Left ventricular outflow tract obstruction

Echocardiography

V. Tomek, J. Marek, O. Reich, 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
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)
Prenatal diagnosis

Septal buldge
Fibrous shelf

RV

LV

AO

LA
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

A

B
SUBVALVAR STENOSIS

VSD, posterior deviation/ muscular outlet IVS
SUBVALVAR STENOSIS

A-V septal defect

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

Normal heart

AVSD
AVSD, fibrous ring
SUBVALVAR STENOSIS

subAO connus  (VIF/IVS)
RT – 3DE: Subaortic conus
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: prevention of recurrence
secondary progressive aortic valve disease
When to operate?

Mean LVOT gradient > 30 mmHg

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

Aim: prevention 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

80.000 children - aortic stenosis

Kardiocentrum, Prague 1991-2006

**AS = 1229**

**Neonates = 525**

critical - 12%
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
A. Normal volume of LV < normal function dysfunction
B. Dilated LV, decreased EF

Classification by Doppler-derived gradient is not valid
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?

Intracardiac and AO arch repair so that left ventricle supports systemic circulation

Univentricular?

• “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

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

Rhodes LA: Circulation 1991;84:2325-35
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/
Aortic valve anatomy

- Mono-cuspid
- Bicuspid
- Functionally bicuspid
- Tricuspid
Aortic valvuloplasty. Kardiocentrum, Prague 1987 - 2005

328 procedures in 316 patients

- 48% Newborns (<4w), N=95
- 30% Infants (4w-1y), N=69
- 22% Children (>1y), N=152

95 newborns
## Aortic valvuloplasty in newborns

### INDICATIONS

<table>
<thead>
<tr>
<th>Indication</th>
<th>N</th>
<th>%</th>
</tr>
</thead>
<tbody>
<tr>
<td>Peak gradient &gt;70 mmHg</td>
<td>52</td>
<td>56.5</td>
</tr>
<tr>
<td>Left ventricular failure</td>
<td>34</td>
<td>37.0</td>
</tr>
<tr>
<td>PDA dependency</td>
<td>6</td>
<td>6.5</td>
</tr>
<tr>
<td><strong>Total</strong></td>
<td>92</td>
<td>100.0</td>
</tr>
</tbody>
</table>
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%)

<table>
<thead>
<tr>
<th>Valve</th>
<th>N</th>
<th>%</th>
</tr>
</thead>
<tbody>
<tr>
<td>Unicuspid</td>
<td>36</td>
<td>39.1</td>
</tr>
<tr>
<td>Bicuspid</td>
<td>45</td>
<td>48.9</td>
</tr>
<tr>
<td>Tricuspid</td>
<td>11</td>
<td>12.0</td>
</tr>
<tr>
<td>Total</td>
<td>92</td>
<td>100</td>
</tr>
</tbody>
</table>
Aortic valvuloplasty in newborns

AORTIC ANNULUS GROWTH

- Aortic annulus (mm)
- BSA (m²)

Normal range

- Before VPL
- Last follow-up
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)

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

*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**

![Graph showing actuarial probabilities with age and survival rates.](image)
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 Sitiwangkul, 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.
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 cm²/m²
- moderate 1.0-1.4 cm²/m²
- severe < 1.0 cm²/m²

Continuity equation: Zoghbi WA, Circulation 1986
3D: Suradi H, Echocardiography 2010
Always consider clinical condition, ECG…
SUPRavalvar Aortic Stenosis

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

1-2% of AS

„hour glass“ variety
RT – 3DE: Supravalvar obstruction
SUPRAVALVULAR AORTIC STENOSIS

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

1-2% of AS

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

Aortic regurgitation – Reversal flow

Moderate AR:

Significant AR

<table>
<thead>
<tr>
<th>Parameter</th>
<th>Value</th>
</tr>
</thead>
<tbody>
<tr>
<td>Sensitivity</td>
<td>100%</td>
</tr>
<tr>
<td>Specificity</td>
<td>87%</td>
</tr>
<tr>
<td>Positive p.v.</td>
<td>67%</td>
</tr>
<tr>
<td>Negative p.v.</td>
<td>100% (versus angio)</td>
</tr>
</tbody>
</table>

*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

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

Tribouilloy CM, Circulation 2000
Aortic regurgitation – Jet area

- < 0.35: Mild
- 0.35 – 0.50: Moderate
- > 0.50: Severe
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

Mild AR
Slope = 230 cm/s²
P1/2t = 560msec

Severe AR
• Rapid deceleration
• LV pressure rises rapidly
• Aortic diastolic pressure drops

Severe AR
Slope = 460 cm/s²
P1/2t = 180msec

Velocity decay is linearly related to the severity of AR

<table>
<thead>
<tr>
<th>Severity</th>
<th>Slope Range</th>
<th>P1/2t Range</th>
</tr>
</thead>
<tbody>
<tr>
<td>Mild AR</td>
<td>Slope &lt; 200 cm/s²</td>
<td>P1/2t &gt; 400msec</td>
</tr>
<tr>
<td>Moderate AR</td>
<td>Slope 200-400 cm/s²</td>
<td>P1/2 400 – 200msec</td>
</tr>
<tr>
<td>Severe AR</td>
<td>Slope &gt; 400 cm/s²</td>
<td>P1/2 &lt; 200msec</td>
</tr>
</tbody>
</table>
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:

<table>
<thead>
<tr>
<th>Preoperative</th>
<th>NYHA III-IV</th>
<th>Postop EF</th>
<th>Postop LVEDd</th>
</tr>
</thead>
<tbody>
<tr>
<td>LVEF &lt; 50%</td>
<td>0.03</td>
<td>0.008</td>
<td>0.01</td>
</tr>
<tr>
<td>LVEDd &gt; 4</td>
<td>ns</td>
<td>0.05</td>
<td>0.003</td>
</tr>
<tr>
<td>LVESd &gt; 4</td>
<td>0.02</td>
<td>0.05</td>
<td>0.007</td>
</tr>
</tbody>
</table>

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