



The Special Echo Case

Michel Romanens, Olten

15 year old male subject with
a systolic murmur



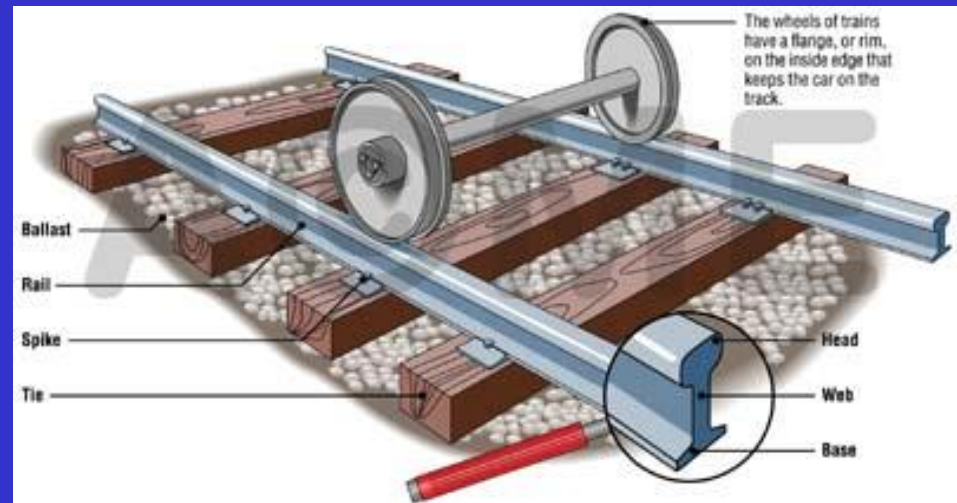
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History: unremarkable

15 years old in search of work
Planning to become an apprentice for
train track building
in the statal Swiss Railway
Company

as usual in Switzerland, the boy had
a check up performed by the
Railway Doctor ...



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Clinical Findings: hypertension

181 cm 68 kg

systolic 2/6 murmur

Regular Heart Beat

Blood pressure at rest:

right arm 142/88, left arm 134/98 mm Hg

Referral of the subject to Echocardiography



The Special Echo Case



Clinical Findings: hypertension

181 cm 68 kg

late systolic 2/6 murmur

Regular Heart Beat

Blood pressure at rest:

right arm 142/88, left arm 134/98 mm Hg

Blood pressure at peak exercise (left arm)

212/120 mm Hg

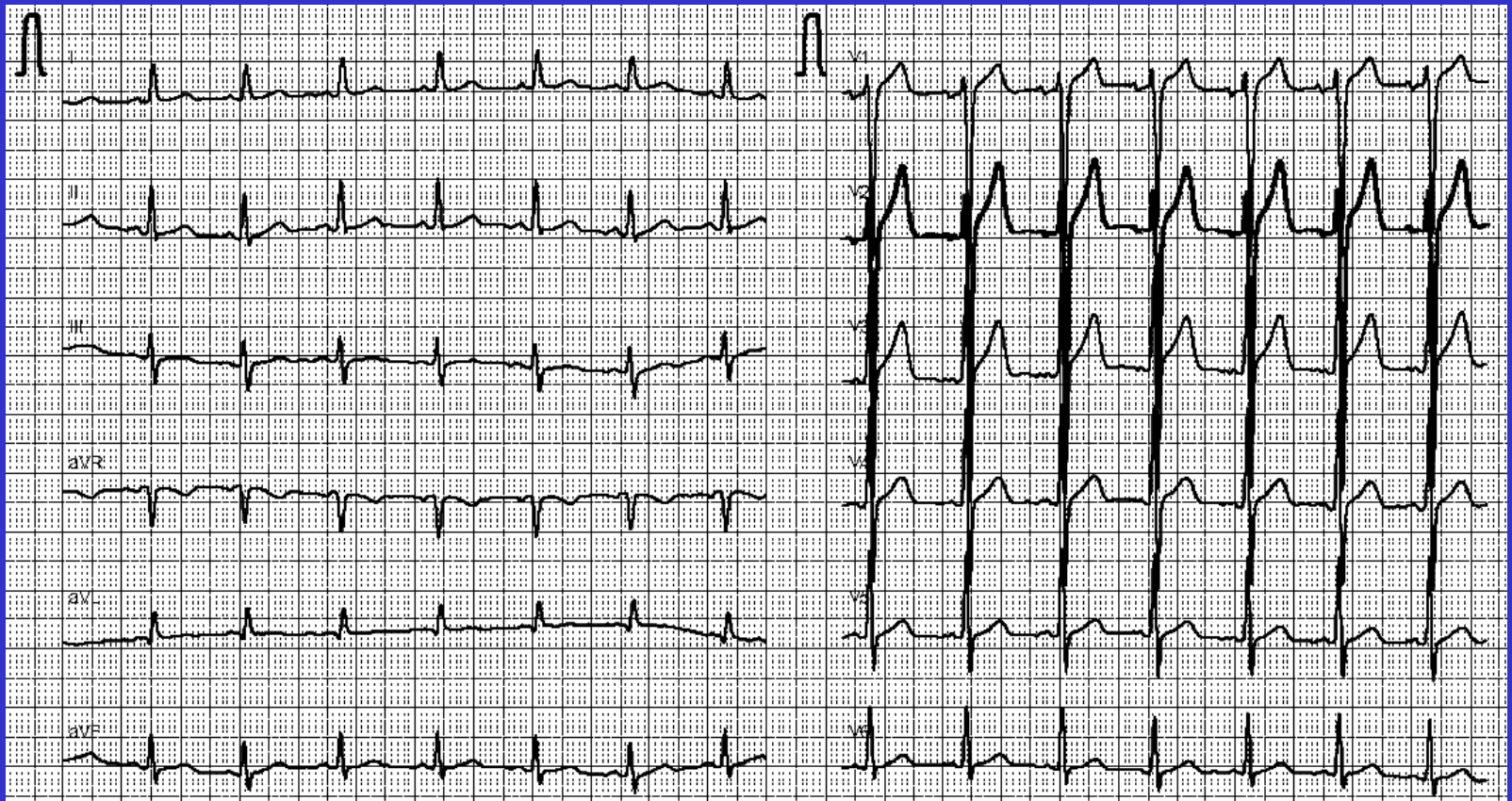


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ECG

Sinus rhythm, left ventricular hypertrophy
no strain pattern / slight axis deviation to the left



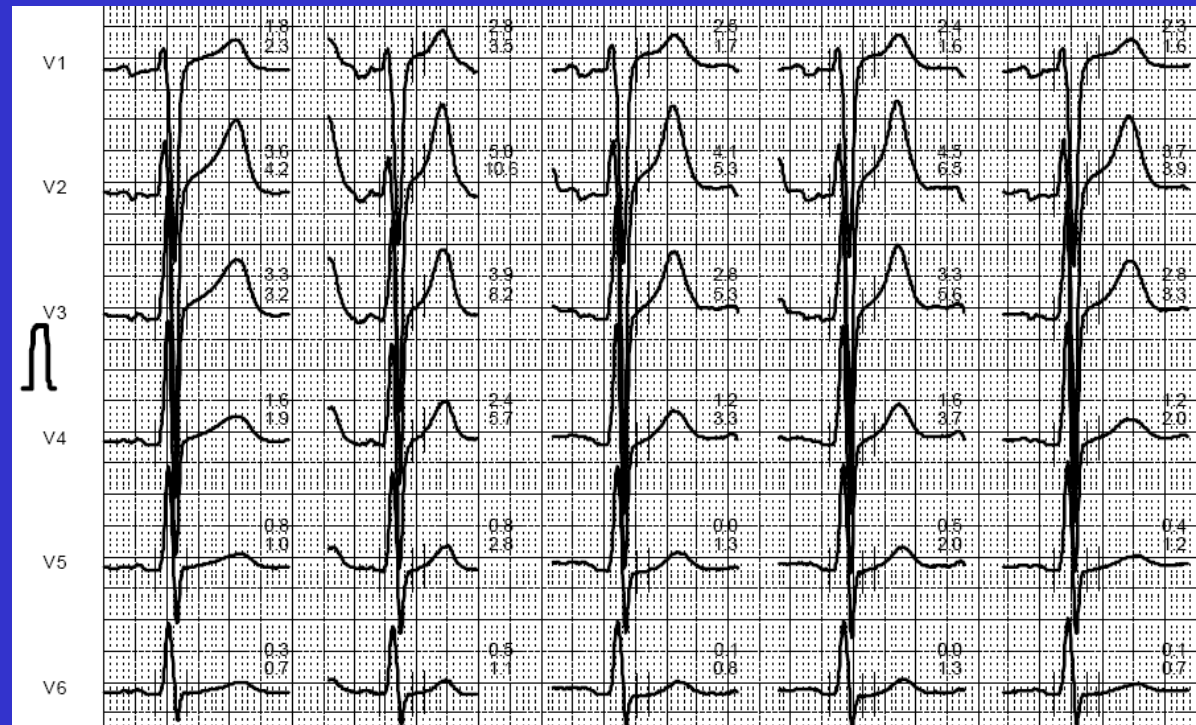
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Exercise Test

Normal, but reduced exercise capacity (71%)
Stopped for leg fatigue

Duration 6 Minutes
ST Segments Normal
Peak BP 212/120 mm Hg

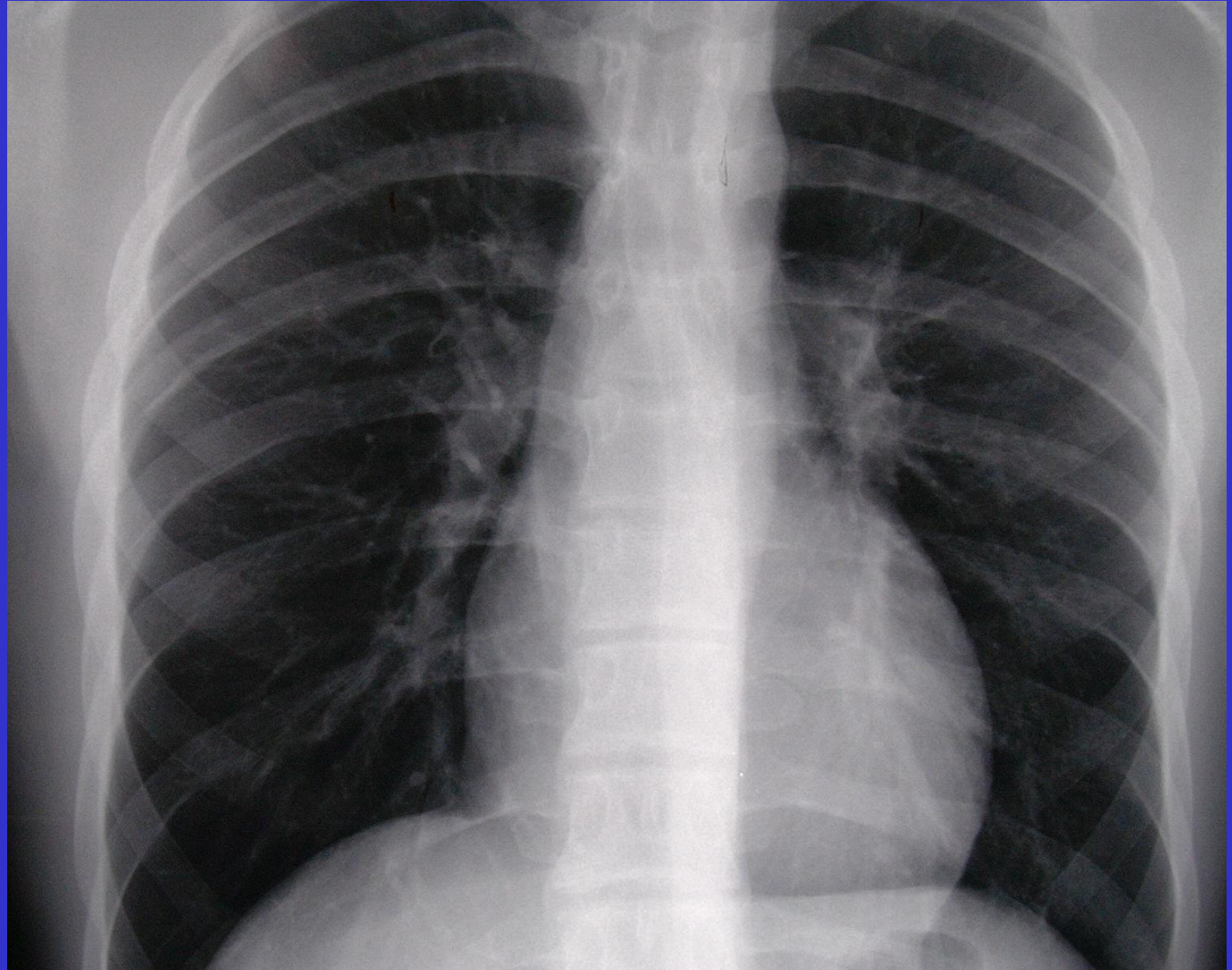


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Chest Radiograph

Normal

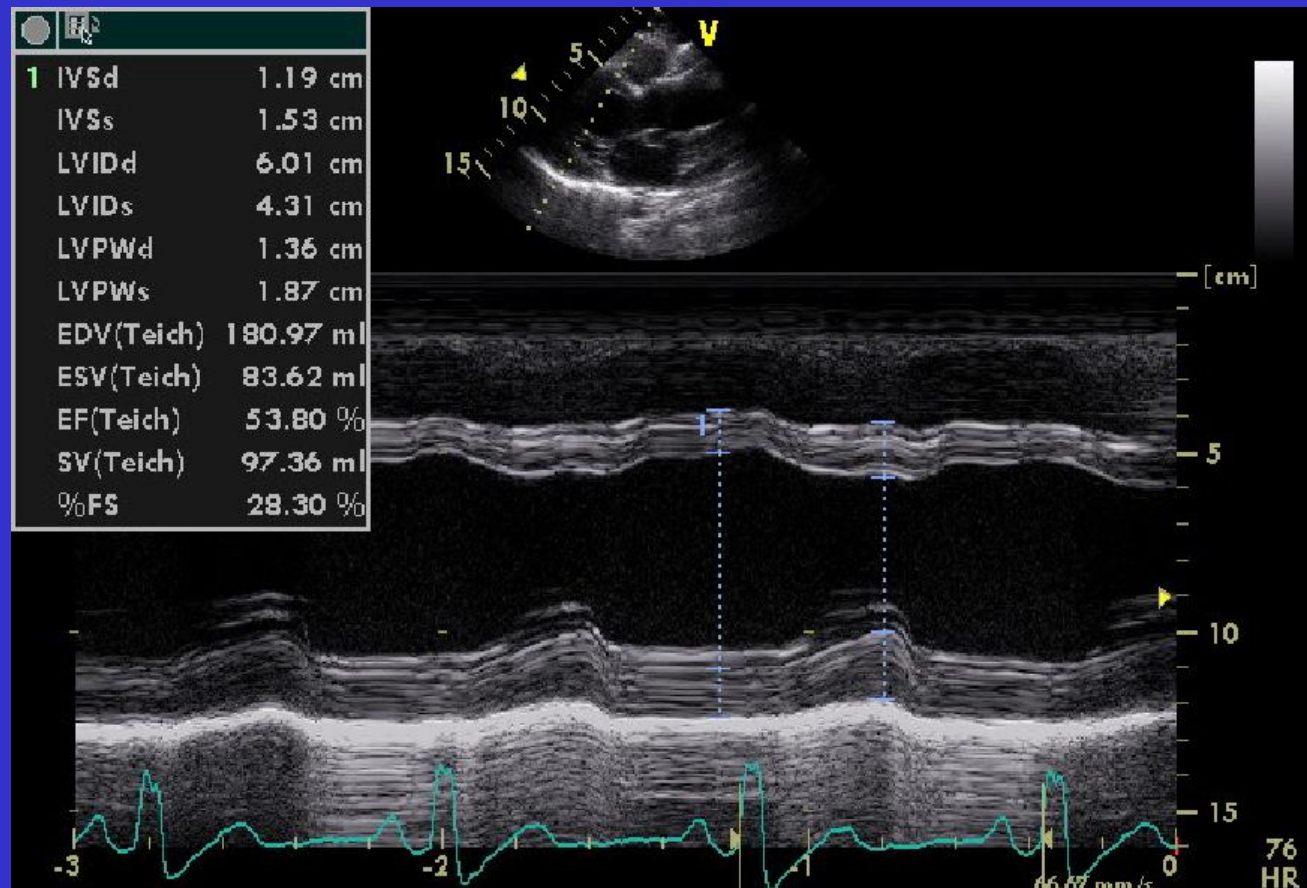


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Echocardiogram

Borderline LV Dilatation, Slight Wall Hypertrophy
Fractional Shortening 28 % / LV Mass: 181 g/m²

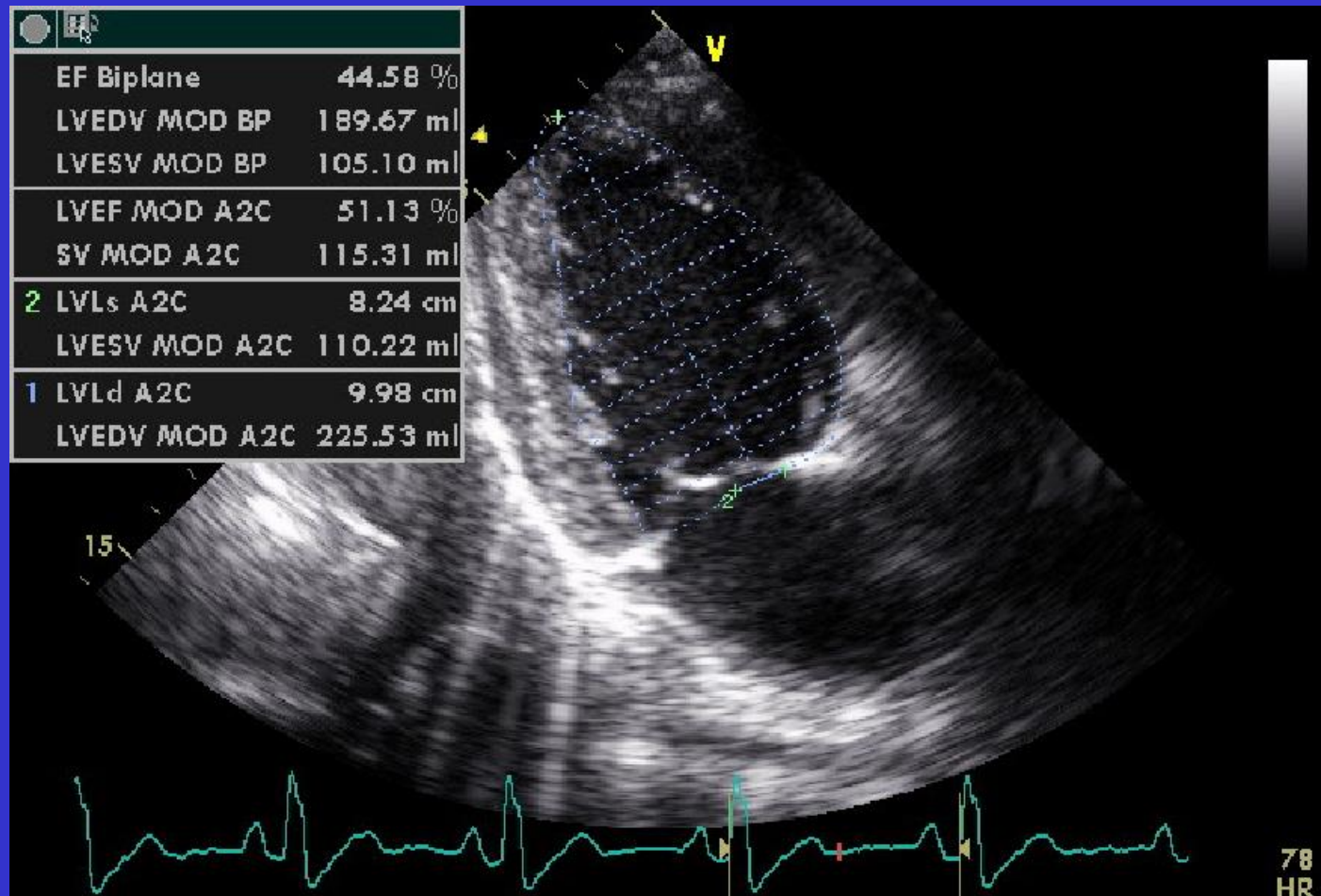


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Echocardiography

Slight reduction of left ventricular function (LVEF 45%)

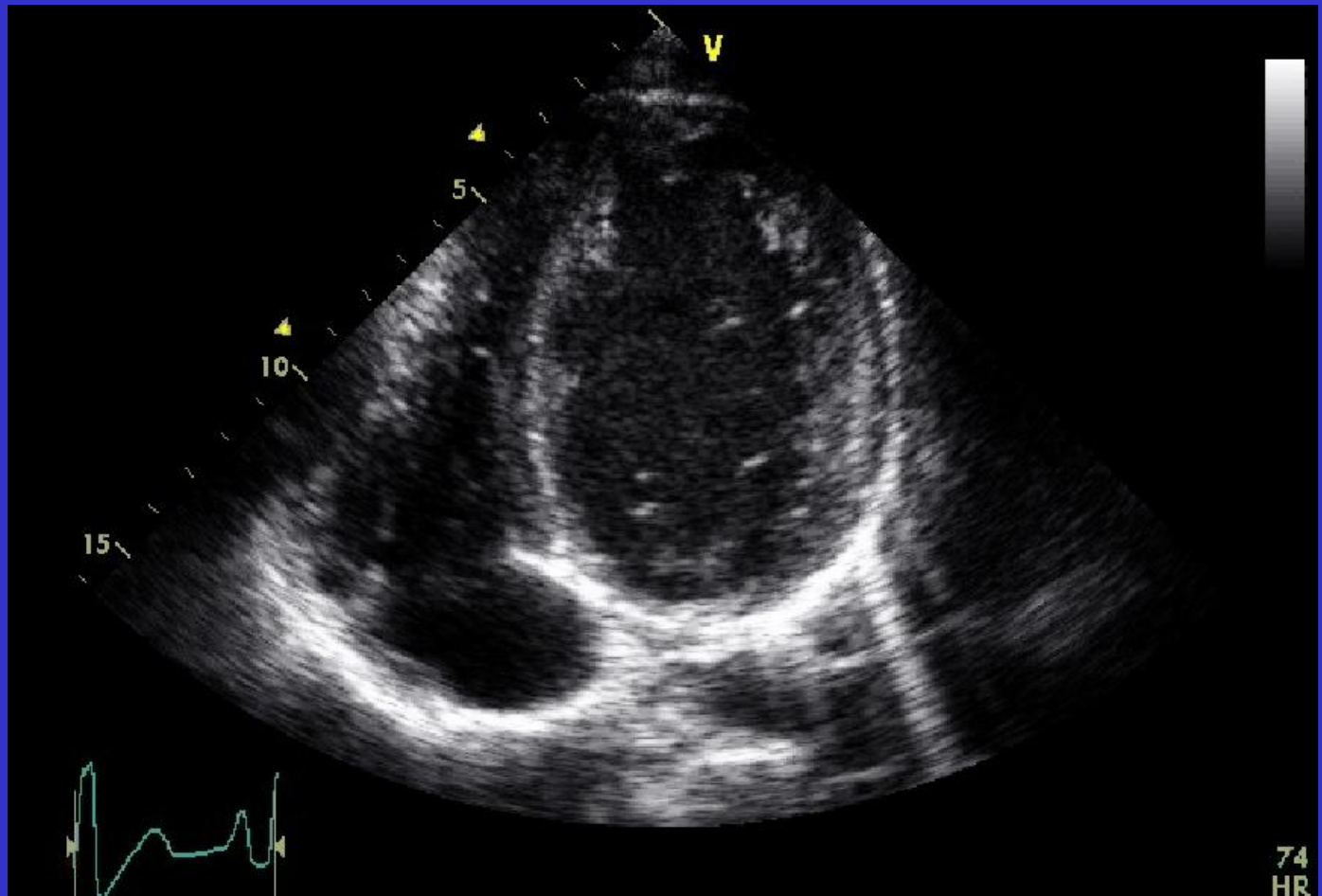




Echocardiography

4 CH VIEW

apical dysfunction ?



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Echocardiography

Apical Close Up 4 CH VIEW
Apical Thinning ?

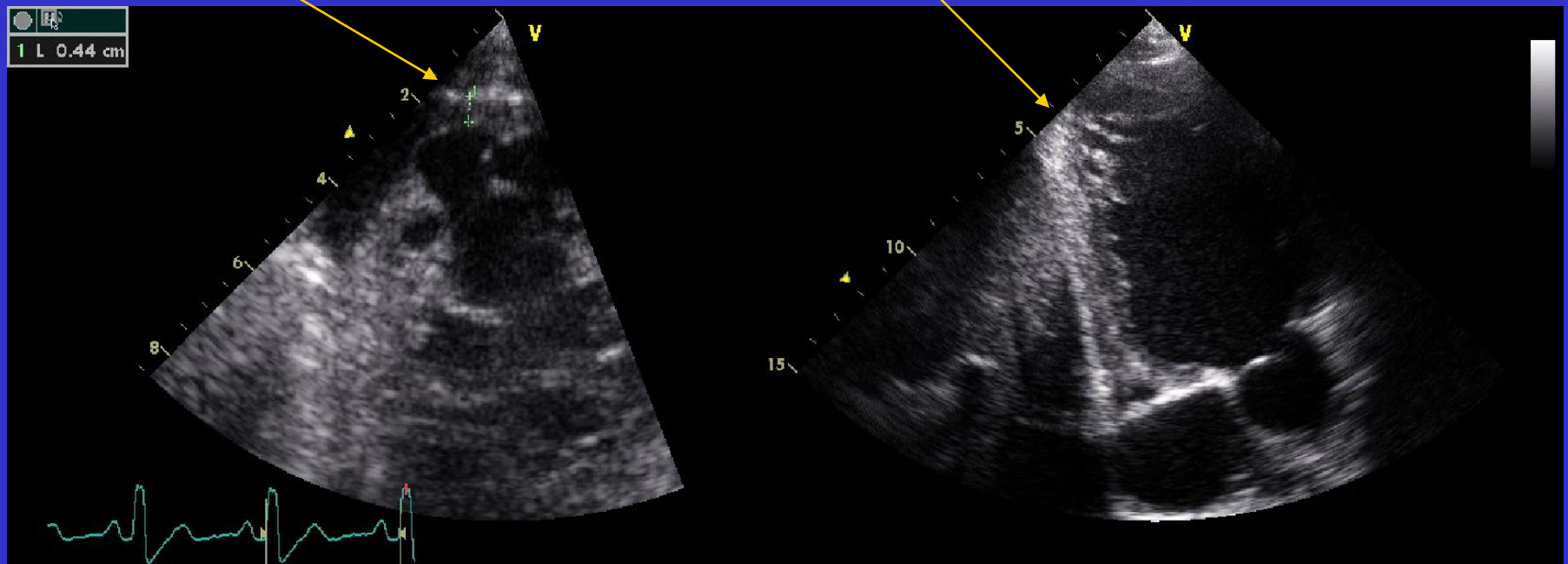


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Echocardiography

Apical Thinning (4mm) and Trabeculations

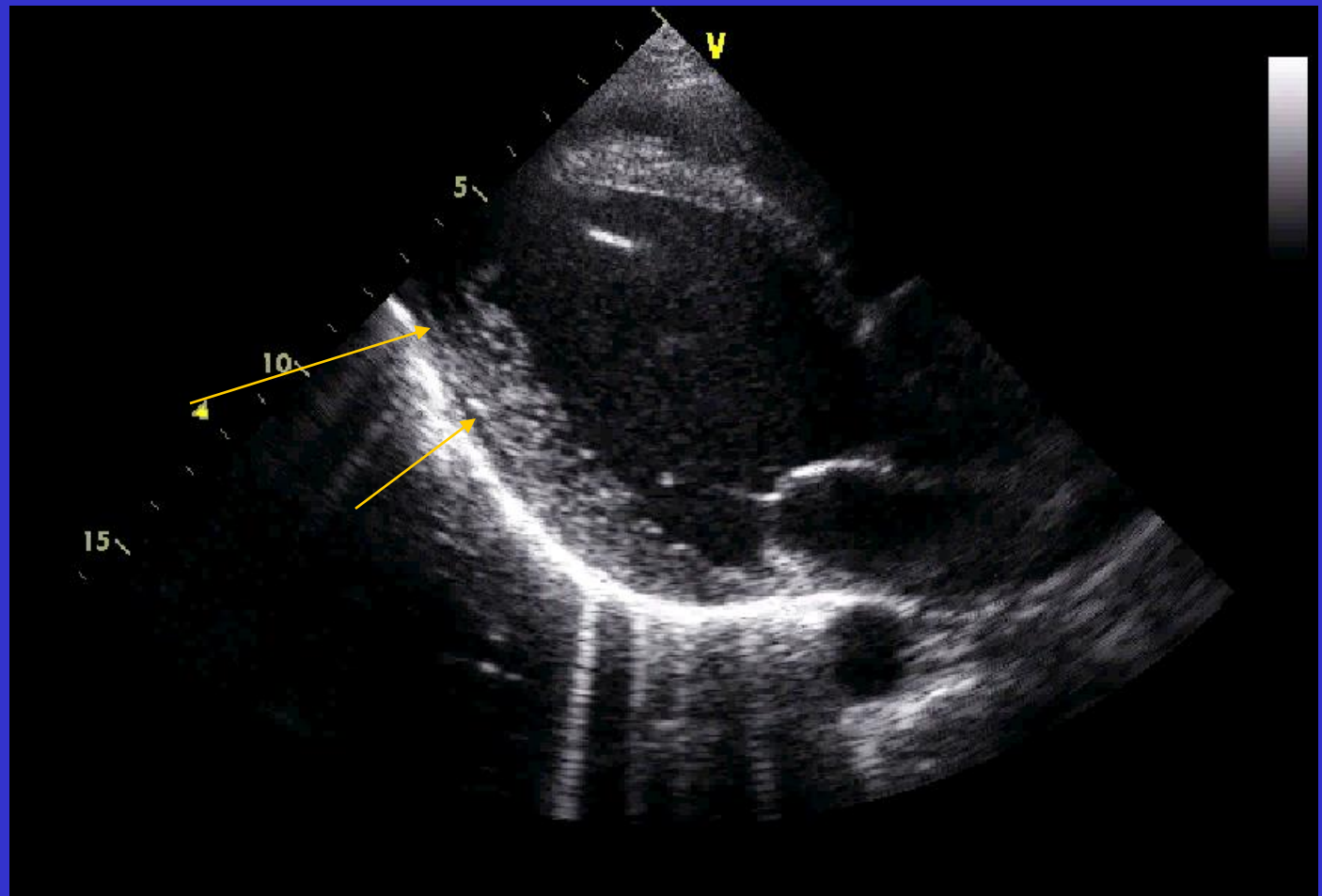


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Echocardiography

Trabeculations extending to the inferior wall

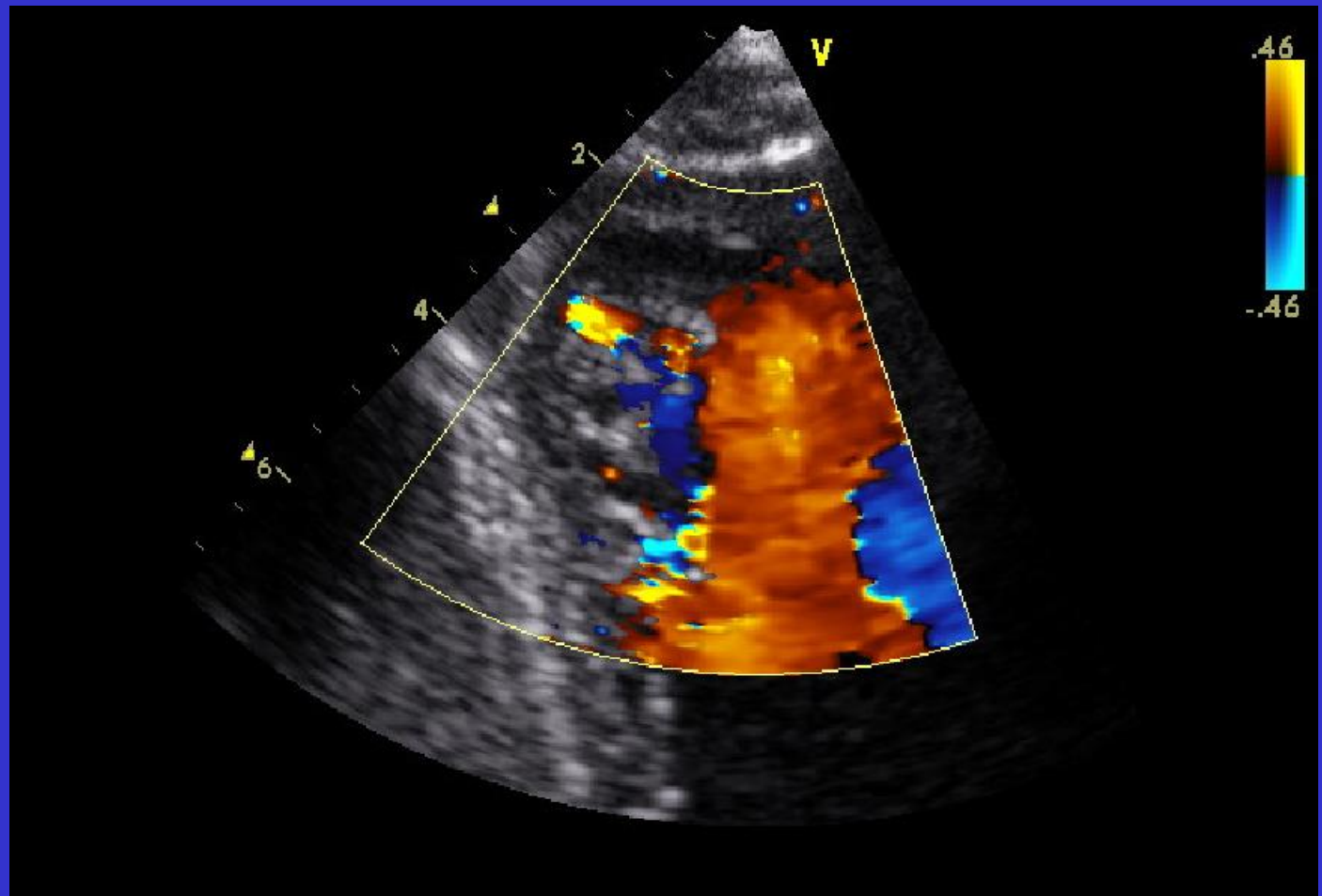


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Echocardiography

Trabeculations on Color Doppler

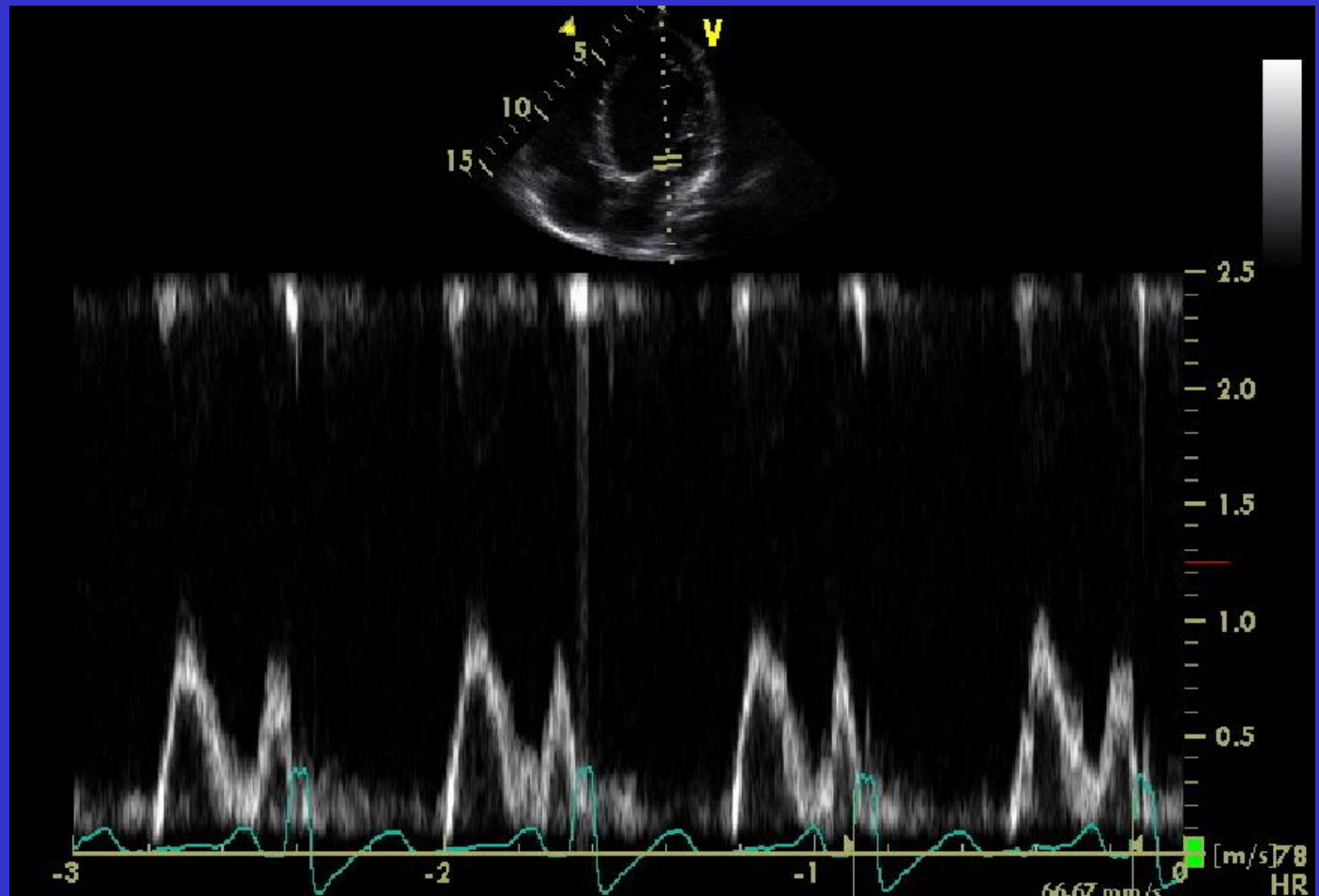


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Echocardiography

Normal diastolic function / no heart valve abnormalities noted



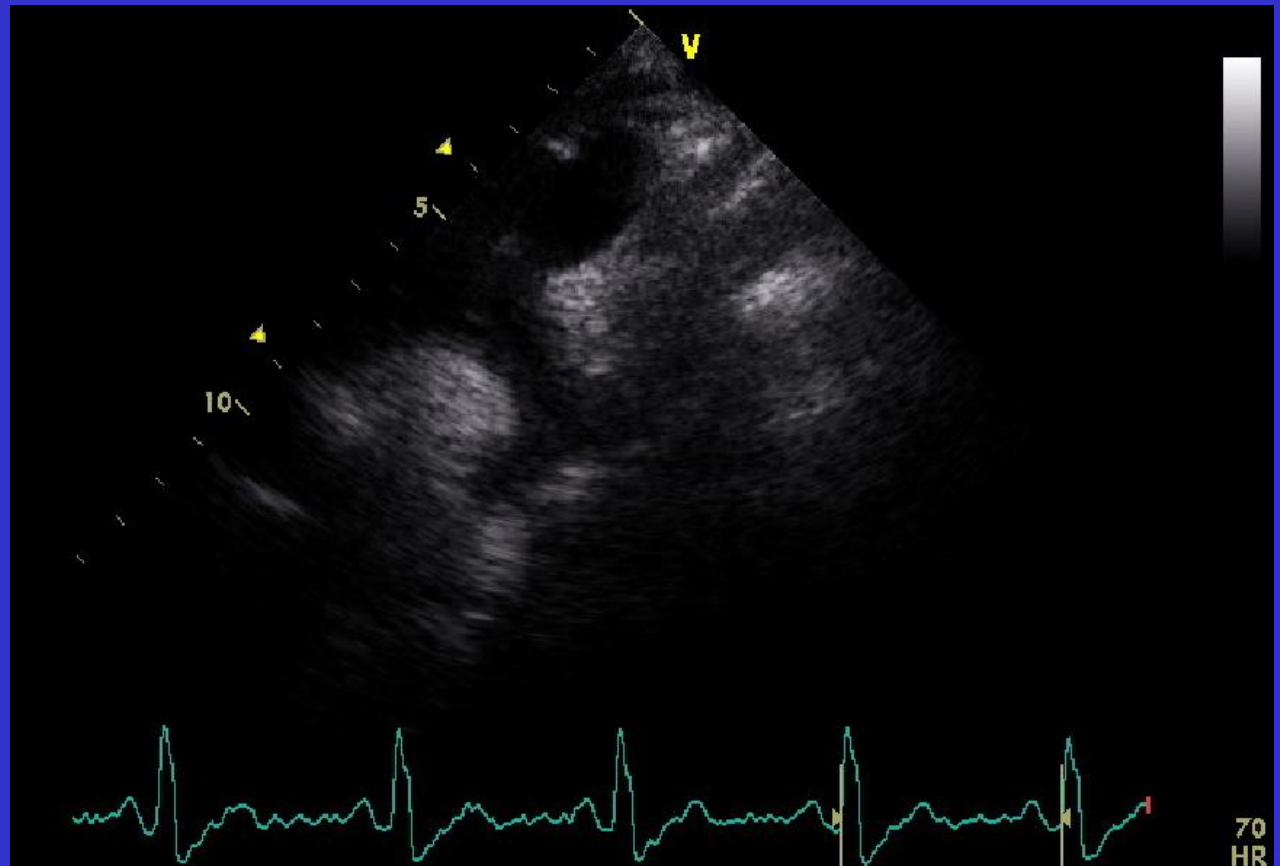
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Echocardiography

Small Aortic Arch (18 mm), gross dilation of brachiocephalic trunc (19 mm)

Aortic Arch Anomaly ?
Double Aortic Arch ?
Vascular Ring ?



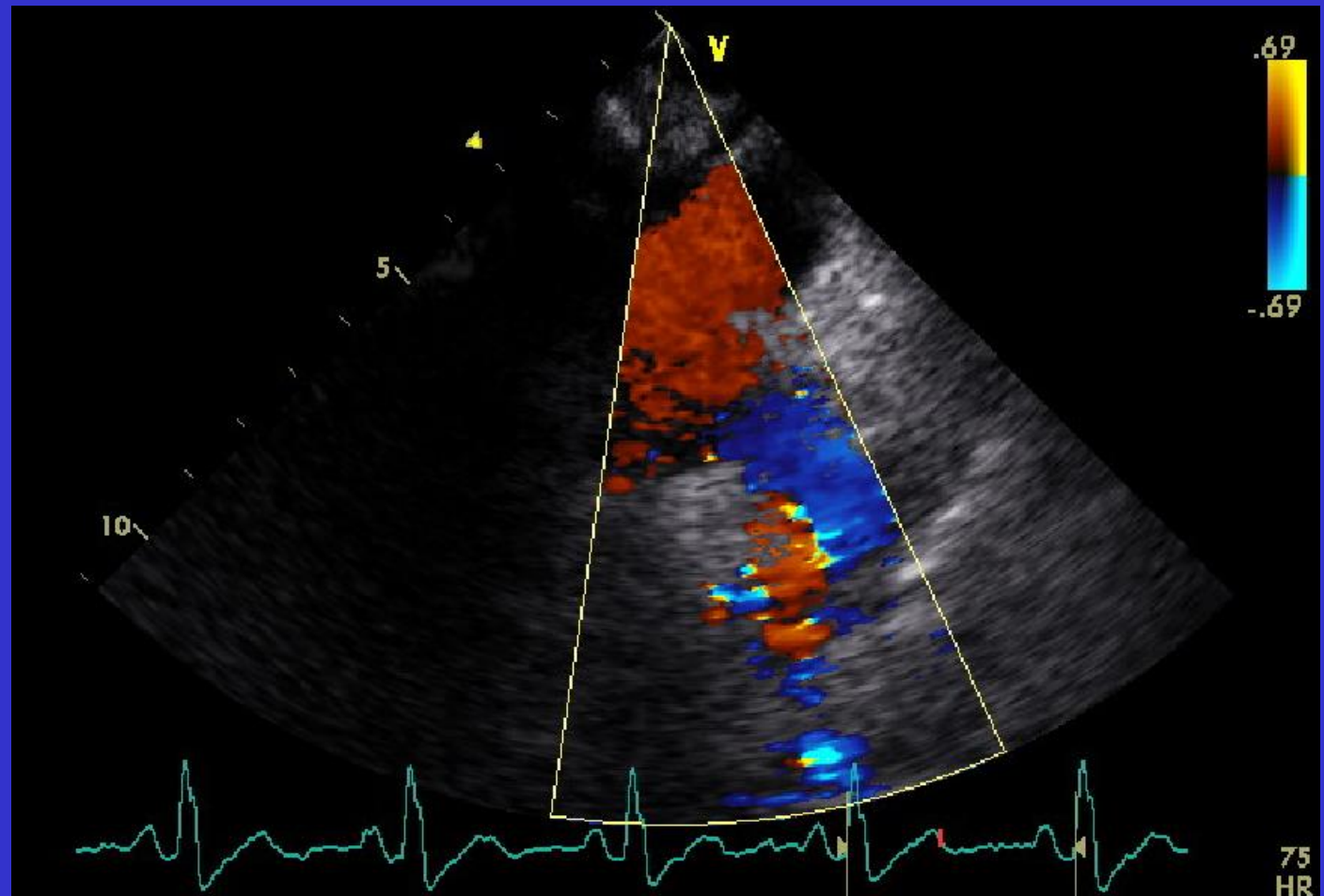
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Echocardiography

Post ductal aliasing at 69 cm/s

Additional
Aortic
Isthmus
Stenosis ?



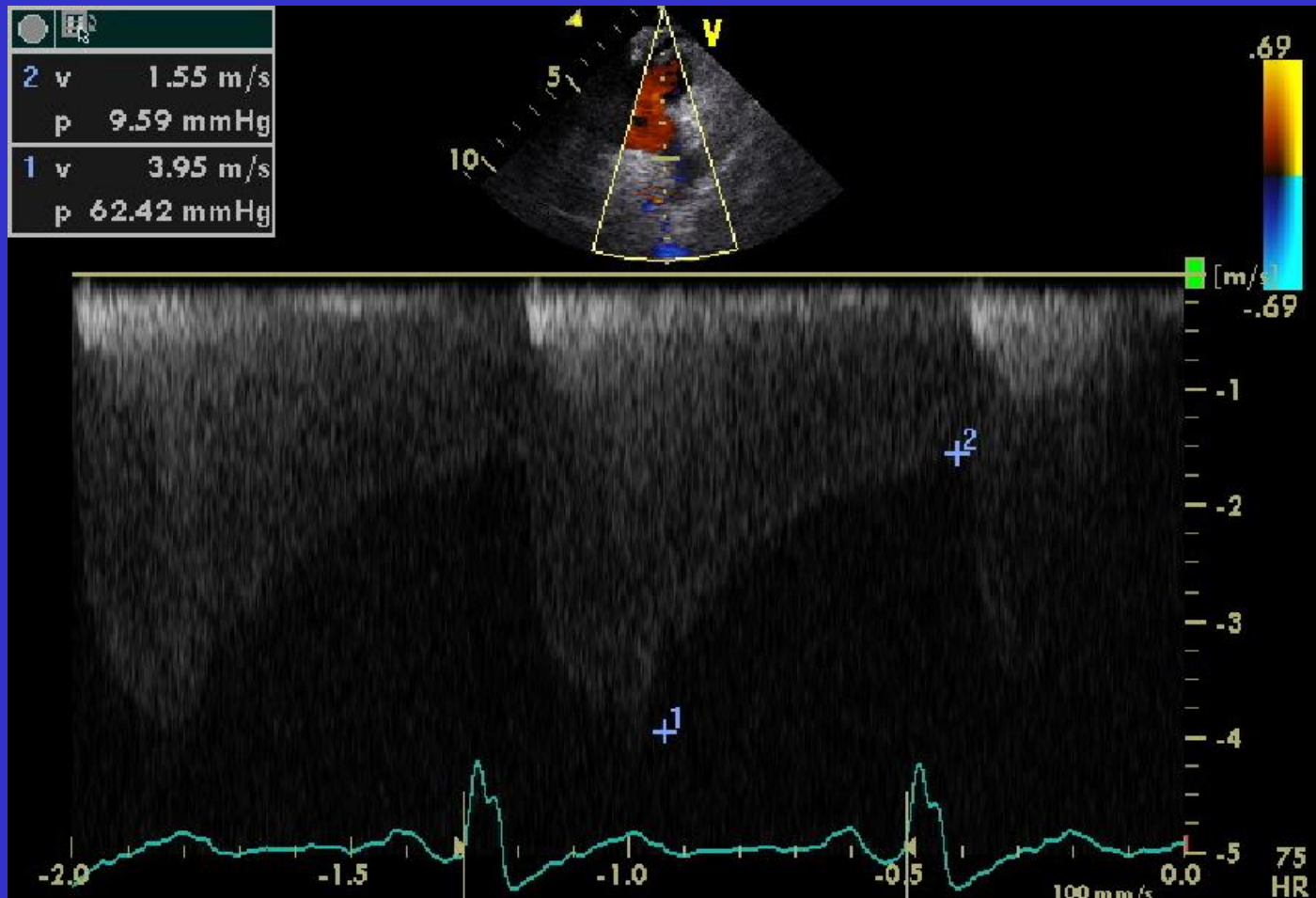
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Echocardiography

Pressure Gradient 62 mm Hg Descending Aorta

High diastolic flow
Suspicion of
post ductal
aortic stenosis



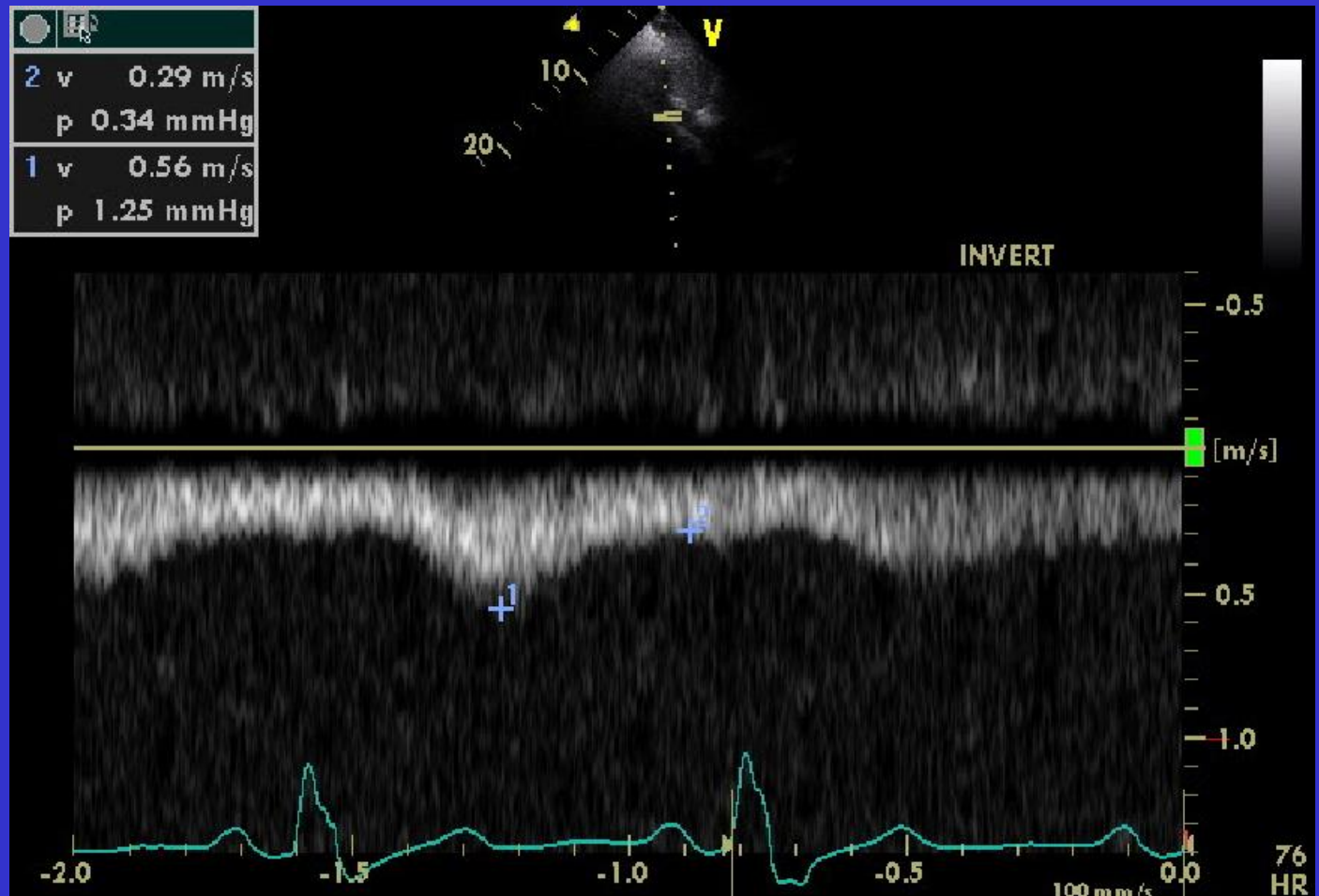
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Echocardiography

Flow pattern in the abdominal aorta

virtual elimination
of systolic flow



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Ankle Blood Pressure

No pulse on palpation

No systolic bruit during Doppler examination





Diagnosis

Severe Postductal Coarctation

Additional abnormalities

left ventricular dysfunction (hypertensive heart disease with BP of 140 mm Hg ?)
apical thinning (unusual finding in hypertension !)
left ventricular hypertrophy without diastolic dysfunction ?
aortic arch abnormality ?

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Preoperative Cardiovascular Magnetic Resonance



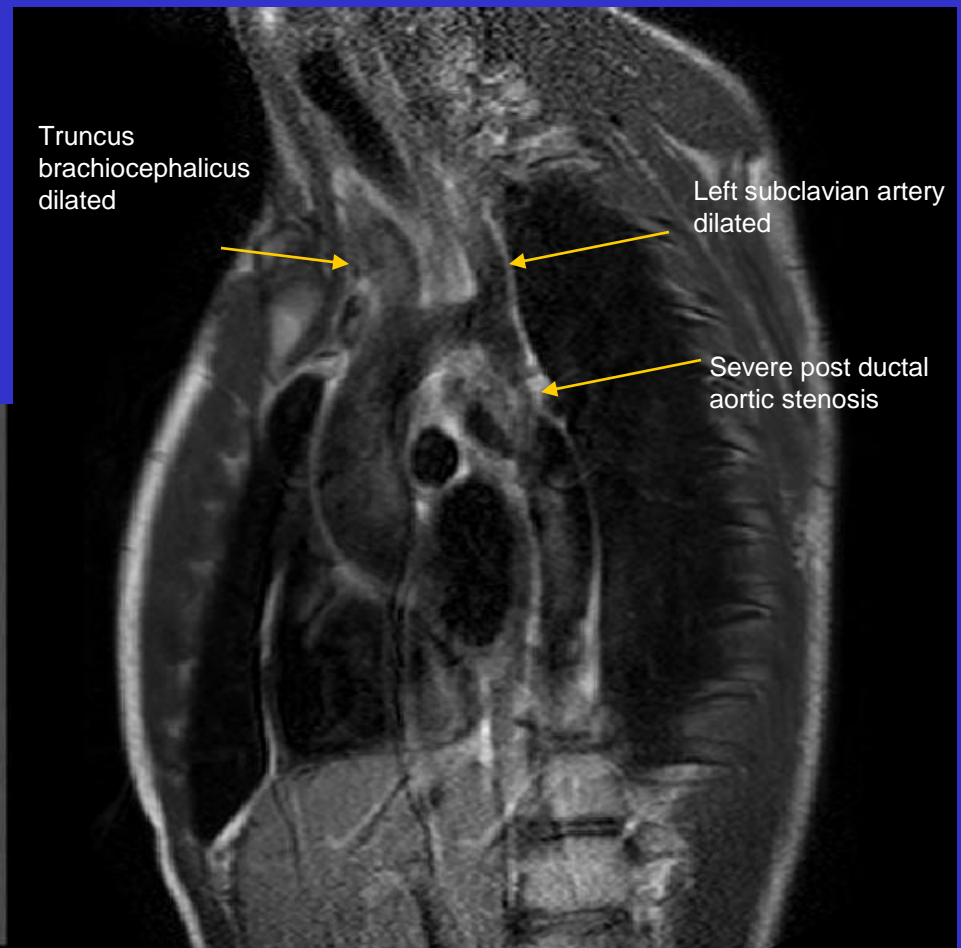
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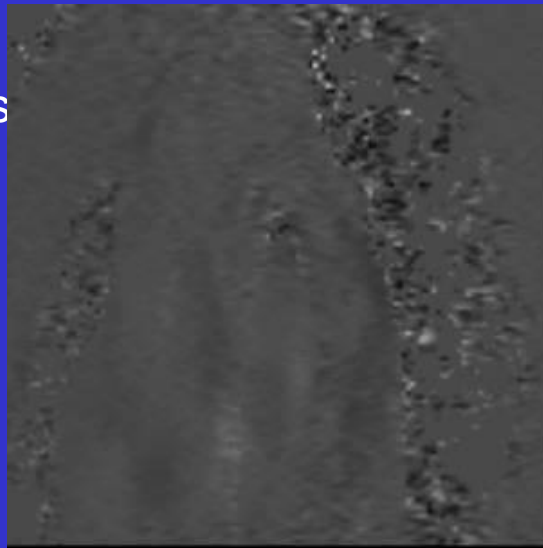
Cardiovascular Magnetic Resonance

Severe Postductal Coarctation

T1 Spin Echo



Quant Flow
VENC at 4 m/s



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Cardiovascular Magnetic Resonance

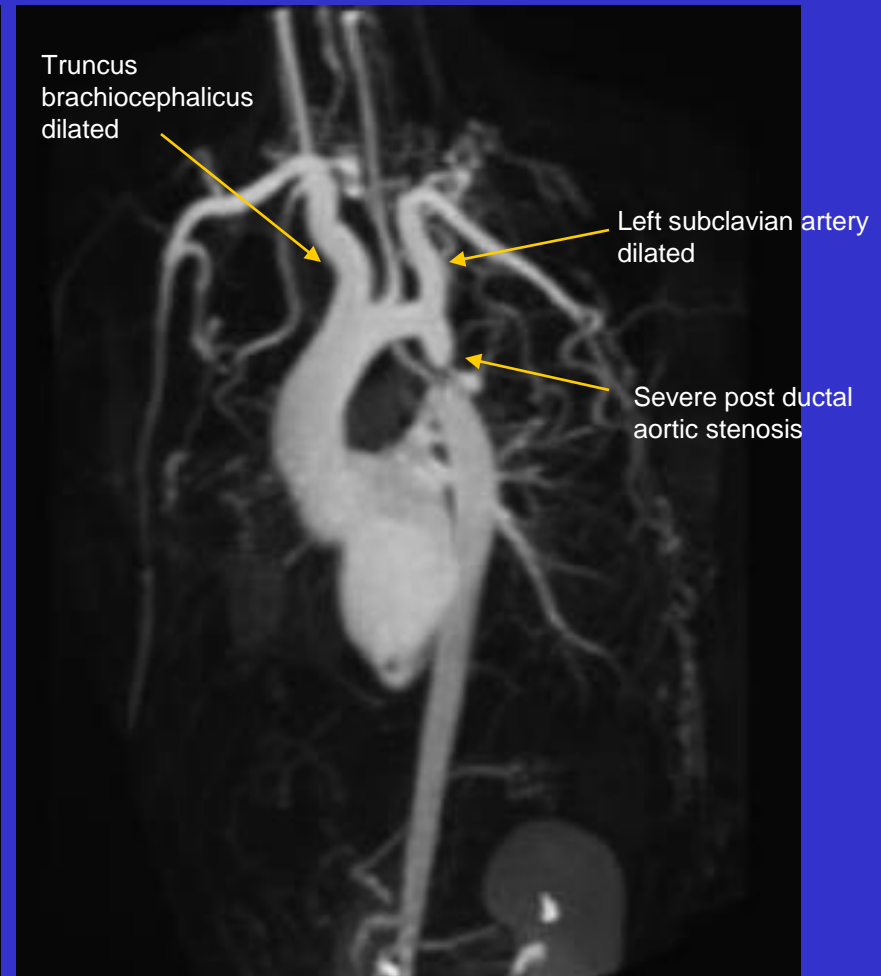
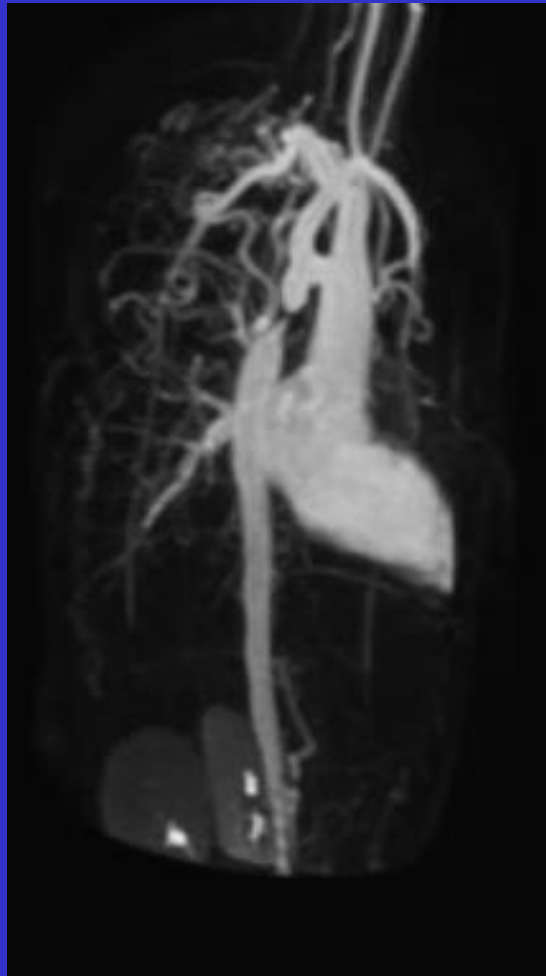
Severe Postductal Coarctation (length 28 mm)

3 D Gd

Collaterals:

Aa. mammae
paravertebral
arteries

no aortic arch
anomaly



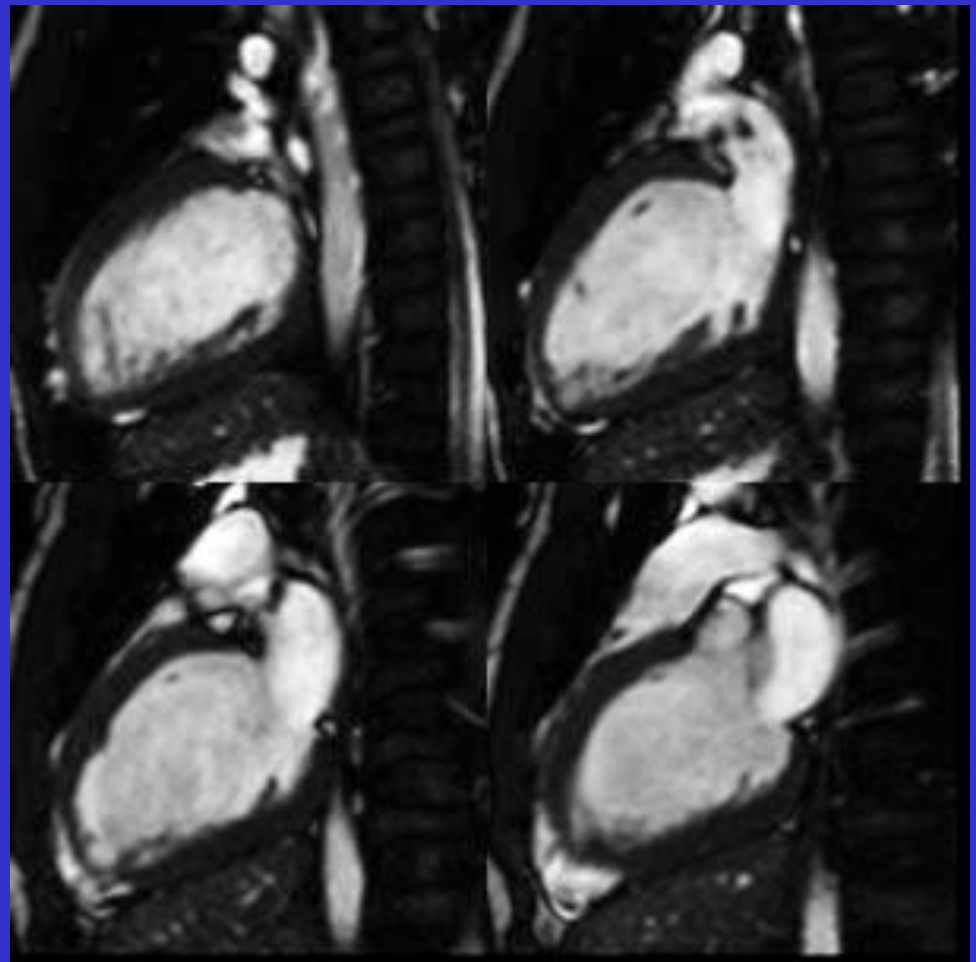
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Cardiovascular Magnetic Resonance

Left ventricular dysfunction (LVEF 37.6 %, LVEDV 209.4 ml)

Cine MRI 2 Chamber View



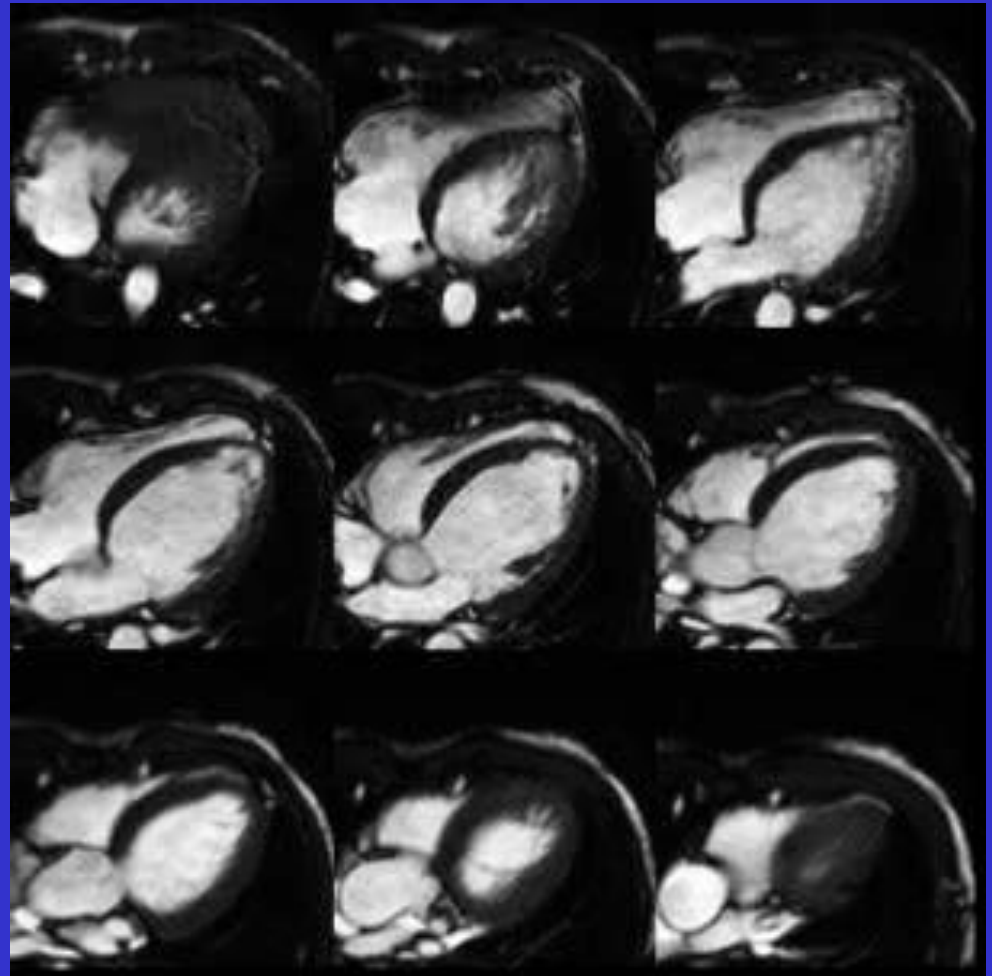
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Cardiovascular Magnetic Resonance

Left ventricular dysfunction (LVEF 37.6 %, LVEDV 209.4 ml)

Cine MRI 4 Chamber View



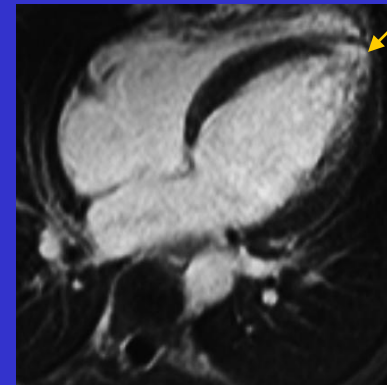
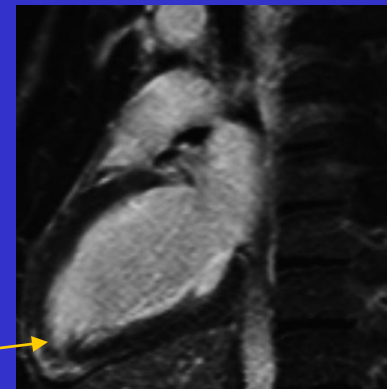
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Cardiovascular Magnetic Resonance

Left ventricular dysfunction (LVEF 37.6 %, LVEDV 209.4 ml)

Delayed Imaging: no contrast enhancement





Coarctation of the Aorta

Typically a discrete narrowing of the aortic lumen

Incidence: 1/6000 live births

Often occurs in pts with Turner's syndrome, is associated with ventricular septal defect, Shone syndrome, and cerebral aneurysms in the circle of Willis

Postductal form (98%)

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20 year survival after operation:

Operation performed before age 14:	91%
Operation performed after age 14:	79%

V. Fuster et al: Circulation 80:840-845, 1989.

Major complications: aortic rupture



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Coarctation of the Aorta in association with left ventricular noncompaction:

J Am Soc Echocardiogr. 2002 Dec;15(12):1523-8.

12 patients with ventricular noncompaction

Mean age 3.5 years.

N=5: isolated noncompaction

N=3: subnormal left ventricular systolic function

N=1: noncompaction + coarctation



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Conclusion:

Unusual presentation of a severe postductal Coarctation with

virtual absence of symptoms

left ventricular hypertrophy and dysfunction

apical thinning and increased trabeculations

non compacted myocardium ?

Follow-up necessary to rule out suspicion of lv noncompaction



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What's next ?

Patient is scheduled for Operation 21 January 2005

Resection of the Coarctation,
eventually implantation of a short aortic prosthesis



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Thank You





Question:

Was there a progressive increase in stenosis severity during life, allowing for adaptation of the organism and explaining some of the unusual findings ?

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Echocardiography

Flow pattern in the abdominal aorta

virtual elimination
of systolic flow





Unusual Findings for Severe Coarctation

relatively low blood pressure in the upper extremities
no rib erosions on chest radiograph, no 3 sign
no symptoms (exercise capacity 76%, untrained smoker)

Key Findings

Late systolic murmur
no ankle pulse