Bicuspid aortic valve

BAV

ZBIGNIEW GĄSIOR
KATEDRA i KLINIKA KARDIOLOGII SUM KATOWICE

BAV

- Pathogenesis of the BAV is unknown
- NOTCH-1 mutations
- Unproper structure of the extracellular protein matrix, fibrilin-1 i fibulin deficiency, accelerated apoptosis of the smoth muscle cells, ↑ metaloproteinase, impaired arterial wall structure
- Familial prevalence (possible – 9%): autosomal dominant pattern with with reduced penetrance

BAV - prevalence

- Approximately 1% of the population
- M / K 3 : 1
- Symptoms usually late (after 40 ys)

Diagnostics:

- TTE sensitivity 78%; specificity 96%
- TEE sensitivity 87%; specificity 91%

BAV and associated cv conditions

Condition	Incidence of BAV	Comments
Coarctation of the aorta	50	Increased risk of aortic complications
Turner syndrome	30	Most frequent cardiac abnormality
Sinus of Valsalva aneurysm	15-20	Frequently asympt; most commonly RCC
Ventricular septal defect	30	Significant aortic regurg
Shone`s complex	60-85	Left-ided obstructive lesion
Ascending aortic dilatation	common	BAV most common

Prognosis

90±3% during 20 yrs (Olmstead study – 212 pat)

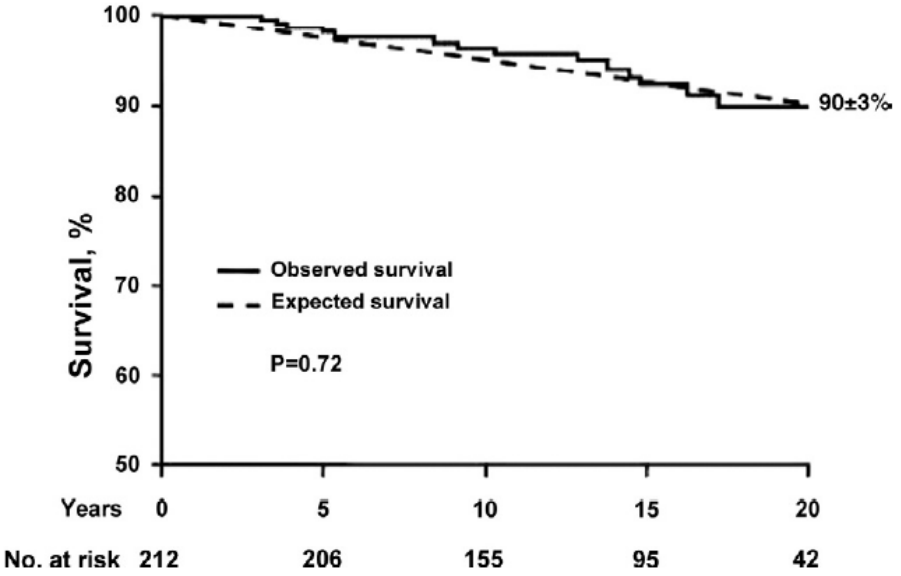
96±1% during 10 yrs (Toronto study – 642 pat)

Complications 25-40% persons in the age 44-52 and severe/moderate stenosis or insufficiency

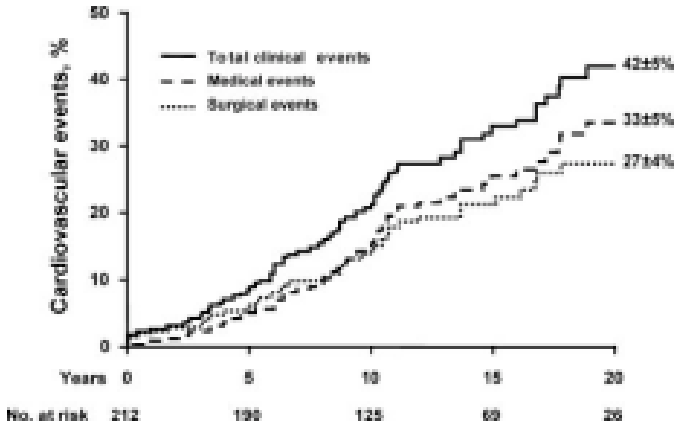
Prognosis in BAV

N=212 asymptomatic BAV, LVEF >50%, AR or AS absent or mild

20-year survival after diagnosis (90%)
– similar to the general population



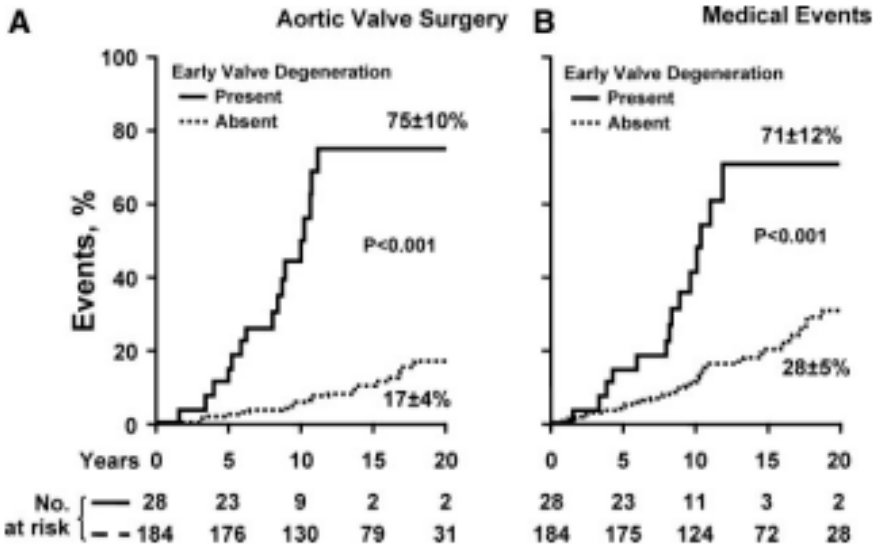
Independent predictors of CVE:
age ≥ 50 years,
valve degeneration after diagnosis



Baseline ascending aorta ≥ 40mm independently predicted surgery for aorta dilatation

Michelena HI et al. Circulation 2008; 117: 2776-84.

Early valve degeneration – risk factor for aortic valve surgery and medical events



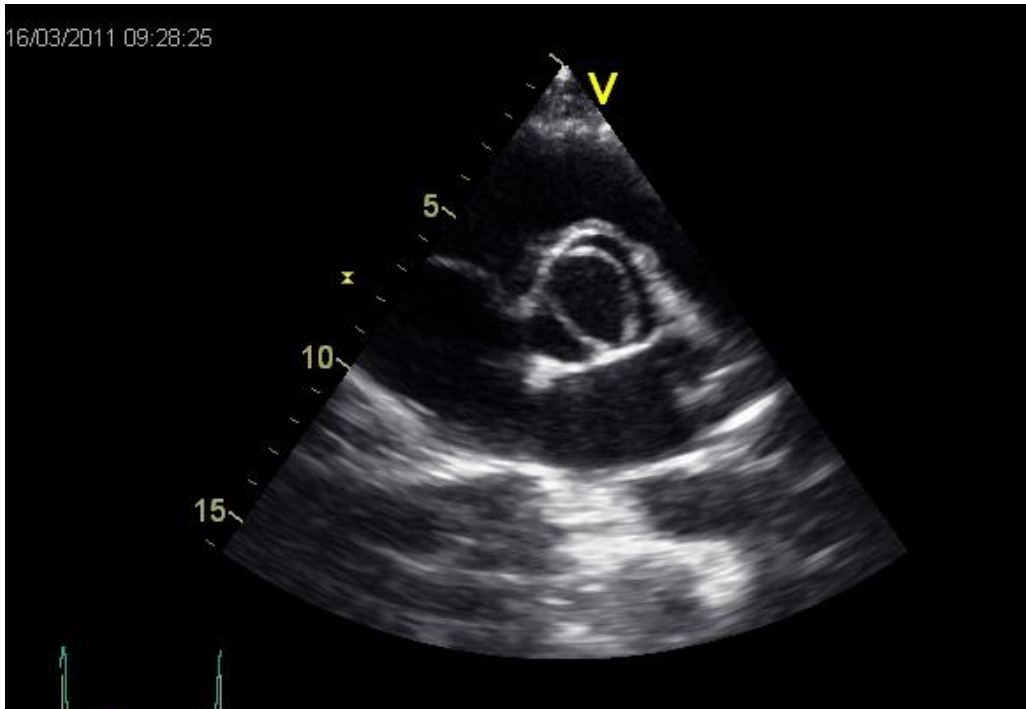
Baseline ascending aorta ≥ 40mm independently predicted surgery for aorta dilatation

N=212 asymptomatic BAV, LVEF >50%, AR or AS absent or mild

Michelena HI et al. Circulation 2008; 117: 2776-84.

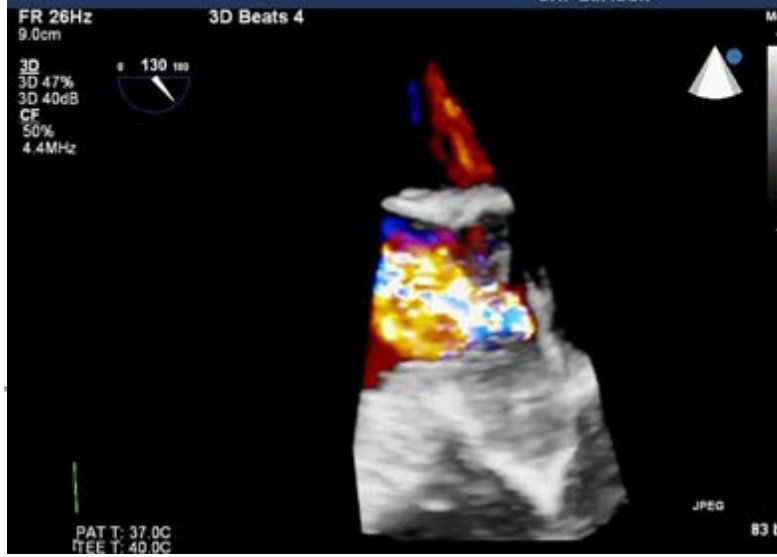
Clinical forms of the BAV

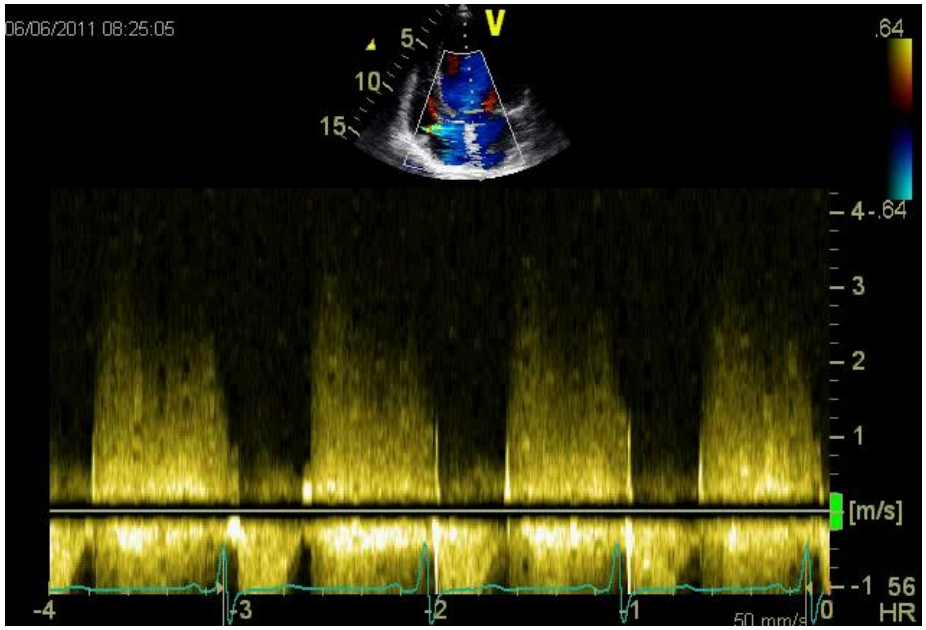
- BAV without disturbances of flow
- Aortic regurgitation
- Valvular stenosis
- Aortic dilatation



Clinical forms of the BAV

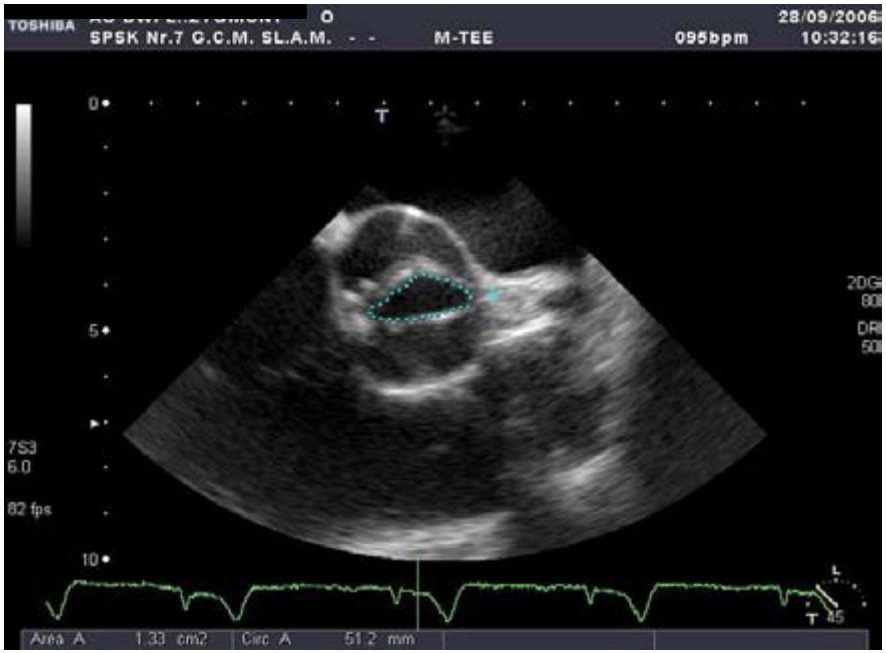
- BAV without disturbances o
- Aortic regurgitation
- Valvular stenosis
- Aortic dilatation





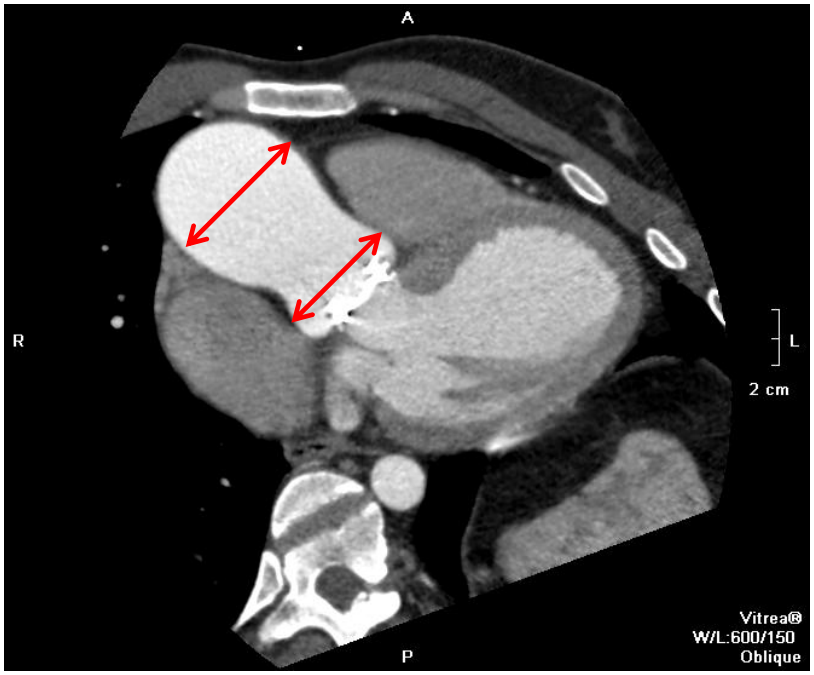
Clinical forms of the BAV

- BAV without disturbances of flow
- Aortic regurgitation
- Valvular stenosis
- Aortic dilatation



Clinical forms of the BAV

MSCT



Aortic root dilatation – max. 44mm
asc. ao. – max. 67mm

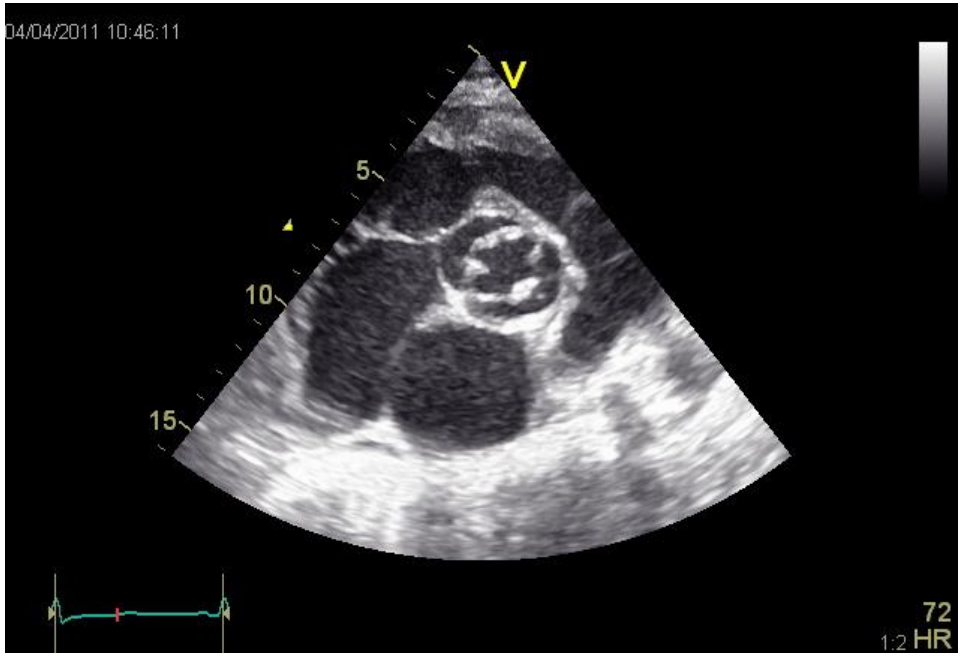
BAV: risk of aortic complication

- Expansion rate > 5mm/year
- Aortic stenosis
- Severe valvular dysfunction
- Hypertension
- Aortic coarctation
- Family history of aortic aneurysm complications

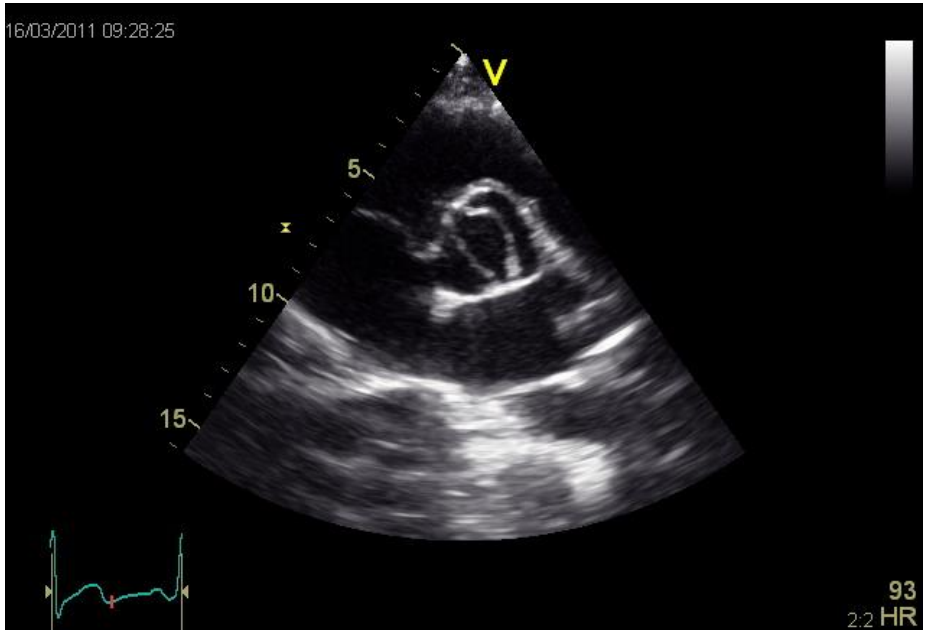
Morfologic pattern BAV

Right – left (12%)

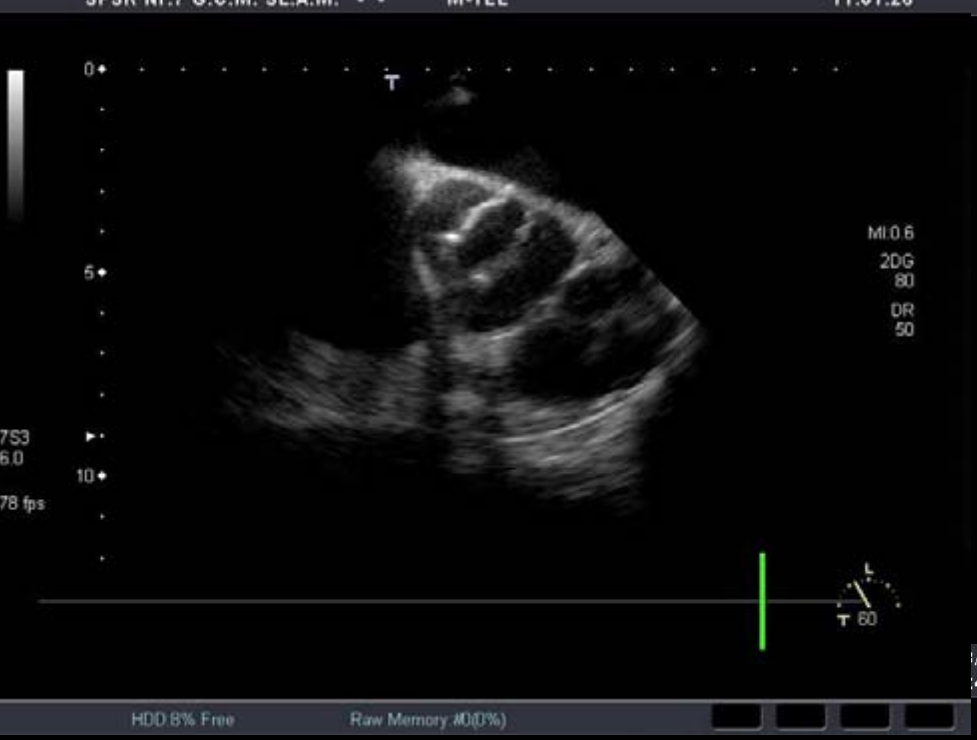
Anterior – posterior (86%)

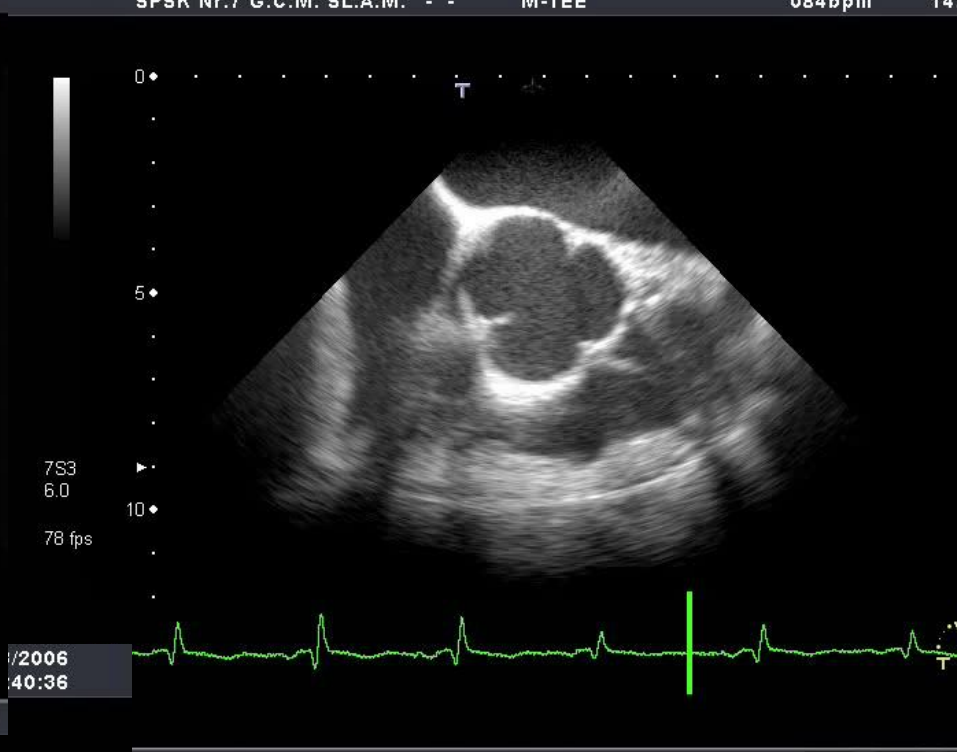


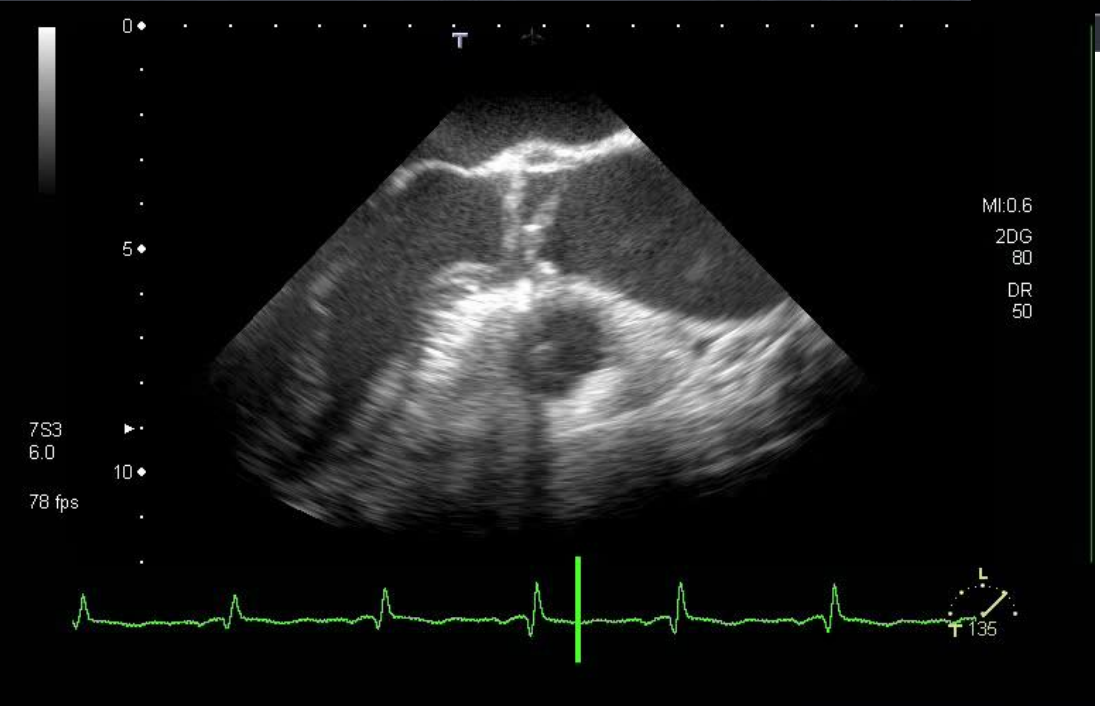
- **Right-left pattern**



- **Anterior-posterior pattern**



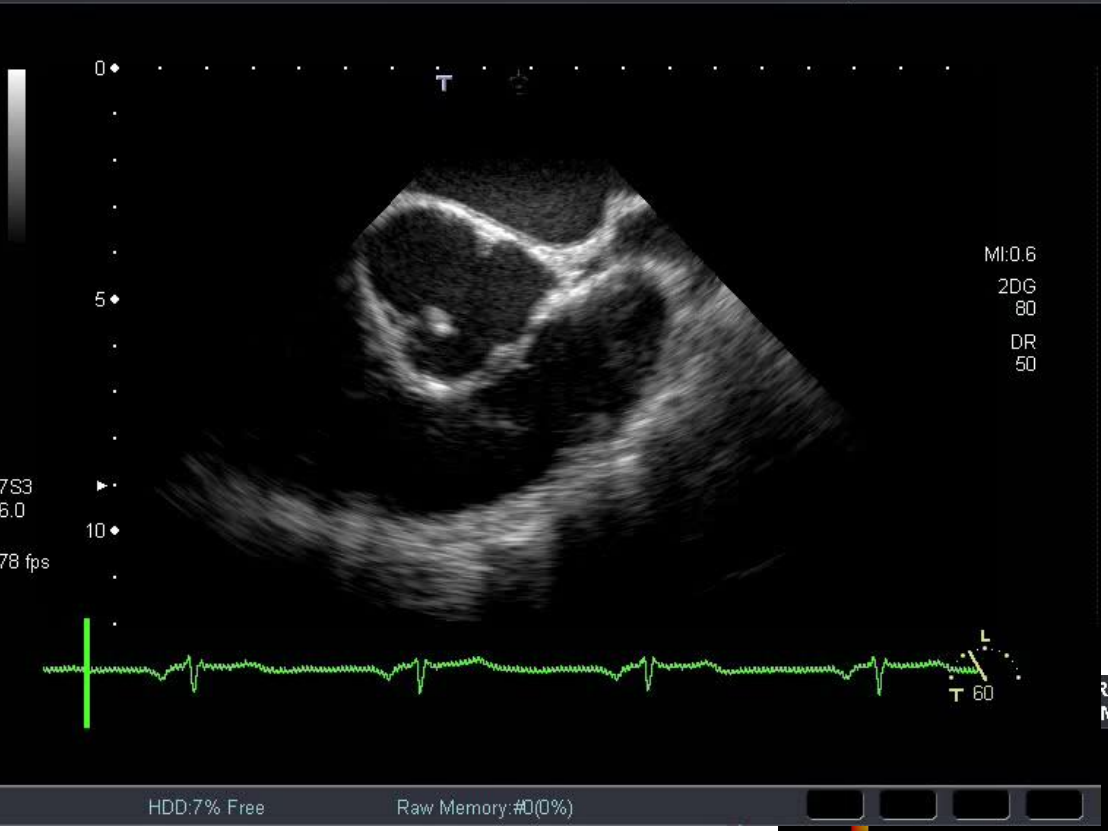


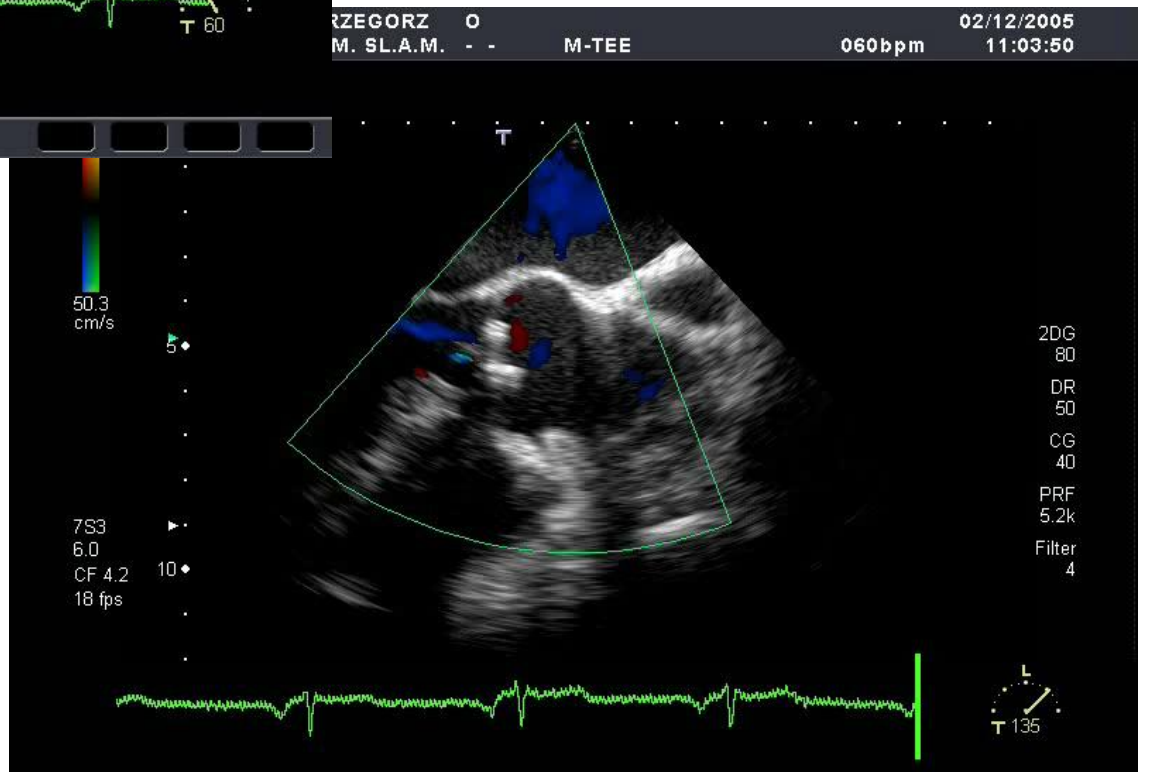


Anterior-posterior pattern

TEE

Right-left pattern TEE AS + AR





Summary

- BAV – one of the most common CHD affecting approximately 1% of the population
- Familial occurence in 9% of first-degree relatives
- Most pts with BAV will require surgical therapy for the valve and/or the aorta during their lifetime
- After BAV replacemment the patient is still at risk for aortic root complications