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, fibrillin-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

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

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

Clinical forms of the BAV

- BAV without disturbances of flow
- Aortic regurgitation
- Valvular stenosis
- Aortic dilatation
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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
Morphologic 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 occurrence 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 replacement the patient is still at risk for aortic root complications