LOW-FLOW LOW-GRADIENT PULMONARY STENOSIS IN REPAIRED TOF: FACT OR FICTION?

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Abbott
Medtronic

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Edwards Lifesciences
Flow Dependence of Transvalvular Gradient

AVA (cm²):
- 2.0
- 1.0
- 0.7
- 0.5
- 0.3

Assumptions:
- HR: 88 b/min
- SEP: 0.32 sec

Grossman W: Cardiac Catheterization and Angiography
## Assessment of Aortic Valve Stenosis

### Table 3  Recommendations for grading of AS severity

<table>
<thead>
<tr>
<th></th>
<th>Aortic sclerosis</th>
<th>Mild</th>
<th>Moderate</th>
<th>Severe</th>
</tr>
</thead>
<tbody>
<tr>
<td>Peak velocity (m/s)</td>
<td>&lt;2.5 m/s</td>
<td>2.6–2.9</td>
<td>3.0–4.0</td>
<td>≥4.0</td>
</tr>
<tr>
<td>Mean gradient (mmHg)</td>
<td>–</td>
<td>&lt;20</td>
<td>20–40</td>
<td>≥40</td>
</tr>
<tr>
<td>AVA (cm²)</td>
<td>–</td>
<td>&gt;1.5</td>
<td>1.0–1.5</td>
<td>&lt;1.0</td>
</tr>
<tr>
<td>Indexed AVA (cm²/m²)</td>
<td>–</td>
<td>&gt;0.85</td>
<td>0.60–0.85</td>
<td>&lt;0.6</td>
</tr>
<tr>
<td>Velocity ratio</td>
<td>–</td>
<td>&gt;0.50</td>
<td>0.25–0.50</td>
<td>&lt;0.25</td>
</tr>
</tbody>
</table>

*EACVI and ASE Recommendations 2017*

*Baumgartner H et al  Eur Heart J CVI 2017;18:254-275*
Assessment of Pulmonary Valve Stenosis

GUIDELINES AND STANDARDS

Echocardiographic Assessment of Valve Stenosis: EAE/ASE Recommendations for Clinical Practice

Helmut Baumgartner, MD, Judy Hung, MD, Javier Bermejo, MD, PhD, John B. Chambers, MD, Arturo Evangelista, MD, Brian P. Griffin, MD, Bernard Iung, MD, Catherine M. Otto, MD, Patricia A. Pellikka, MD, and Miguel Quiñones, MD

Pulmonic stenosis severity
Quantitative assessment of pulmonary stenosis severity is based mainly on the transpulmonary pressure gradient. Calculation of pulmonic valve area by planimetry is not possible since the required image plane is in general not available. Continuity equation or proximal isovelocity surface area method, although feasible in principle, has not been validated in pulmonary stenosis and is rarely performed.

EAE and ASE Recommendations 2009
Assessment of Pulmonary Valve Stenosis

GUIDELINES AND STANDARDS

Echocardiographic Assessment of Valve Stenosis: EAE/ASE Recommendations for Clinical Practice

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Table 11 Grading of pulmonary stenosis

<table>
<thead>
<tr>
<th></th>
<th>Mild</th>
<th>Moderate</th>
<th>Severe</th>
</tr>
</thead>
<tbody>
<tr>
<td>Peak velocity (m/s)</td>
<td>&lt;3</td>
<td>3–4</td>
<td>&gt;4</td>
</tr>
<tr>
<td>Peak gradient (mmHg)</td>
<td>&lt;36</td>
<td>36–64</td>
<td>&gt;64</td>
</tr>
</tbody>
</table>
Transvalvular Doppler gradient is generally used to classify the severity of PS in children. …

Good correlation with catheterization derived gradients. …

“Our analysis reveals marked heterogeneity in the classification of PS severity from Doppler gradients, especially in mild to moderate forms. …

# Table 13  Indications for intervention in right ventricular outflow tract obstruction

<table>
<thead>
<tr>
<th>Indications</th>
<th>Class</th>
<th>Level</th>
</tr>
</thead>
<tbody>
<tr>
<td>RVOTO at any level should be repaired regardless of symptoms when Doppler peak gradient is &gt;64 mmHg (peak velocity &gt;4 m/s), provided that RV function is normal and no valve substitute is required</td>
<td>I</td>
<td>C</td>
</tr>
<tr>
<td>In valvular PS, balloon valvotomy should be the intervention of choice</td>
<td>I</td>
<td>C</td>
</tr>
<tr>
<td>In asymptomatic patients in whom balloon valvotomy is ineffective and surgical valve replacement is the only option, surgery should be performed in the presence of a systolic RV pressure &gt;80 mmHg (TR velocity &gt;4.3 m/s)</td>
<td>I</td>
<td>C</td>
</tr>
</tbody>
</table>

Intervention in patients with gradient <64 mmHg should be considered in the presence of:
- symptoms related to PS or,
- double-chambered RV (which is usually progressive) or,
- important arrhythmias or,
- right-to-left shunting via an ASD or VSD.

Peripheral PS, regardless of symptoms, should be considered for repair if >50% diameter narrowing and RV systolic pressure >50 mmHg and/or lung perfusion abnormalities are present.

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*Baumgartner H et al ESC GL for the management of grown-up congenital heart disease 2010*
Low-Gradient PS?

- Female, 32 years old (1985)
- Tetralogy of Fallot
- 1990 BT shunt
- 1991 Repair (transannular patch)
- 2002 Pulmonary valve replacement (Contegra graft 22mm) for severe PR
- 2007 BVT for xenograft stenosis
- 2009 successful resuscitation (VF), ICD
- 2009 VT ablation
Low-Gradient PS?

- Female, 32 years old (1985), TOF repair

PRESENTATION 2013:

- Recent decrease in exercise capacity
- 4 adequate ICD shocks since 2009 documented increase of arrhythmias
- CPET:
  125 W (75%)
  \( V_{\text{O}_2 \text{max}} \) 19.8 ml/kg/min (69%)
- NT-proBNP 1800 pg/ml
Low-Gradient PS?  F32yrs
Valve morphology by echocardiography suspicious of aortic stenosis

Step 1 Assess velocity/gradient

LOW GRADIENT AS
Vmax < 4m/s ΔPm < 40mmHg

HIGH GRADIENT AS
Vmax ≥ 4m/s ΔPm ≥ 40mmHg

Step 2 Assess AVA

AVA ≤ 1.0 cm²

AVA >1.0 cm² -> moderate AS

High flow status excluded

no

yes

Severe high gradient AS (normal flow / low flow) (normal EF /low EF)

Define whether high flow status is reversible

Step 3 Exclude measurement errors that may cause gradient / flow / AVA underestimation!!

Step 4 Define flow status (SV index)

Low flow (SVI ≤35ml/m²)

Normal flow (SVI >35ml/m²) -> severe AS unlikely

Not reversible -> severe AS

Reversible -> re-assess at restored normal flow

EACVI and ASE Recommendations 2017
Baumgartner H et al  Eur Heart J CVI 2017;18:254-275
Assessment of Low Flow, Low Gradient Aortic Valve Stenosis

Step 5
Low flow (SVI ≤ 35 ml/m²)
Assess LVEF

LVEF < 50%

Step 6
Dobutamine echo
Flow reserve
pseudosevere AS
true severe AS

LVEF ≥ 50%

Integrated approach (table 5)

Step 7
Calcium Score by CT (see table 5)
Table 5  Criteria that increase the likelihood of severe AS in patients with AVA <1.0 cm² and mean gradient <40 mmHg in the presence of preserved EF

<table>
<thead>
<tr>
<th>(1) Clinical criteria:</th>
</tr>
</thead>
<tbody>
<tr>
<td>Physical examination consistent with severe aortic stenosis</td>
</tr>
<tr>
<td>Typical symptoms without other explanation</td>
</tr>
<tr>
<td>Elderly patient (&gt;70 years)</td>
</tr>
</tbody>
</table>

<table>
<thead>
<tr>
<th>(2) Qualitative imaging data:</th>
</tr>
</thead>
<tbody>
<tr>
<td>LVH (additional history of hypertension to be considered)</td>
</tr>
<tr>
<td>Reduced LV longitudinal function without other explanation</td>
</tr>
</tbody>
</table>

<table>
<thead>
<tr>
<th>(3) Quantitative imaging data:</th>
</tr>
</thead>
<tbody>
<tr>
<td>Mean gradient 30–40 mmHg*</td>
</tr>
<tr>
<td>AVA ≤0.8 cm²</td>
</tr>
<tr>
<td>Low flow (SVi &lt;35 mL/m²) confirmed by other techniques than standard</td>
</tr>
<tr>
<td>Doppler technique (LVOT measurement by 3D TEE or MSCT; CMR, invasive data)</td>
</tr>
<tr>
<td>Calcium score by MSCT**</td>
</tr>
<tr>
<td>Severe AS likely: men ≥2000  women ≥1200</td>
</tr>
<tr>
<td>Severe AS very likely: men ≥ 3000  women ≥1600</td>
</tr>
<tr>
<td>Severe AS unlikely: men &lt;1600  women &lt;800</td>
</tr>
</tbody>
</table>

*Haemodynamics measured when the patient is normotensive

**Values are given in arbitrary units using Agatston method for quantification of valve calcification

EACVI and ASE Recommendations for assessment of AS 2017

Baumgartner H et al  Eur Heart J CVI 2017;18:254-275
Assessment of Low-Flow, Low-Gradient Pulmonary Stenosis?

- No validated techniques to measure "valve area"
- No validated techniques to measure transvalvular flow
- Limitations of echocardiography with regard to quantification of RV function (gold standard CMR; ICD!)
- RV contractile reserve?
RV Contractile Reserve in Patients with Repaired Tetralogy of Fallot

Ishii H et al Am J Cardiol 2005;95:1338-43
Van den Berg J et al Int J Cardiol 2009;133:364-70
Parish V et al Int J Cardiol 2013;166:96-105

- Exercise (Ex) or Dobutamine stress (DS)
- Tissue Doppler imaging (TDI) indices, TAPSE and fractional area change (FAC) of the RV
- Comparison with healthy controls
  Predictor of exercise capacity and outcome
  Early detection of RV dysfunction

Ex echo TDI
DSE TDI
DS CMR RVEF
DS CMR
Ex echo FAC, TAPSE
Responders and nonresponders by Ex echo FAC

- Nonresponders had lower LVEF and higher LVESV and a higher (n.s.) event rate
- RV pressures increased in both groups

Low-Gradient PS? F32yrs
Low-Gradient PS? \hspace{2cm} F32yrs

BEFORE
Percutaneous Pulmonary Valve Implantation

AFTER
Low-Gradient PS?

BEFORE
Percutaneous Pulmonary Valve Implantation

AFTER
F32yrs
Low-Gradient PS?  F32yrs

- Female, 32 years old (1985), ToF repair
- 1990 BT shunt, 1991 Repair
- 2002 Pulmonary valve replacement
- 2009 Successful resuscitation (VF), ICD
- 2013 PPVI
- 2014: good exercise capacity (150W, 94%) less documented arrhythmias
  NT-proBNP 1800 -> 1300 pg/ml mild PR, Vmax 2m/s, LVF still reduced
- 2017: stable results
Low-Flow Low Gradient PS in Repaired TOF: Fact or Fiction?

• Fact! But unfortunately insufficiently studied!

• Low flow that may result in a low gradient despite severe RVOTO may primarily be due to RV dysfunction and/or severe TR

• Exercise or dobutamine stress may be used to increase flow and study the gradient response

• Further research is required to help identifying severe PS in case of a low gradient
EuroGUCH &

11th Advanced Symposium on Congenital Heart Disease in the Adult

Save the date: 20-21. April 2018
Münster, Germany
11th Advanced Symposium on Congenital Heart Disease in the Adult

Course Directors:
Michael Gatzoulis, Gerhard Diller, Helmut Baumgartner
Thank you for your attention!