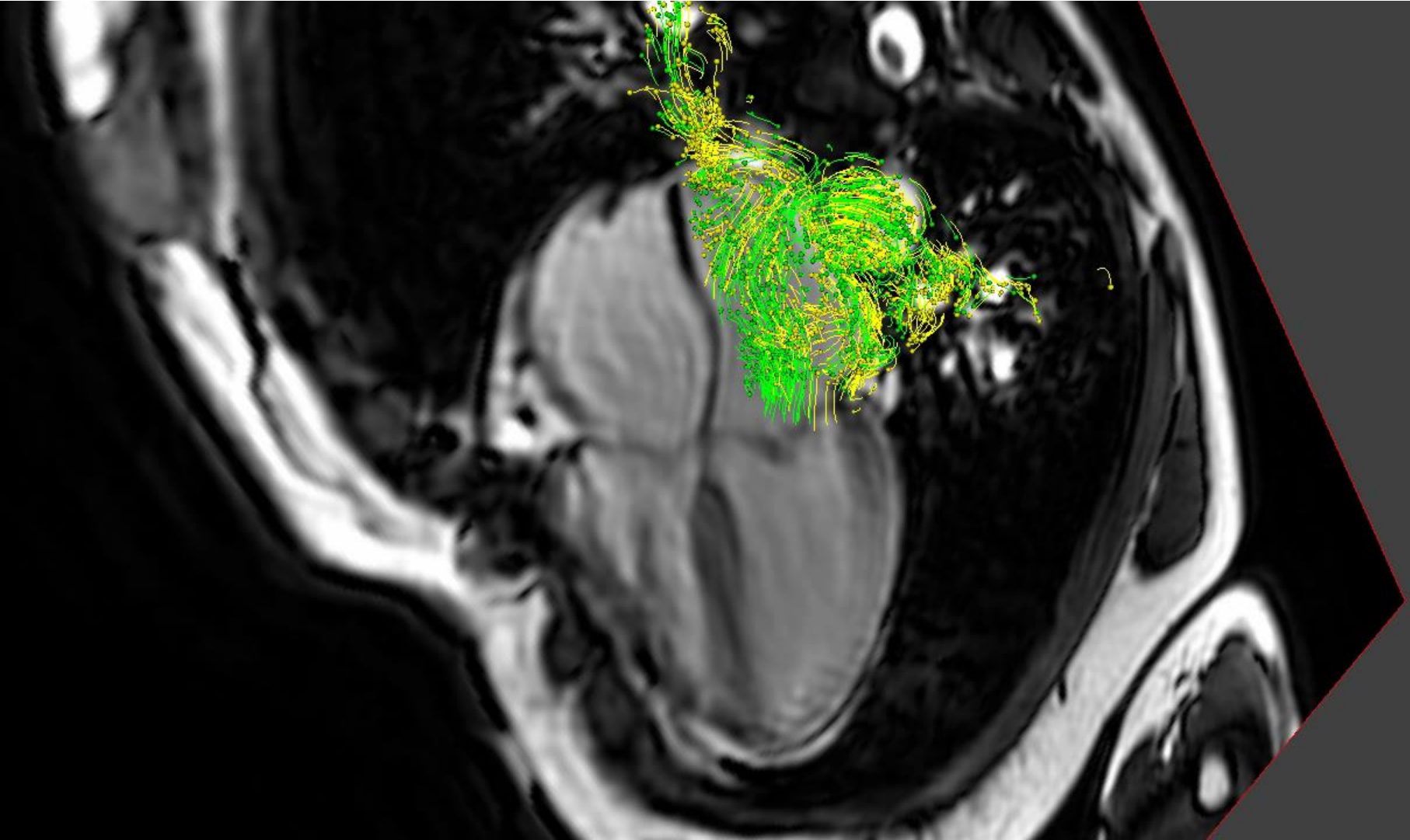
Atrioventricular septal defect-complete

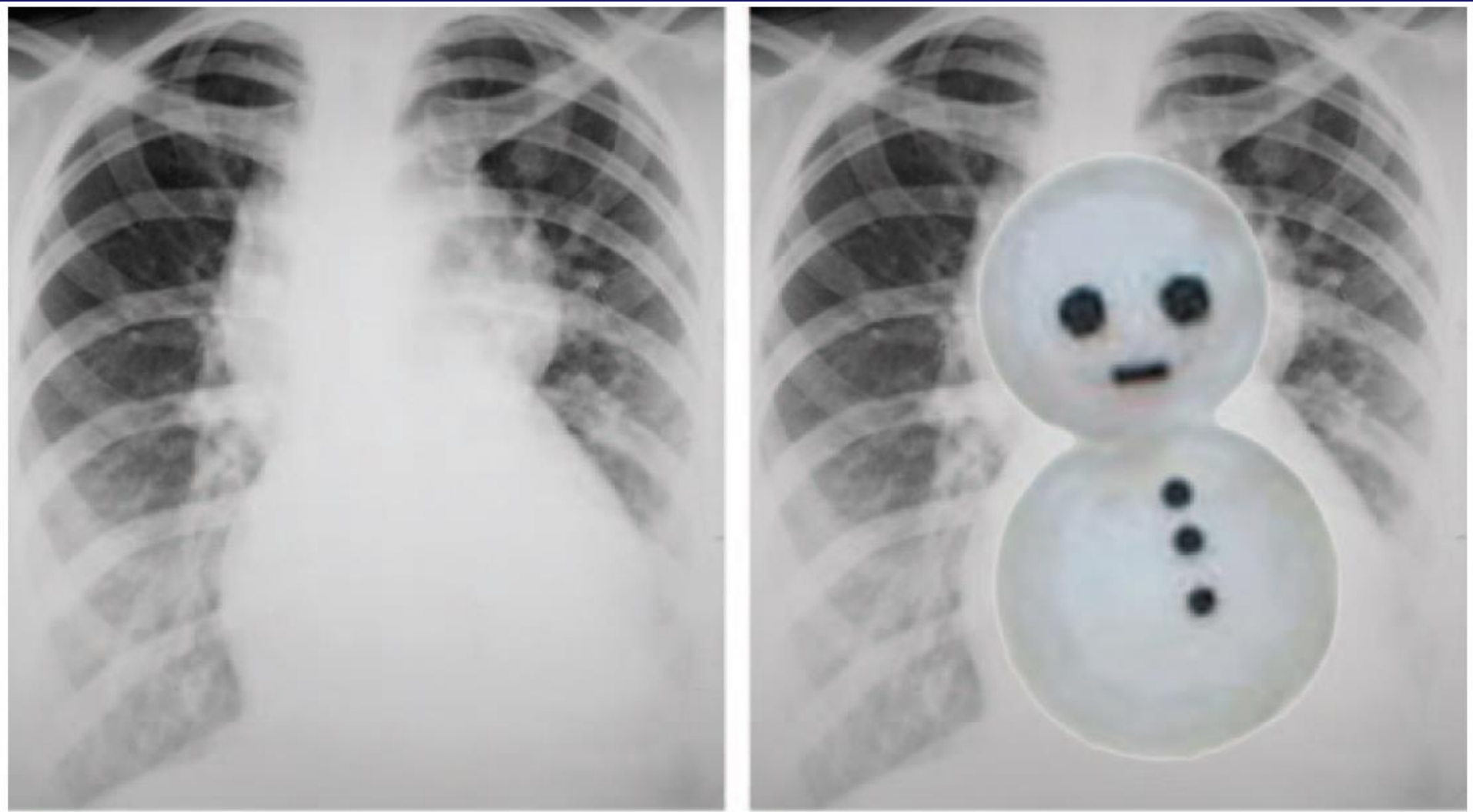


- Treatment options:
 - Closure ASD-VSD components
 - Reconstruction AV-valves
- Follow-up:
 - AV-valve regurgitation
 - AV-valve stenosis
 - Residual shunt
 - Arrhythmias-AV block



4D flow na AVSD correctie



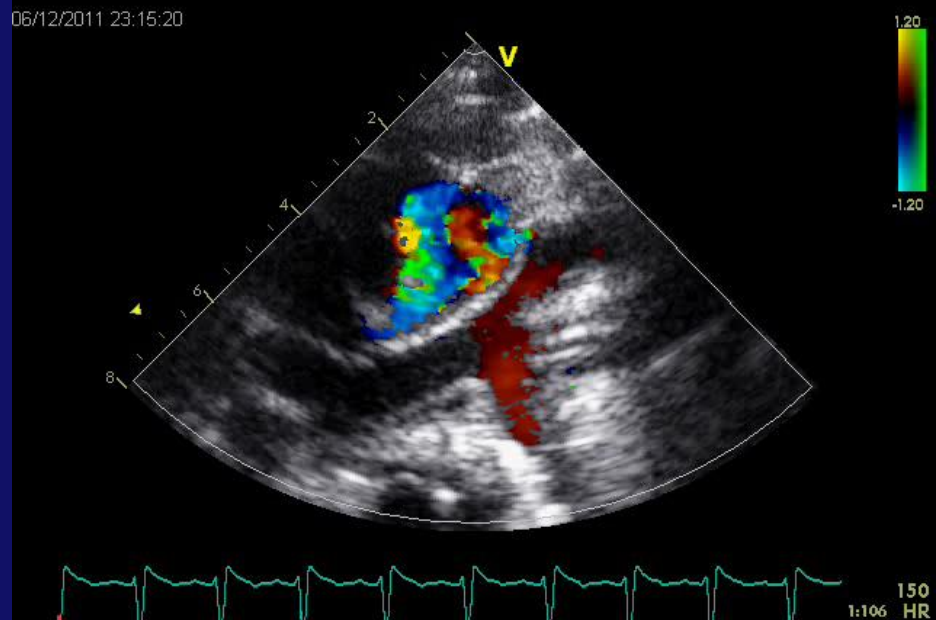
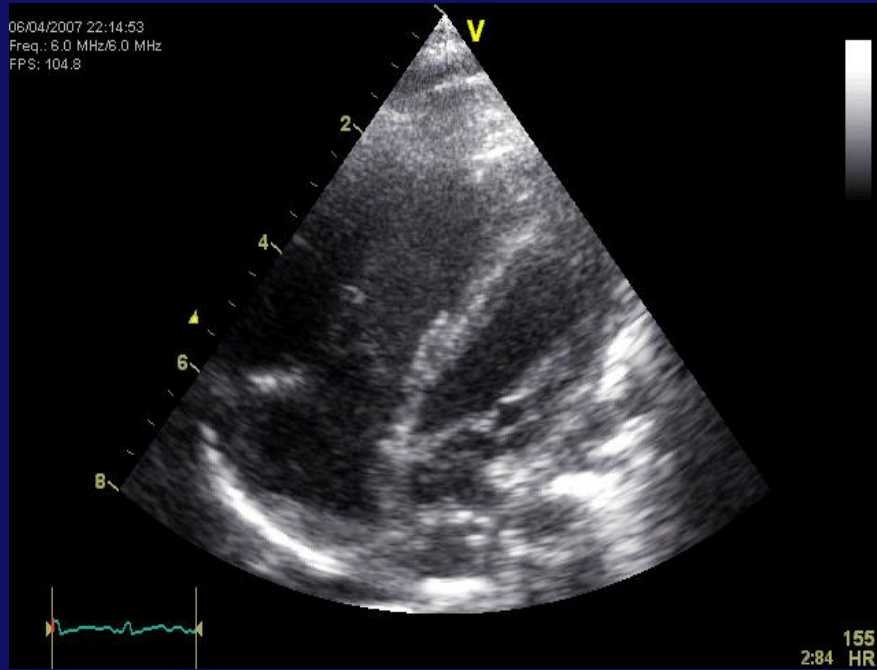
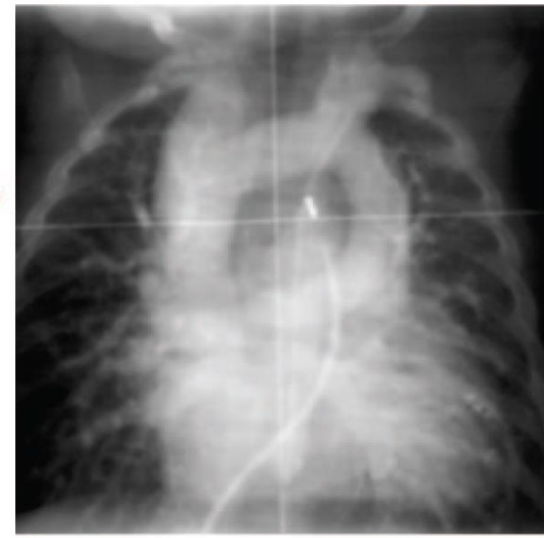
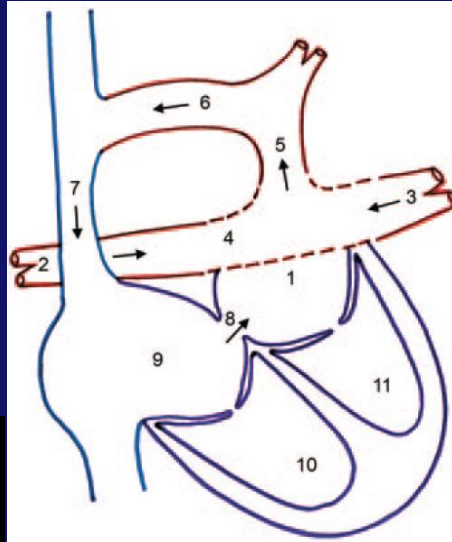
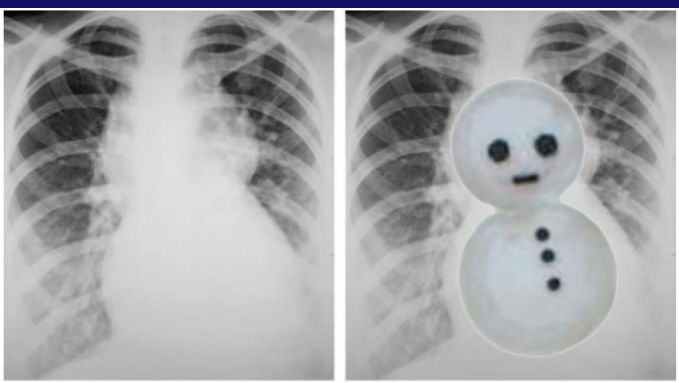


Welke hartafwijking

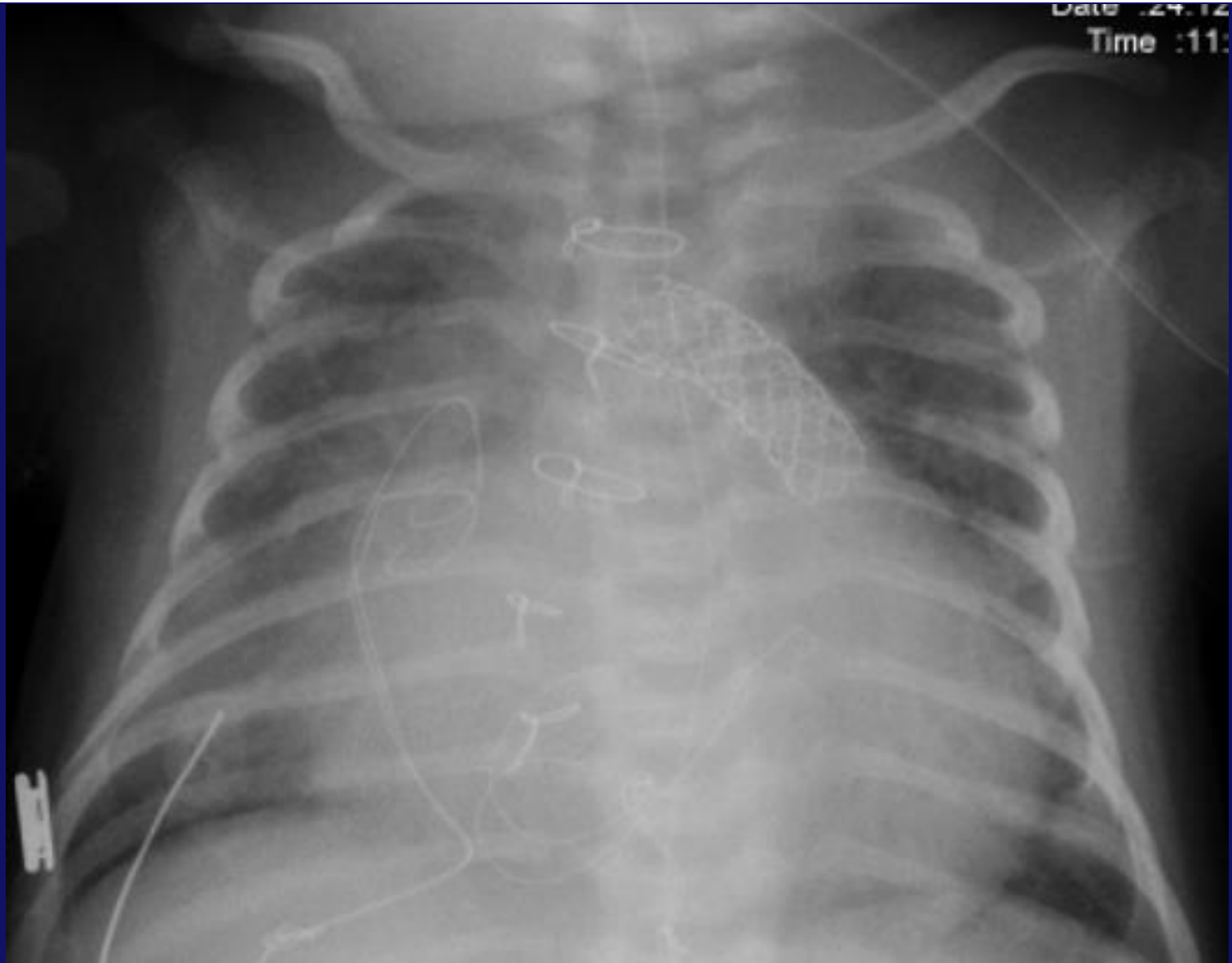
- Totaal abnormaal pulmonaal veneuze retour
- Hypoplastisch linker hart syndroom
- Aortklep stenose
- Coarctatio aortae

Welke hartafwijking

- Totaal abnormaal pulmonaal veneuze retour
- Hypoplastisch linker hart syndroom
- Aortklep stenose
- Coarctatio aortae



En waar zit deze stent?



- Stent in aorta
- Stent in arteria pulmonalis
- Stent in ductus
- Stent in vena cava superior

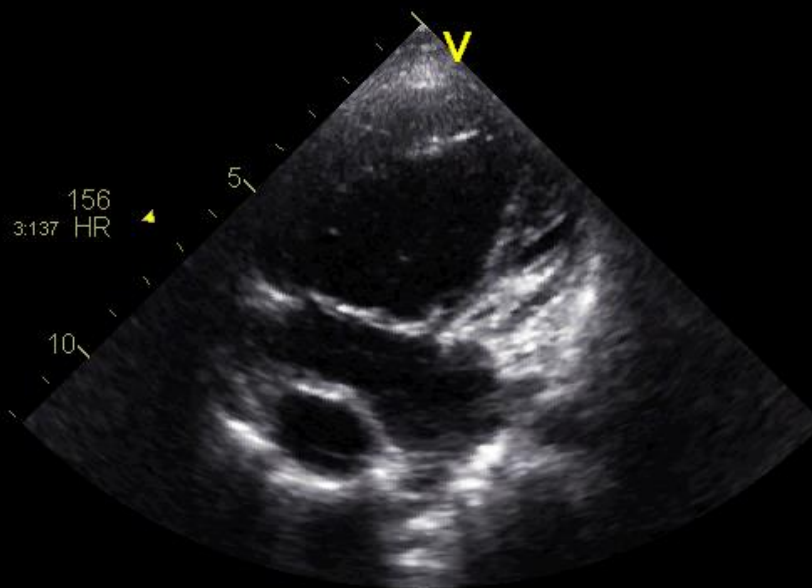
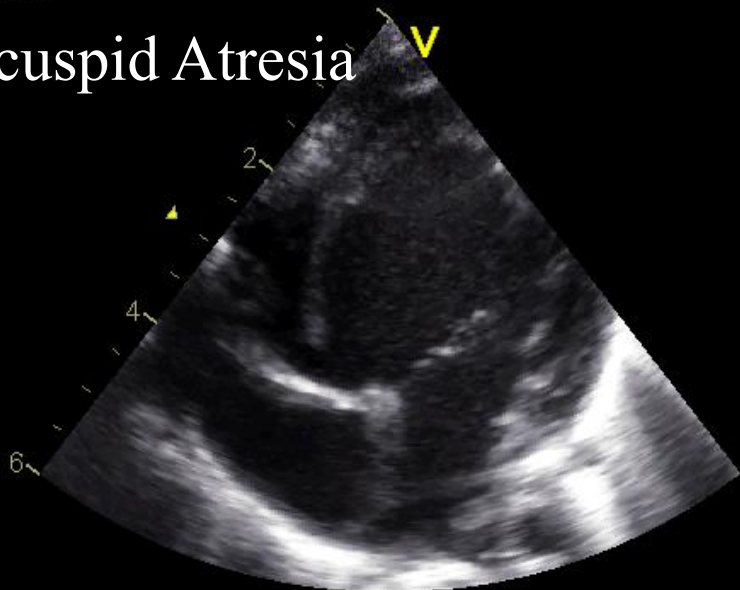
- Stent in aorta
- Stent in arteria pulmonalis
- Stent in ductus
- Stent in vena cava superior

- Types:
 - Tricuspid atresia
 - Hypoplastic left/right heart syndrome
 - Unbalanced AVSD
 - Etc
- Presentation:
 - Depending on defect
 - Cyanosis-duct dependency

Univentricular heart-TA & HLHS

30/09/2010 09:59:38

Tricuspid Atresia



Hypoplastic Left Heart

Fontan circulation

- First step: Bidirectional Glenn anastomosis
- Second step: Fontan circulation
 - Classic: Connection between right atrium or caval veins and pulmonary circulation
 - Now: total cavo-pulmonary circulation
 - ICV to pulmonary circulation: extracardiac-lateral tunnel
- Follow-up
 - Ventricular failure
 - Arrhythmias
 - Protein losing enteropathy

Fontan circulation

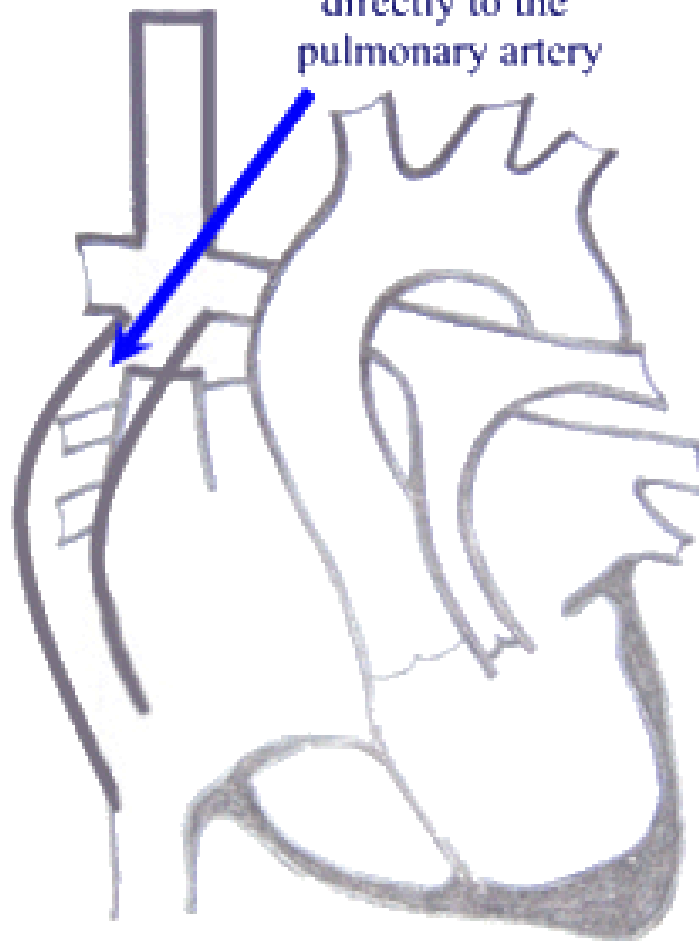
Glenn Operation

SVC is connected
to the pulmonary artery



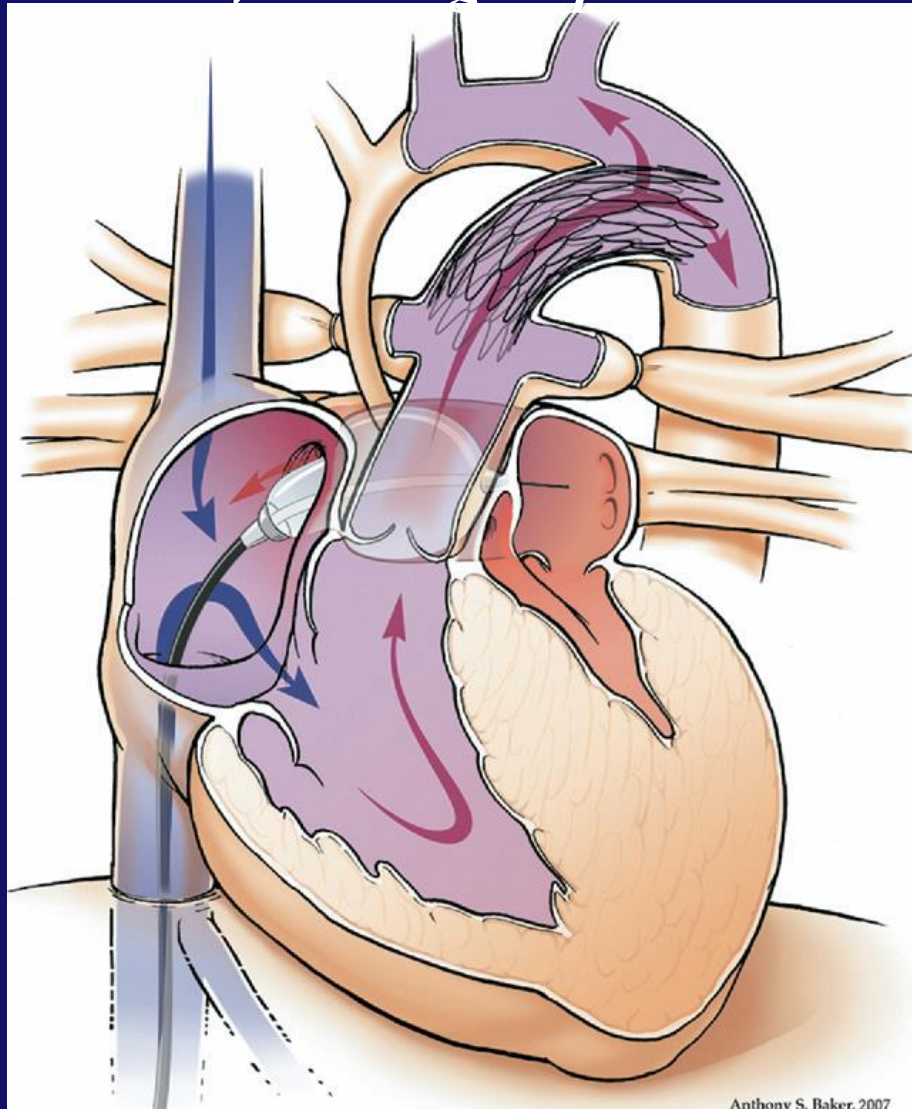
Fontan Operation

Additional tube to take
blood from IVC
directly to the
pulmonary artery



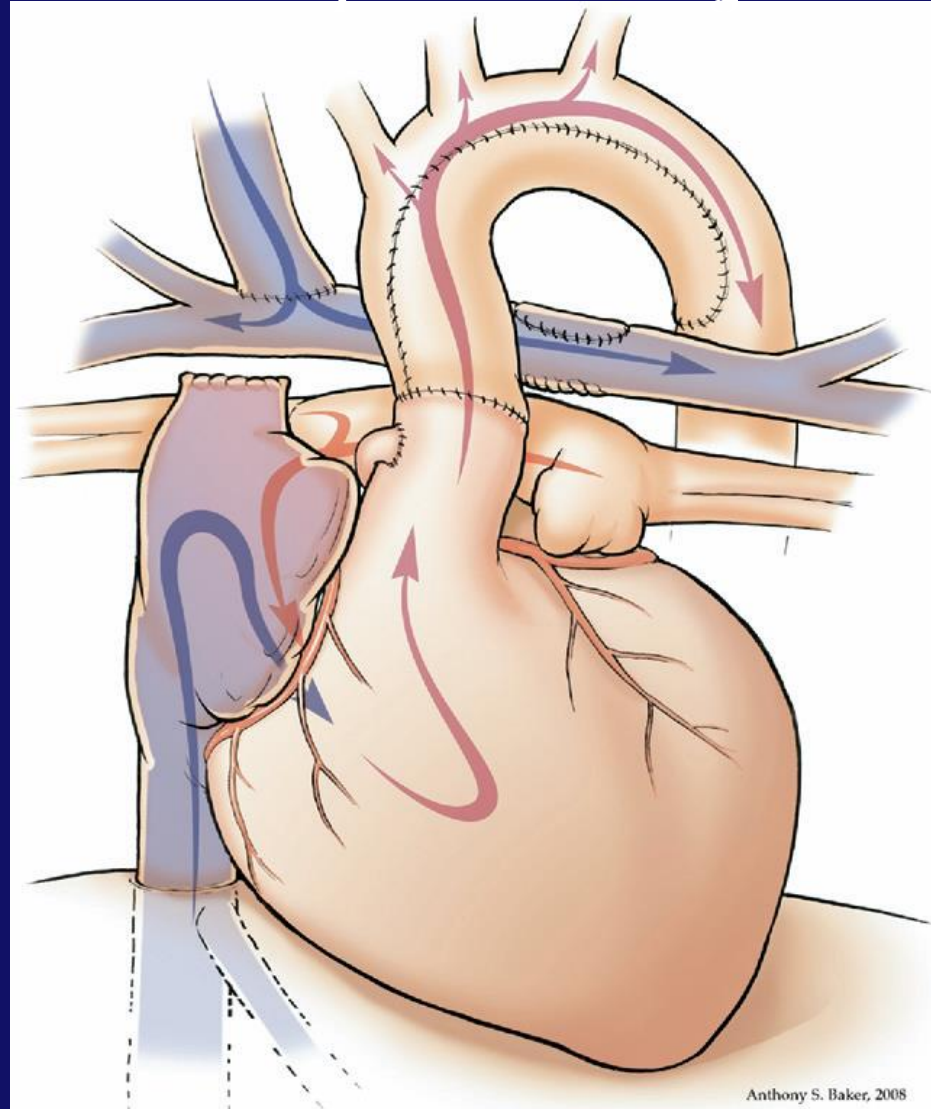
Hybrid Approach for Hypoplastic Left Heart Syndrome

The hybrid stage 1 palliation



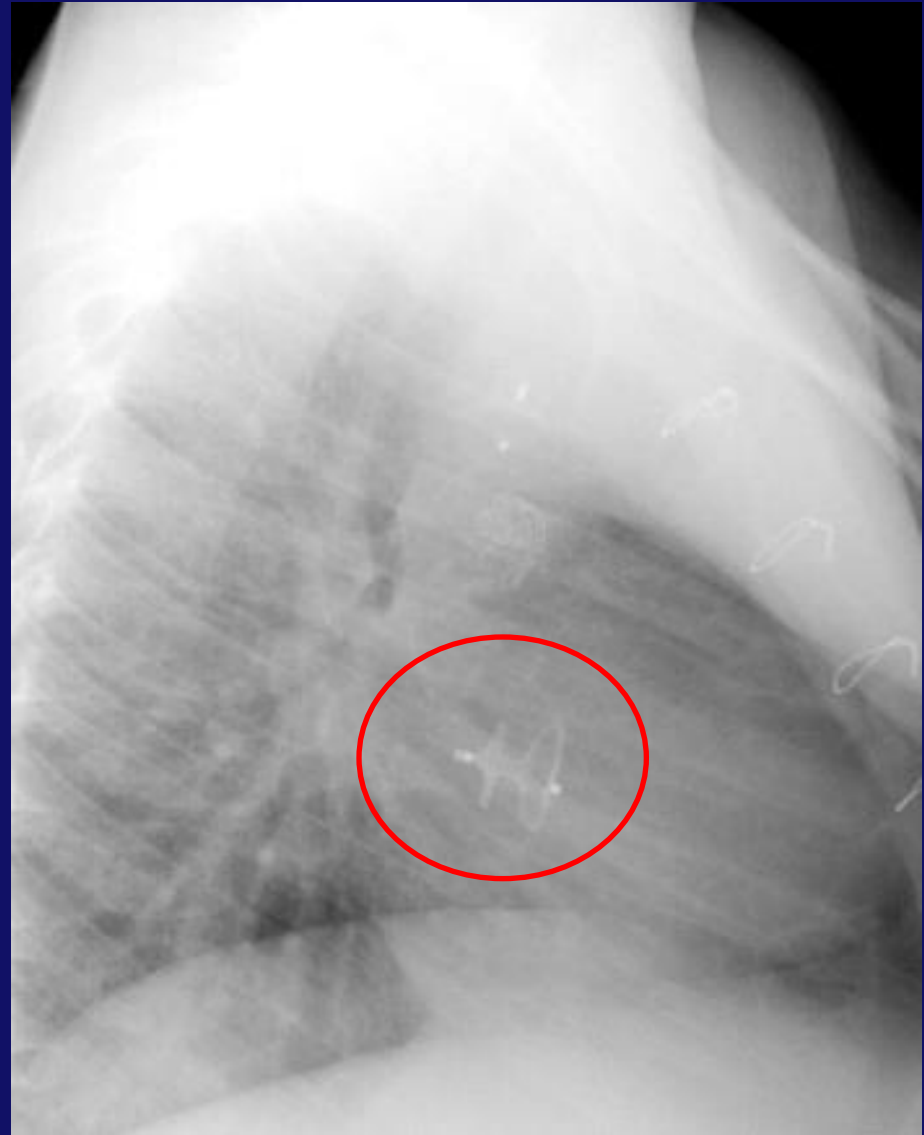
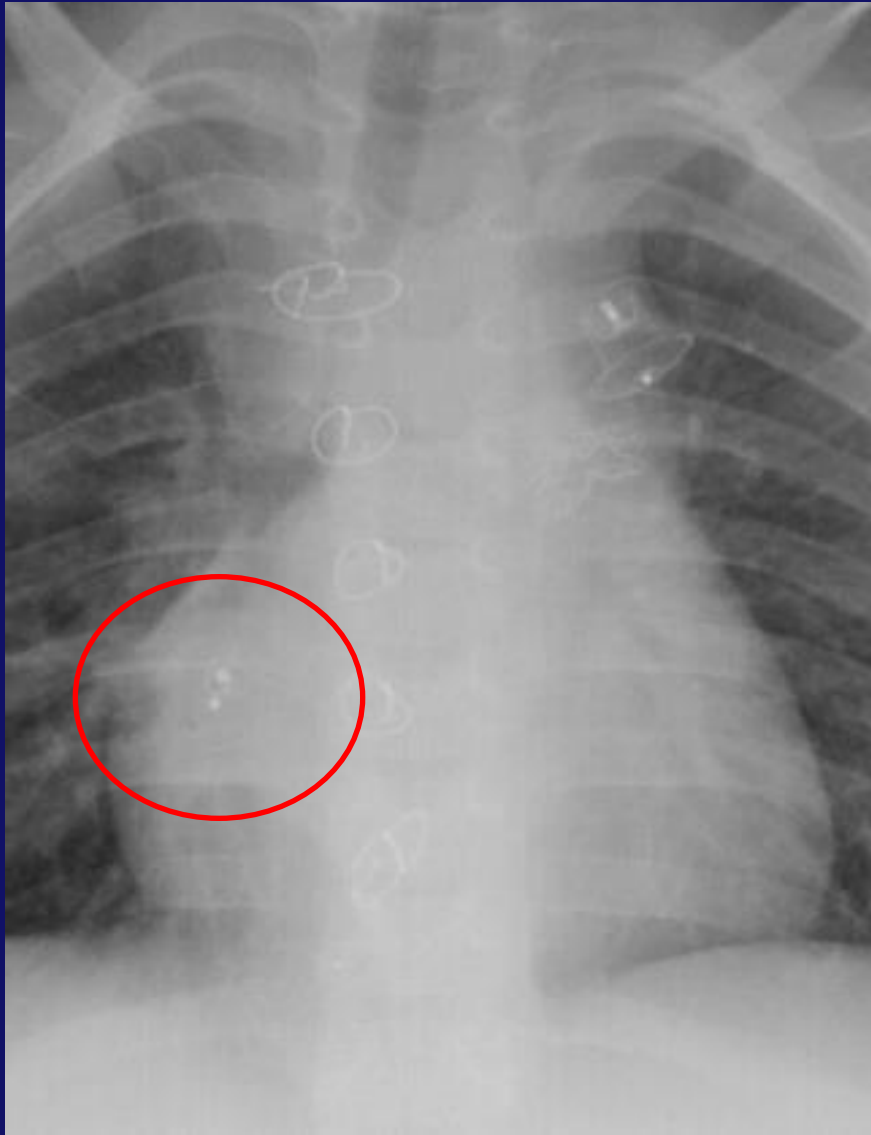
Anthony S. Baker, 2007

The comprehensive stage 2



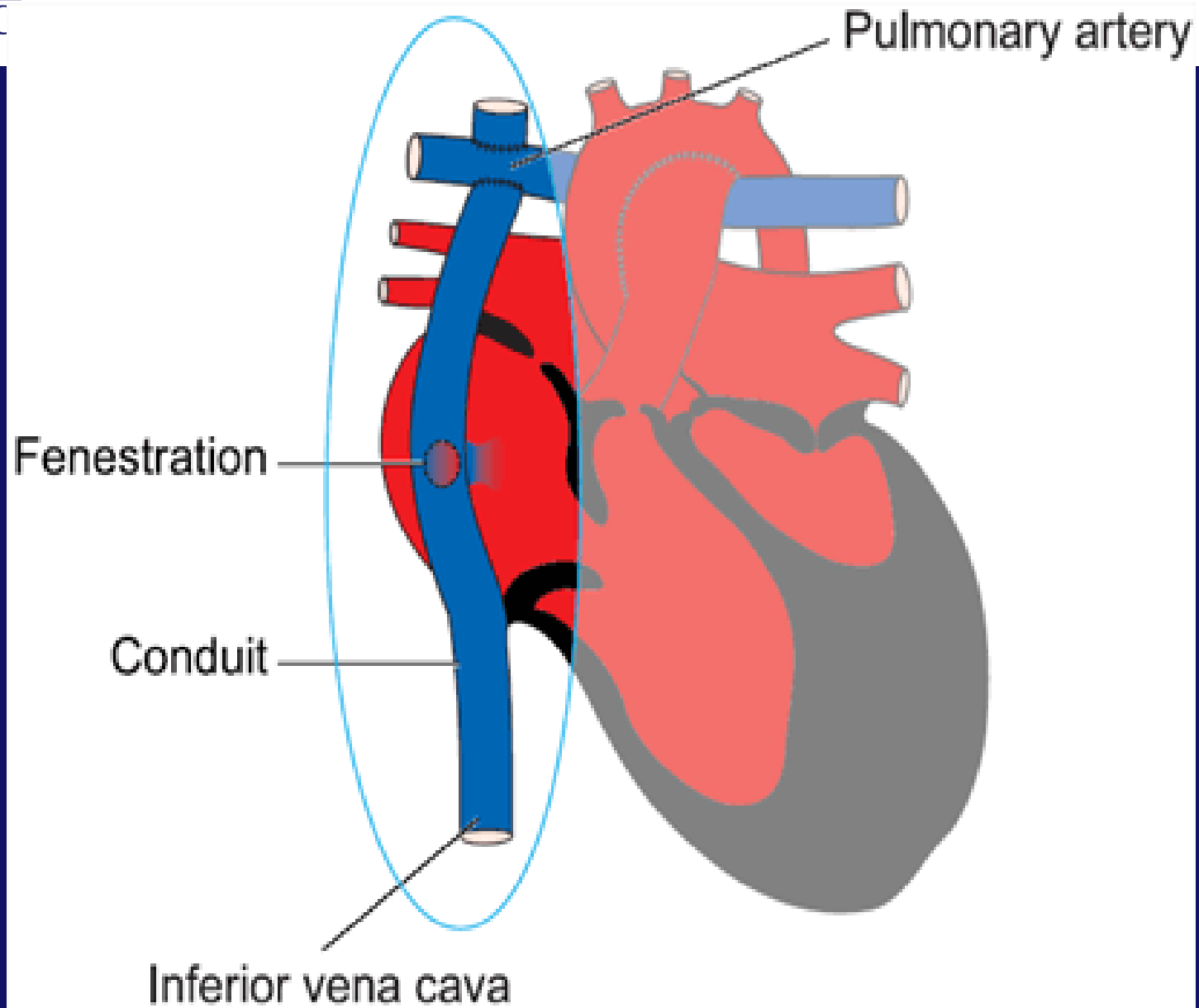
Anthony S. Baker, 2008

Jongen, na completeren Fontan circulatie



- Plug in ductus
- Device in ASD
- Device in fenestratie
- Stent in vena cava inferior

- Plug in ductus
- Device in ASD
- Device in fenestratie
- Stent in vena cava inferior



Waarom fenestratie?

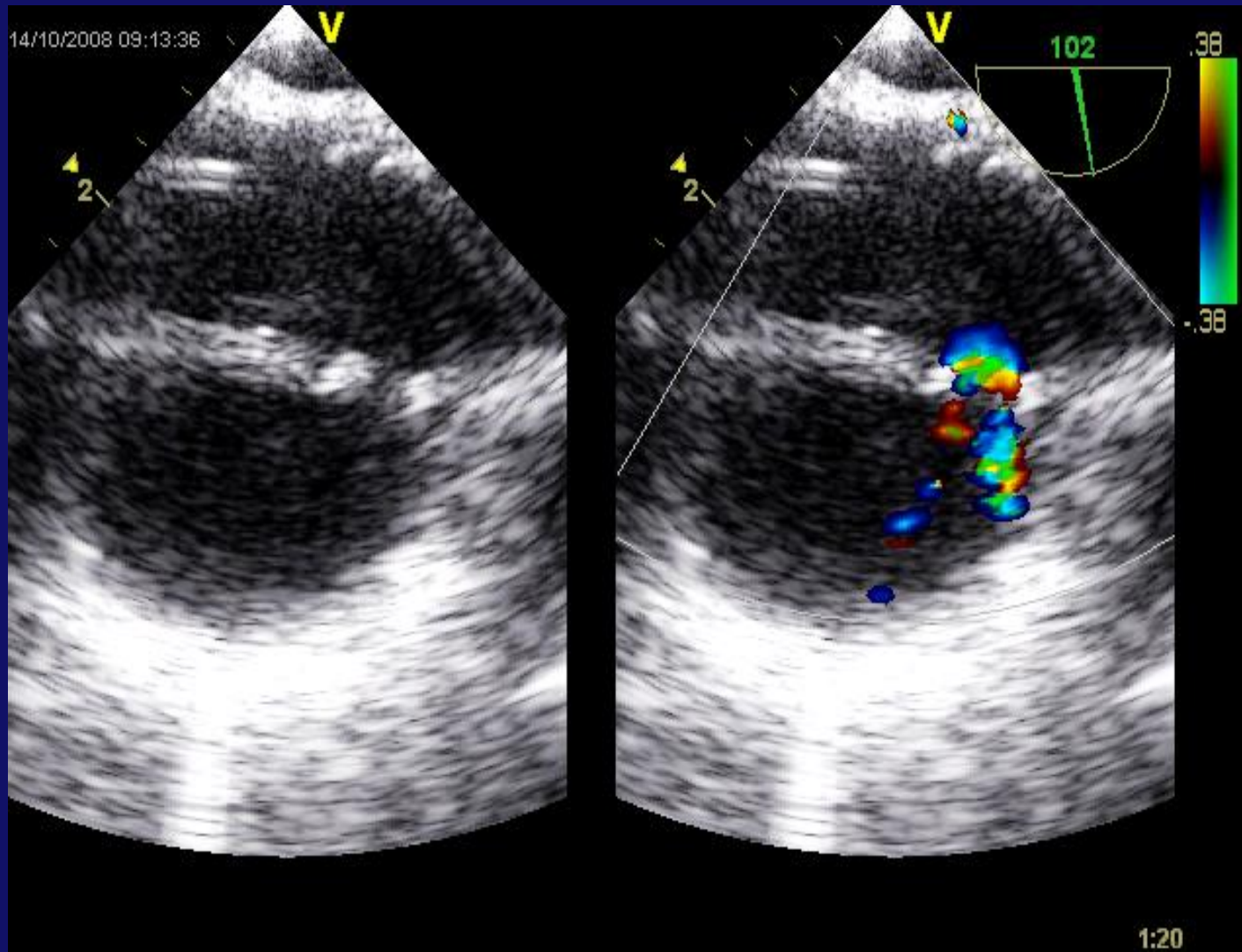
- In hoog risico patienten*:
 - Lage mortaliteit
 - Minder pleuravocht
 - Kortere opname duur
- Elective fenestratie#: ter discussie@
 - Mogelijk verminderd Fontan-falen
 - Mogelijk minder pleuravocht

*Bridges et al. *Circulation* 1992;86:1762-1769

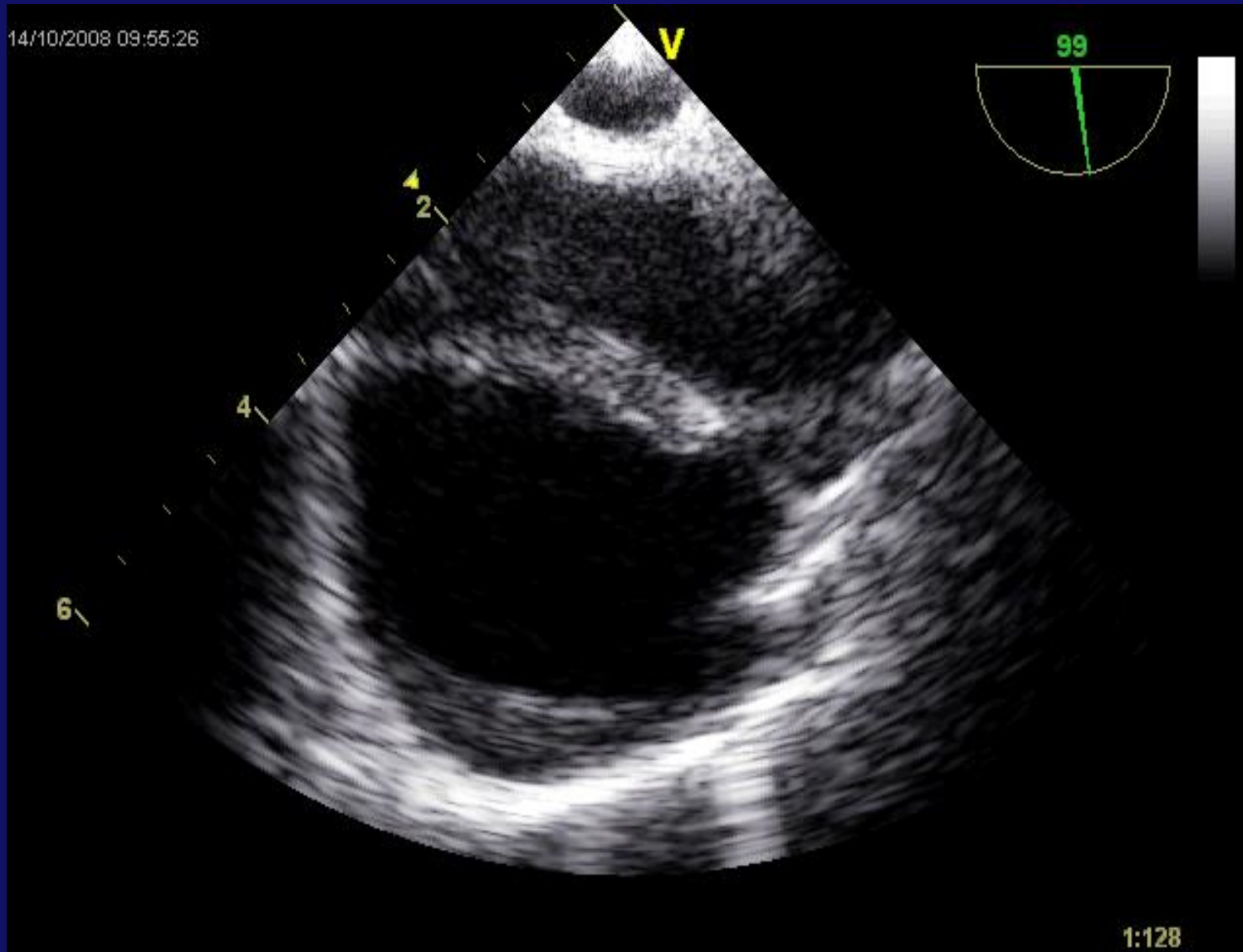
#Airam et al. *Ann Thorac Surg* 2000;69:1900-1906

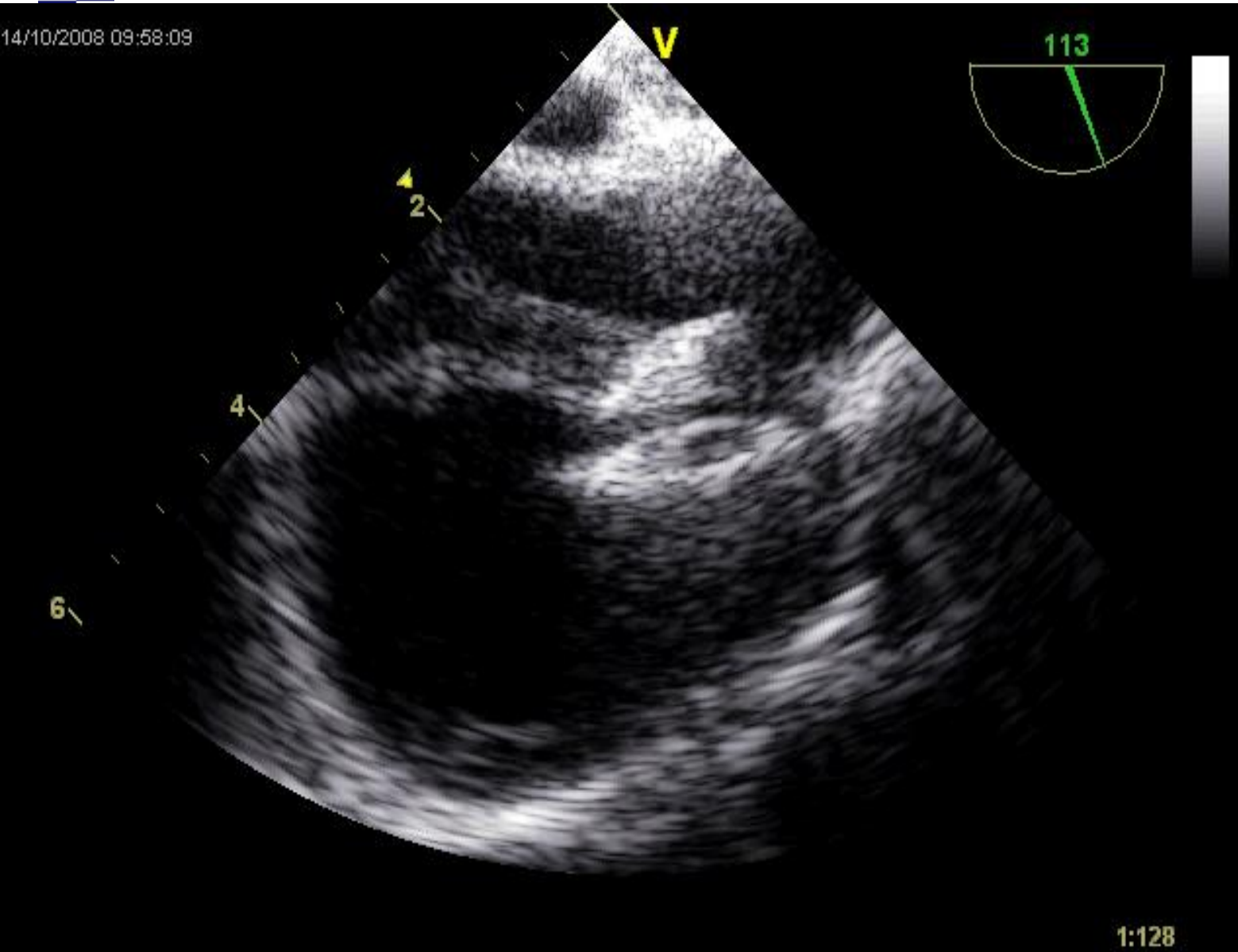
@Hosein et al. *Eur J Thorac Surg* 2007;31:344-353

- Toch weer R-L shunt met lagere saturaties
- Door verbinding van veneus lage snelheid bloedstroom met systeem arteriele circulatie verhoogde kans op paradoxe thrombo-embolische complicaties
- Noodzaak voor antistollingstherapie
- Na bepaalde periode: cathetersluiting

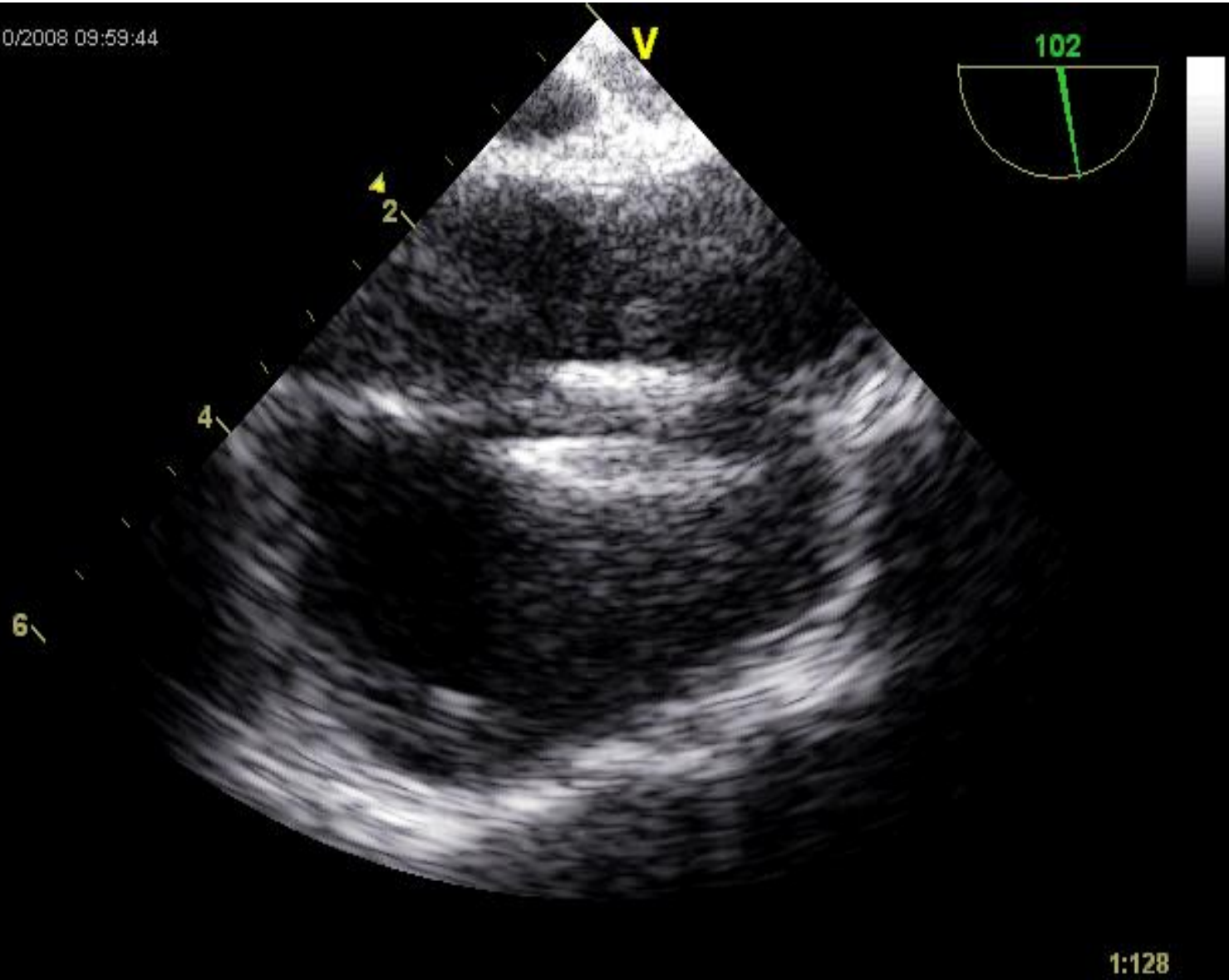


14/10/2008 09:55:26

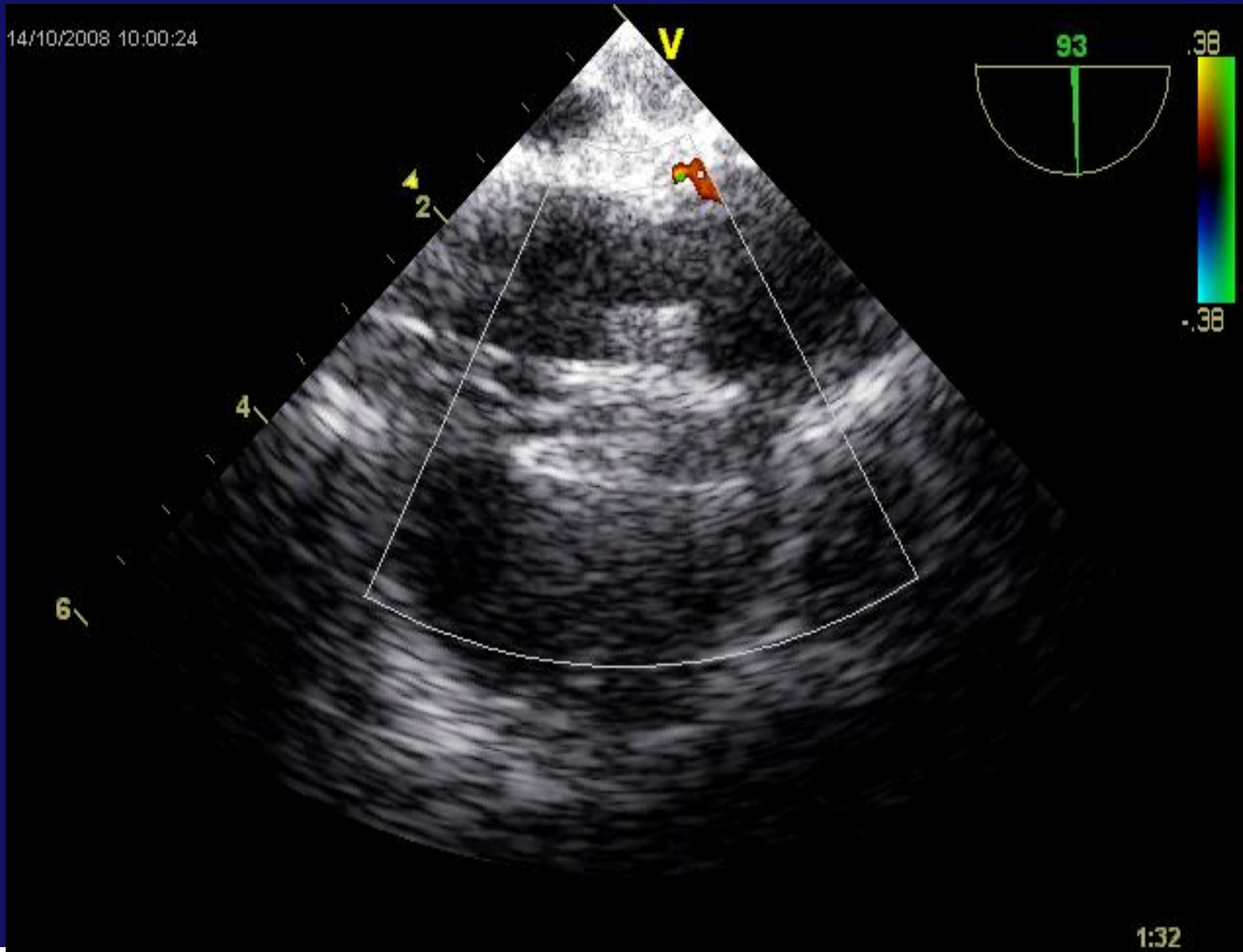




14/10/2008 09:59:44



14/10/2008 10:00:24



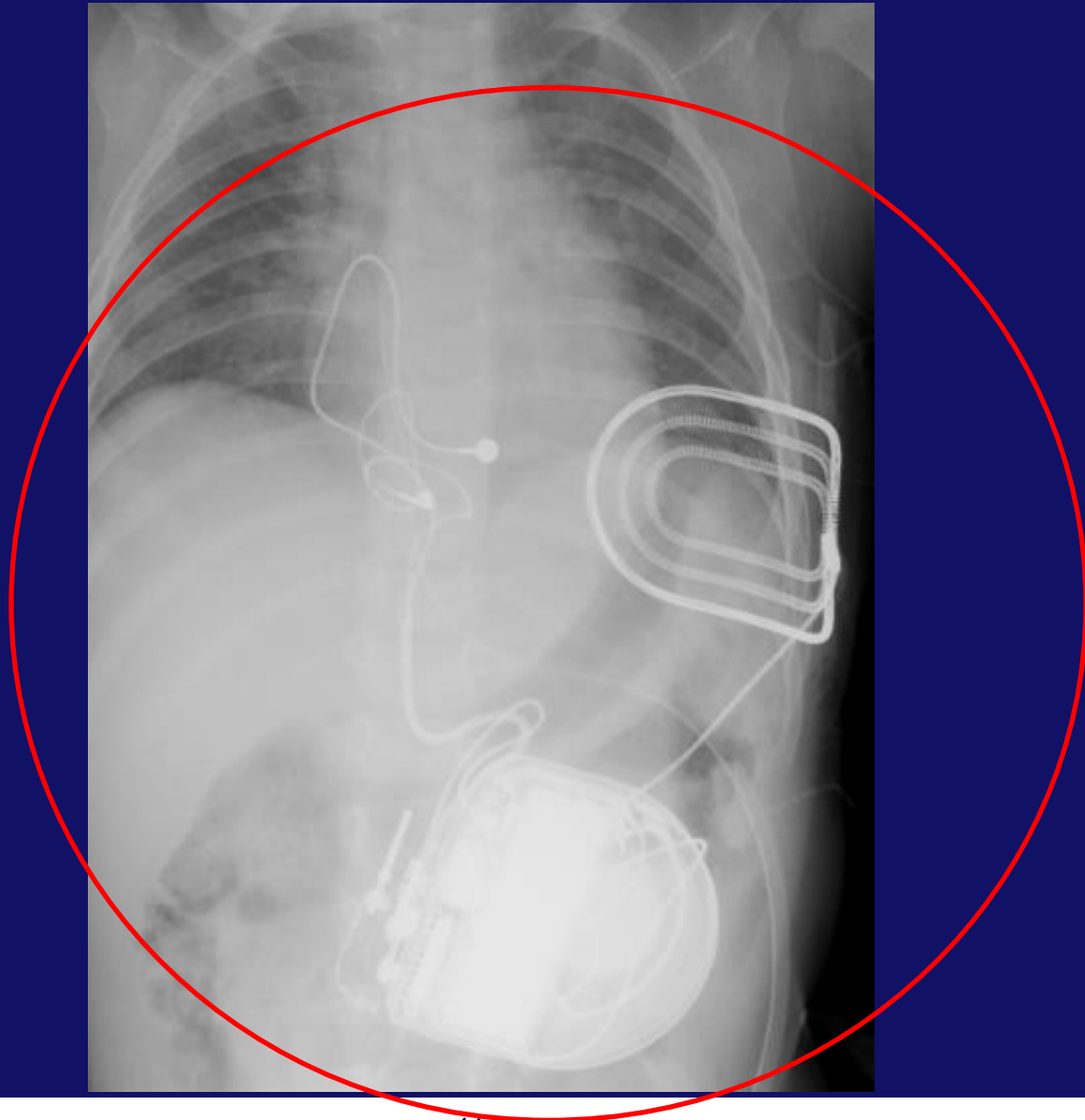
1:32

Conclusies: aangeboren hartafwijkingen

- Meest voorkomende aangeboren afwijking
- Manier van presentatie:
 - Prenataal
 - Cyanose: Tetralogie of Fallot, TGA, Univentriculaire harten
 - Pulmonale overflow: Groot VSD, ASD, AVSD
 - Hartfalen: Critical PS, AoS, coarctation of aorta
 - Hartgeruis: Small VSD/ASD/ODB
- Meestal wat aan te doen
- Geven raadselachtige afwijkingen op de x-thorax...

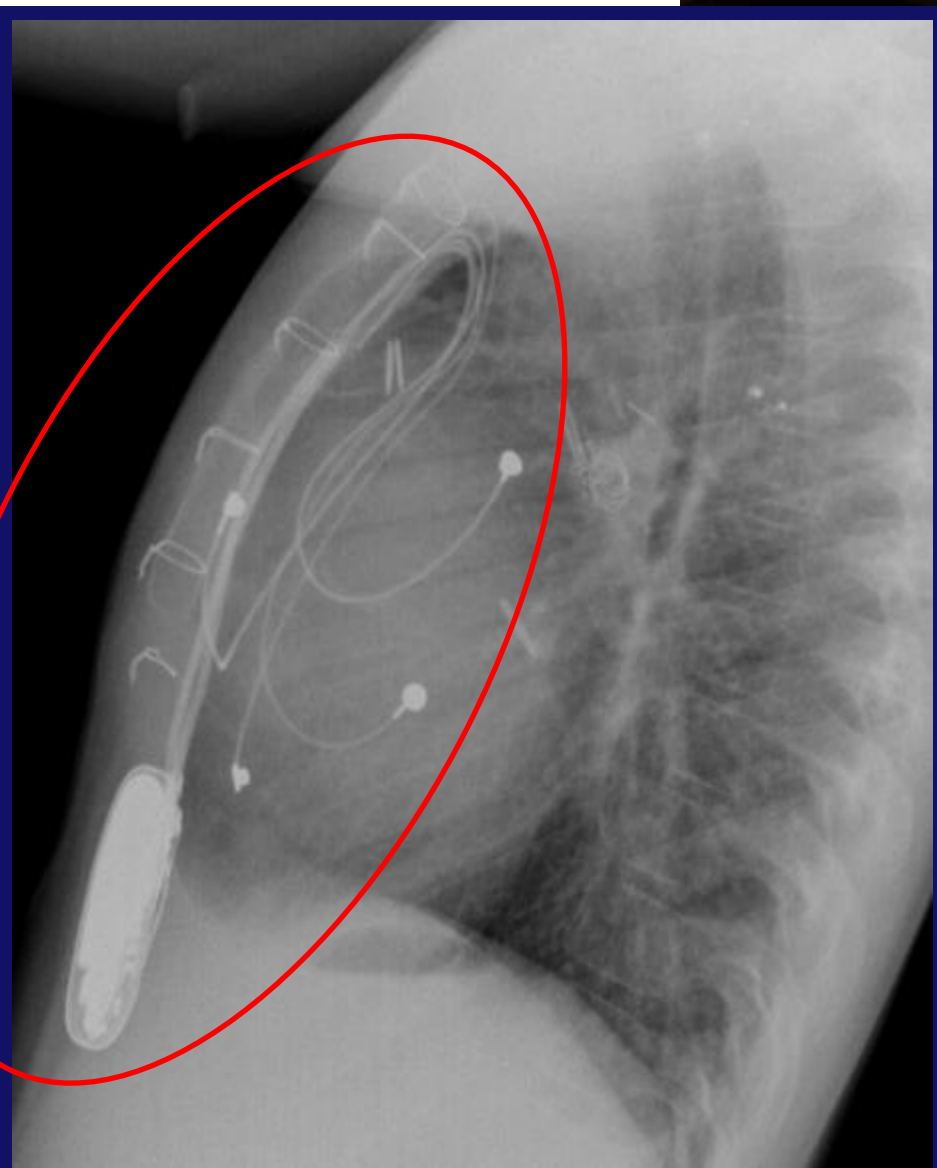
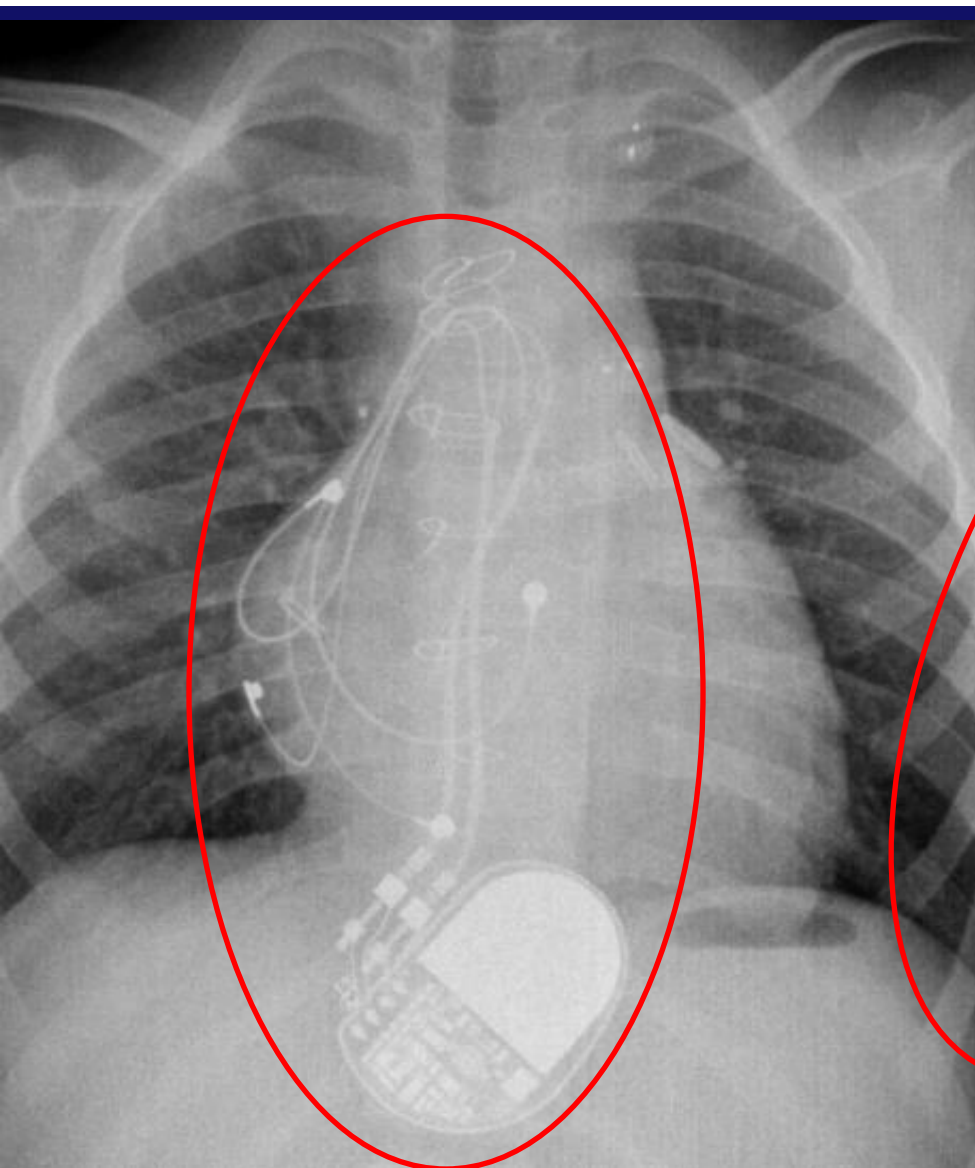
Dank voor de aandacht





- Een biventriculaire pacemaker
- Een transveneuze pacemaker
- Een epicardiale pacemaker
- Een epicardiale ICD

- Een biventriculaire pacemaker
- Een transveneuze pacemaker
- Een epicardiale pacemaker
- Een epicardiale ICD



- Een DDD epicardiale pacemaker
- Een VVI epicardiale pacemaker
- Een DDD transveneuze pacemaker
- Een VVI transveneuze pacemaker

- Een DDD epicardiale pacemaker
- Een VVI epicardiale pacemaker
- Een DDD transveneuze pacemaker
- Een VVI transveneuze pacemaker

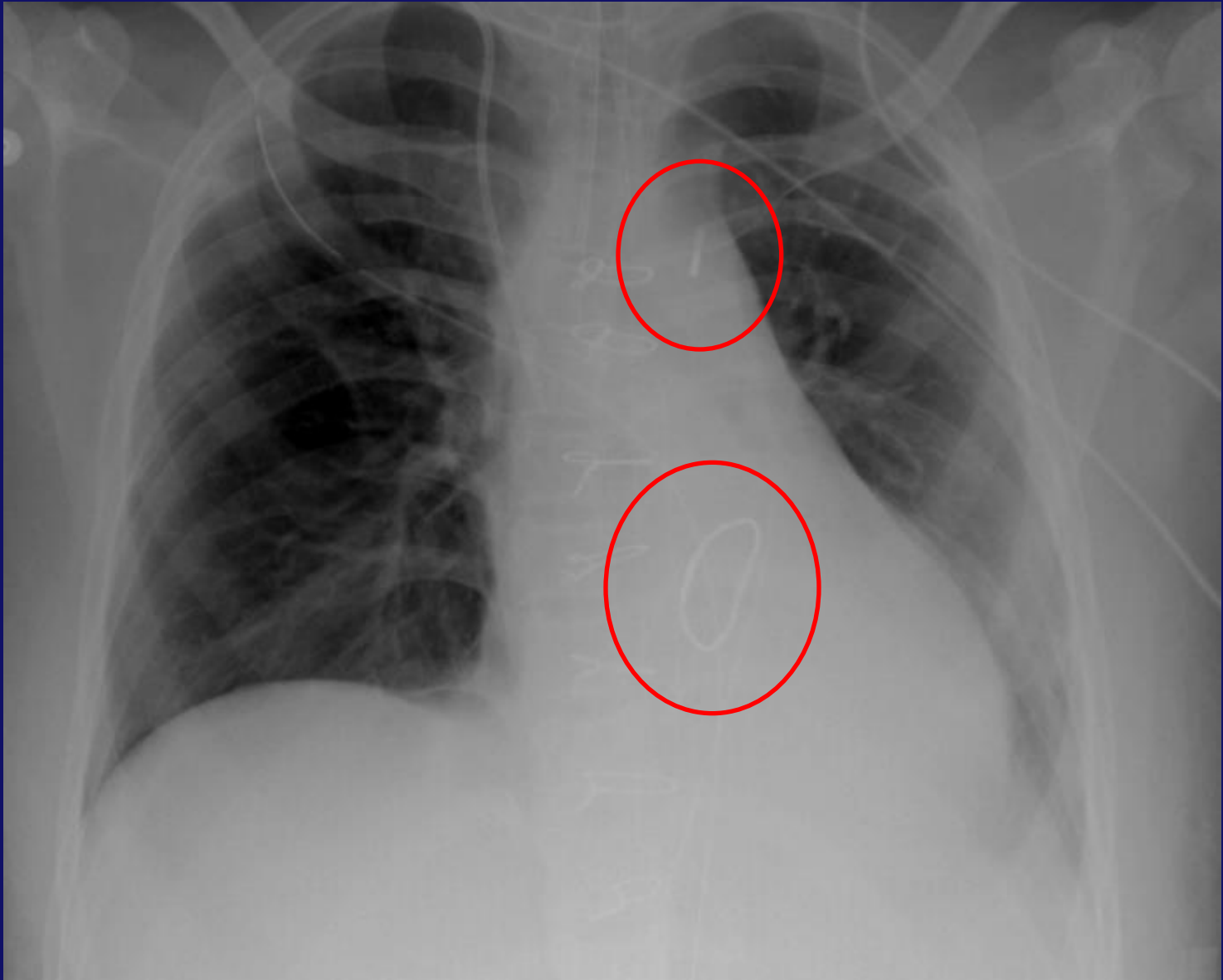
Category	Chamber(s) Paced	Chamber(s) Sensed	Response to Sensing	Programmability Rate Modulation	Antitachyarrhythmia Function(s)
	O = None A = Atrium V = Ventricle D = Dual (A+V)	O = None A = Atrium V = Ventricle D = Dual (A+V)	O = None T = Triggered I = Inhibited D = Dual (T+I)	O = None P = Simple Programmable M = Multiprogrammable C = Communicating R = Rate Modulation	O = None P = Pacing S = Shock D = Dual (P+S)
	S = Single (A or V)	S = Single (A or V)			

Note: Positions I through III are used exclusively for antibradyarrhythmia function

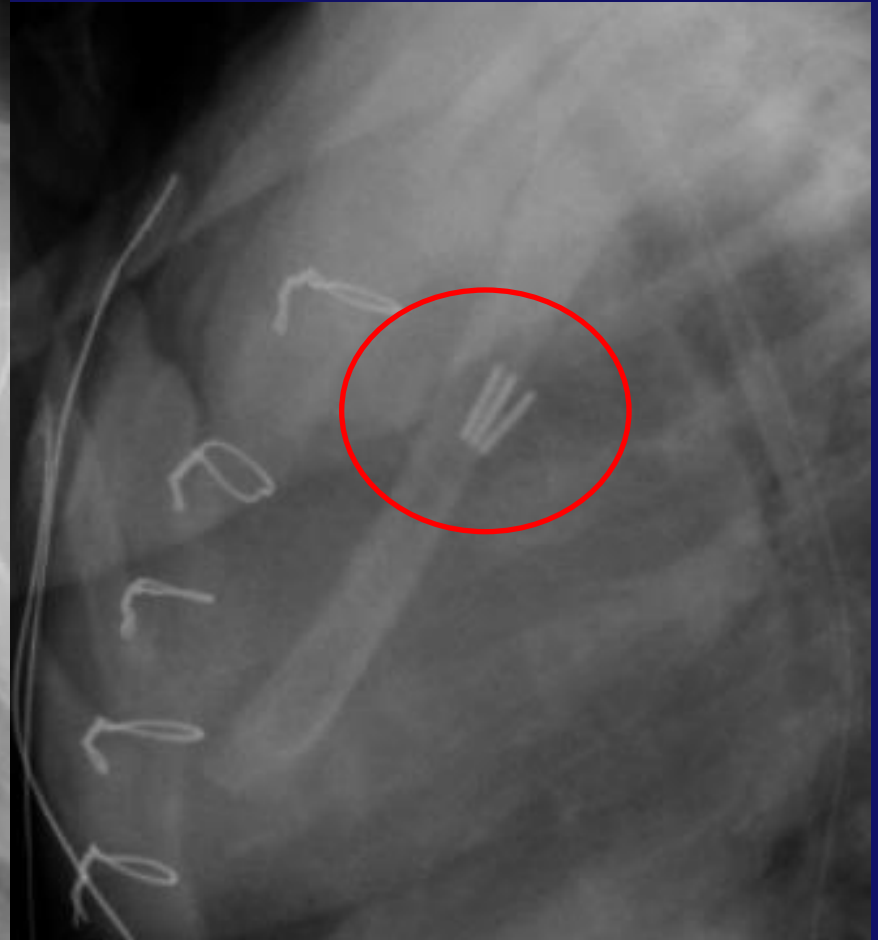
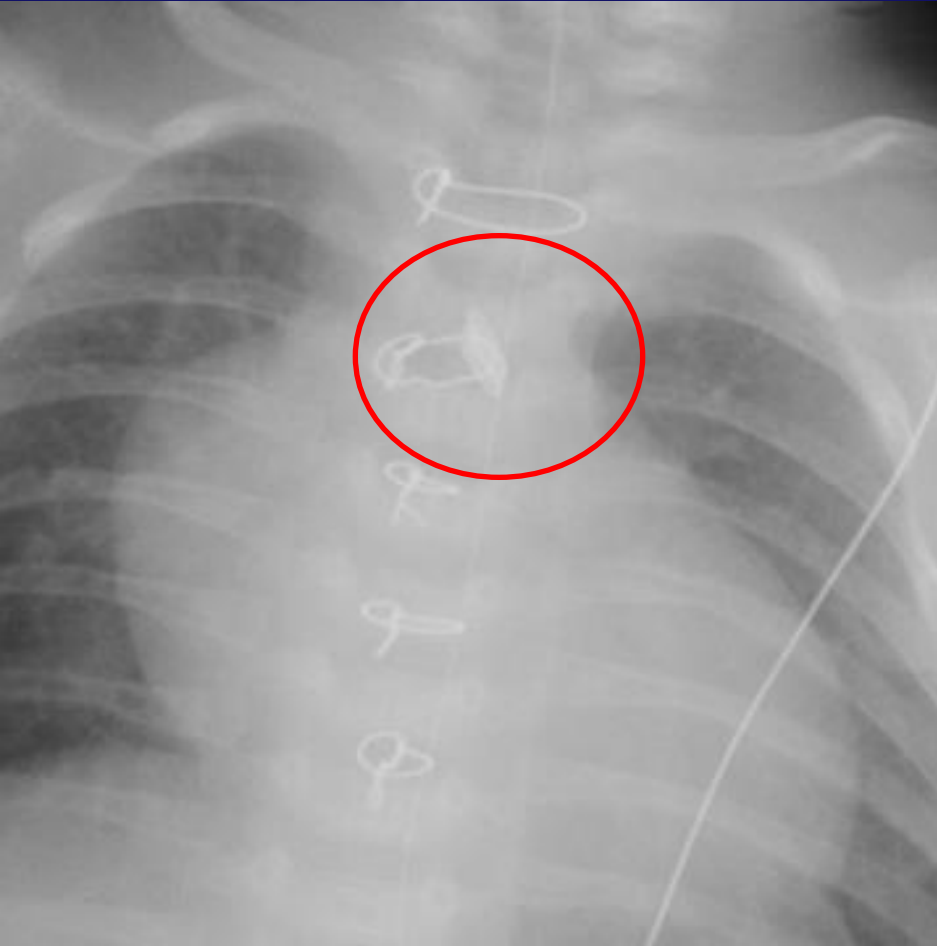
Vascular plug



AMPLATZER® Vascular Plug III
© AGA Medical Corporation

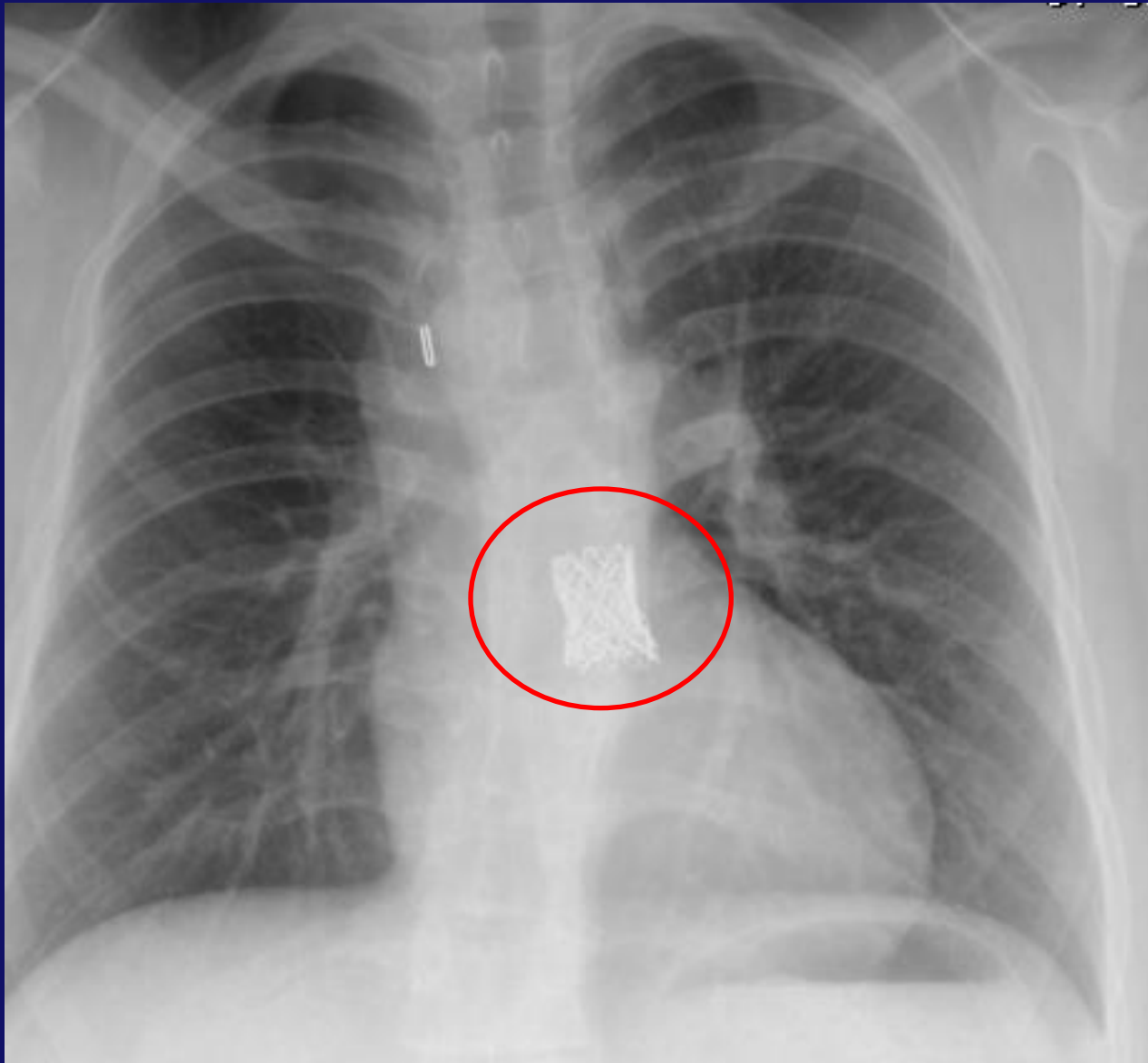


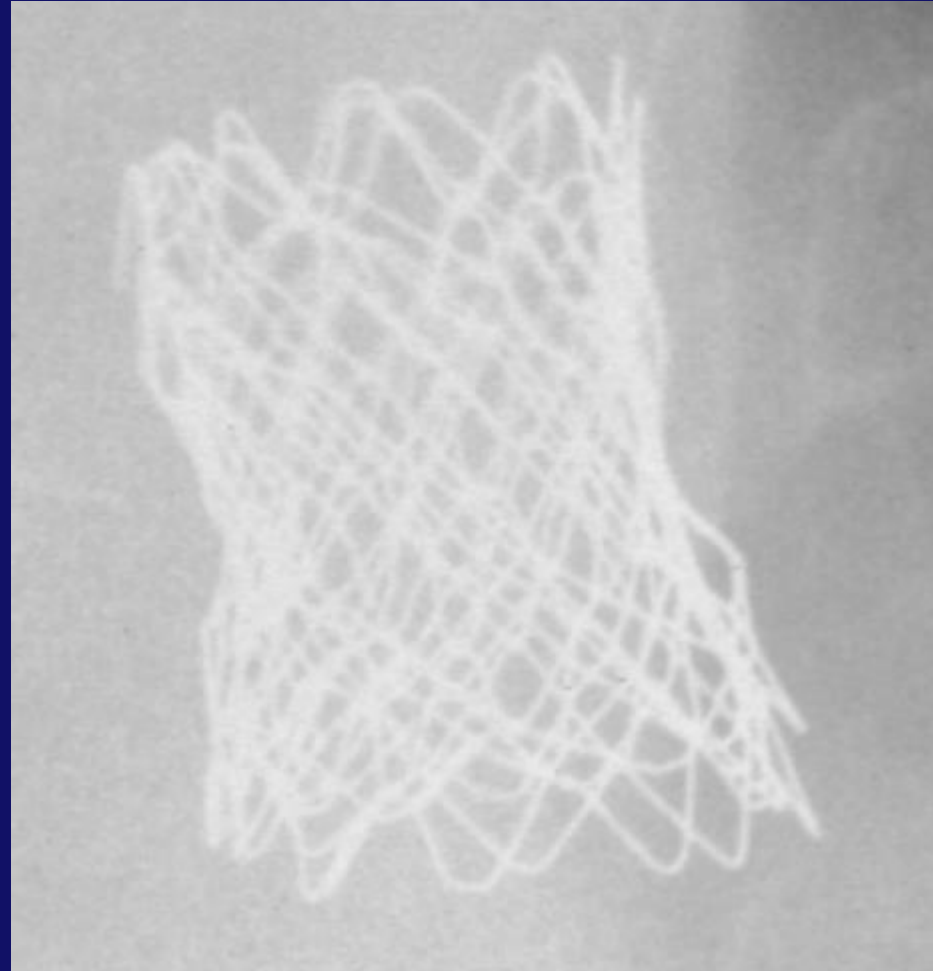
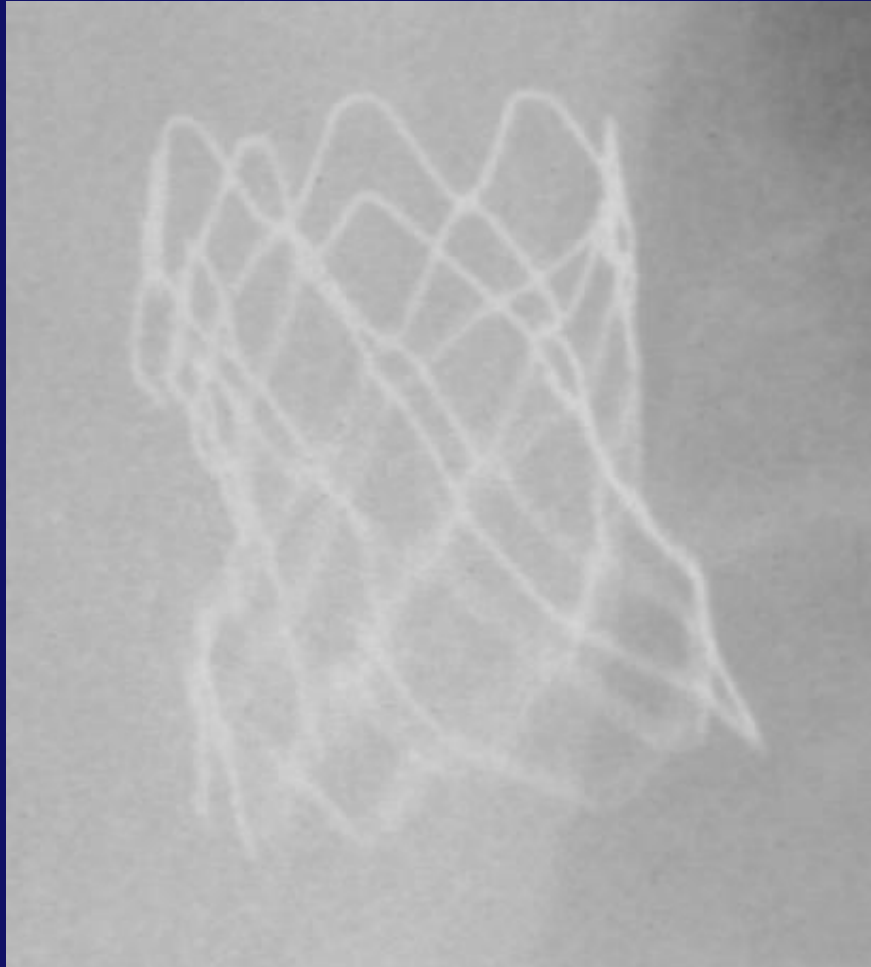
Jongen, 1,5 mnd met DORV



- Bij te veel longflow en waarbij het onderliggend defect niet meteen is te corrigeren
- Voorbeelden:
 - Double outlet right ventricle zonder PS
 - VSD(s), dat/die niet in 1 keer te sluiten zijn

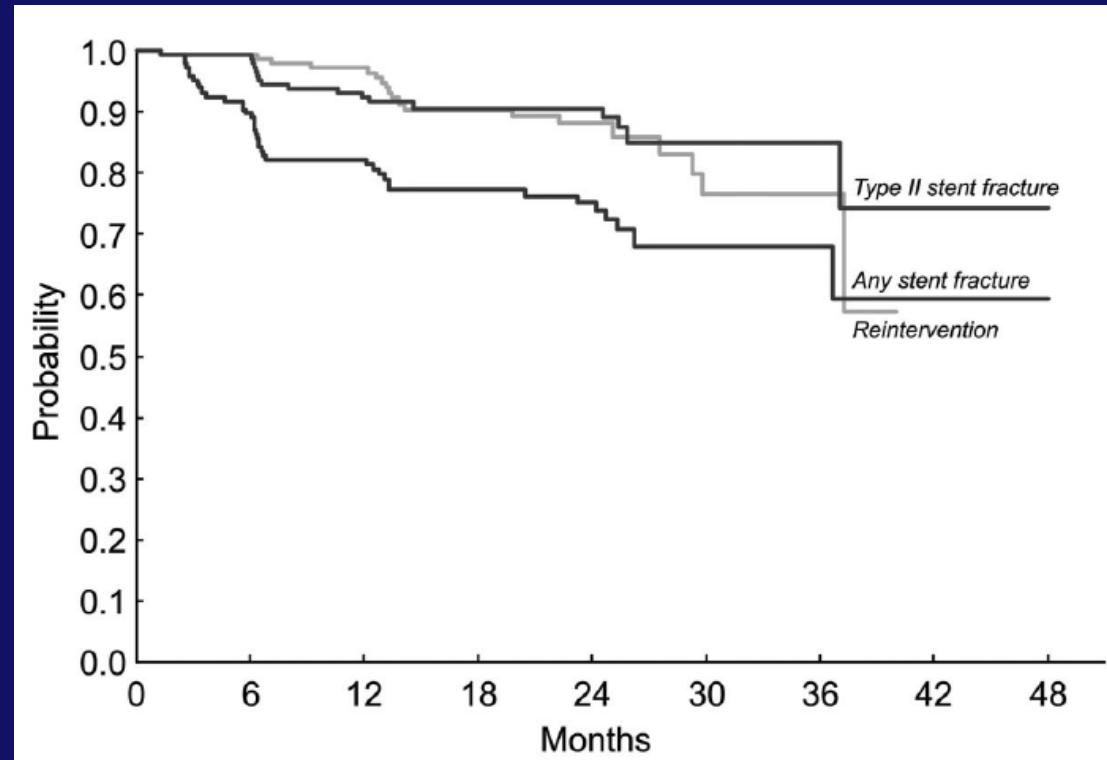
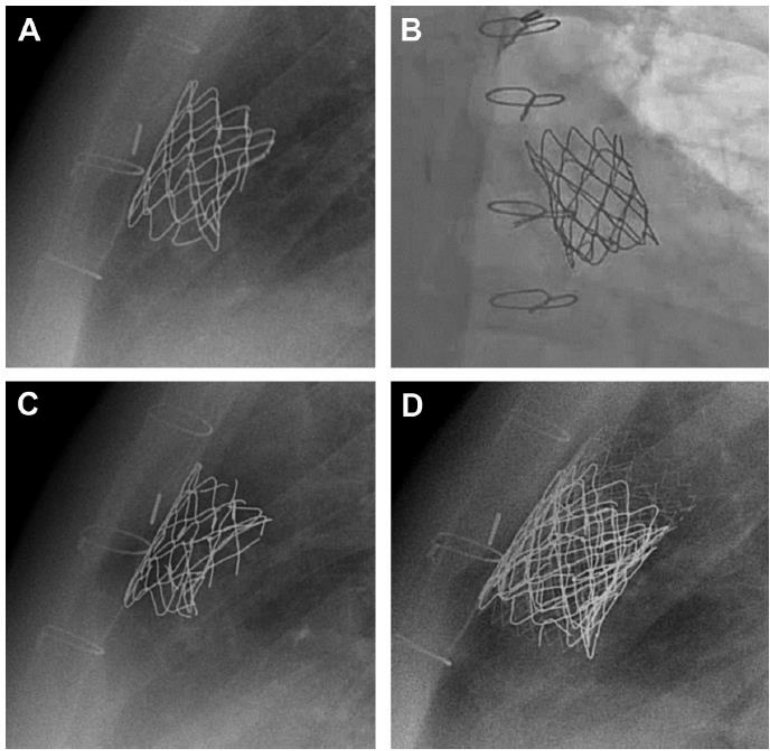
Meisje, 15-4-1994,





**Stent Fracture, Valve Dysfunction, and Right Ventricular
Outflow Tract Reintervention After Transcatheter
Pulmonary Valve Implantation**
Patient-Related and Procedural Risk Factors in the US Melody
Valve Trial

Doff B. McElhinney, MD; John P. Cheatham, MD; Thomas K. Jones, MD; James E. Lock, MD;
Julie A. Vincent, MD; Evan M. Zahn, MD; William E. Hellenbrand, MD



Conclusions—MSF was common after TPV implant in this multicenter experience and was more likely in patients with severely obstructed RVOT conduits and when the TPV was directly behind the anterior chest wall and/or clearly compressed. A TPV implant site protected by a prestent or bioprosthesis valve was associated with lower risk of MSF and reintervention.