The Swiss pulmonary hypertension registry: what does it learn us?

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Why do we need a pulmonary hypertension registry?
PH: diagnostic algorithm

Eur Heart J 2009; 30(20):2493-537
Eur Respir J 2009; 34:1219-1263

ESC / ERS Guidelines for the diagnosis and treatment of pulmonary hypertension

The Task Force for the Diagnosis and Treatment of Pulmonary Hypertension of the European Society of Cardiology (ESC) and the European Respiratory Society (ERS)
Pulmonary arterial hypertension (PAH)

PAH is a rare disease = orphan disease

“orphan disease”: affect < 1:2000 individuals

USA: <200’000
EU: < 5/10’000
AUS: < 2000
J: < 50’000
WHO: < 6.5-10/10’000

Why a PH registry?

To yield informations on:

- diagnosis / subgroups
- prognosis
- demographics
- hemodynamics
- treatment modalities
- treatment outcome
- survival
The swiss PAH – registry

initiated 1999 - retrospective analysis 1990-1999
- prospectively since 1999

collect information into:
• prevalence
• epidemiology
• characteristics
• therapy
• outcome

of PAH patients in Switzerland
**Swiss PH registry: patients characteristics 1999**
(retrospective data 1991-1999)

<table>
<thead>
<tr>
<th></th>
<th>N</th>
<th>surviving</th>
<th>dead</th>
<th>p</th>
</tr>
</thead>
<tbody>
<tr>
<td><strong>6' walk test (m)</strong></td>
<td>62</td>
<td>363 ±135</td>
<td>235 ±155</td>
<td>0.002</td>
</tr>
<tr>
<td><strong>SvO2(%)</strong></td>
<td>66</td>
<td>66.5 ± 9.5</td>
<td>57.9 ± 10.1</td>
<td>0.006</td>
</tr>
<tr>
<td><strong>PAPm (mm Hg)</strong></td>
<td>82</td>
<td>53 ± 12</td>
<td>59 ± 13</td>
<td>0.06</td>
</tr>
<tr>
<td><strong>PAPs (mm Hg)</strong></td>
<td>82</td>
<td>81 ± 19</td>
<td>84 ± 21</td>
<td>0.05</td>
</tr>
<tr>
<td><strong>PVR (dyn.sec.cm⁻⁵)</strong></td>
<td>67</td>
<td>892 ± 419</td>
<td>959 ± 455</td>
<td>0.11</td>
</tr>
<tr>
<td><strong>PaO2(kPa)</strong></td>
<td>72</td>
<td>9.1 ± 1.8</td>
<td>9.2 ± 2.3</td>
<td>0.9</td>
</tr>
</tbody>
</table>

**NYHA class II vs III/IV(n)** 94: 18 vs 44, 2 vs 18, 0.015
Swiss PH registry: number of patients

Swiss PH registry: number of patients

Source: Alabus / Swiss PH Registry
Swiss PH registry: age distribution

![Age distribution chart]

- **black**: retrospective data 1991 - 1999
- **white**: prospective data 1999 - 2004

Stricker H et al, swiss med wkly 2001; 131:346-350
Tueller T et al, swiss med wkly 2008; 138:379-384
Swiss PH registry: demographics 2004

250 patients
Females 147 (59%)
median age
F 59y (46-70), M 63y (45-70)
median follow-up
18.8 months (9-31)
median time between visits
3.0 months (2-5)
most patients:
IPAH or CTEPH
NYHA III-IV

Prevalence
all PAH: 55 / million adult inhabitants
IPAH: 8.6 / million

Incidence
all PAH: 3.5 / million

Tueller T et al, swiss med wkly 2008; 138:379-384
Swiss PH registry: patients characteristics 2004 (prospective data 1999-2004)

Tueller T et al, swiss med wkly 2008; 138:379-384
Swiss PH registry: survival 2004

Fischler M et al., swiss med wkly 2008; 138:371-378
Swiss PH registry: survival CH vs. NIH-formula and literature data (IPAH)

Fischler M et al., swiss med wkly 2008; 138:371-378
Swiss PH registry: evolution over time

Fischler M et al., swiss med wkly 2008; 138:371-378
Swiss PH registry: modality of treatment

<table>
<thead>
<tr>
<th></th>
<th>Oxygen (%)</th>
<th>Diuretics (%)</th>
<th>OAK (%)</th>
</tr>
</thead>
<tbody>
<tr>
<td></td>
<td>IPAH</td>
<td>CTEPH</td>
<td>IPAH</td>
</tr>
<tr>
<td>Baseline</td>
<td>25</td>
<td>22</td>
<td>33</td>
</tr>
<tr>
<td>Best</td>
<td>41*</td>
<td>30</td>
<td>43</td>
</tr>
<tr>
<td>Last</td>
<td>42</td>
<td>38</td>
<td>49</td>
</tr>
</tbody>
</table>

IPAH: Idiopathic pulmonary artery hypertension, CTEPH: Chronic thromboembolic pulmonary hypertension, OAK = oral anticoagulation. *p < 0.05; **p < 0.001

<table>
<thead>
<tr>
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<th>CTEPH</th>
</tr>
</thead>
<tbody>
<tr>
<td></td>
<td>BL</td>
<td>Best</td>
</tr>
<tr>
<td>Specific Therapy</td>
<td>26 of 83 (31%)</td>
<td>63 of 75 (84%)</td>
</tr>
<tr>
<td>Single drug</td>
<td></td>
<td></td>
</tr>
<tr>
<td>CCB (%)</td>
<td>8</td>
<td>9</td>
</tr>
<tr>
<td>Ila inh (%)</td>
<td>50</td>
<td>22</td>
</tr>
<tr>
<td>Ila iv (%)</td>
<td>8</td>
<td>6</td>
</tr>
<tr>
<td>Bas (%)</td>
<td>50</td>
<td>37</td>
</tr>
<tr>
<td>Sil (%)</td>
<td>0</td>
<td>4</td>
</tr>
<tr>
<td>Multiple drugs</td>
<td></td>
<td></td>
</tr>
<tr>
<td>Bas-Sil (%)</td>
<td>0</td>
<td>2</td>
</tr>
<tr>
<td>Bas-Do (%)</td>
<td>4</td>
<td>19</td>
</tr>
<tr>
<td>Ila+Sil (%)</td>
<td>0</td>
<td>2</td>
</tr>
<tr>
<td>&gt; (5)</td>
<td>0</td>
<td>6</td>
</tr>
</tbody>
</table>

Fischler M et al., swiss med wkly 2008; 138:371-378
**PH: therapy**

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ESC / ERS Guidelines for the diagnosis and treatment of pulmonary hypertension

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Swiss PH registry: number of patients (12.2009)

- Prevalence 2009: 118 / million inhab. = 1:8475
- Prevalence 2009 - IPAH (n=213): 30 / million inhab.

Total patients: 832
Swiss PH registry: PAH by aetiology
(according to WHO classification, status 12.2009)

- 1.1. Idiopathic (IPAH)
- 1.3.1. Collagen vascular disease
- 1.3.2. Congenital L->R shunt
- 1.3.3. Portal hypertension
- 1.3.4. HIV infection
- 1.3.5. Drugs
- 1.3.6. Other PAH
- 1.4.1. Pulmonary VOD
- 2. PH with left heart disease
- 3. PH with lung diseases or hypoxemia
- 4. CTEPH
- 5. Miscellaneous
- Not classified

Source: Swiss PH Registry
Swiss PH registry: PAH by aetiology
(according to WHO classification, status 12.2009)

Source: Swiss PH Registry
Swiss PH registry: demographic status (12.2009)

Source: Alabus / Swiss PH Registry
Swiss PH registry:
baseline right heart catheter (12.2009)

Source: Alabus / Swiss PH Registry
PAH pathogenesis: relevant pathways

ACCF/AHA 2009 Expert Consensus Document on PAH
Mc Laughing V et al, J Am Coll Cardiol; 53:17,2009
Conclusion / 1

- pulmonary arterial hypertension is a rare disease

- a national PAH register is crucial for a better knowledge of the disease

- the PAH-register provide us information on:
  - epidemiology and prevalence of PAH (different subgroups) in Switzerland
  - clinical course of the disease
  - treatement outcome
  - (time to clinical worsening)
  - survival
Conclusion / 2

Data from PAH-registers have limitation and need correlation to RCT

Is a good quality control tool:
- comparaison with data from other national registries or from major international centers
- comparaison between swiss centers

Knowledge on improvements during time, specially in the era of specific PH therapies
Conclusion / 3

All PAH patients should be included in a national registry.

The SSHP-centres have access to the swiss PAH-registry.

All swiss PAH patients should be referred to a SSPH-centre:
• for the initial work-up if PAH is suspected
• to establish indication for treatment
• for follow-up visits and register-update (at least once a year)
MALATTIA REUMATOLOGICA
AR, scleroderma, lupus, SS, ...

Interstiziopatia?
Ipertensione polmonare?
BILANCIO SPECIALISTICO!

SINTOMI RESPIRATORI
dispnea allo sforzo
tosse
Updated Classification of PH

Grazie per l’attenzione!

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