A pulmonary vascular resistance of 8 Woods units per meter squared defines operablity in congenital heart disease…”

RMF Berger
Beatrix Children’s Hospital
University Medical Center Groningen The Netherlands
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To be considered….

- Clinical characteristics
- Age
- PVR (PVR/SVR ratio)
- Qp:Qs
- Cardiac morphology
- Associated anomalies
Pulmonary Arterial Hypertension

PAH in CHD
assessment of reversibility

- Clinical characteristics
- Hemodynamics and pulmonary vasoreactivity
- Lung biopsy
- Pulmonary arterial wedge angiography
- Pre-treatment?

Berger, Eur Heart J 2000
Perioperative risks


Survival in Eisenmenger syndrome vs corrected VSD

*Figure 6* Actuarial survival curves: Simple (0) vs Complex (1) congenital heart defect ($P=0.001$).
Survival in Eisenmenger syndrome
Based on type of heart defect, age

Eisenmenger's syndrome: marked restriction in exercise capacity

Mean ± SD
28.7 ± 10.4
25.5 ± 9.1
23.4 ± 8.9
23.3 ± 7.4
22.7 ± 7.6
20.8 ± 4.2
20.1 ± 6.5
19.8 ± 5.8
19.2 ± 6.2
18.6 ± 6.9
14.6 ± 4.7
11.5 ± 3.6

Correlation between pre-op biopsy findings and pulmonary arterial pressure after operation

There was no correlation between the pulmonary arterial pressure or resistance values recorded in the preoperative study and those obtained 1 year after surgery. For example, postoperative elevation in pulmonary vascular resistance was present in three of 21 patients with preoperative elevations and in five of 35 with normal preoperative values.

late postoperative pulmonary hemodynamics. It was reassuring to observe that patients who underwent surgical correction in infancy (first 8 months of life) had normal pulmonary hemodynamics 1 year after repair despite marked abnormalities at the time of repair with regard to structural remodeling and growth of the pulmonary arteries and/or intimal hyperplasia. The norm-

Ikawa et al, Circulation 1984

PULMONARY VASCULAR RESISTANCE DURING EXERCISE LATE AFTER REPAIR OF LARGE VENTRICULAR SEPTAL DEFECTS

In conclusion, this study demonstrated that a large VSD with PVR of more than an Rp/Rs of 0.5 should be closed while the patient is younger than 1 year old. This is also preferable in those with an Rp/Rs less than 0.5, but even when operation is delayed to the age of 4 years normal pulmonary vascular responses to moderate exercise are usually present late after operation.

Fig. 1. Relationship between postoperative PVR during exercise and preoperative Rp/Rs. There was significant correlation between these (r = 0.58, p < 0.001). Exercise

Follow up 9-21 yrs (mean 14.5 yrs)

Ikawa et al, J Thorac Cardiovasc Surg 1995
Regression of Pulmonary Vascular Disease

- At young age (< 9 mo – 1 yr) Rabinovitch et al, Circulation 1984
- After PA-banding Wagenvoort et al, J Thorac Cardiovasc Surg 1984
  Batista et al, Arq Bras Cardiol. 1997
- “Antiproliferative” drugs

Long-term outcome of patients operated for large ventricular septal defects with increased pulmonary vascular resistance
Kannan BR et al, Indian Heart J 2003

- Cohort of patients with non-restrictive VSD and PVR > 6 WU.m2
- 38 patients, age 6 months – 27 years (median 7.5 yrs)
- Operated between 1985-1996
- Pre-op hemodynamic variables:
  - PVR 7.6 ± 1.8 Wood’s units
  - Qp : Qs 1.9 ± 0.48
  - PVR/SVR 0.41 ± 0.12
- Mean follow-up 8.7 yrs
- 30 patients (80%) had a good outcome:
  - asymptomatic, significant reduction in PAP
- 8 patients (20%) poor outcome:
  - 5 immediate post-op deaths, 1 late death
  - 2 surviving with persistent severe PH
Long-term outcome of patients operated for large ventricular septal defects with increased pulmonary vascular resistance

Kannan BR et al, Indian Heart J. 2003

n=11

AsD with pulmonary vascular disease
Prediction of outcome after surgical correction

- 702 patients
- 40 (6%) pulm vasc disease
  TPR > 7 WU.m2
- 26 surgical closure
  14 medical treatment
- Median follow up 12 yrs

Steele et al., Circulation 1987
ASD with pulmonary vascular disease
Prediction of outcome after surgical correction

Surgical treatment

Medical treatment

FIGURE 3. Observed survival to 10 years in 26 surgically treated patients with atrial septal defect and pulmonary vascular obstructive disease. Percentile of patients which had observation at 3, 5, and 10 years.

FIGURE 4. Observed survival to 10 years in 14 medically treated patients with atrial septal defect and pulmonary vascular obstructive disease. Percentile of patients which had observation at 3, 5, and 10 years.


Rp during preoperative testing according to outcome following surgery

TABLE 3. Outcome and the Results of Hemodynamic Testing

<table>
<thead>
<tr>
<th>Rp units · m⁻² Oxygen Alone</th>
<th>Good Outcome</th>
<th>Poor Outcome</th>
</tr>
</thead>
<tbody>
<tr>
<td>Age, months</td>
<td>52±9</td>
<td>25±10</td>
</tr>
<tr>
<td>Rp 1, dyn × cm⁻² · m⁻²</td>
<td>872±64</td>
<td>172±272</td>
</tr>
<tr>
<td>units·m⁻²</td>
<td>(3.9±0.8)</td>
<td>(2.4±3.6)</td>
</tr>
<tr>
<td>Rp 2, dyn × cm⁻² · m⁻²</td>
<td>472±46</td>
<td>1015±2004</td>
</tr>
<tr>
<td>units·m⁻²</td>
<td>(5.9±0.5)</td>
<td>(12.7±2.5)</td>
</tr>
<tr>
<td>Rp 3, dyn × cm⁻² · m⁻²</td>
<td>322±241</td>
<td>741±1721</td>
</tr>
<tr>
<td>units·m⁻²</td>
<td>(4.1±0.3)</td>
<td>(9.3±2.2)</td>
</tr>
<tr>
<td>Rp 1, dyn × cm⁻² · m⁻²</td>
<td>1400±36</td>
<td>1400±246</td>
</tr>
<tr>
<td>units·m⁻²</td>
<td>(18.9±1.0)</td>
<td>(17.5±3.1)</td>
</tr>
<tr>
<td>Rp 2, dyn × cm⁻² · m⁻²</td>
<td>1756±36</td>
<td>1520±246</td>
</tr>
<tr>
<td>units·m⁻²</td>
<td>(21.7±1.0)</td>
<td>(19.0±3.1)</td>
</tr>
<tr>
<td>Rp 3, dyn × cm⁻² · m⁻²</td>
<td>1844±38</td>
<td>1784±236</td>
</tr>
<tr>
<td>units·m⁻²</td>
<td>(23.1±1.1)</td>
<td>(22.3±1.7)</td>
</tr>
<tr>
<td>Rp/Rp 1</td>
<td>0.60±0.11</td>
<td>1.98±0.72</td>
</tr>
<tr>
<td>Rp/Rp 2</td>
<td>0.28±0.02</td>
<td>0.94±0.34</td>
</tr>
<tr>
<td>Rp/Rp 3</td>
<td>0.19±0.01</td>
<td>0.43±0.07</td>
</tr>
</tbody>
</table>

1) baseline; 2) 100% oxygen; 3) 100% oxygen and nitric oxide; Rp pulmonary vascular resistance index; Rp systolic vascular resistance index. *P<0.05 in comparison to baseline.
†P<0.05 in comparison to 100% oxygen.
‡P<0.05 in comparison to patients who had a good outcome.
“Would you deny surgical repair to a patient with a pulmonary vascular resistance of 8 Woods units per meter squared ...?”
All old data........??
Anzheng Hospital Beijing, China; Gan H-L et al

1212 patients with cong. syst-to-pulm shunt and advanced pulmonary hypertension (1990-2008)

the patients were entered into 2 groups respectively: non-surgical group (n=297 cases) and surgical group (n= 915).

the age when the closure procedure was undertaken ranged from 13 months to 37 yrs old.

The mean follow-up period was 97.2±57.36 months

propensity scores were estimated using unconditional logistic regression to determine the predicted probability of inclusion into the surgical group for each of the 1212 patients.

Gan et al, WCPCCS, Cairns 2009
**Definition of a propensity score**

**Clinical features**
- Age / Sex
- Clinical right heart failure
- Hemoptysis
- Cyanosis at rest / on exertion
- Clubbing fingers and toes
- Systolic heart murmur ≥ II/VI
- Hemoglobin
- Shunt level
- NYHA functional class ≥ III
- 6MWD

**Echocardiography**
- LVEF
- Bidirectional shunt on TTE
- Severe tricuspid regurgitation

**Hemodynamics**
- Right atrium pressure
- PVR
- QP/QS
- SPAP
- SPAP/SBP
- mPAP
- SaO2 / PaO2
- Positive reaction to NO / O2

Gan et al, WCPCCS, Cairns 2009

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**Figure 2. The Kaplan-Meier survival curve for the non-surgical therapy group and surgical therapy group when the PVR < 16 Wood (survival years)**

Gan et al, WCPCCS, Cairns 2009
PVRi < 8 WU.m2

A pulmonary vascular resistance of 8 Woods units per meter squared defines operability in congenital heart disease, unless…”