Left ventricular outflow tract obstruction

Echocardiography

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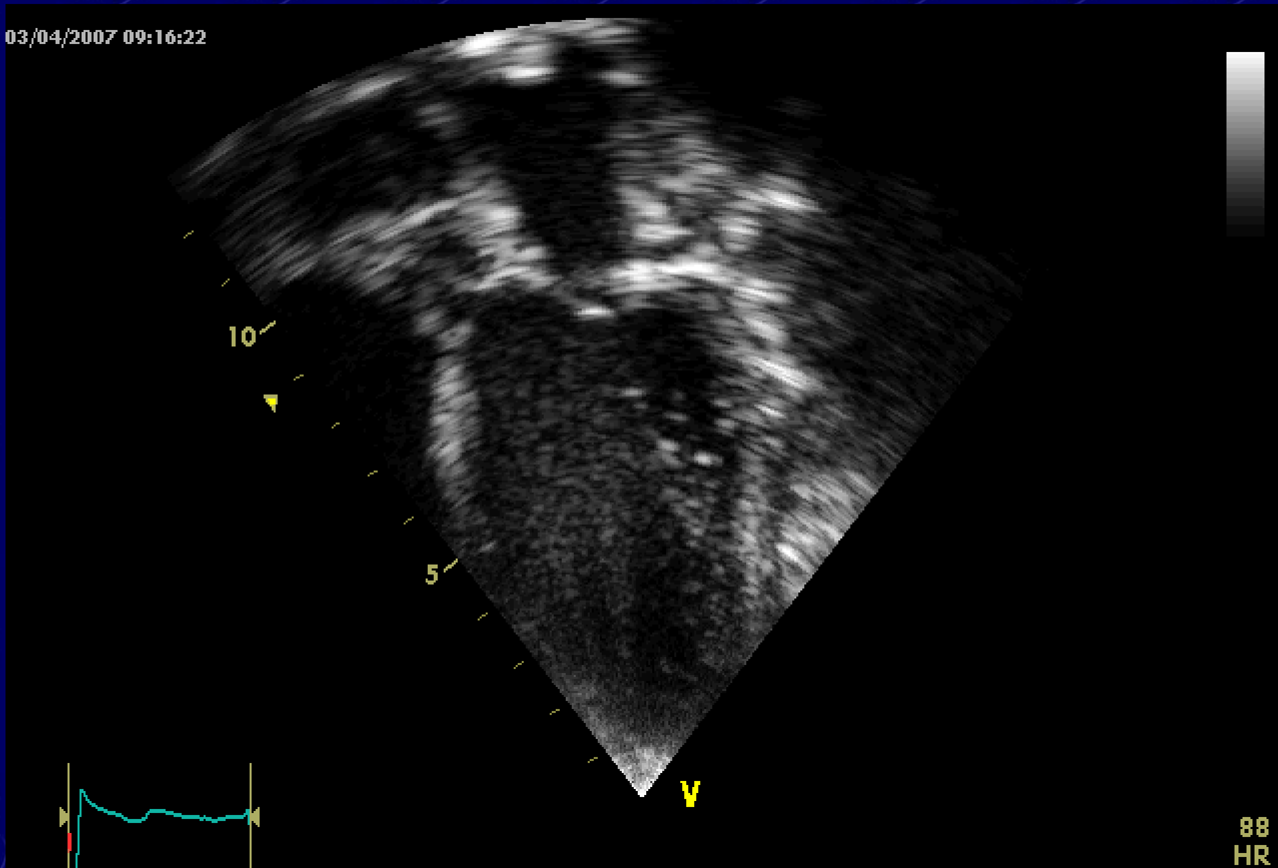
J. Škovránek, P.Tax,
J. Gilík

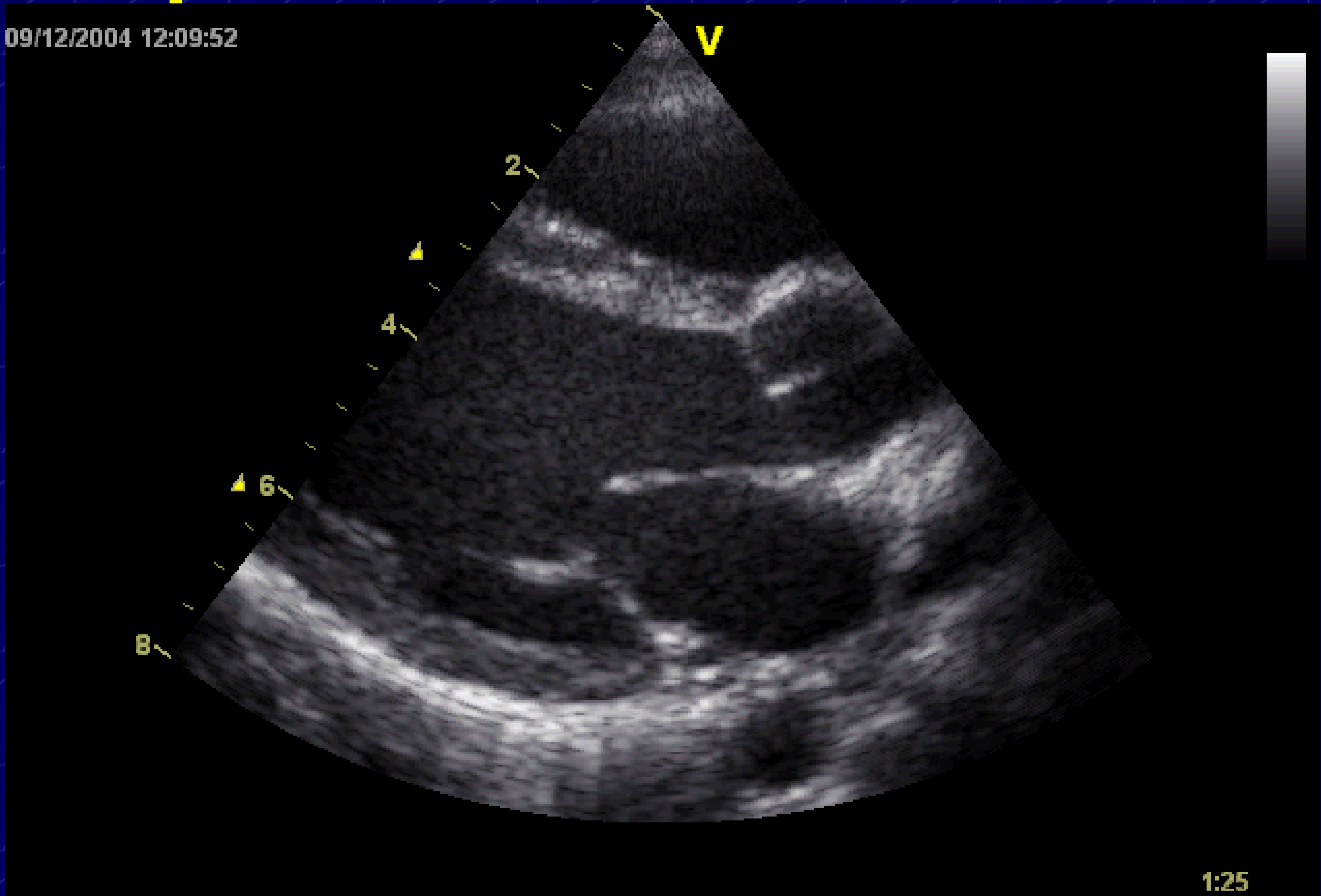
Kardiocentrum,
University Hospital Motol,
Prague, Czech Republic

No disclosures

Normal anatomy of LVOT
„apical modif. 4 – chamber view“



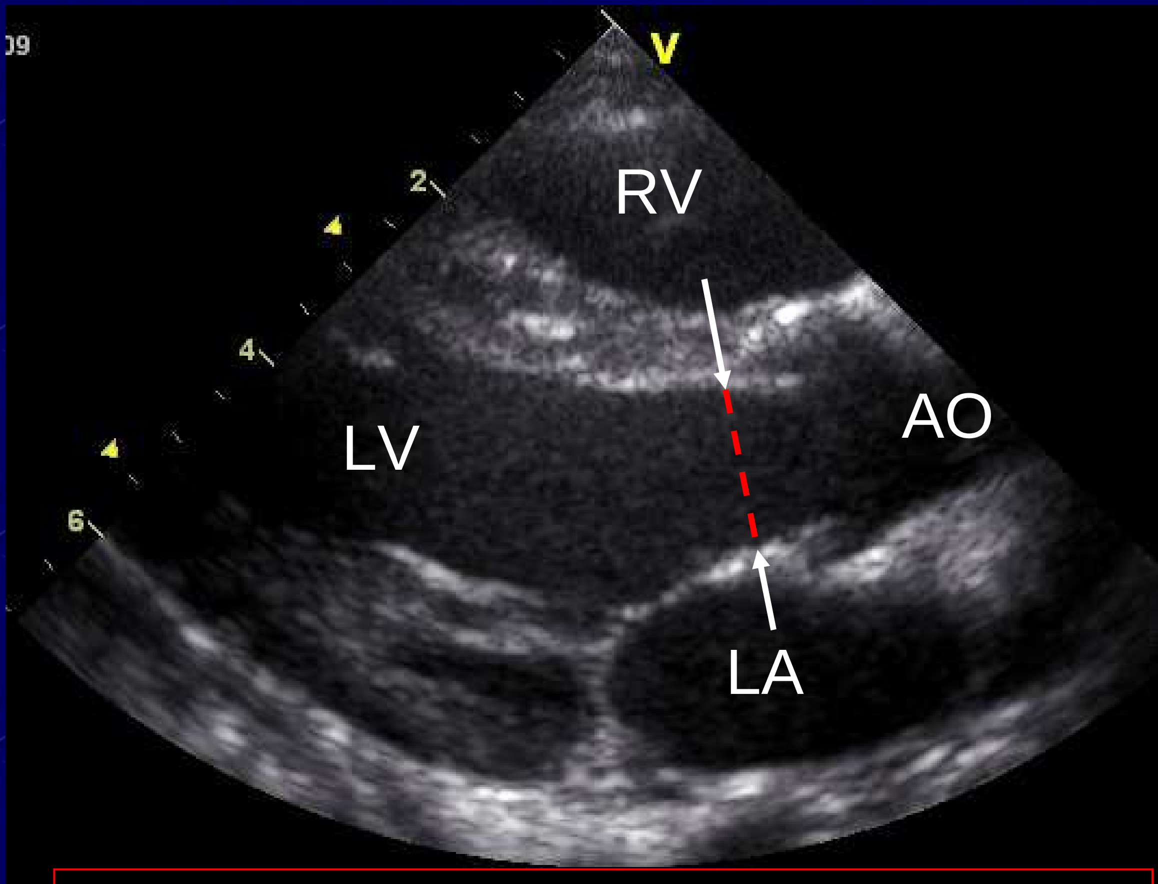
Normal anatomy of LVOT parasternal LAX view



Valve mobility and cusp separation
AO-MV, LVW thickness, SF, FE
AO root dimensions

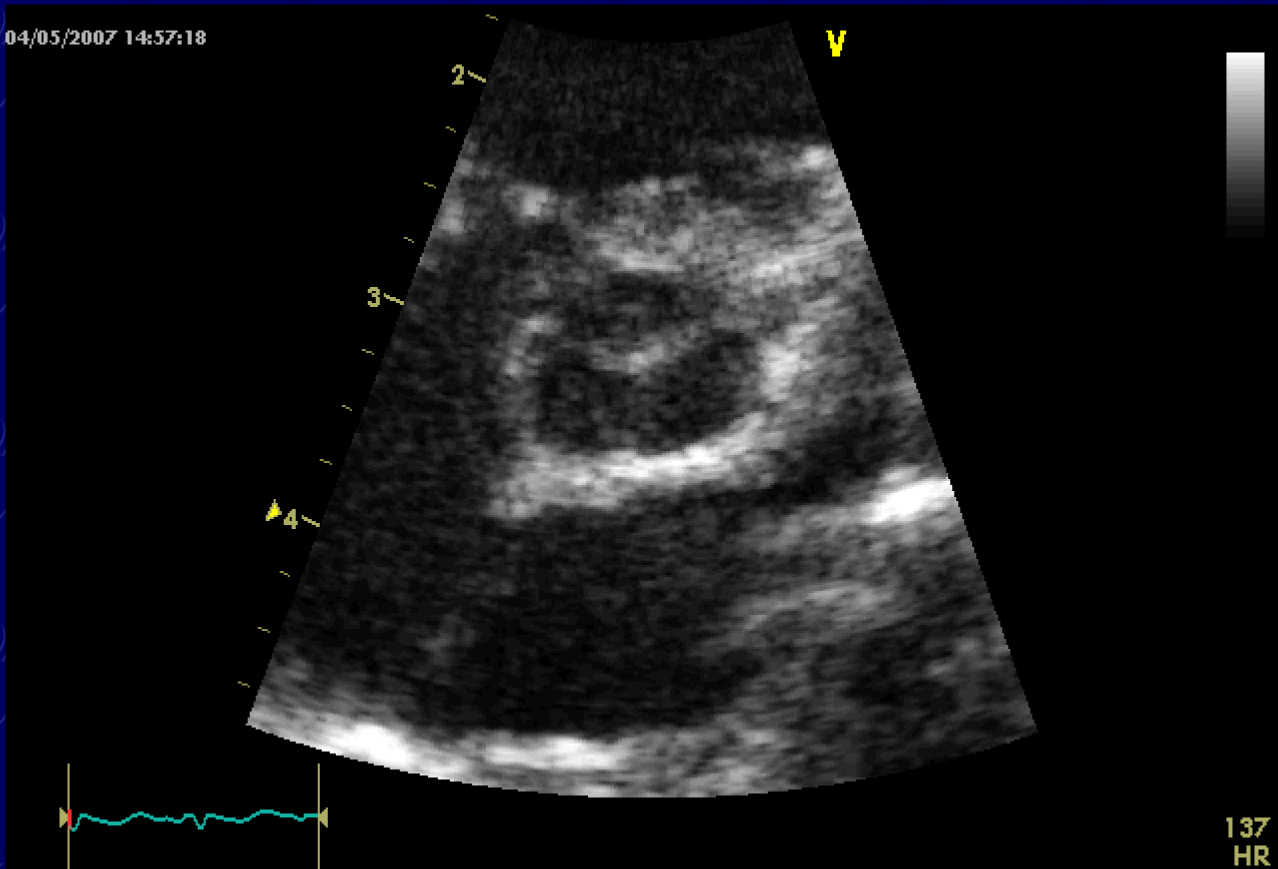
Normal anatomy of LVOT

parasternal LAX view



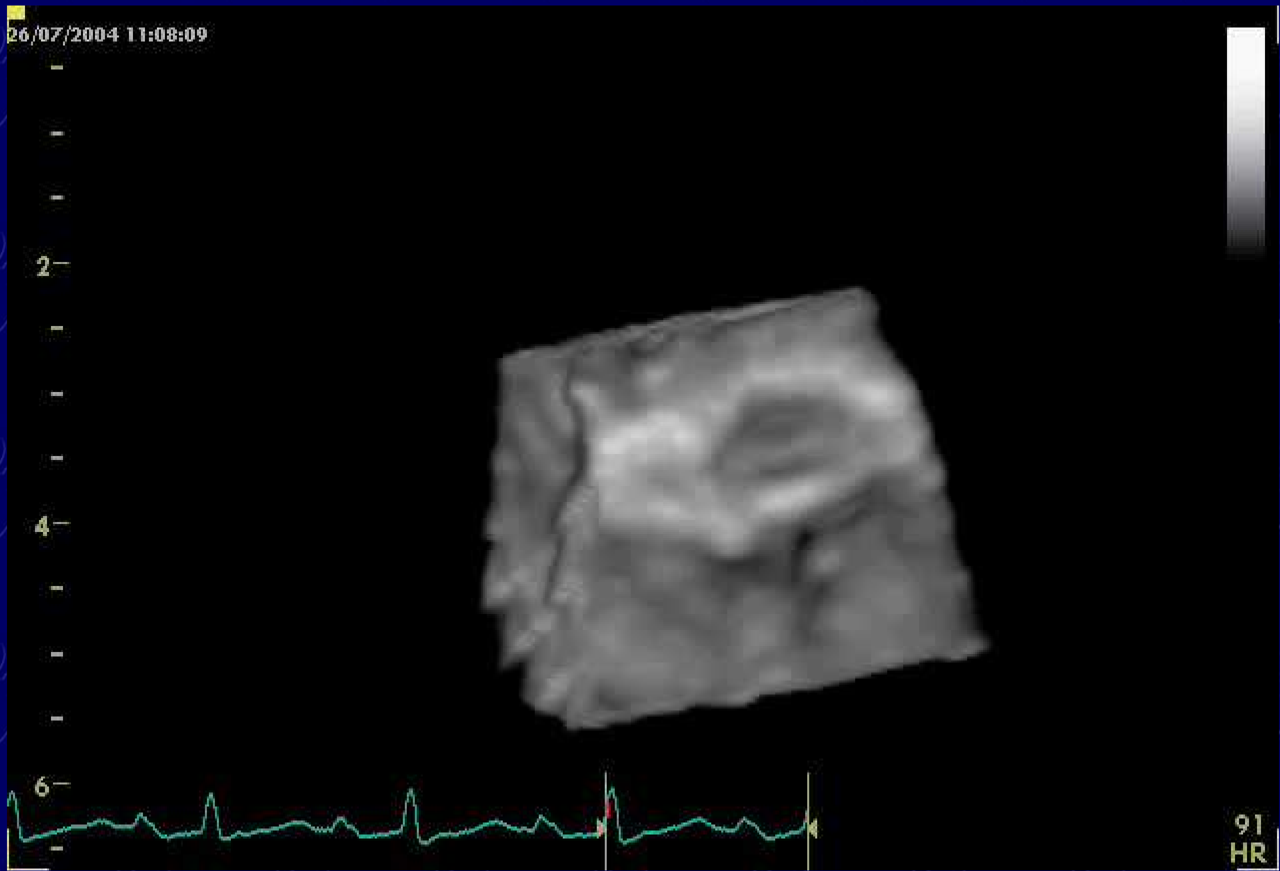
V-A junction – semilunar hinge points

Normal anatomy of LVOT
parasternal SAX view



Face of AOV
Commissures
Cusp No

Normal anatomy of LVOT parasternal SAX view



AORTIC VALVAR STENOSIS

SUPRAVALVAR AORTIC STENOSIS

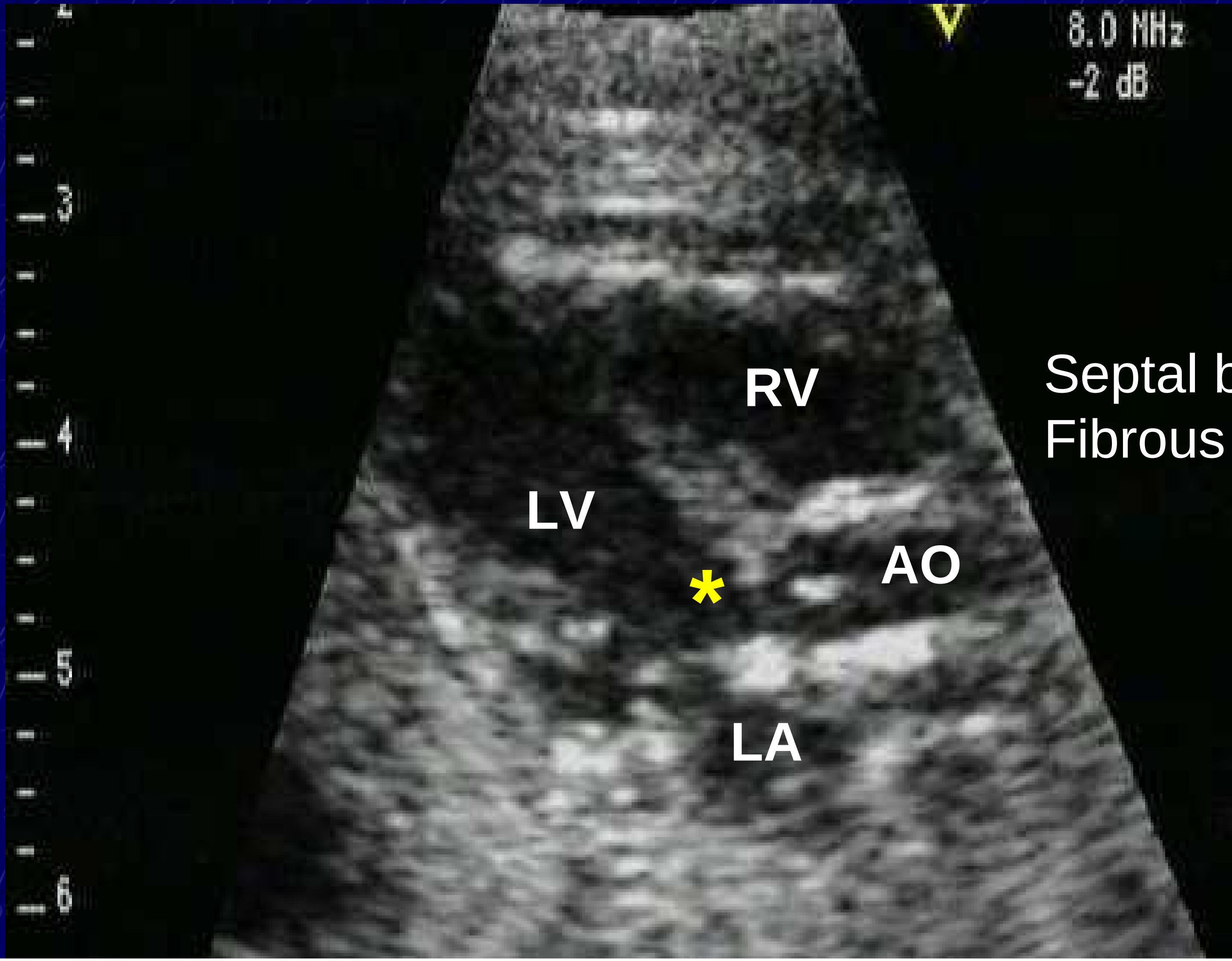
SUBVALVAR STENOSIS

SUBVALVAR STENOSIS

- ✓ intact ventricular septum - fibrous membrane
  - fibromuscular ridge
- ✓ + VSD (posterior deviation of outlet IVS/ short segment obstr.)
  (32% of SAS, Kitchiner 1994, Br Heart J)
- ✓ accessory tissue tags - anomalous attachements of AV valve
- ✓ hypertrophy of IVS (HOCMP)

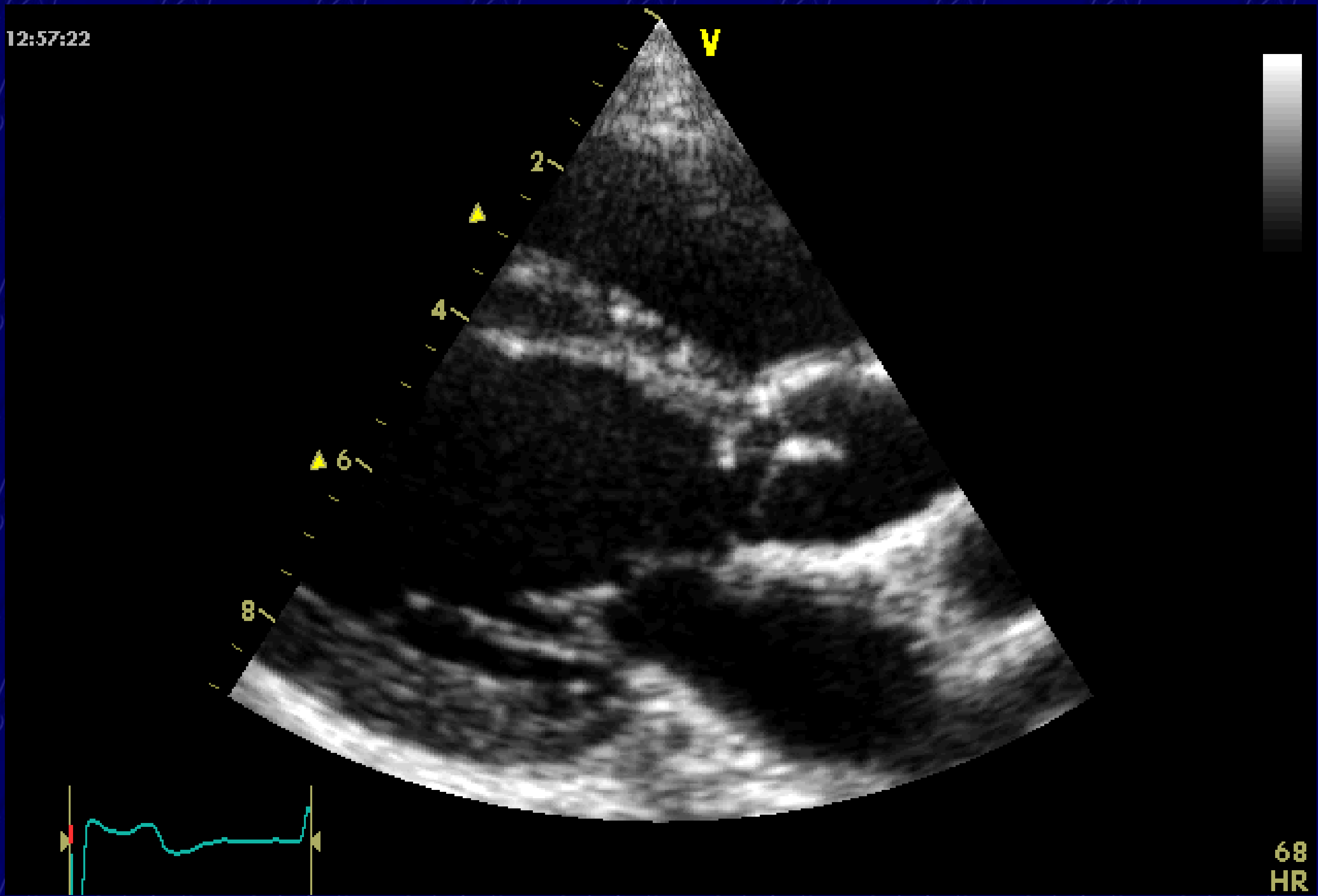
SUBVALVAR STENOSIS

Prenatal diagnosis



Septal buldge
Fibrous shelf

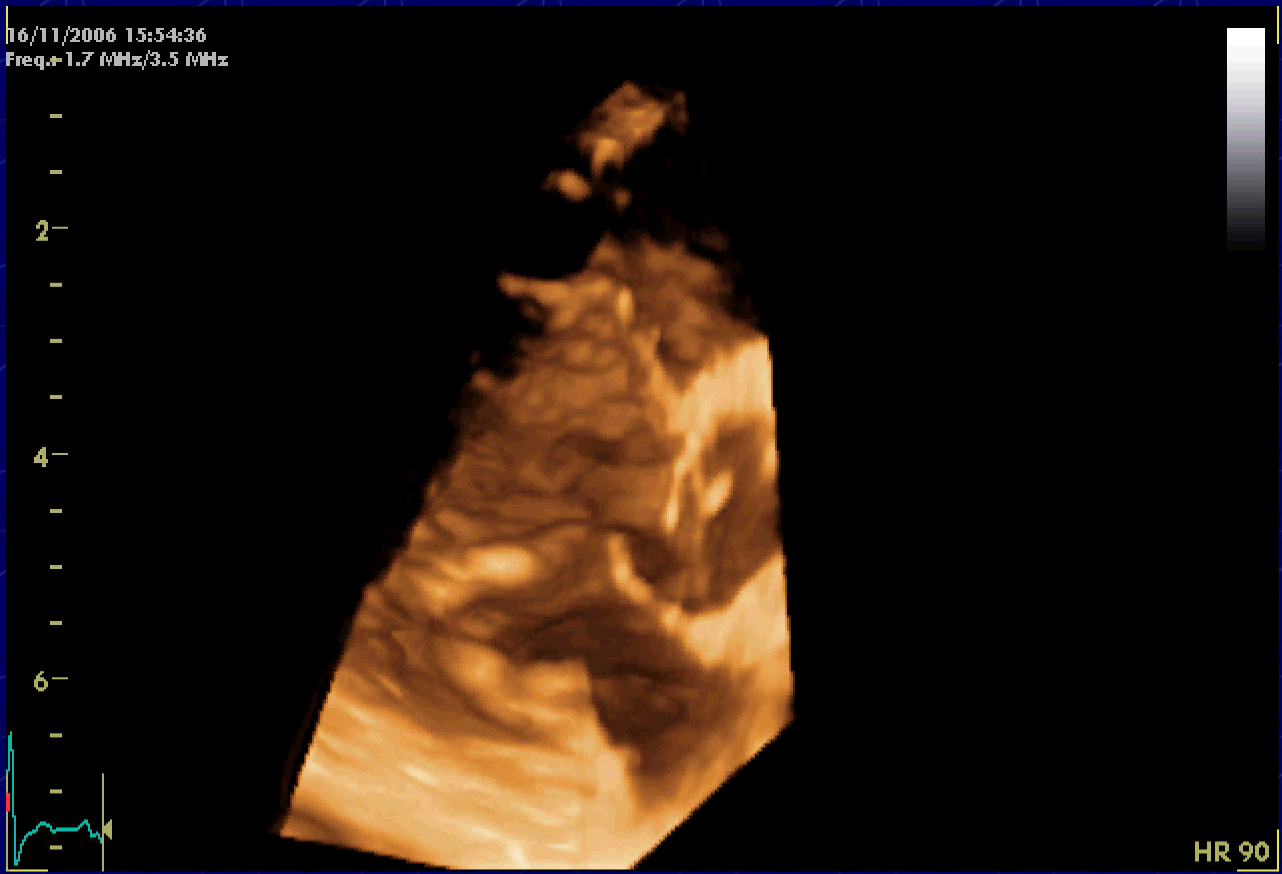
SUBVALVAR STENOSIS



subvalvar ridge

SUBVALVAR STENOSIS

3-D echocardiography

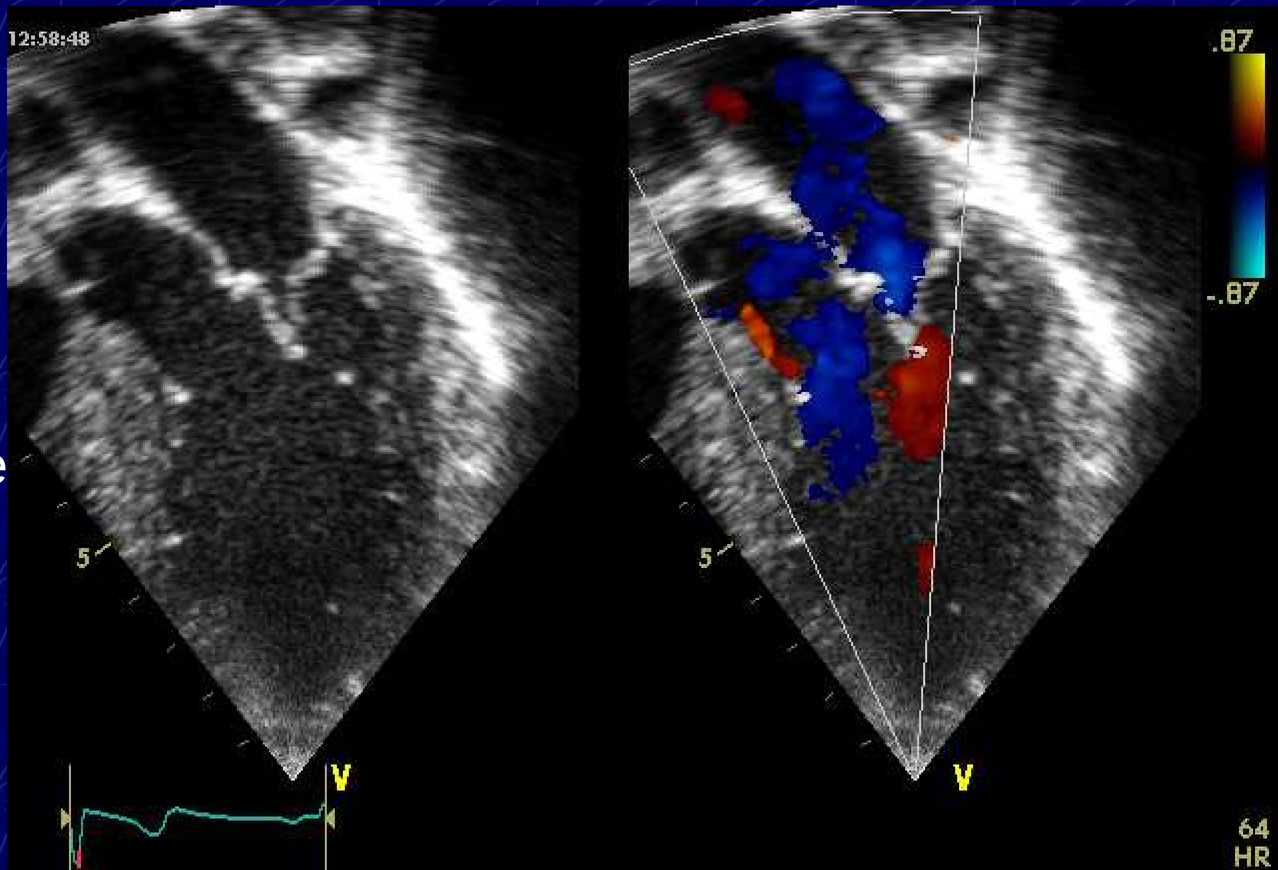


Courtesy of Jan Marek, GOSH London

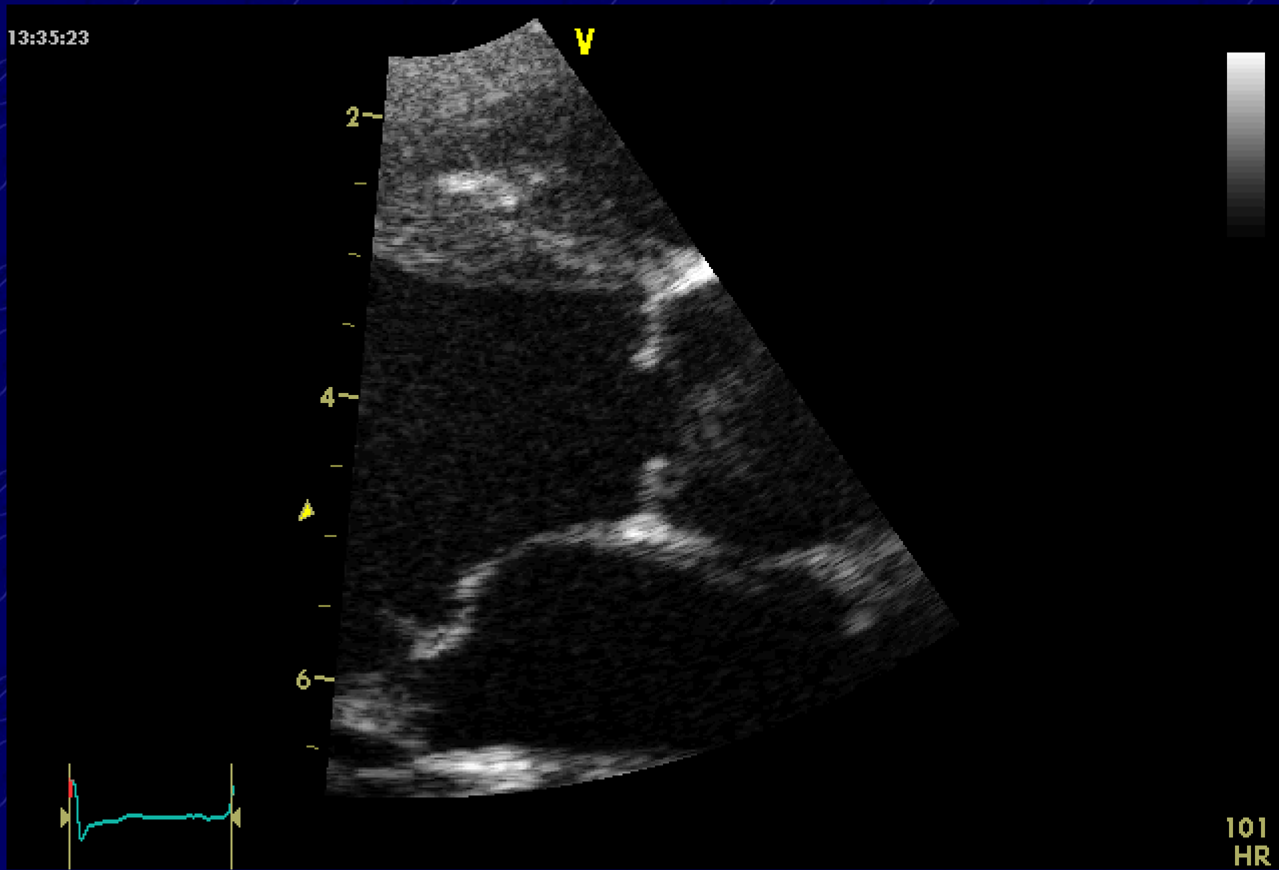
SUBVALVAR STENOSIS

Fibromuscular ridge

Accessory valvar tissue



SUBVALVAR STENOSIS

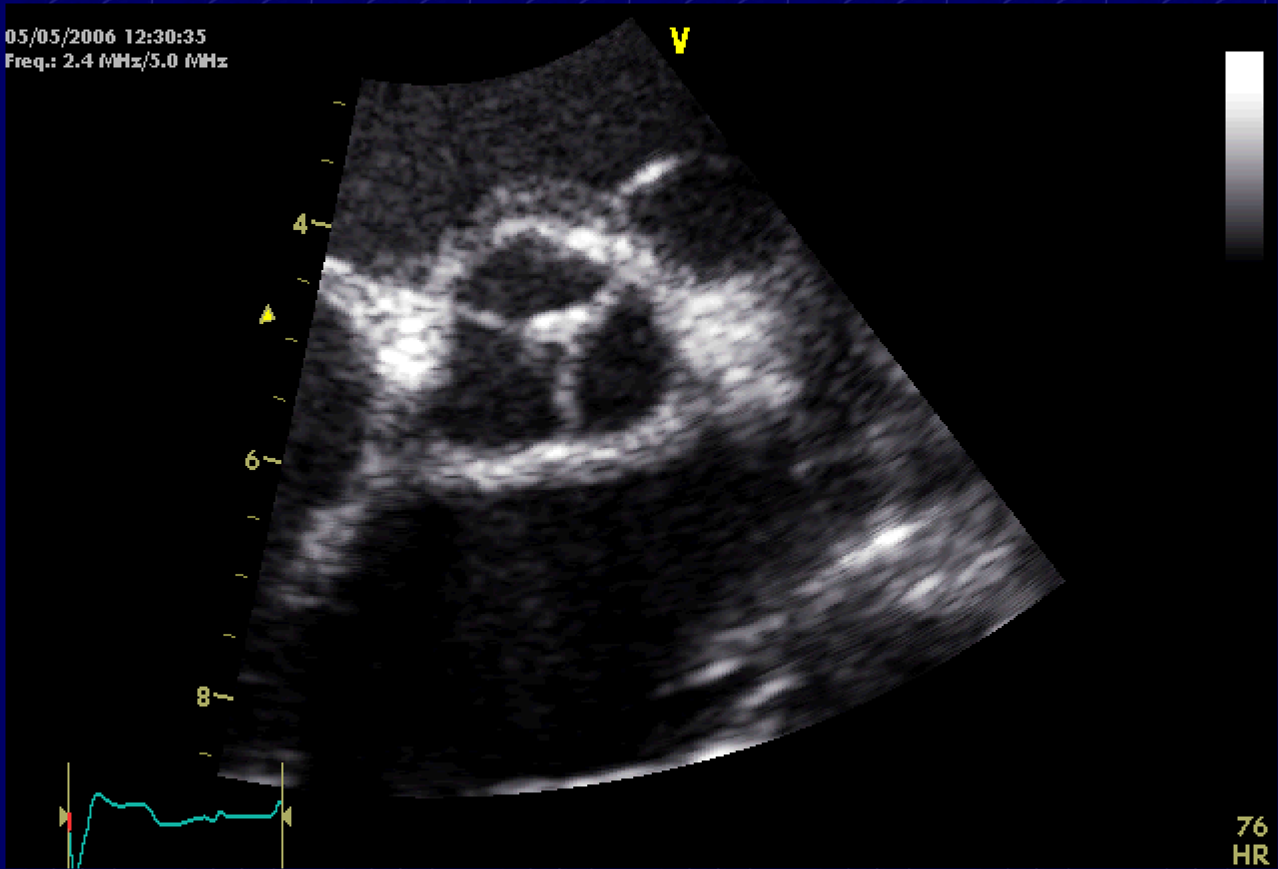


fibrous membrane

SUBVALVAR STENOSIS

Echocardiographist should be more accurate

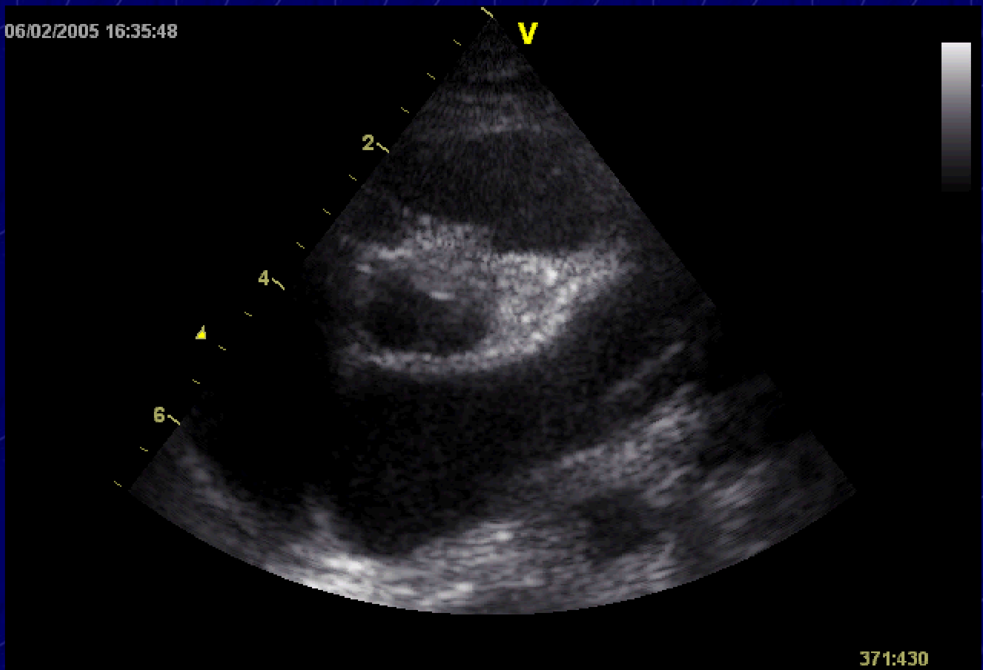
SUBVALVAR STENOSIS

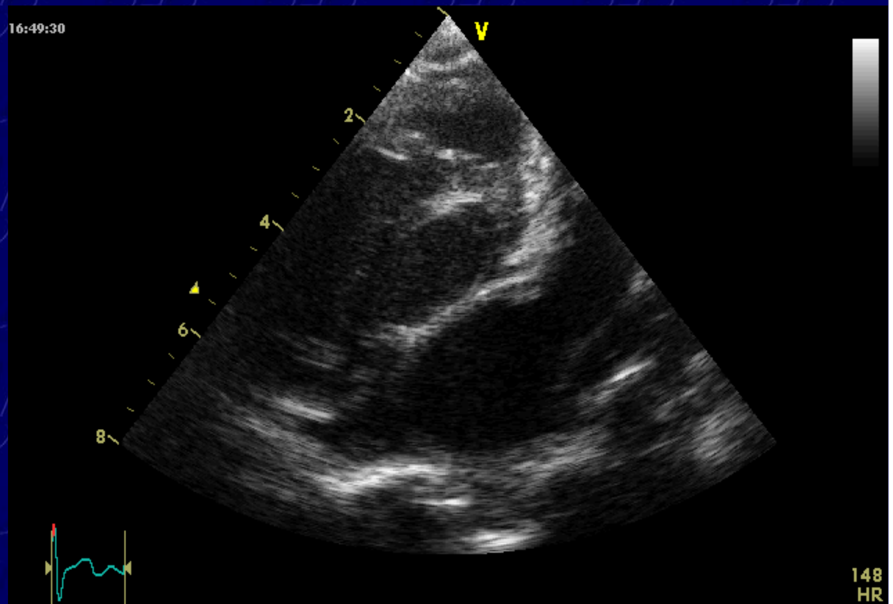


fibrous membrane

SUBVALVAR STENOSIS

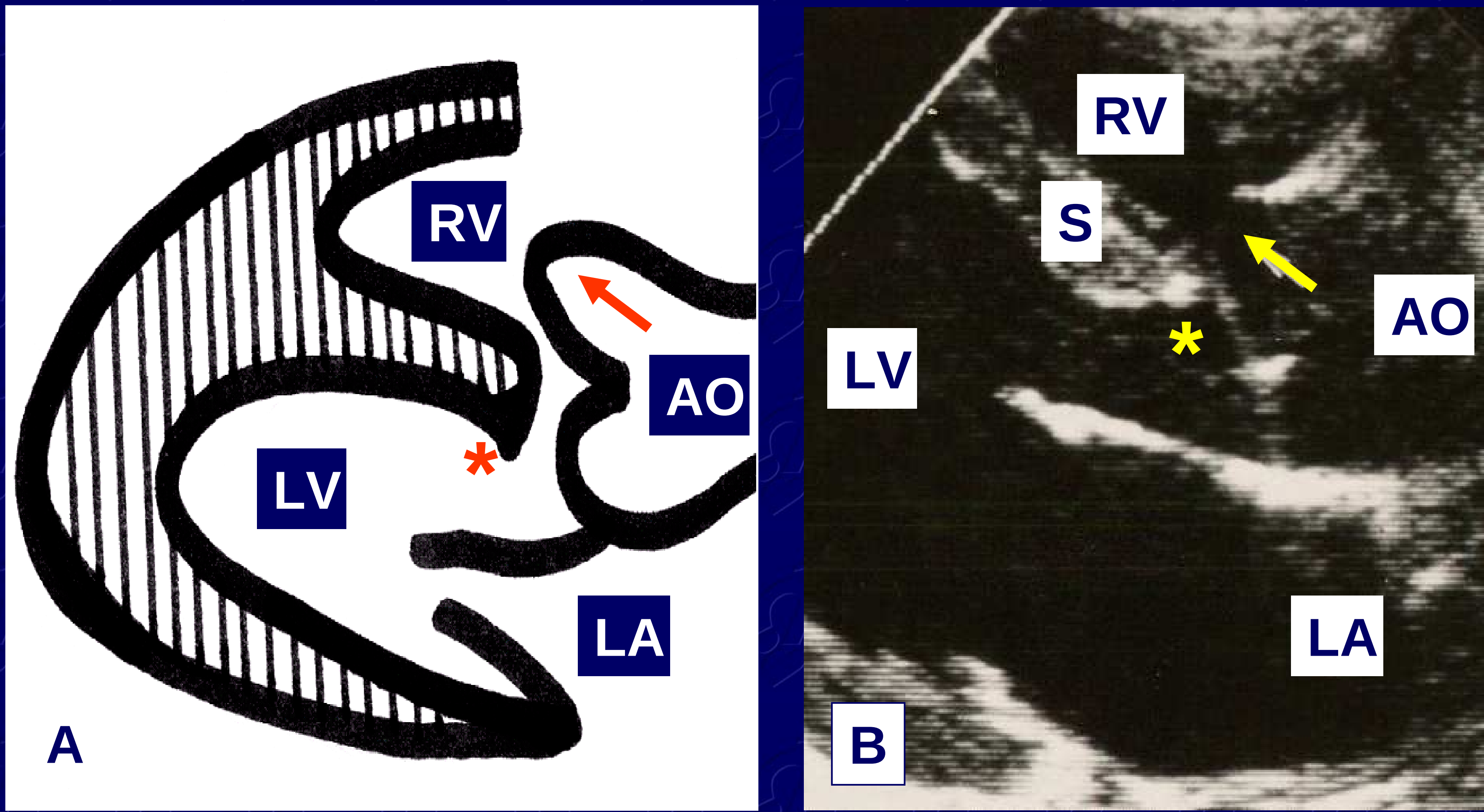
SAX – ridge/ diaphragmatic fashion





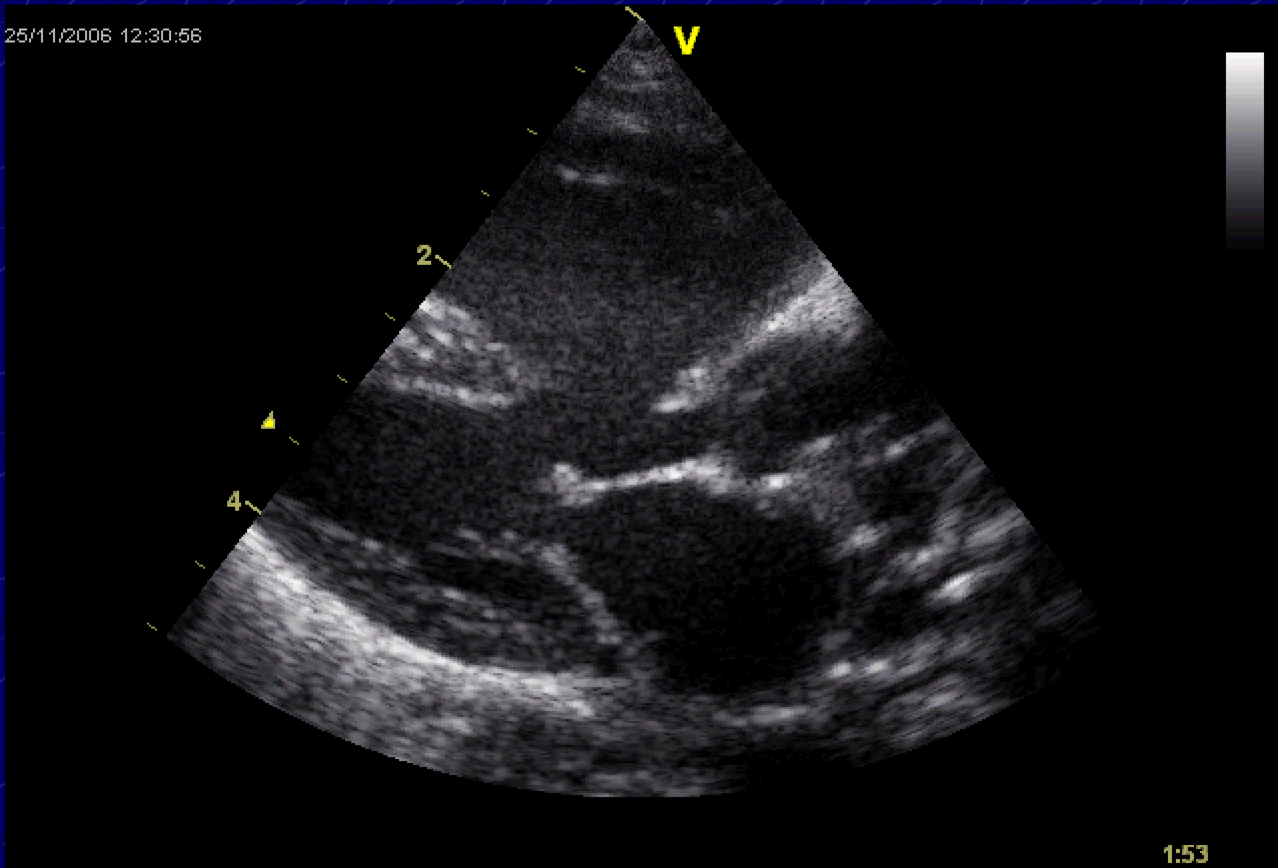
SUBVALVAR STENOSIS

VSD, AOVP



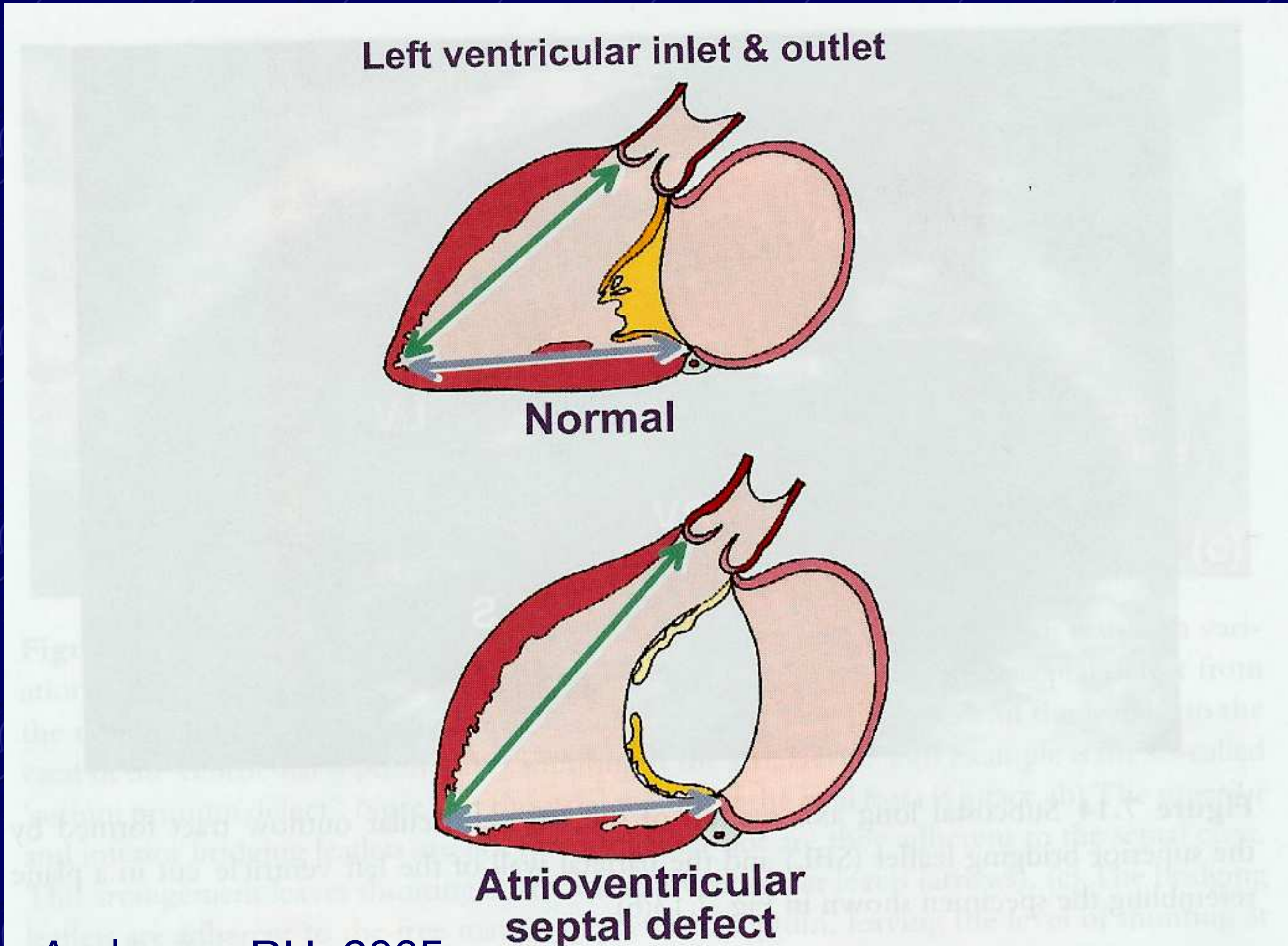
SUBVALVAR STENOSIS

VSD, posterior deviation/ muscular outlet IVS



SUBVALVAR STENOSIS

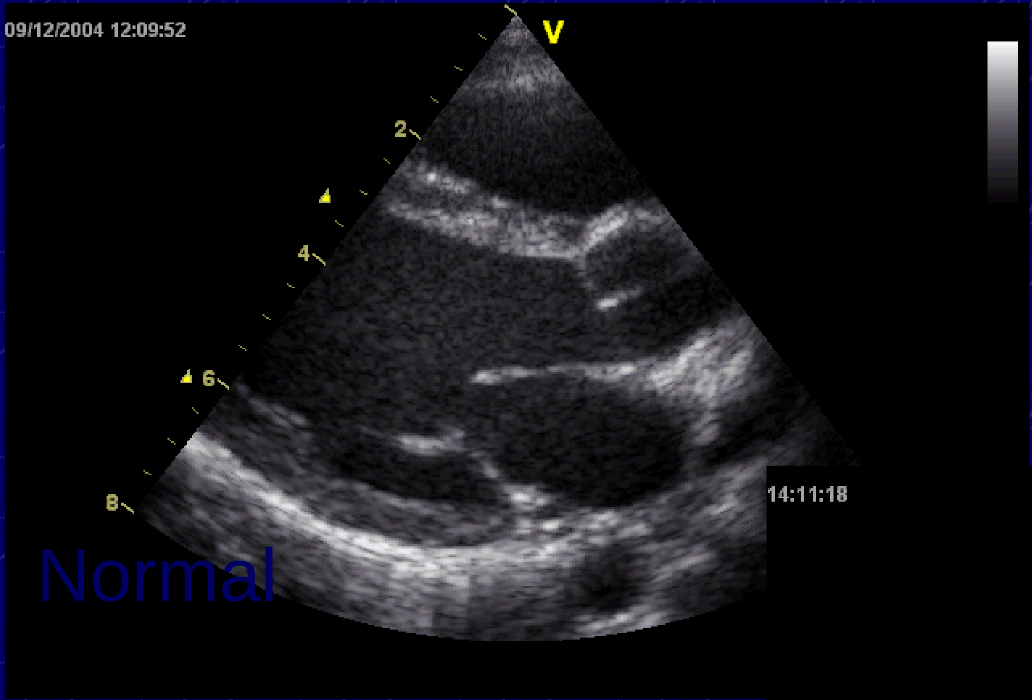
A-V septal defect



Anderson RH, 2005

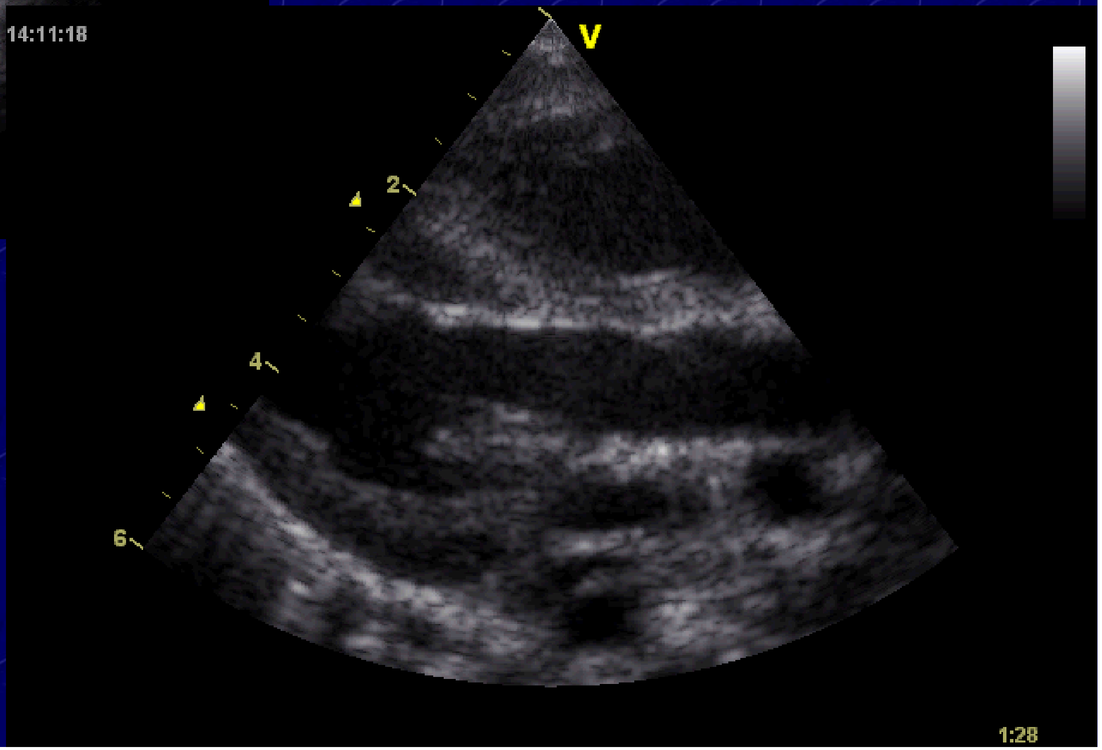
- LVOT – no interpose LAVV/septum
- unwedge AO
- LA wall „inherniated“

SUBVALVAR STENOSIS



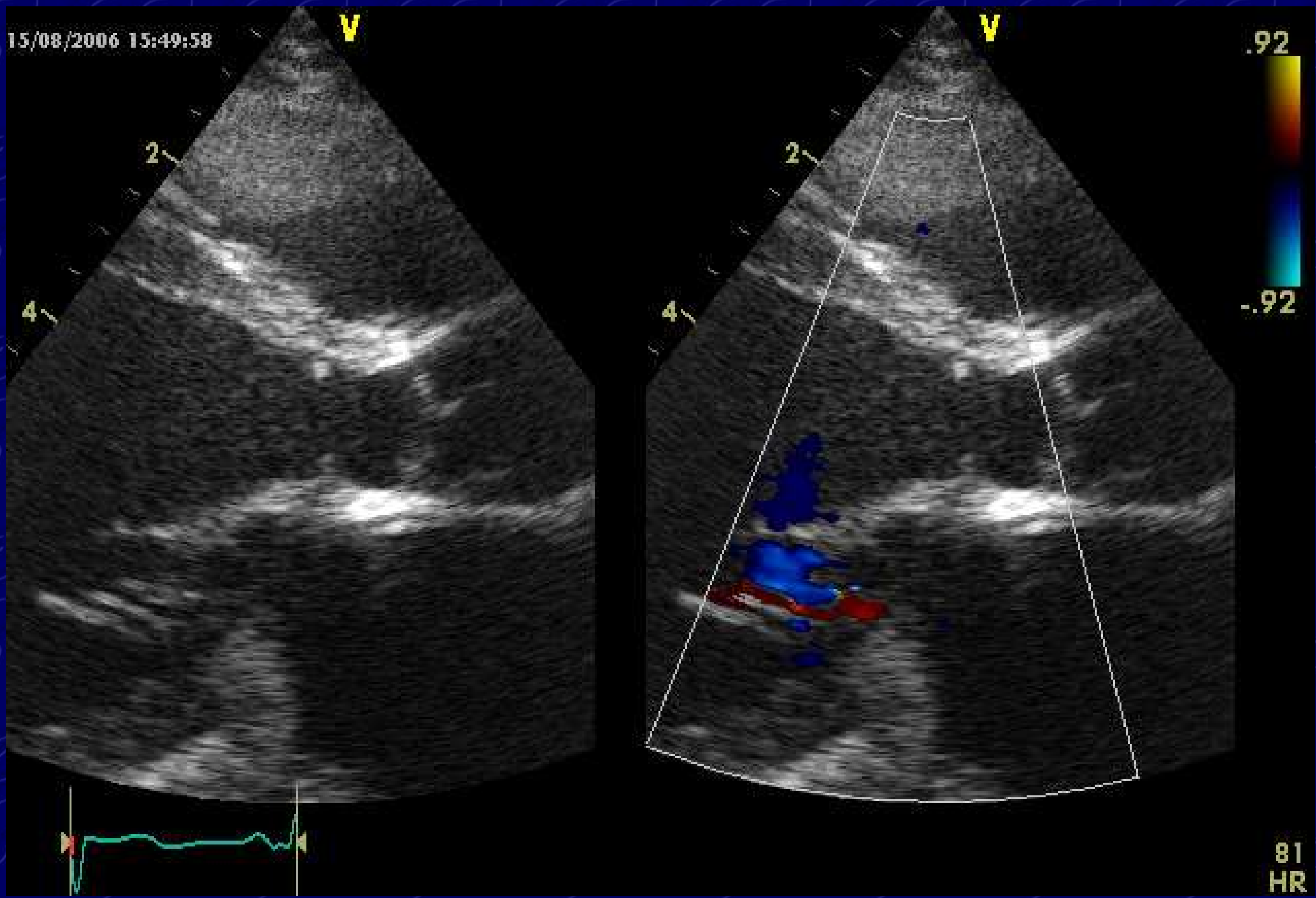
normal heart

AVSD



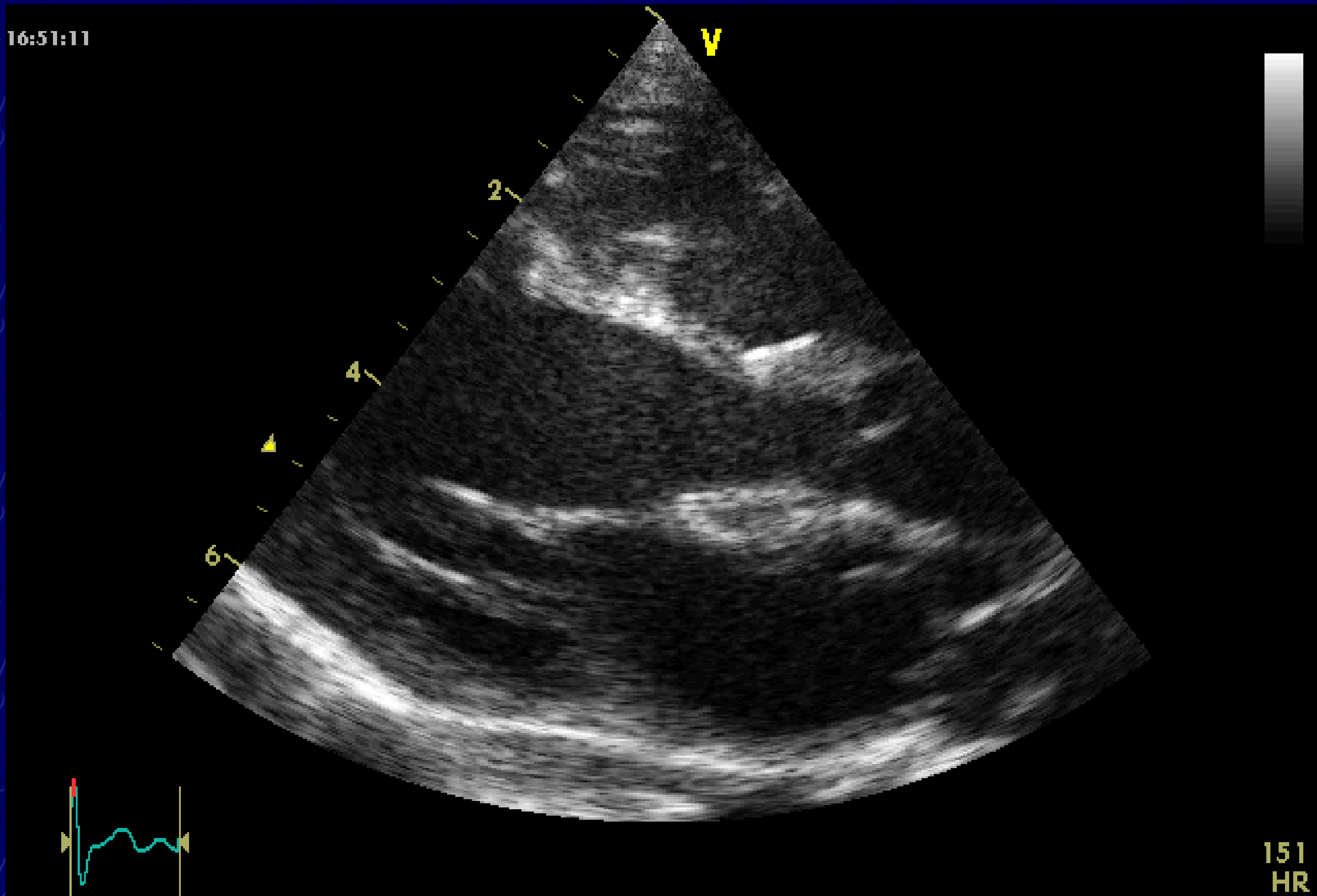
SUBVALVAR STENOSIS

AVSD, fibrous ring

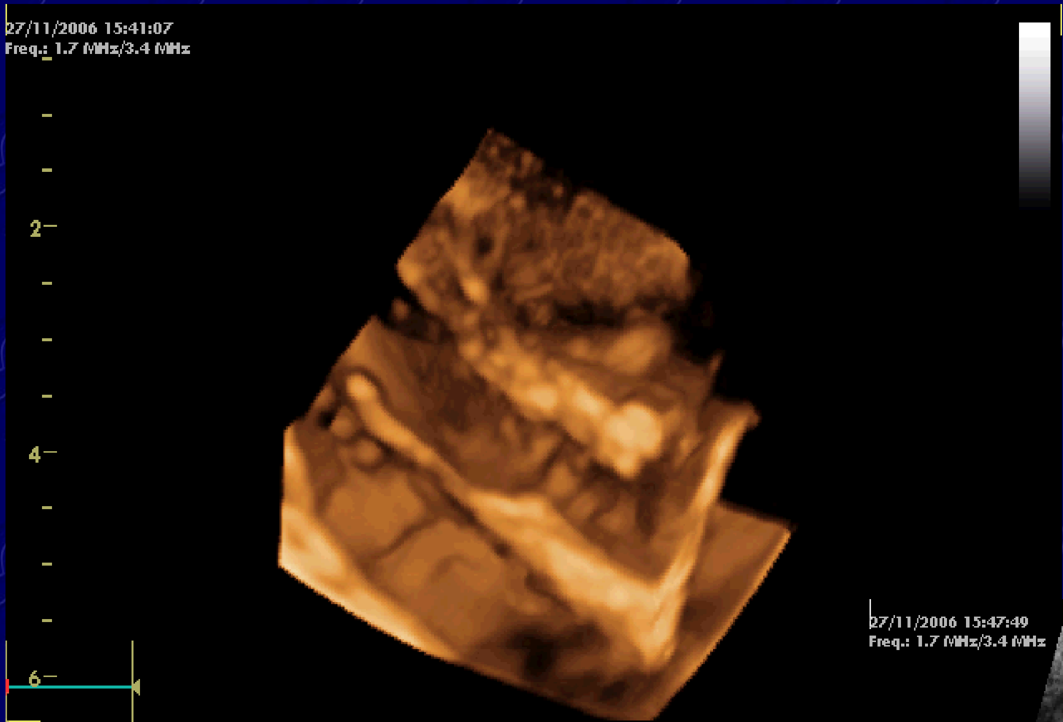


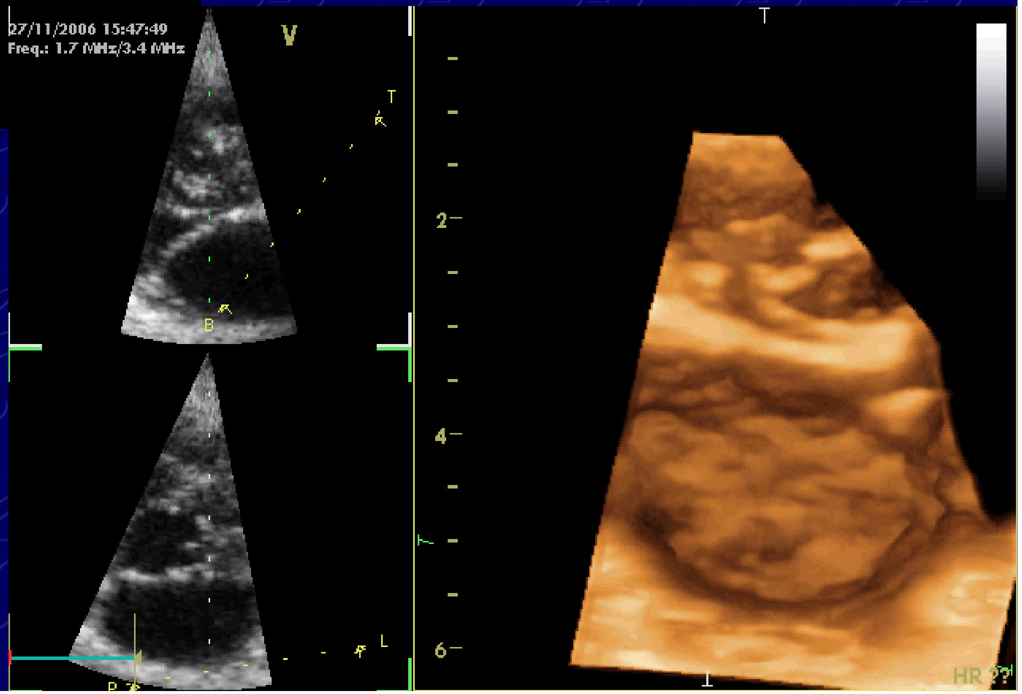
SUBVALVAR STENOSIS

subAO connus (VIF/IVS)



RT – 3DE: Subaortic conus





SUBVALVAR STENOSIS

When to operate?

Mean LVOT gradient > 30 mmHg

D.Coleman, et al., JACC 1994

Peak LVOT grad > 40 mmHg

Brauner, et al., JACC 1997

Aim: prevence of recurrence

secondary progressive aortic valve disease

SUBVALVAR STENOSIS

When to operate?

Mean LVOT gradient > 30 mmHg
Karamlou, et al., Ann Thorac Surg 2007

Peak LVOT grad > 40 mmHg
Brauner, et al., JACC 1997

Aim: prevence of recurrence
secondary progressive aortic valve disease

AORTIC VALVAR STENOSIS

- ✓ CRITICAL NEONATAL AS
- ✓ DOMED AOV IN INFANCY
- ✓ BICUSPID AOV

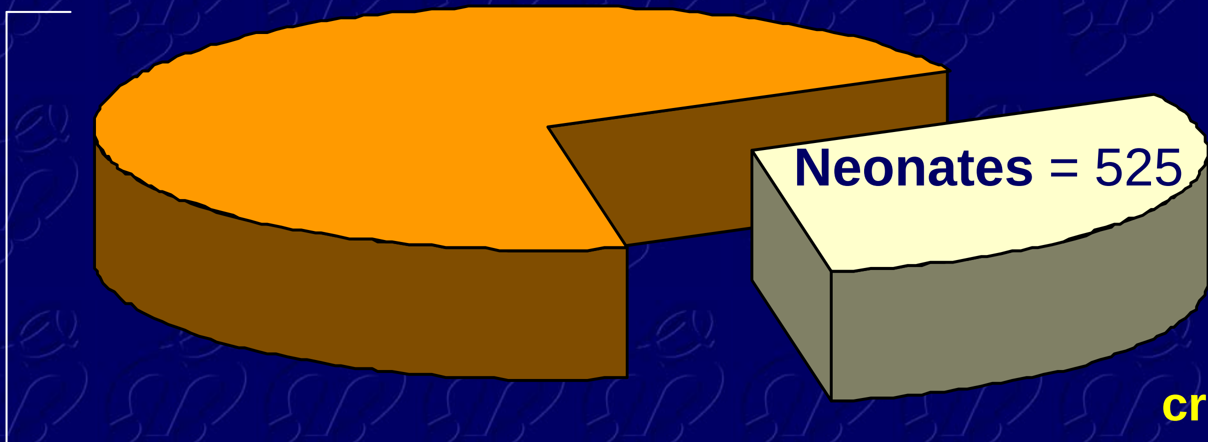
CRITICAL NEONATAL AS

2005 yr natality (WHO): 136 mil.
810.000 congenital heart defects

↓ 7,8%

80.000 children - aortic stenosis

Kardiocentrum,Prague 1991- 2006



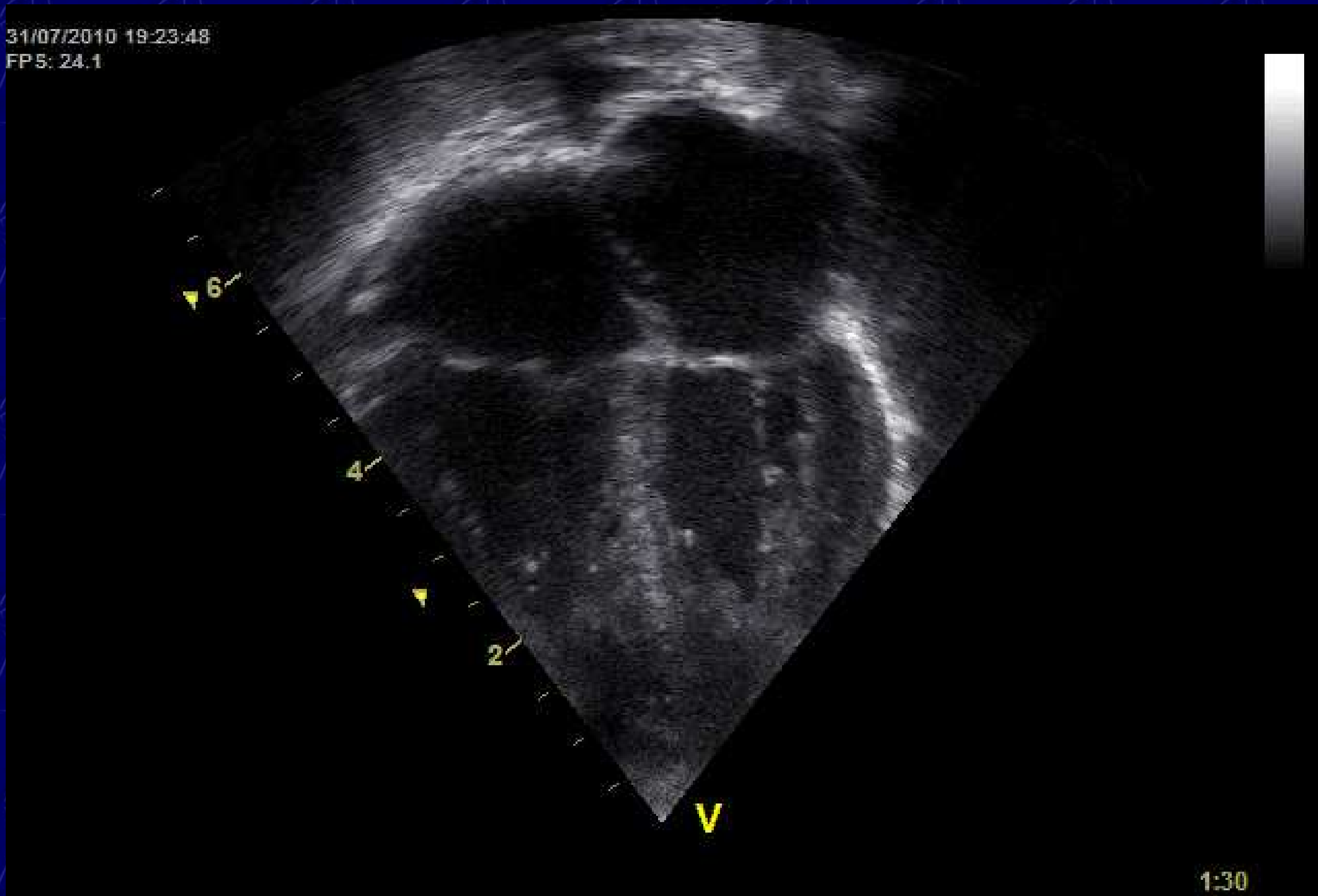
CRITICAL NEONATAL AS

unique entity, differs to "adult form" AS

- clinical feature – left heart failure, ductal dependency
- ↑ morbidity/ mortality , ↑ re-interventions
- ↓ aortic „annulus"
- ↑ co-morbidity – MS, COA . . . (Shone sy)
- endocardial fibroelastosis
- LV morphology - 3 forms

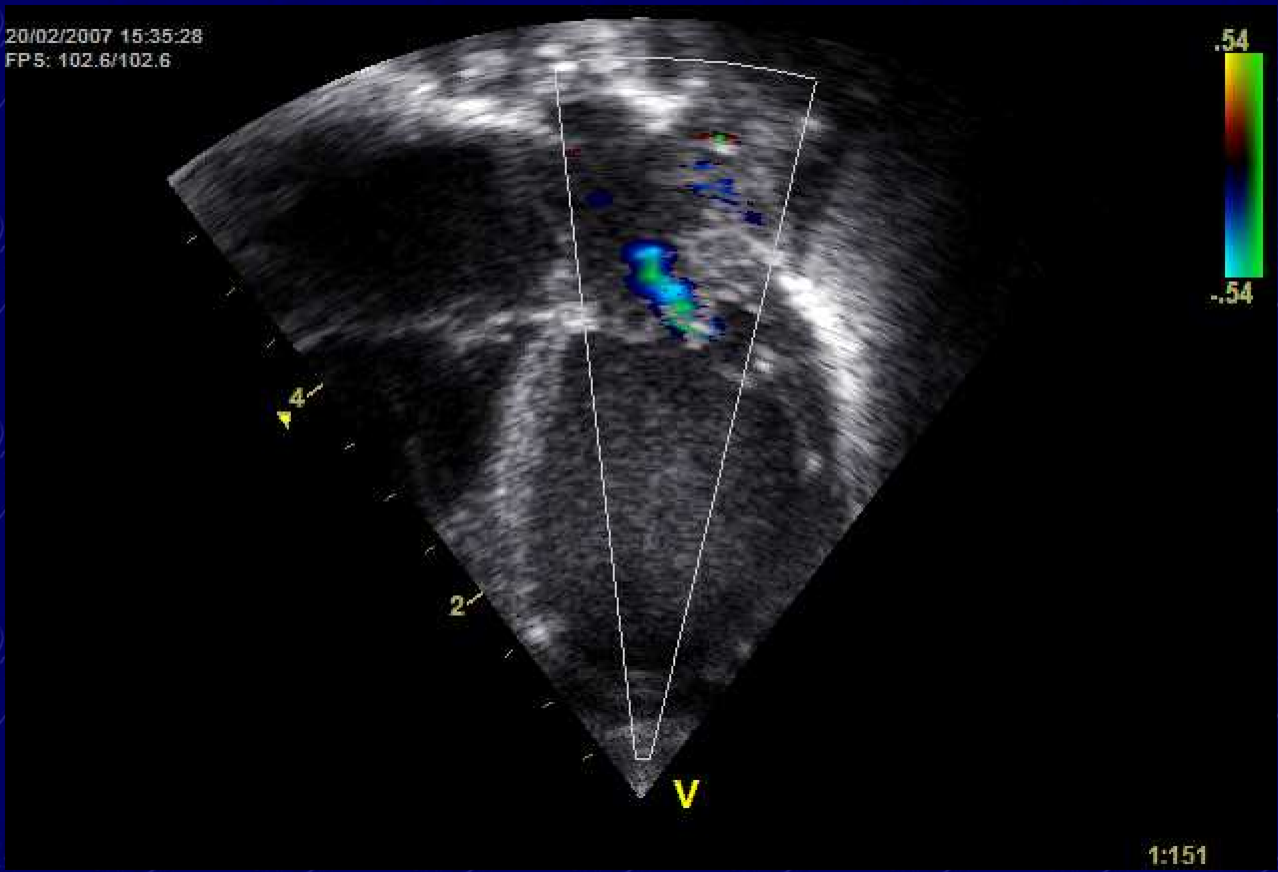
CRITICAL NEONATAL AS

A. Normal volume of LV — normal function / dysfunction



CRITICAL NEONATAL AS

B. Dilated LV, decreased EF



Classification by Doppler-derived gradient is not valid

CRITICAL NEONATAL AS

C. “small left ventricle”

CRITICAL NEONATAL AS

Small ventricle in critical aortic stenosis – How small?

Optimal treatment for best survival and long-term prognosis?

Biventricular?	Univentricular?
Intracardiac and AO arch repair so that left ventricle supports systemic circulation	•"Fontan" palliative operation (Norwood I, BCA, TCPC) •Neonatal heart transplantation

Can we predict the long term outcome? Not always!

CRITICAL NEONATAL AS

ECHO criteria for biventricular repair

LV / RV length	> 0.8
AO annulus	> 3.5 cm/m2
MV area	> 4.75 cm2/m2
LV mass	> 35 g/m2

Rhodes LA: Circulation 1991;84:2325-35

LV length	> 25mm
AO annulus	> 5mm
MV orifice	> 9mm
LV vol.	> 20 ml/m2

Leung MP: J Thorac Cardiovasc Surg 1991;101:526-35

CRITICAL AORTIC STENOSIS IN THE NEONATE: A MULTI-INSTITUTIONAL STUDY OF MANAGEMENT, OUTCOMES, AND RISK FACTORS

Initially intended BIV (116) - 5yr survival rate 70%

Initially UNIV (179) - 5yr survival rate 60%

Independent risk factors associated with greater survival benefit for UNIV versus BIV:

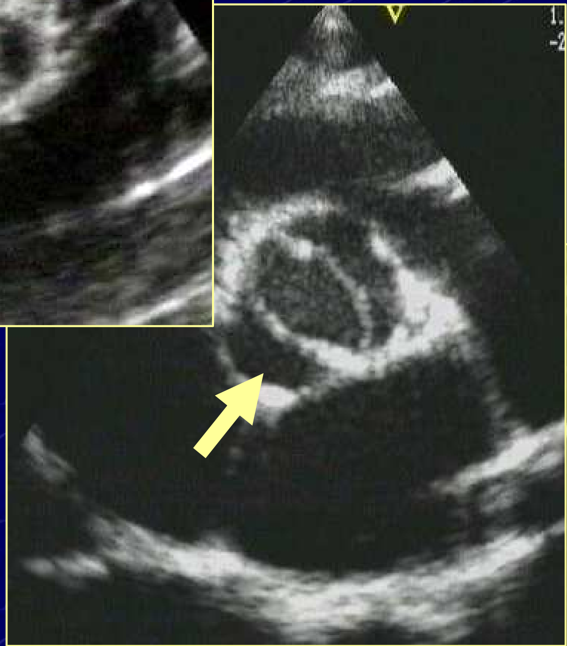
- Younger age
- Lower AOV Z-score
- Shorter LV length
- Higher EFE
- Absence TR
- Larger ascendent AO

Lofland, GK, J Thorac Cardiovasc Surg 2001

http://www.ctsnet.org/aortic_stenosis_calc/ ← Calculator

Aortic valve anatomy

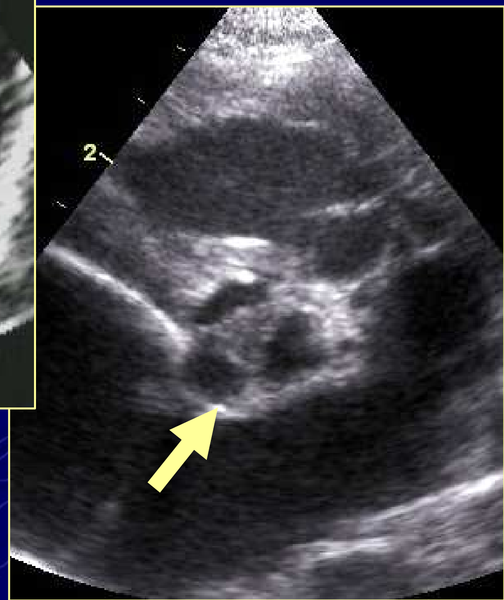
Mono-
cuspid



Bicuspid

Functionally
bicuspid

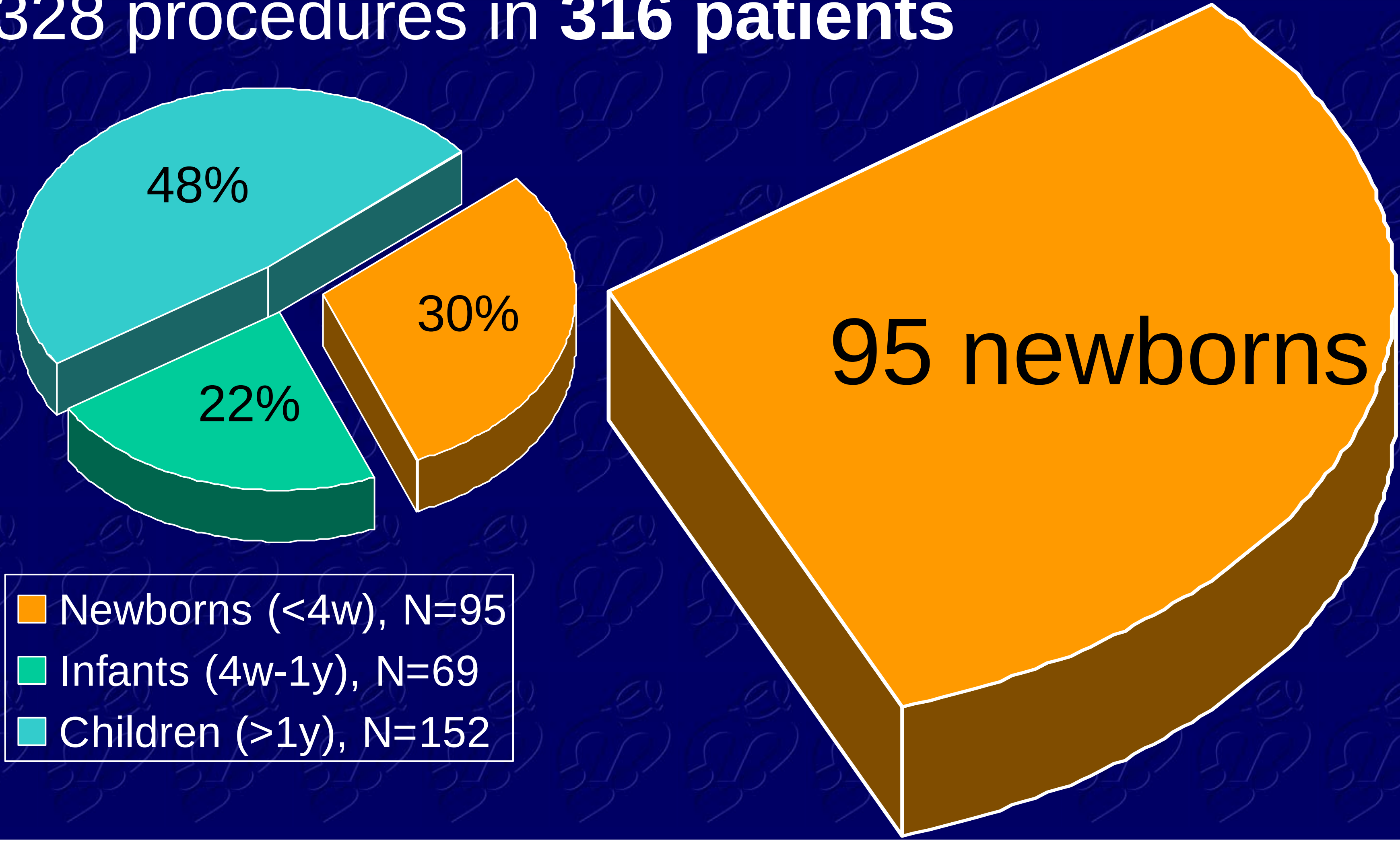
Tricuspid



CRITICAL NEONATAL AS

Aortic valvuloplasty. Kardiocentrum, Prague
1987 - 2005

328 procedures in **316 patients**



Aortic valvuloplasty in newborns

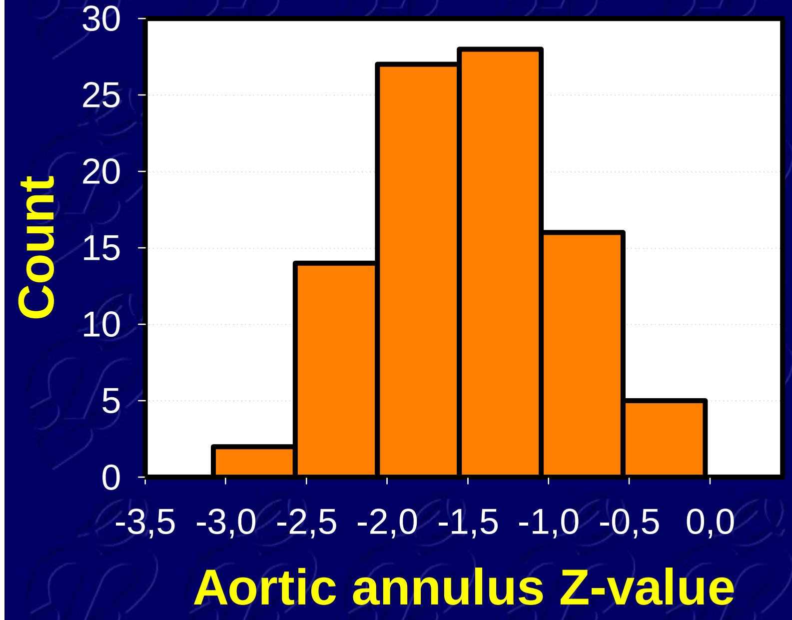
INDICATIONS

Indication	N	%
Peak gradient >70 mmHg	52	56.5
Left ventricular failure	34	37.0
PDA dependency	6	6.5
Total	92	100.0

Aortic valvuloplasty in newborns

AORTIC VALVE ANATOMY

Aortic annulus diameter: -3.08 to -0.02 Z (-1.51 ± 0.57 Z)

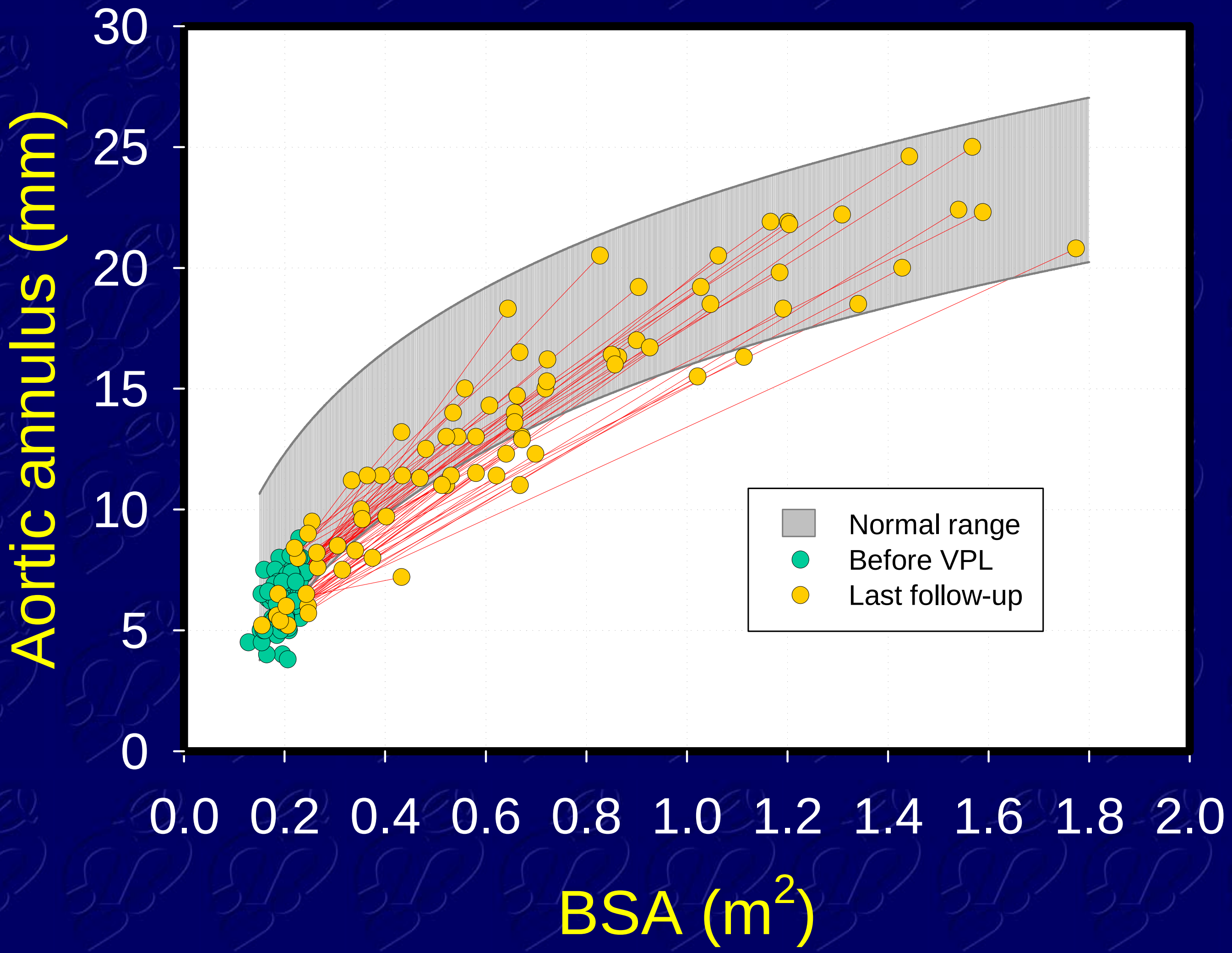


< -2 Z in 23 patients (25%)

Valve	N	%
Unicuspid	36	39.1
Bicuspid	45	48.9
Tricuspid	11	12.0
Total	92	100

Aortic valvuloplasty in newborns

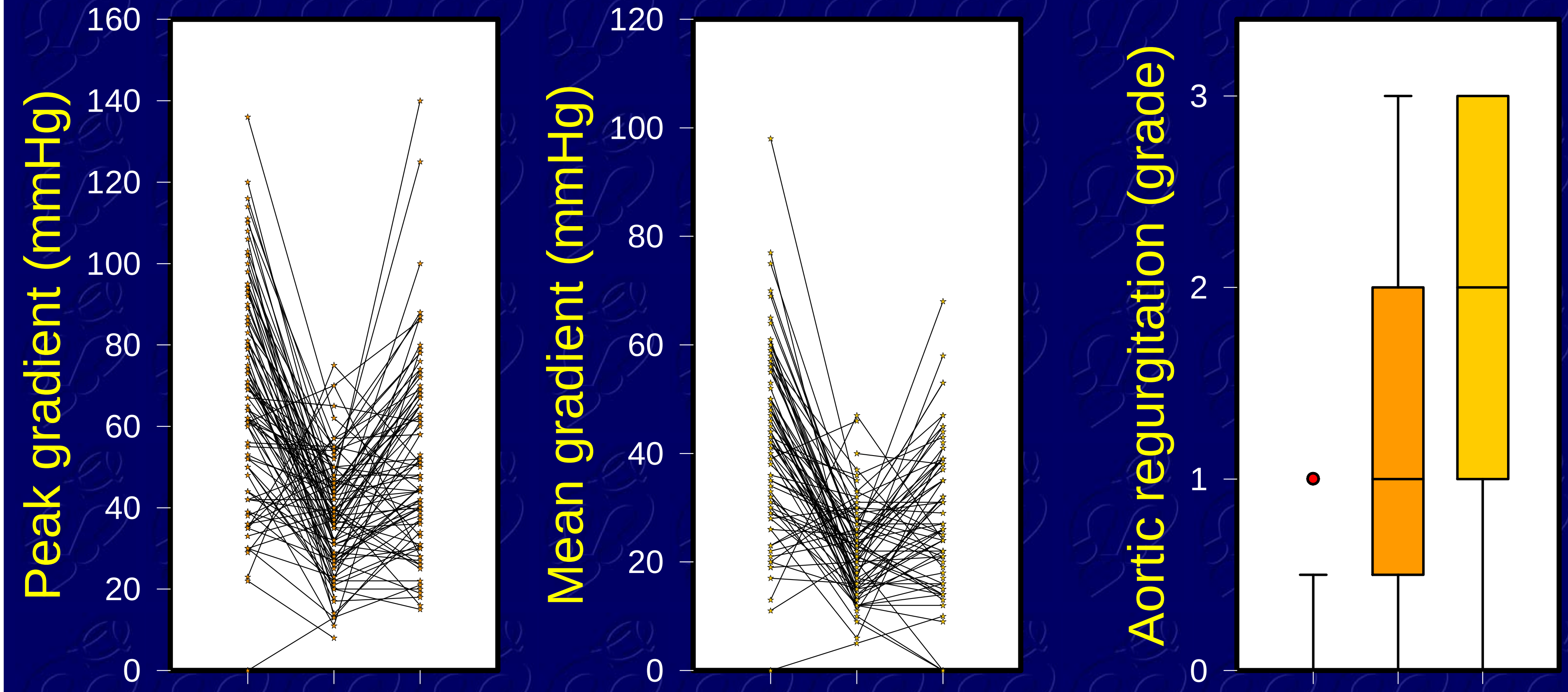
AORTIC ANNULUS GROWTH



Aortic valvuloplasty in newborns

AO GRADIENT & REGURGITATION

Before VPL, after VPL, and at latest follow-up



All differences significant ($p<0.05$)

Aortic valvuloplasty in newborns

INDEPENDENT RISK FACTORS

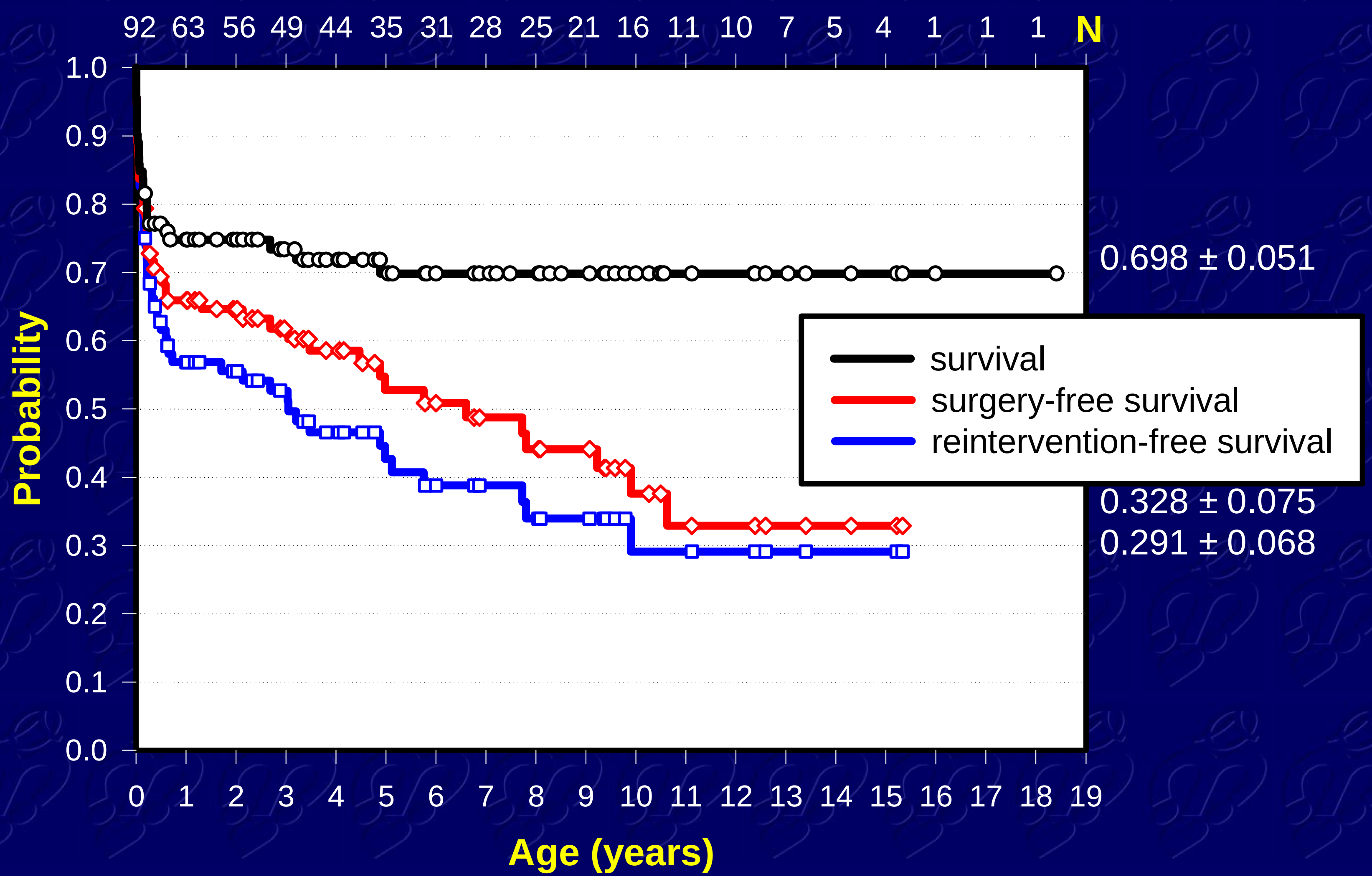
(multiple logistic regression)

End-point	Risk factor*	Odds (95%CI)	P
Death	LVF / PDA depend.	3.83 (2.11 – 6.96)	<0.001
Death or reintervention	LVF / PDA depend.	1.96 (1.08 – 3.54)	0.026
	AO annulus Z	0.19 (0.07 – 0.58)	0.003
	Mitral insufficiency	1.87 (1.11 – 3.15)	0.019

*Potential risk factors tested: sequential # of VPL, doctor, BSA, indication for VPL (gradient or LVF/PDA dependency), FE, COA, MS, PH, AO annulus Z, # of cusps, balloon to annulus ratio, LV volume Z, mitral annulus Z, LVSF, AI grade, MI grade

Aortic valvuloplasty in newborns

ACTUARIAL PROBABILITIES

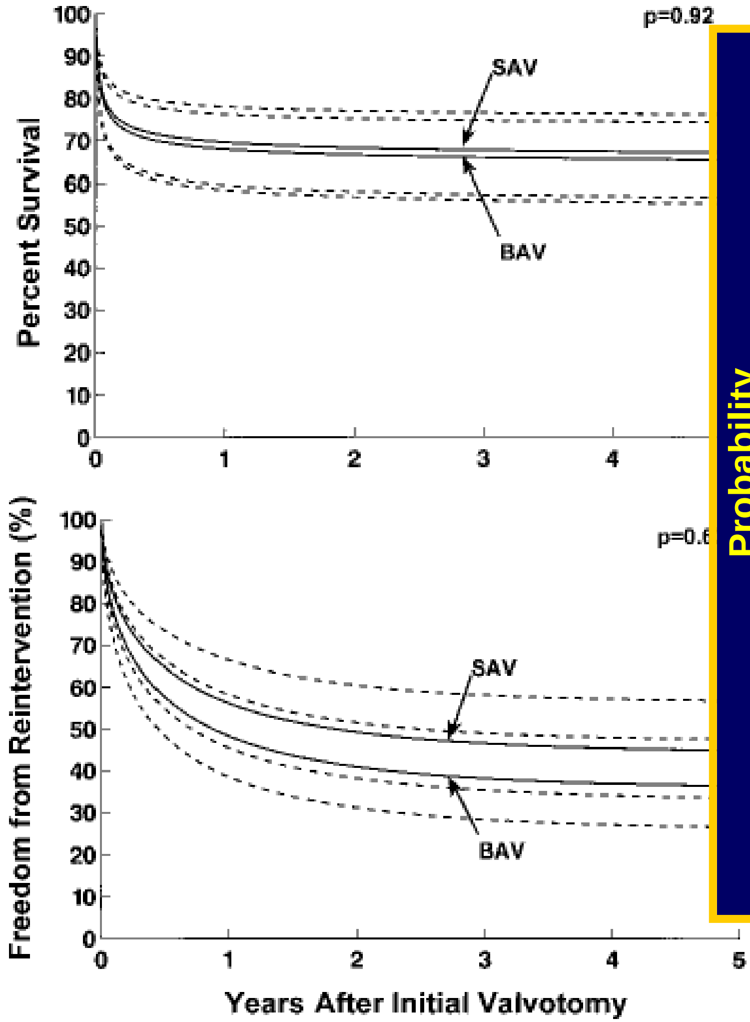


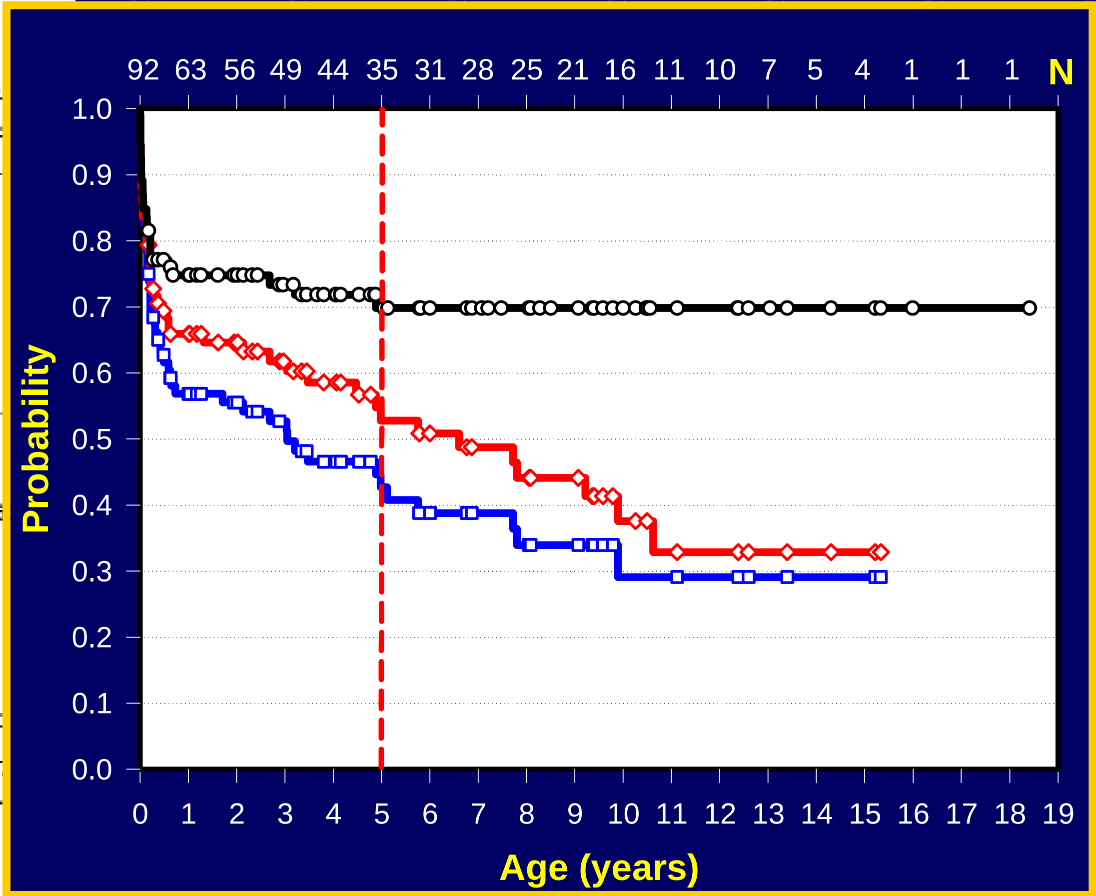
Are Outcomes of Surgical Versus Transcatheter Balloon Valvotomy Equivalent in Neonatal Critical Aortic Stenosis?

Brian W. McCrindle, MD, MPH; Eugene H. Blackstone, MD; William G. Williams, MD; Rekwan Sittiwangkul, MD; Thomas L. Spray, MD; Anthony Azakie, MD; Richard A. Jonas, MD; and the members of the Congenital Heart Surgeons Society

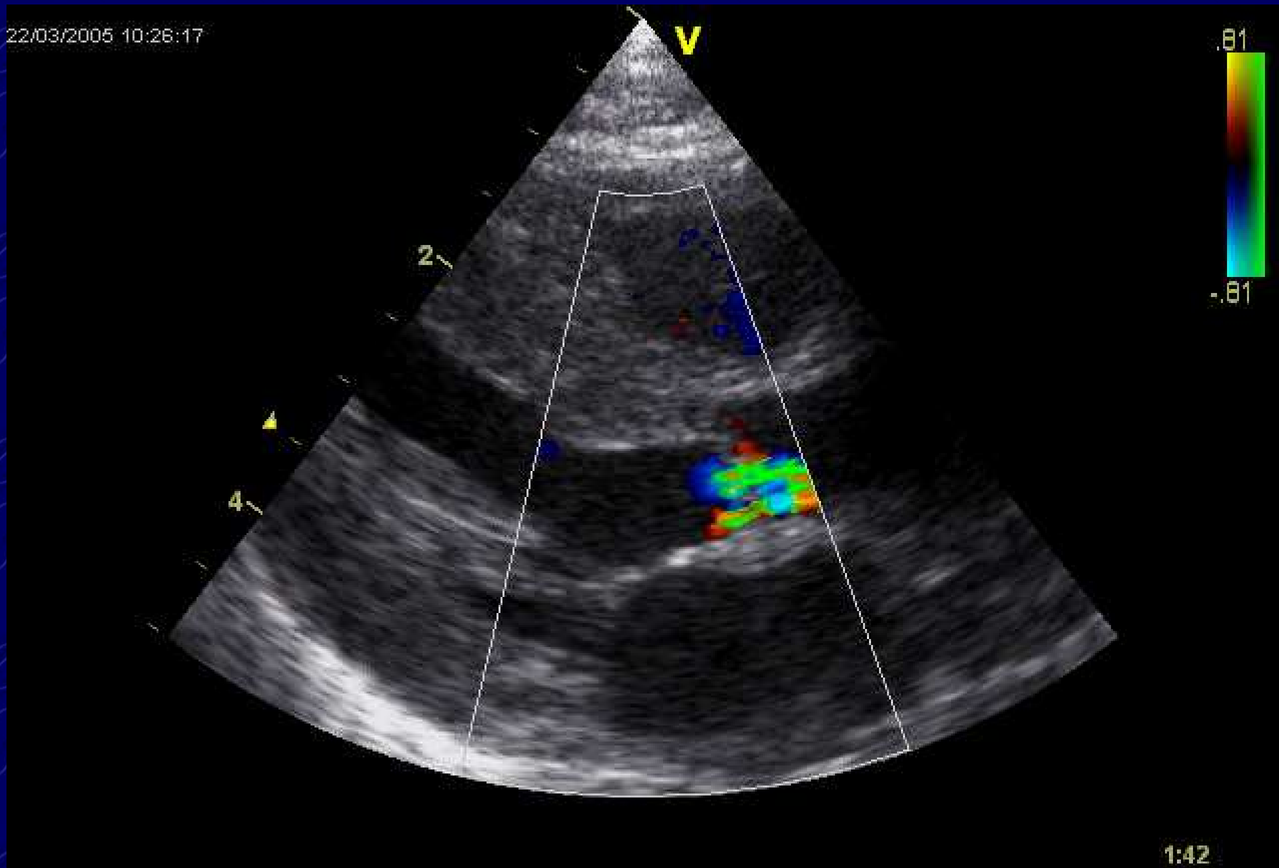
(*Circulation*. 2001;104[suppl I]:I-152-I-158.)

University of Toronto, Hospital for Sick Children, Toronto, Cleveland Clinic Foundation, Children's Hospital of Philadelphia, Children's Hospital, Boston

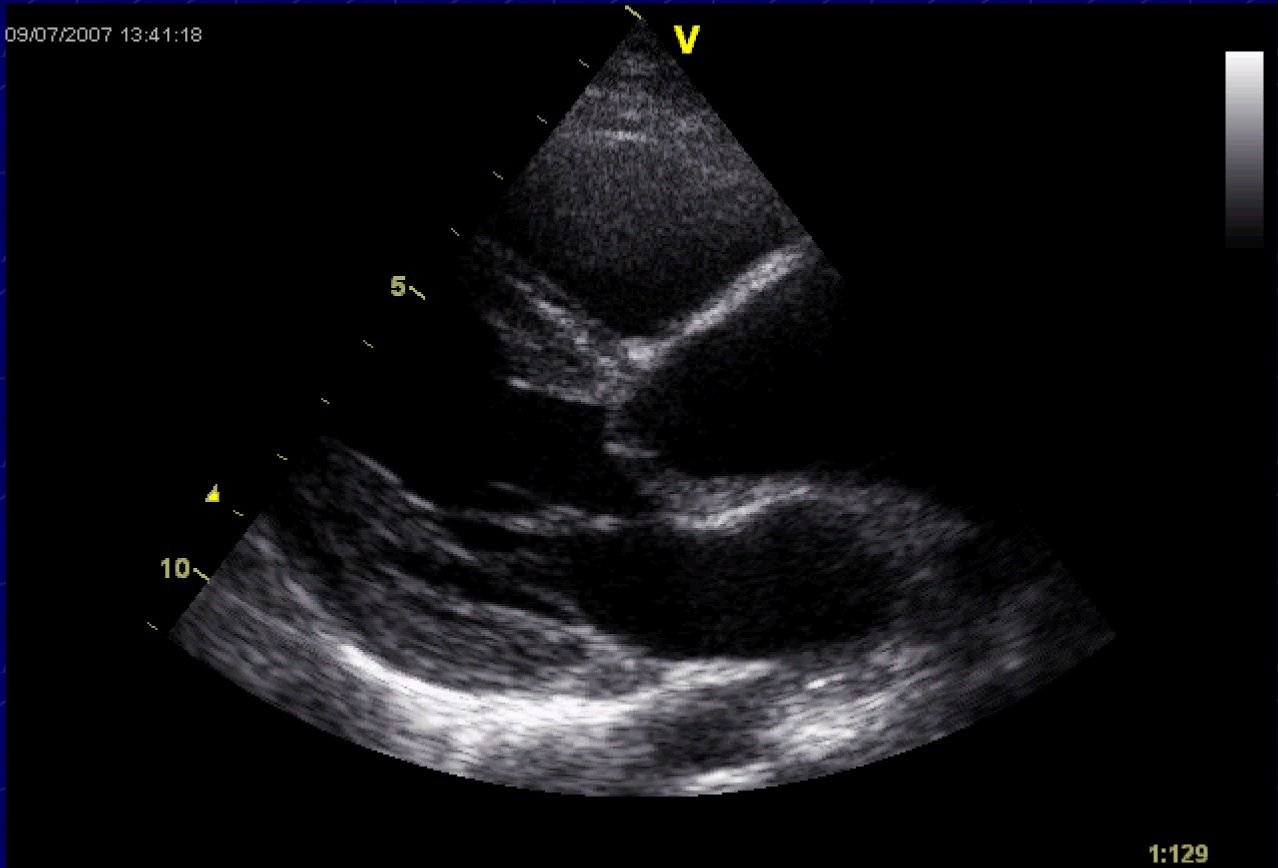




DOMED AORTIC VALVE IN INFANCY



DOMED AORTIC VALVE IN INFANCY

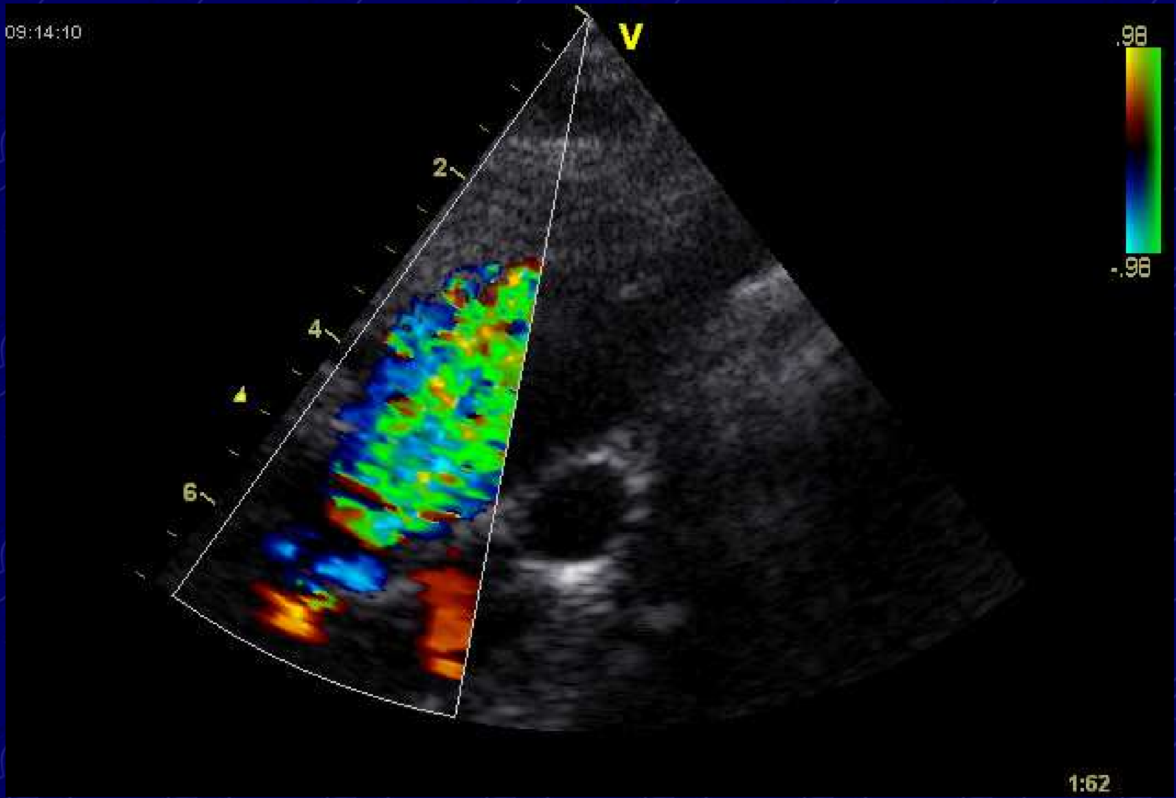


Dilation of AO root → turbulent flow
→ developmental abnorm.

DOMED AORTIC VALVE IN INFANCY

CW Doppler

Rule: US beam nearly parallel to the direction of flow



Transducer position:

- Suprasternal notch
- Apical „5-ch“
- Right parasternal

Underestimation of the true velocity can occur if the intercept angle > 20 gr
Over/underestimation unpredictably when angle correction is used to calculate the flow velocity.

DOMED AORTIC VALVE IN INFANCY

Classification of AS

By peak Doppler velocity across the AOV

- mild < 3m/s (< 36 mmHg)
- moderate 3-4 m/s (36-64 mmHg)
- severe > 4 m/s (> 64 mmHg)

Sleeping/anxious,output … !

By assessment of valve area

- mild > 1.4 cm^2/m^2
- moderate 1.0-1.4 cm^2/m^2
- severe < 1.0 cm^2/m^2

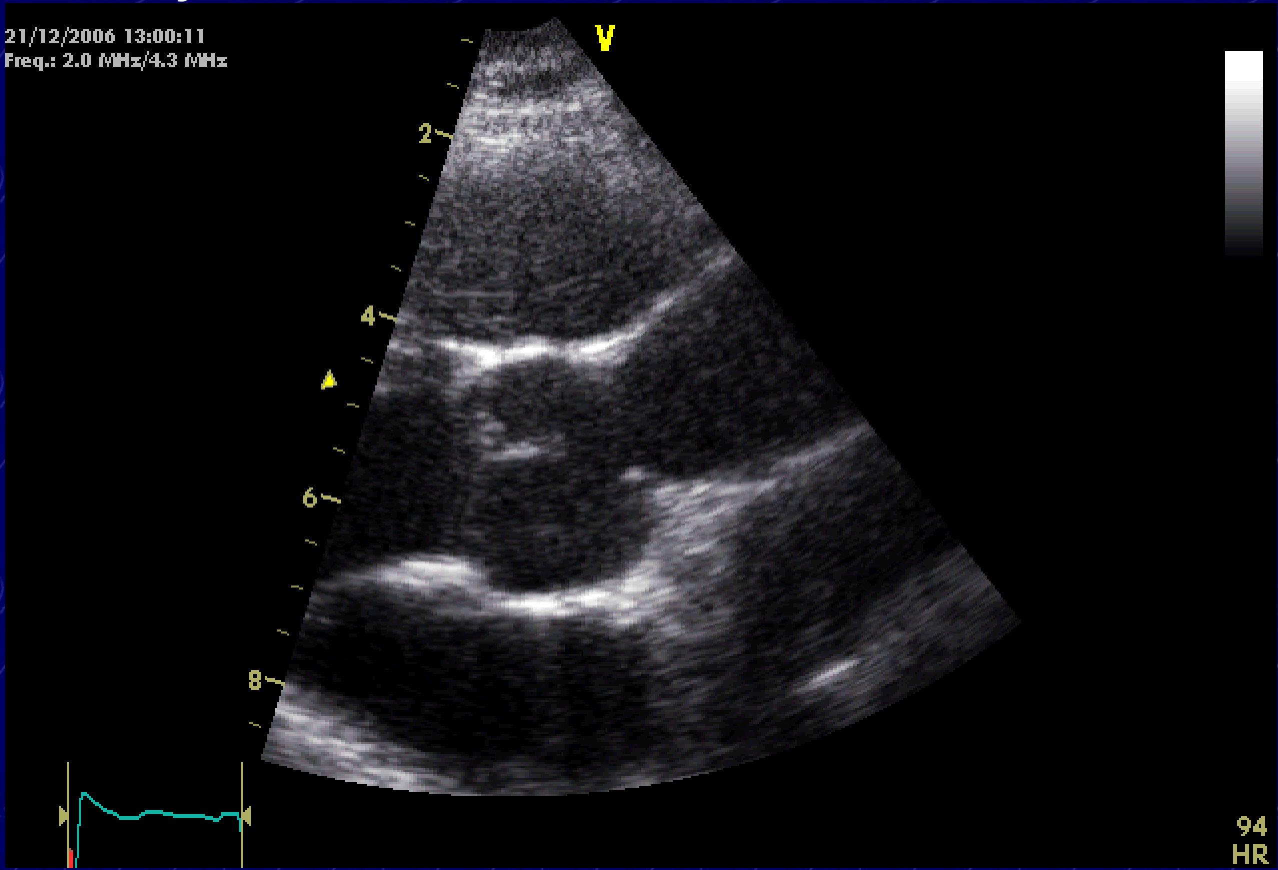
Continuity eqaution: Zoghbi WA, Circulation 1986
3D: Suradi H, Echocardiography 2010

Always consider clinical condition, ECG…

SUPRAVALVAR AORTIC STENOSIS

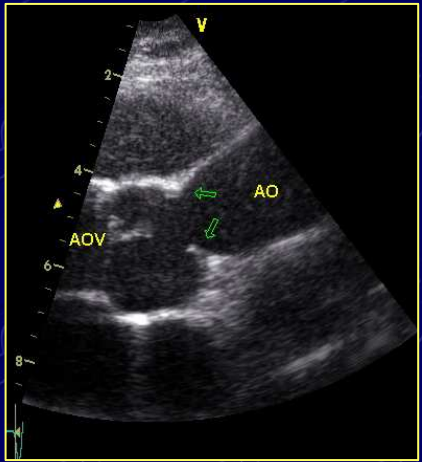
1-2% of AS

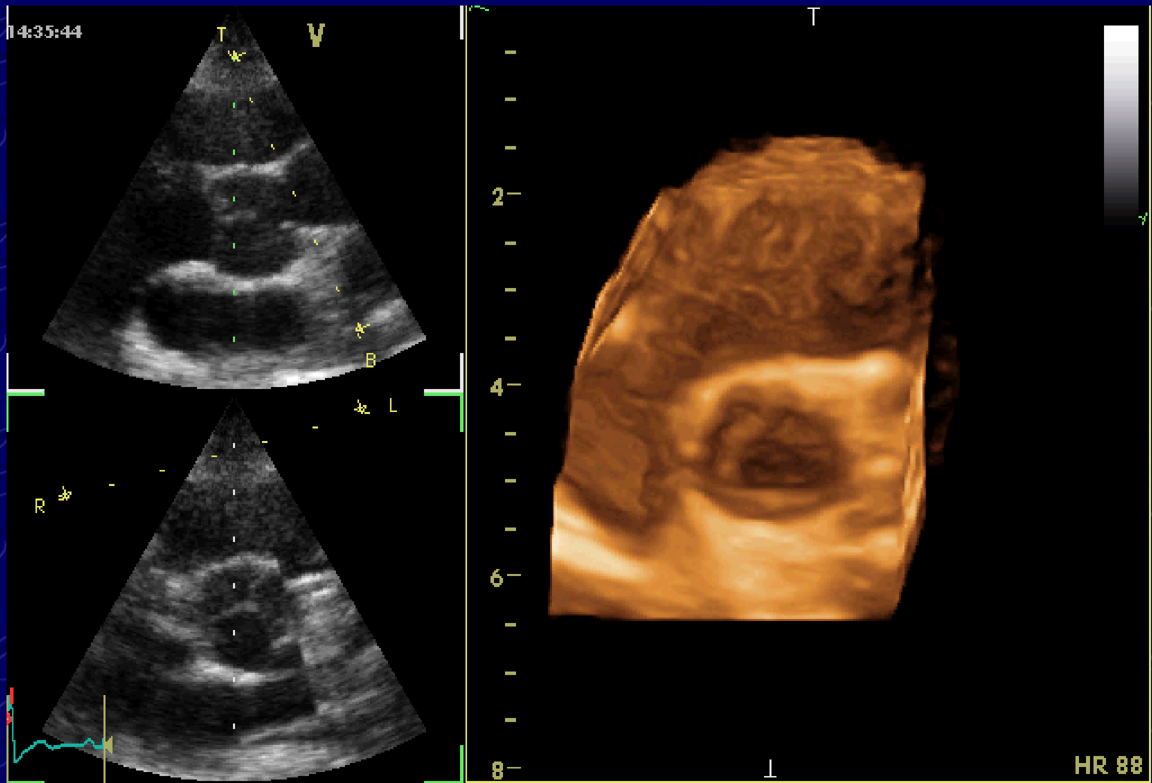
- familial
- disorders of Ca metabolism (Williams sy)
- sporadically in normal children



„hour glass“ variety

RT – 3DE: Supravalvar obstruction



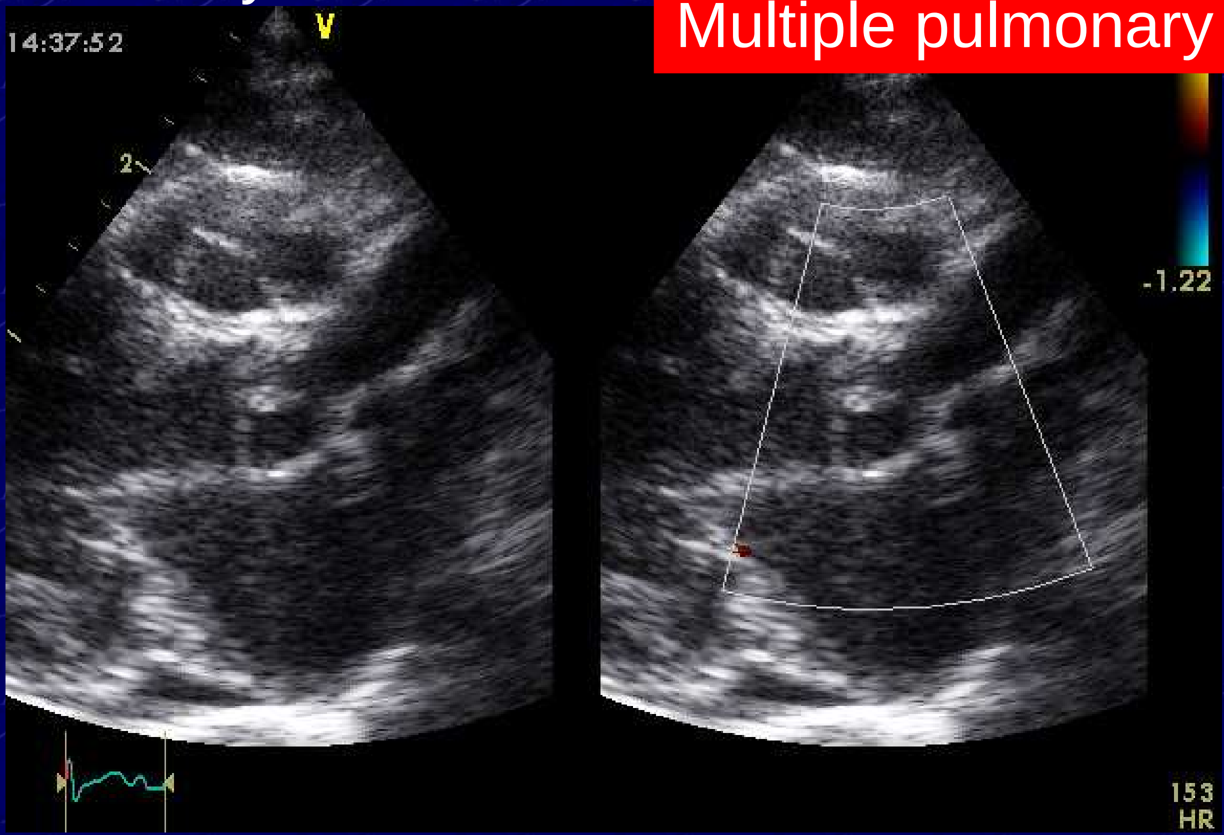


SUPRAVALVAR AORTIC STENOSIS

1-2% of AS

- familial
- disorders of Ca metabolism (Williams sy)
- sporadically in normal children

Multiple pulmonary stenosis – 20%



Tubular/diffuse

Surgery: Doppler grad. > 85 mmHg

Tani LY, Am J Cardiol, 2000

AORTIC REGURGITATION

- Qualitative and/or semi-quantitative assessment

✓ Diastolic run-off: Ascendent … Abdominal AO
✓ Jet in CFM: length, width, area, VC-W
✓ Pressure 1/2 time / Deceleration Slope

- Quantitative assessment

✓ Regurgitation fraction

$$RF = LVOT\ (VTI \times \pi r^2) - MV\ (VTI \times \pi r^2)$$

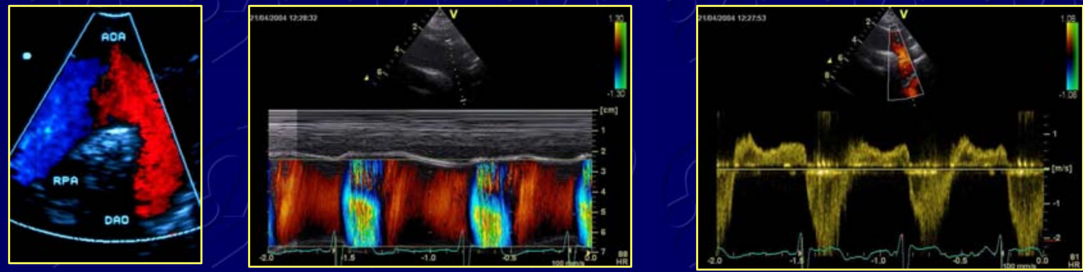
- Left ventricular assessment:

✓ Size / volume, function

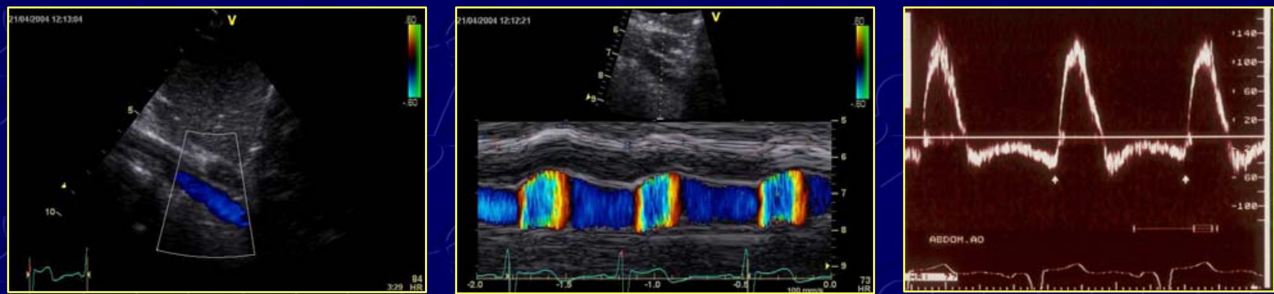
(Zoghbi et al: Guiedeline recommending AR, J Am Soc, 2003)

Aortic regurgitation – Reversal flow

Moderate AR:



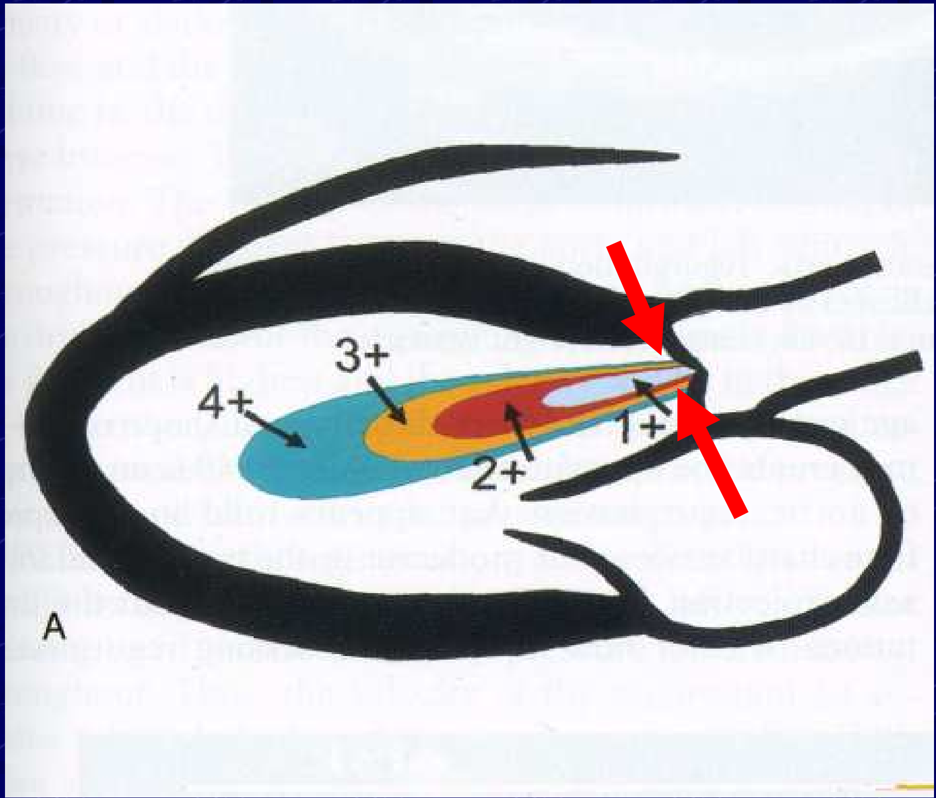
Significant AR

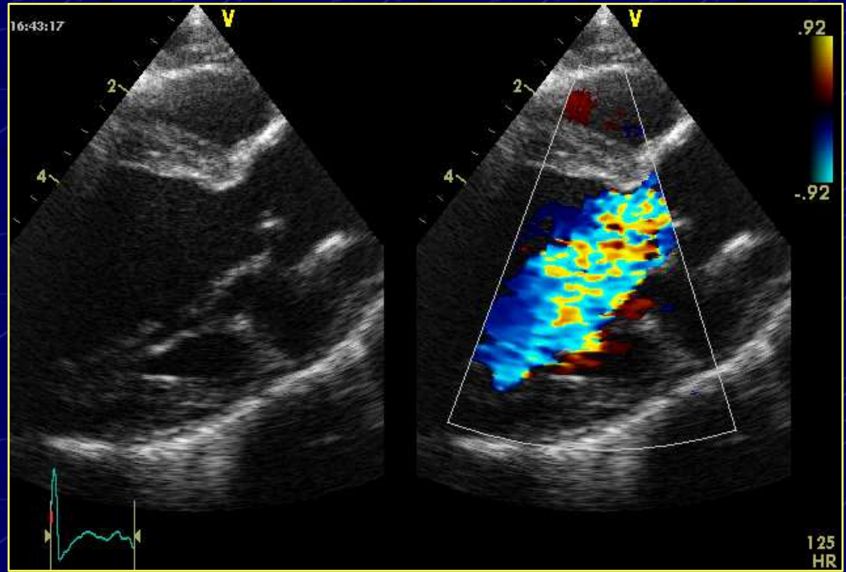


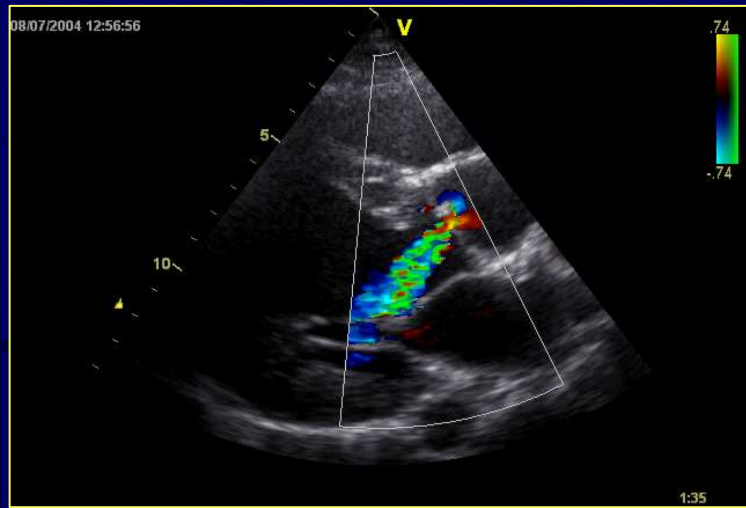
Sensitivity	100%
Specificity	87%
Positive p.v.	67%
Negative p.v.	100% (versus angio)

Tani LY, Am J Cardiol 1997

Aortic regurgitation – Jet width

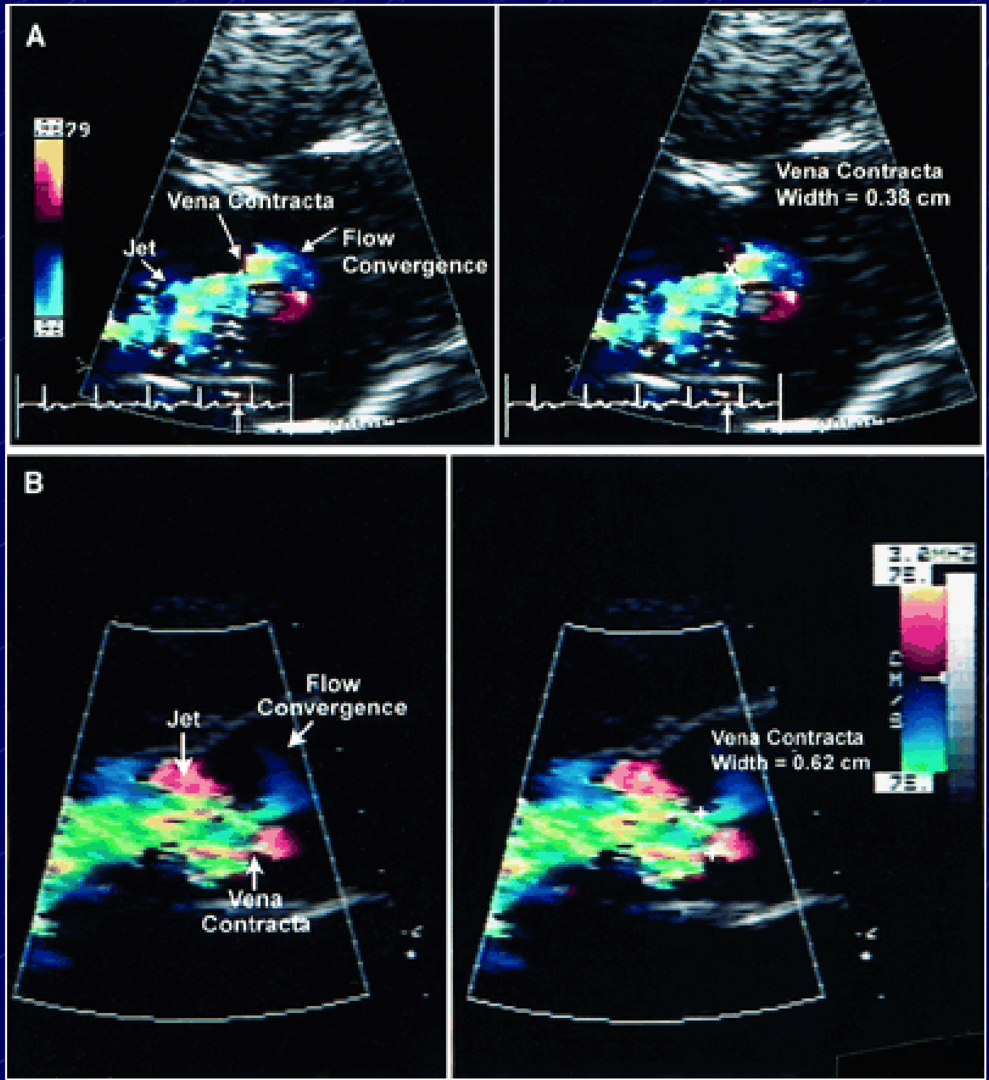






AR jet width < 30%	Mild
AR jet width 30-60%	Moderate
AR jet width > 60%	Severe

Aortic regurgitation – Vena Contracta



Tribouilloy CM, Circulation 2000

Narrowest portion of a jet at the level of the AOV

(Effective regurgitant orifice area)

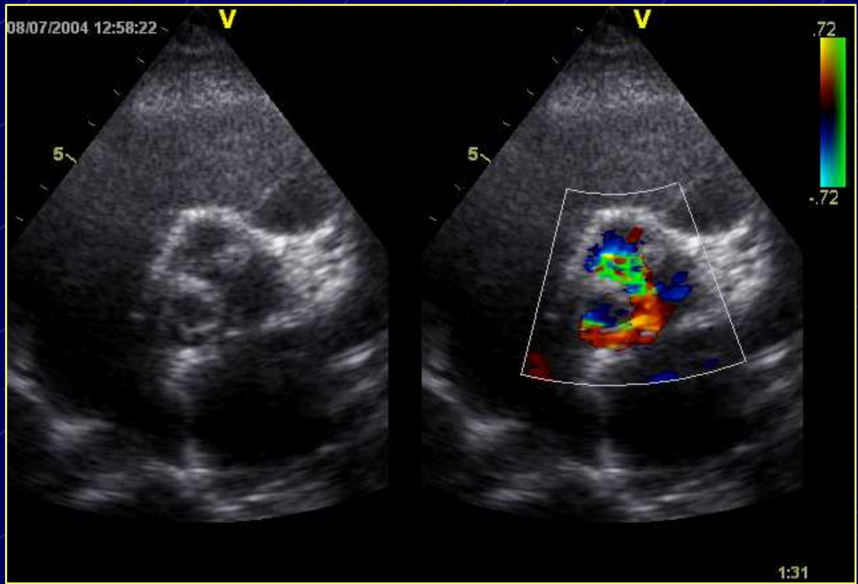
VC-W > 6mm Severe

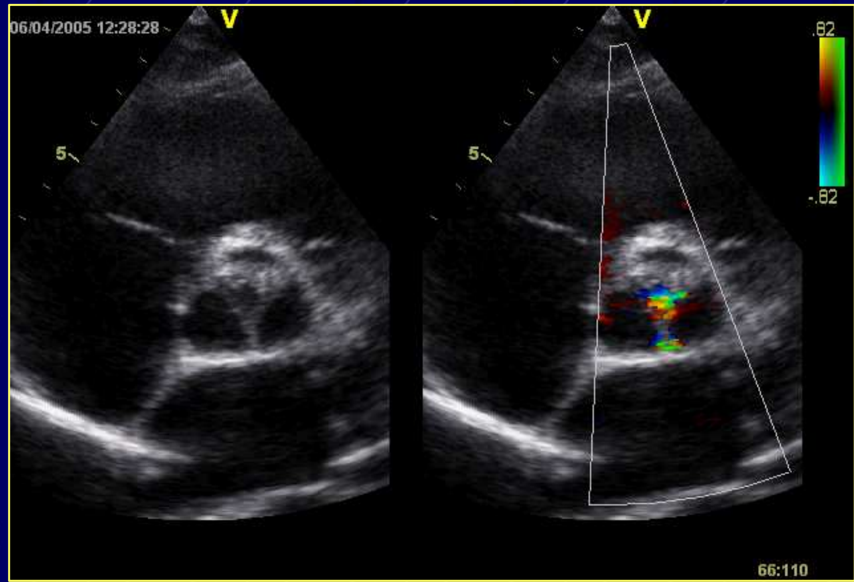
Sensitivity 95%

Specificity 90%

VC-W < 3mm Mild

Aortic regurgitation – Jet area





< 0.35	Mild
0.35 – 0.50	Moderate
> 0.50	Severe

AORTIC REGURGITATION

Limitations colour flow

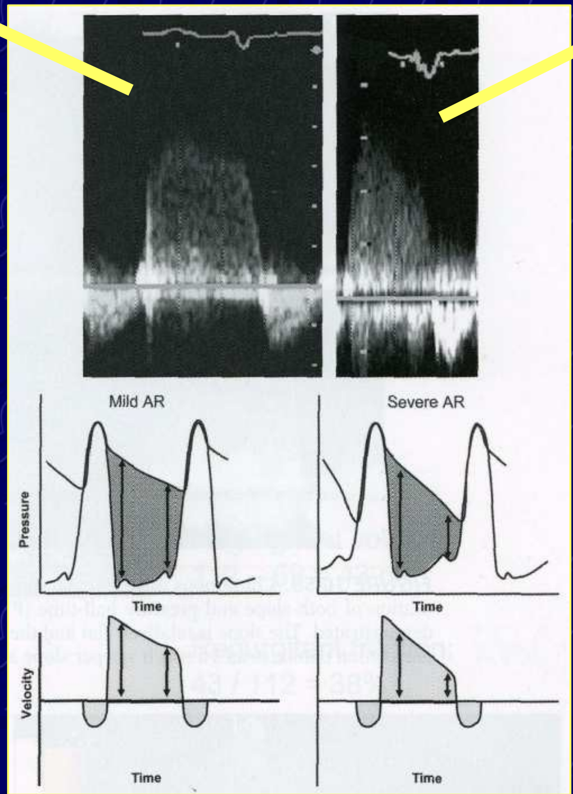
- Eccentric jets
- Failure to appreciate 3D nature jet
- Instrument settings

  → gain, scale, frequency, wall filters.
- Acoustic window (adults)

Aortic regurgitation – Jet deceleration

Mild AR

- Slow deceleration
- LV pressure rises slowly
- Aortic diastolic pressure maintained



Severe AR

- Rapid deceleration
- LV pressure rises rapidly
- Aortic diastolic pressure drops

Mild AR

Slope = 230 cm/s^2

P1/2t = 560msec

Severe AR

Slope = 460cm/s^2

P1/2t = 180msec

Velocity decay is linearly related to the severity of AR

Mild AR	Slope < 200 cm/s^2	P1/2t > 400msec
Moderate AR	Slope 200-400 cm/s^2	P1/2 400 – 200msec
Severe AR	Slope > 400 cm/s^2	P1/2 < 200msec

Chronic AR in asymptomatic patients- - Criteria for intervention:

- LV dysfunction at rest (**EF < 50%)**

- Normal LV systolic function but with severe LV dilatation (**LVEDd >75 mm** or **LVESd >55 mm**)

- Normal systolic function at rest (**ejection fraction >0.50** with **LVEDd <75 mm** or **LVESd <55 mm**) but with decline in ejection fraction during
  - Radionuclide exercise
  - Stress Echocardiography

ACC/AHA Recommendations, Circulation 1998

AR in asymptomatic children

N = 49, mean age 13.9, all AVR (49% Rheumatic)
Mean FU 3.3 (+/-2.1)yrs

I. LVEDd > 4 z-score / II. LVEDd < 4 z-score

Multivariate predictors of clinical outcome:

Preoperative	NYHA III-IV	Postop EF	Postop LVEDd
LVEF < 50%	0.03	0.008	0.01
LVEDd > 4	ns	0.05	0.003
LVESd > 4	0.02	0.05	0.007

Tafreshi Pediatr Cardiol 2005

Chronic AR in asymptomatic patients

…..patients with evidence of LV systolic dysfunction, even if asymptomatic, should undergo AVR before more severe symptoms or more severe ventricular dysfunction develop

ACC/AHA Recommendations, Circulation 1998