LVOTO AND AORTA IN CONGENITAL CARDIOVASCULAR DISEASES

ANNA KLISIEWICZ

ECHOCARDIOGRAPHIC LABORATORY AND DEPARTMENT
OF ADULT CONGENITAL HEART DISEASE
INSTITUTE OF CARDIOLOGY, WARSAW

31.03.2012
Szczecin

European Heart Journal
doi:10.1093/eurheartj/ehq249

ESC GUIDELINES

ESC Guidelines for the management of grown-up congenital heart disease (new version 2010)

The Task Force on the Management of Grown-up Congenital Heart Disease of the European Society of Cardiology (ESC)

Endorsed by the Association for European Paediatric Cardiology (AEPC)

Authors/Task Force Members: Helmut Baumgartner (Chairperson) (Germany)*, Philipp Bonhoeffer (UK), Natasja M. S. De Groot (The Netherlands), Fokko de Haan (Germany), John Erik Deanfield (UK), Nazzareno Galie (Italy), Michael A. Gatzoulis (UK), Christa Gohlke-Baerwolf (Germany), Harald Kaemmerer (Germany), Philip Kilner (UK), Folkert Meijboom (The Netherlands), Barbara J. M. Mulder (The Netherlands), Erwin Oechslin (Canada), Jose M. Oliver (Spain), Alain Serraf (France), Andras Szatmari (Hungary), Erik Thaulow (Norway), Pascal R. Vouhe (France), Edmond Walma (The Netherlands).

Left ventricular outflow tract obstruction (LVOTO)

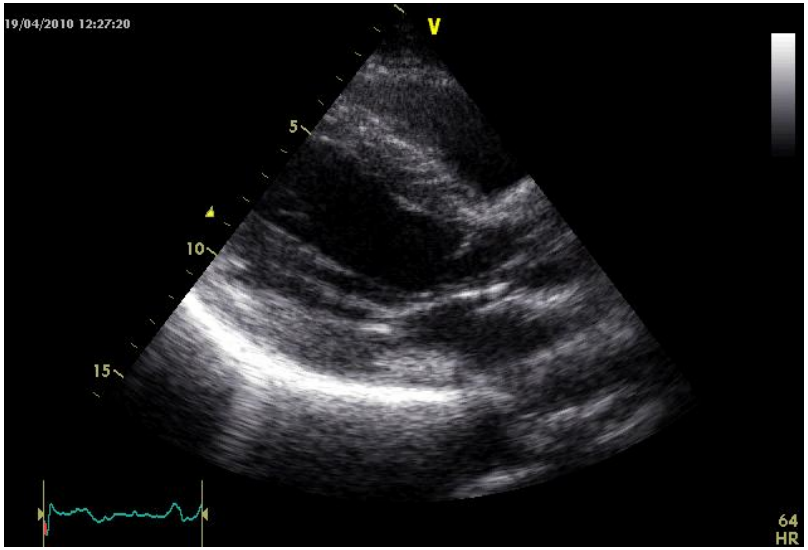
- Valvular (75%)
- Subvalvular
- Supravalvular
- Multiple levels

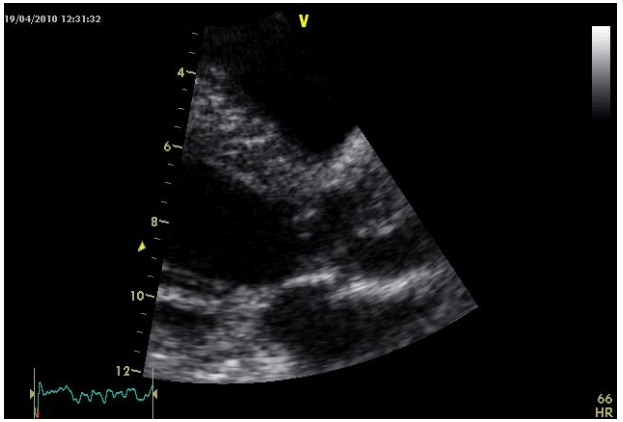
Subvalvular stenosis (subAS)

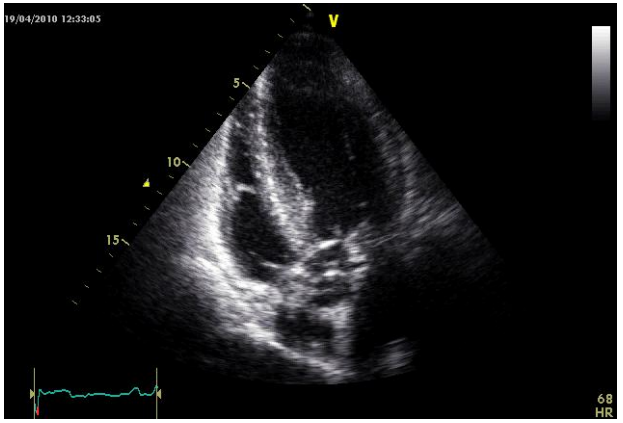
- 6,5% congenital heart diseases
- frequently associated with ASD, VSD and Shone's complex
  (supravalve mitral membrane, parachute mitral valve, subaortic stenosis, aortic coarctation, bicuspd aortic valve)

- fibrous ridge (membrane) – most frequent form of subAS (90% cases)
- fibromuscular narrowing
- additional mitral tissue (casuistic)

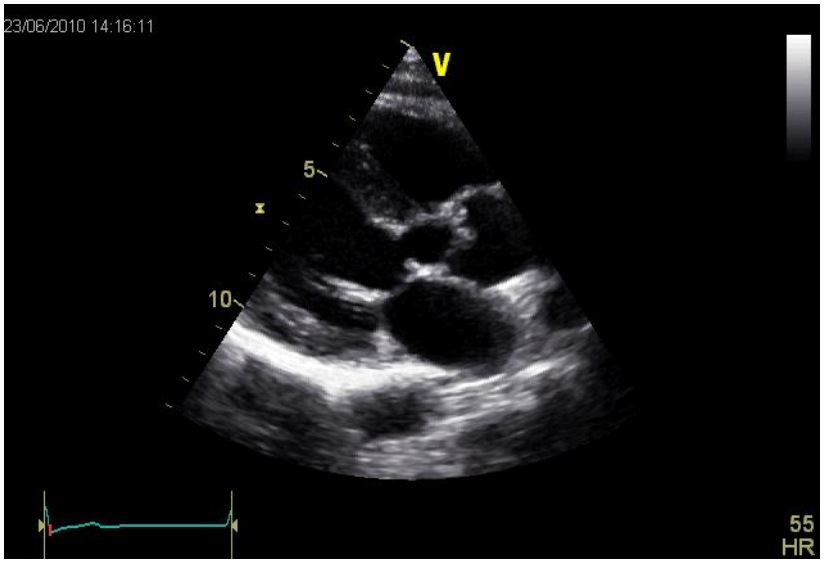
subAo membrane

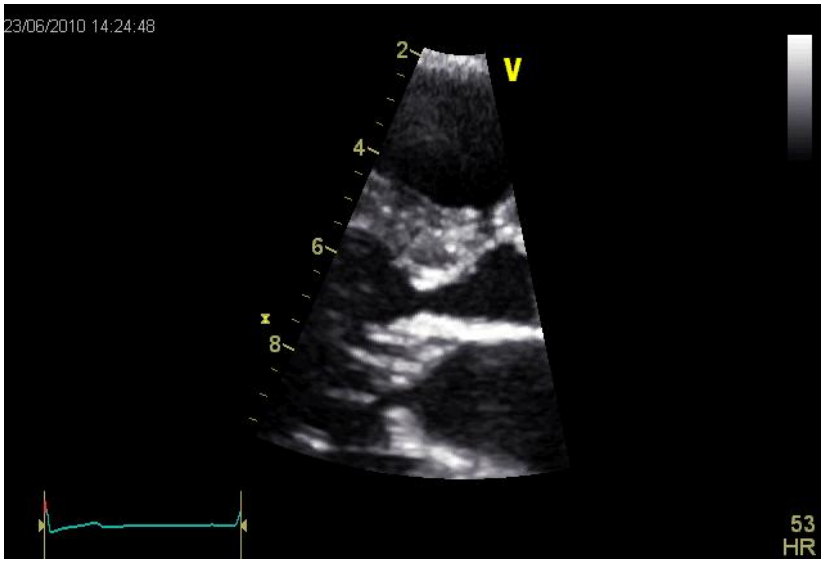




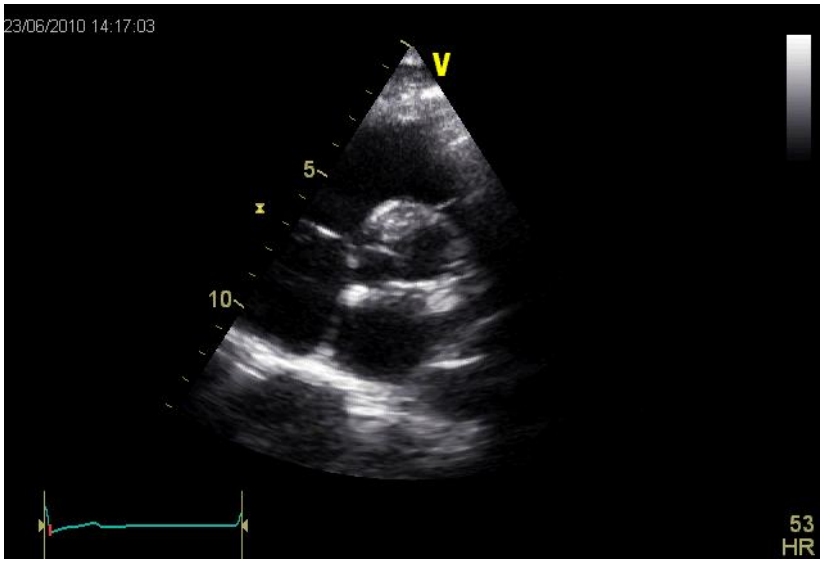


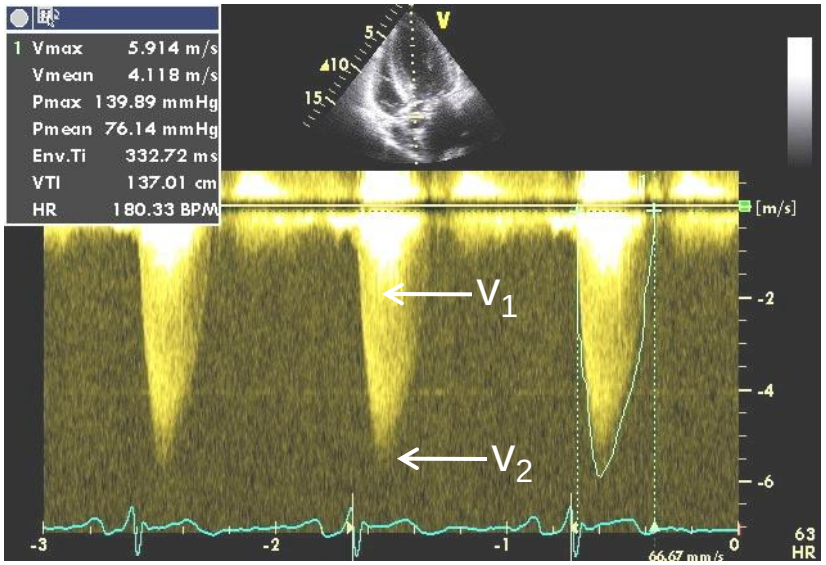
Fibromuscular narrowing





Fibromuscular narrowing





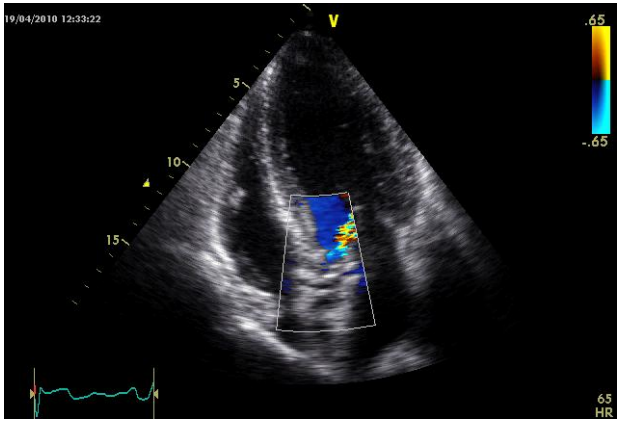
$$\Delta p = 4(v_2^2 - v_1^2)$$

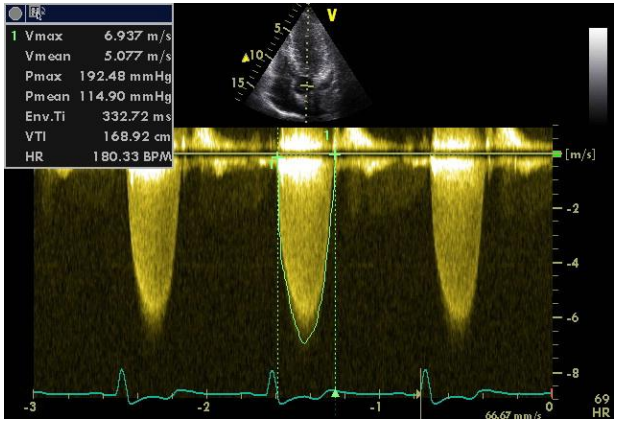
subAS

ESC Guidelines for the management of grown-up congenital heart disease (new version 2010)

Table 10 Indications for intervention in subaortic stenosis

Indications	Class[a]	Level[b]
Symptomatic patients (spontaneous or on exercise test) with a mean Doppler gradient ≥50 mmHg[c] or severe AR should undergo surgery	I	C
Asymptomatic patients should be considered for surgery when:		
• LVEF is <50% (gradient may be <50mmHg due to low flow)	IIa	C
• AR is severe and LVESD >50mm (or 25 mm/m² BSA) and/or EF <50%[d]	IIa	C
• mean Doppler gradient is ≥50 mmHg[c] and LVH marked	IIa	C
• mean Doppler gradient is ≥50 mmHg[c] and blood pressure response is abnormal on exercise testing	IIa	C
Asymptomatic patients may be considered for surgery when:		
• mean Doppler gradient is ≥50 mmHg[c], LV normal, exercise testing normal, and surgical risk low	IIb	C
• progression of AR is documented and AR becomes more than mild (to prevent further progression)	IIb	C





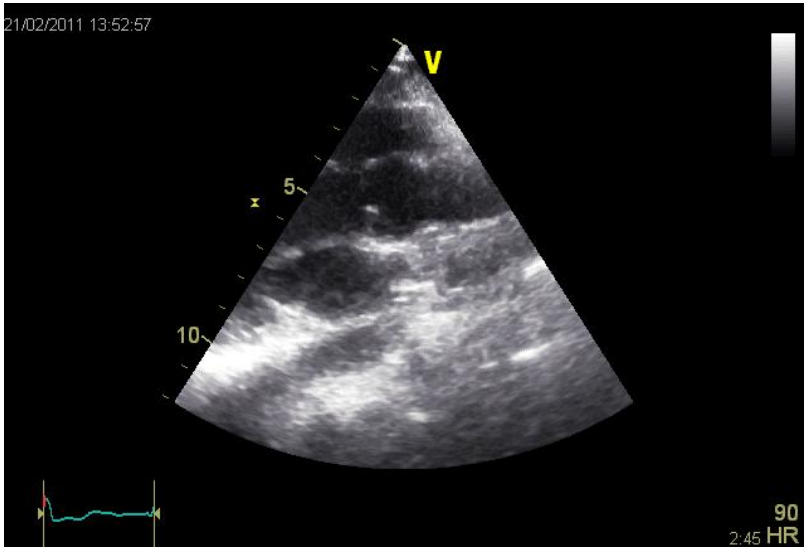
Supravalvular aortic stenosis (supraAS)

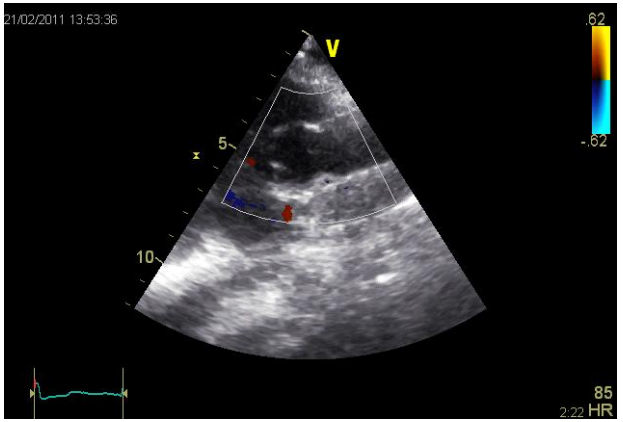
- About 7% of LVOTO
- Frequently associated with Williams-Beu syndrome (up to 80%)

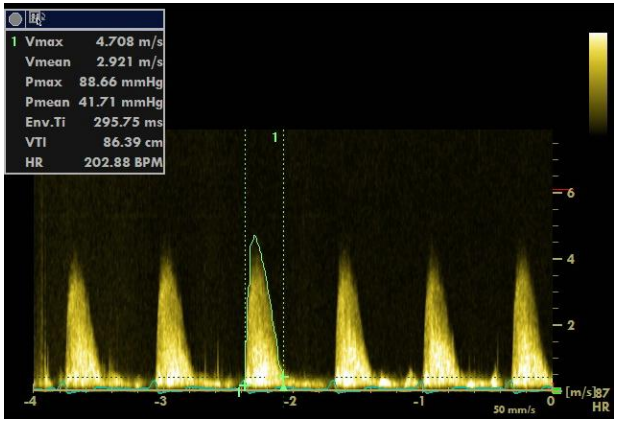
„elfin" face

- Hourglass deformity
- Localized fibrous diaphragm
- Diffuse hypoplasia of ascending aorta

supraAS - TTE


21/02/2011 13:52:57
V
5
10
90
2:45 HR


21/02/2011 13:53:36
.62
-.62
V
5
10
85
2.22 HR

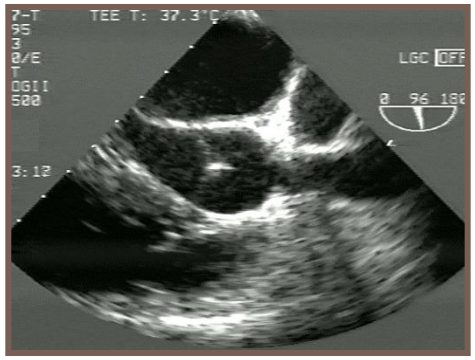

1 Vmax 4.708 m/s
Vmean 2.921 m/s
Pmax 88.66 mmHg
Pmean 41.71 mmHg
Env.Ti 295.75 ms
VTI 86.39 cm
HR 202.88 BPM
1
6
4
2
-4
-3
-2
-1
0
50 mm/s
[m/s]87
HR

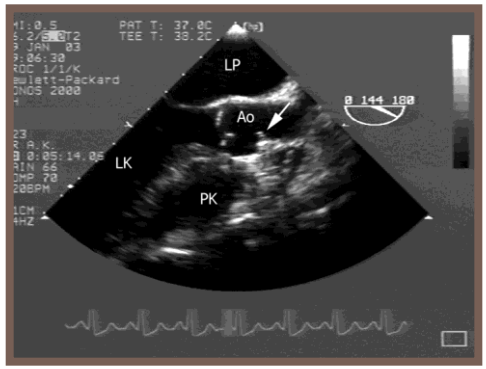
supraAS

ESC Guidelines for the management of grown-up congenital heart disease (new version 2010)

Table 9 Indications for intervention in supravalvular aortic stenosis

Indications	Class[a]	Level[b]
Patients with symptoms (spontaneous or on exercise test) and mean Doppler gradient ≥50 mmHg should undergo surgery	I	C
Patients with mean Doppler gradient <50 mmHg should undergo surgery when they have:		
• symptoms attributable to obstruction (exertional dyspnoea, angina, syncope) and/or	I	C
• LV systolic dysfunction (without other explanation)	I	C
• severe LVH, attributable to obstruction (not related to hypertension)	I	C
• when surgery for significant CAD is required	I	C
Patients with mean Doppler gradient ≥50 mmHg[c] but without symptoms, LV systolic dysfunction, LVH, or abnormal exercise test may be considered for repair when the surgical risk is low	IIb	C



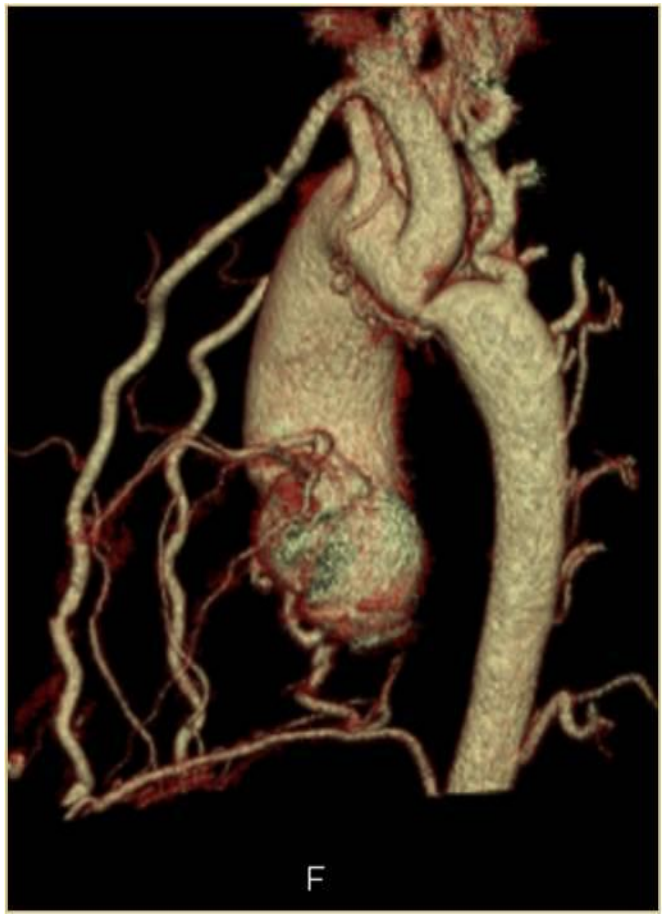


Coarctation of the aorta

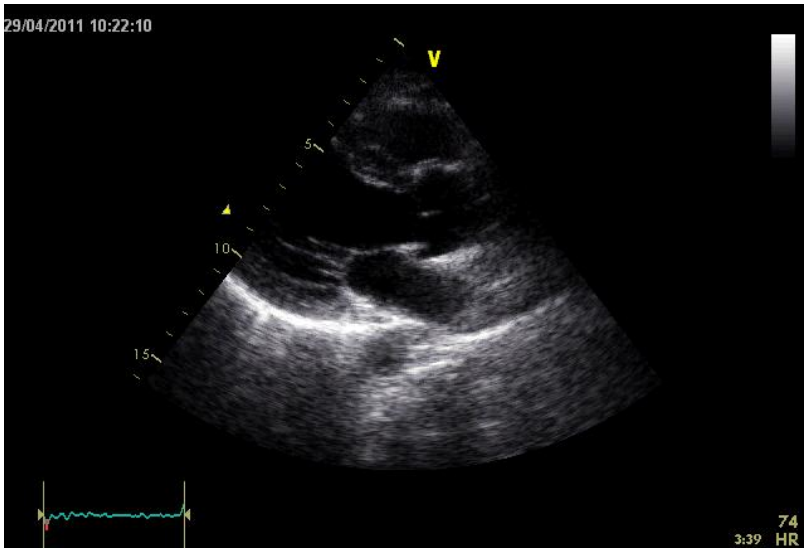
- 5 – 8% all congenial heart defects
- Associated with:
  - BAV
  - Subvalvular, supravalvularAS
  - Turner syndrome
  - Shone's syndrome
  - Congenital rubella syndrome

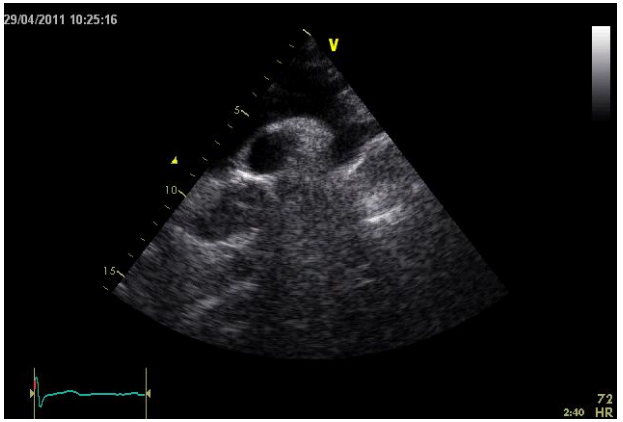
- Hourglass deformity
- Localized fibrous diaphragm
- Diffuse hypoplasia of descending aorta

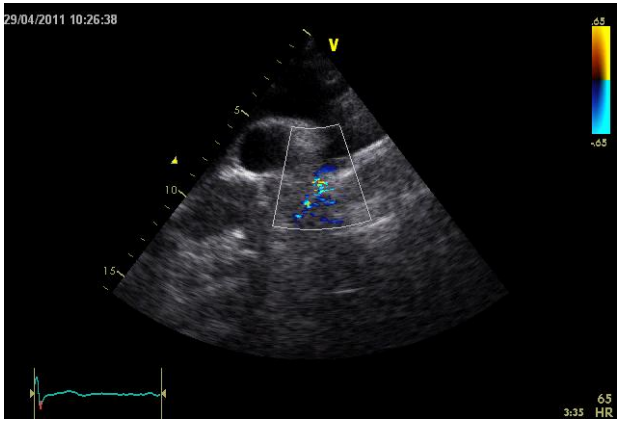
CT in diagnosis of CoA



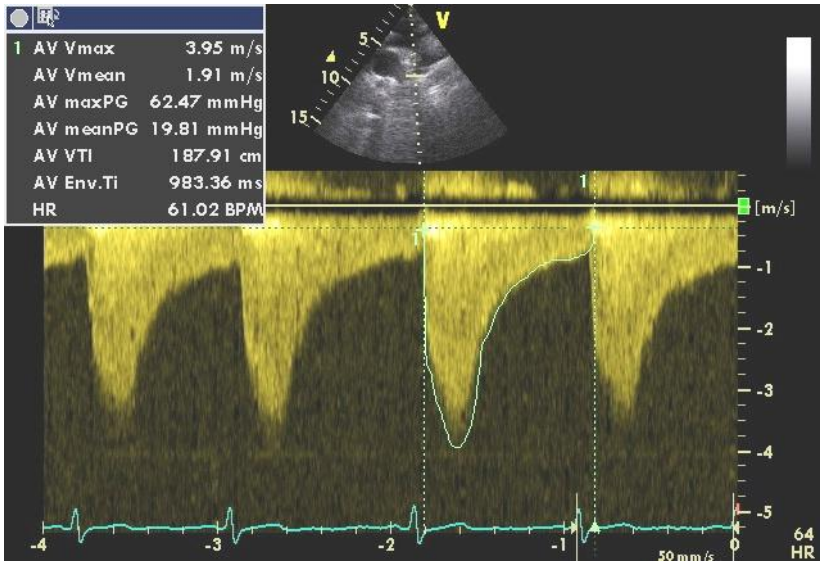
Structural assessment

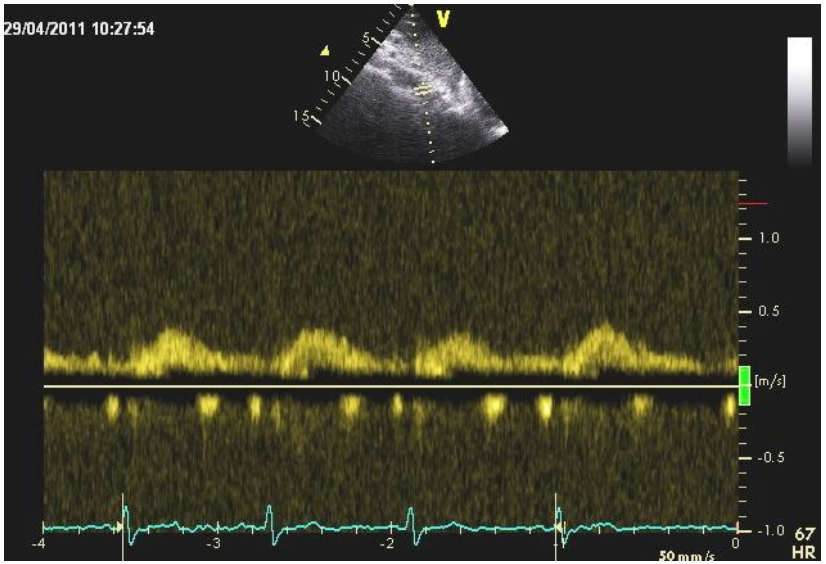






Doppler examination





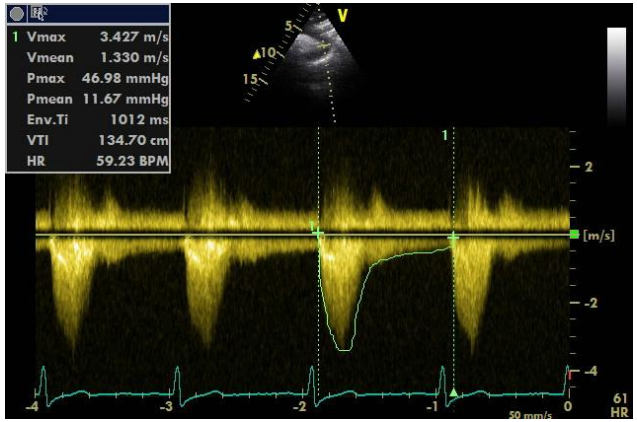
Diastolic ‘run-off’ phenomenon is most reliable sign of significant coarctation

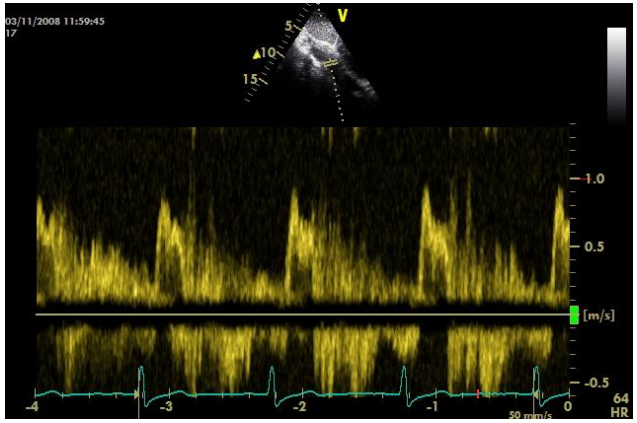
CoAo

ESC Guidelines for the management of grown-up congenital heart disease (new version 2010)

Table 11 Indications for intervention in coarctation of the aorta

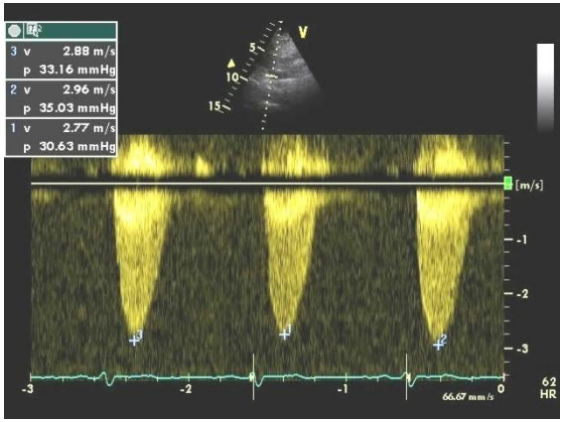
Indications	Class[a]	Level[b]
All patients with a non-invasive pressure difference >20 mmHg between upper and lower limbs, regardless of symptoms but with upper limb hypertension (>140/90 mmHg in adults), pathological blood pressure response during exercise, or significant LVH should have intervention	I	C
Independent of the pressure gradient, hypertensive patients with ≥50% aortic narrowing relative to the aortic diameter at the diaphragm level (on CMR, CT, or invasive angiography) should be considered for intervention	IIa	C
Independent of the pressure gradient and presence of hypertension, patients with ≥50% aortic narrowing relative to the aortic diameter at the diaphragm level (on CMR, CT, or invasive angiography) may be considered for intervention	IIb	C

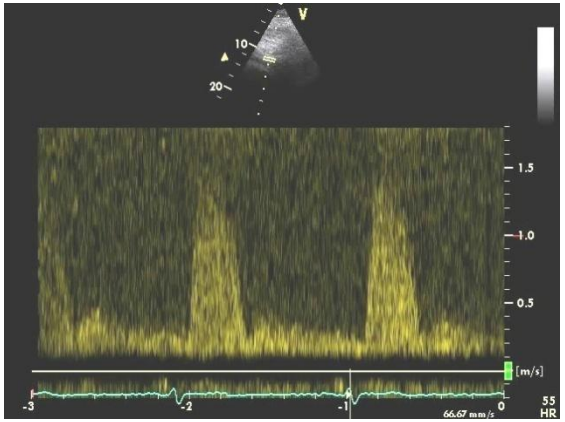




Post-operation assessment

- Stent implantation
- Operation
  - End-to end anastomosis
  - Subclavian flap aortoplasty
  - Prosthetic flap aortoplasty





Increased systolic flow rates may developed due to a lack of aortic compliance

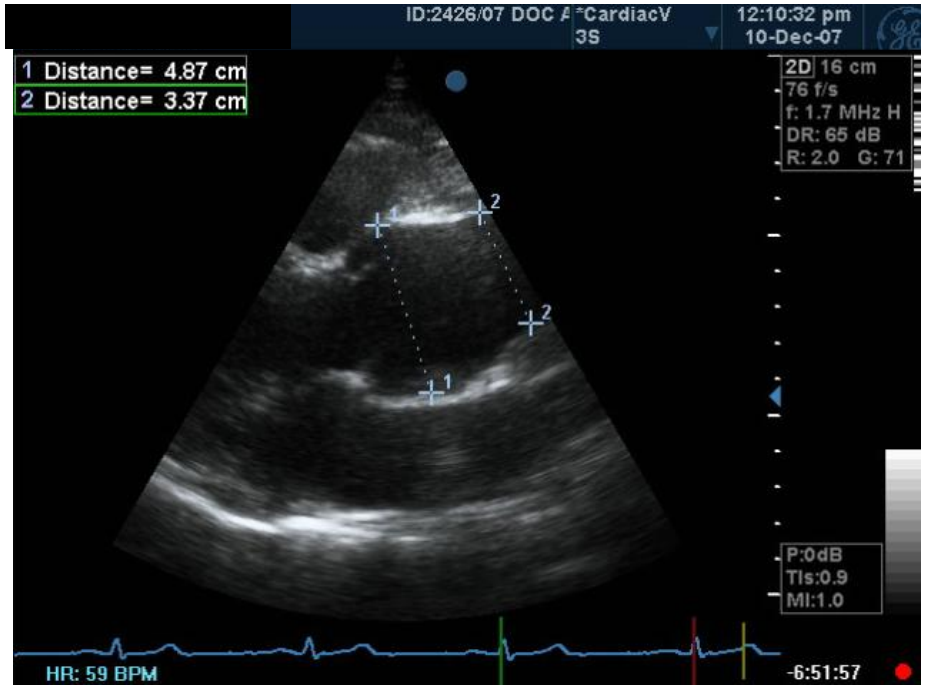
Marfane syndrome

- Autosomal, dominant disorder of connective tissue
- Prevelance 2 – 3 per 10 000
- Mutations in the FBN^{1} gene encoding fibrilline-1(glycoprotein in the extracellular matrix)
- Deficiency of fibrillin may lead to weakening of the supportive tissues
- More than 1000 mutations have been identified
- Progressive dilation of the aorta

How to diagnose Marfan syndrome?

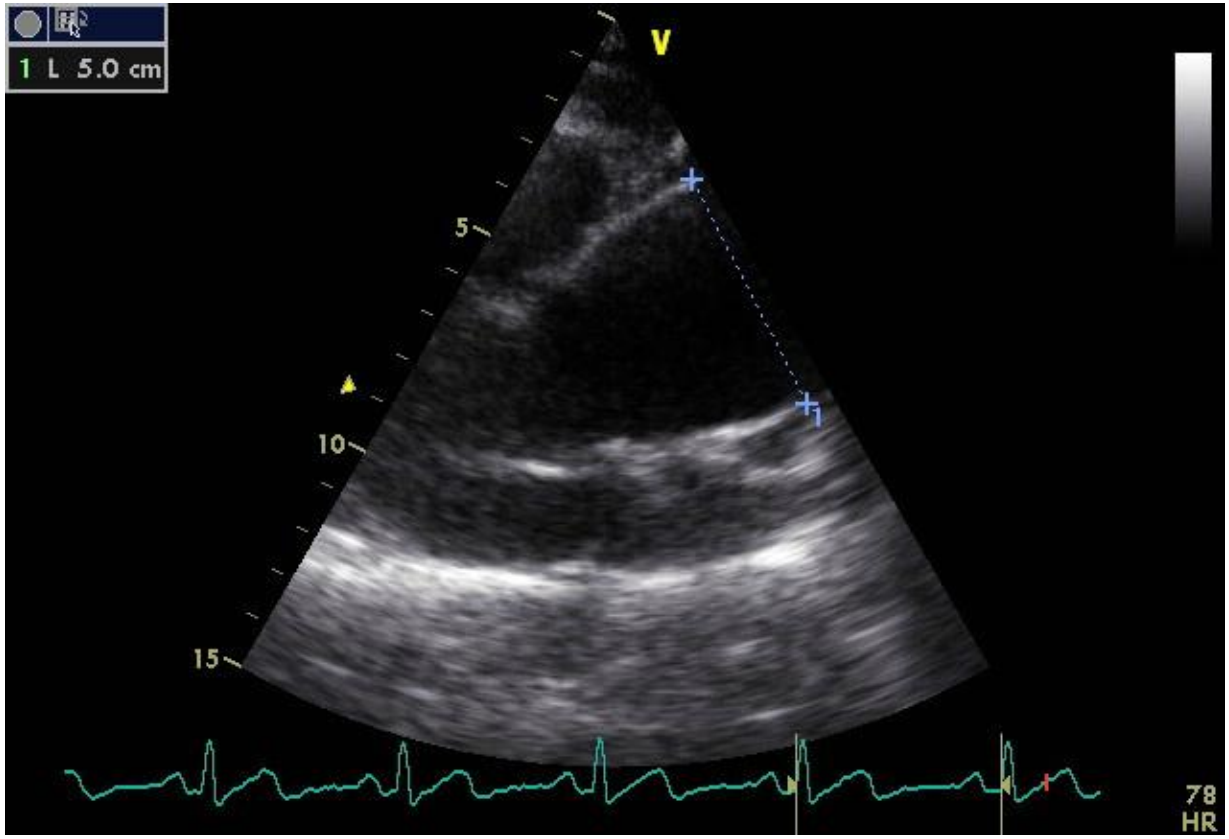
- Family history of heart-related death
- Physical features:
  - Skeletal frame (the ratio of arm/leg size to trunk size)
  - Lens dislocation
  - Aortic and valve abnormality
- Genetic analysis (?)

Dilation of the aortic root in Marfan syndrome



Dilation of the aortic root is found in about 60-80% of patients with Marfan syndrome

Ascending aorta dilation in Marfan syndrome



Marfan syndrome

ESC Guidelines for the management of grown-up congenital heart disease (new version 2010)

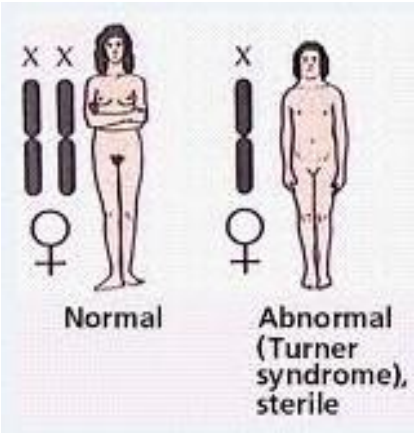
Table 12 Indications for aortic surgery in Marfan syndrome[67]

Indications	Class[a]	Level[b]
Patients should undergo surgery when aortic root maximal diameter is:		
• >50 mm	I	C[c]
• 46–50 mm with		
- family history of dissection or	I	C
- progressive dilation >2 mm/year as confirmed by repeated measurement or	I	C
- severe AR or MR or	I	C
- desire of pregnancy	I	C
• Patients should be considered for surgery when other parts of the aorta >50 mm or dilation is progressive	IIa	C

- Progressive dilation of the aorta leads to aortic dissection or rupture
- Dissection may occur in patients with mild aortic dilation
- Completely safe diameter of the aorta does not exist
- Higher risk of re-dissection and reccurent aneurysm

Turner syndrome

- Chromosomal abnormality in wich one or part of X chromosome is absent
- Prevelance 1 : 2 500 woman
- Characteristic physical abnormalities (short stature, broad chest, low-set ears, webbed neck
- Cardiovascular malformations 17 - 45%
  - Bicuspid aortic valve up to 15%
  - Coarctation of the aorta 5 – 12%
  - Aortic dilation and rupture
  - Hypoplastic left heart syndrome



Aortic dilation and dissection in population with CHD

	Marfan syndrome	Isolated BAV	Coarctation	Turner syndrome
Incidence population	1 : 5 000	> 1 : 100		1 : 2 500 (weman)
Site of dilation	Aortic root	Asc aorta	Root and asc aorta	Asc aorta
Age onset, y	20 - 40	50's		20 - 40
Progressive dilation	yes	+/-	+/-	?
Aortic dissection	50%	0,5%	1% (?)	1 – 2%

Curr Opin Cardiol 2008,23,519

European Heart Journal
doi:10.1093/eurheartj/ehq249

ESC GUIDELINES

ESC Guidelines for the management of grown-up congenital heart disease (new version 2010)

„Echocardiography remains the first-line investigation and continues to evolve"

Philip Kilner (UK), Folkert Meijboom (The Netherlands), Barbara J. M. Mulder (The Netherlands), Erwin Oechslin (Canada), Jose M. Oliver (Spain), Alain Serraf (France), Andras Szatmari (Hungary), Erik Thaulow (Norway), Pascal R. Vouhe (France), Edmond Walma (The Netherlands).