Challenging The 2015 PH Guidelines - comments from the Nurses

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### Recommendations for pulmonary hypertension expert referral centres

<table>
<thead>
<tr>
<th>Recommendations</th>
<th>Class</th>
<th>Level</th>
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<tbody>
<tr>
<td>It is recommended for expert referral centres to provide care by a multiprofessional team (cardiology and respiratory medicine physicians, clinical nurse specialist, radiologists, psychological and social work support, appropriate on-call expertise)</td>
<td>I</td>
<td>C</td>
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<tr>
<td>It is recommended for expert referral centres to have direct links and quick referral patterns to other services (such as CTD, family planning, PEA, lung transplantation, adult congenital heart disease)</td>
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<td>It should be considered that a expert referral centre follow at least 50 patients with PAH or CTEPH and should receive at least two new referrals per month with documented PAH or CTEPH</td>
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<tr>
<td>It should be considered that a expert referral centre perform at least 20 vasoreactivity tests in IPAH, HPAH or DPAH patients per year</td>
<td>IIa</td>
<td>C</td>
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<tr>
<td>Expert referral centres should participate in collaborative clinical research in PAH, including phase II and phase III clinical trials</td>
<td>IIa</td>
<td>C</td>
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</table>

**Pulmonary arterial hypertension therapy**

It is recommended for expert referral centres to provide care by a multi-professional team (cardiology and respiratory medicine physicians, clinical nurse specialist, radiologists, psychological and social work support, appropriate on-call expertise)
12.1 Facilities and skills required for an expert referral centre

Expert referral centres are recommended to provide care by an interprofessional team that should, as a minimum, comprise:

(a) two consultant physicians (normally from either or both cardiology and respiratory medicine) experienced in and with a special interest in PH with dedicated PH clinical sessions for outpatients, inpatients and a multidisciplinary team meeting

(b) clinical nurse specialist

(c) radiologist with expertise in pulmonary hypertension imaging

(d) cardiologist or PH physician with expertise in echocardiography

(e) cardiologist or PH physician with expertise in RHC and vasoreactivity testing

(f) access to psychological and social work support

(g) appropriate on-call cover and expertise
Clinical Nurse Specialists

5 main competency areas¹:

• Clinical expertise
• Research
• Leadership
• Collaboration
• Change agent

Demonstrated by¹:

• Listening, counselling
• Supporting patients and families
• Providing direct care
• Formal education
• Collaboration with the healthcare team
• Advocating for additional resources for patients and family members

The impact of CNSs on patients living with a chronic disease

- Improved quality of life ¹ ² ³ ⁴
- Fewer and shorter hospitalisations ¹ ⁵ ⁶ ⁷
- Decreased costs of care ¹ ⁷ ⁸ ⁹
- Increased patient and health provider satisfaction ¹ ⁸ ⁹ ¹⁰

7. Topp (1998) Nurs Case Manage. 3(4) p 140
Advances in the management of pulmonary hypertension

• Over the past 20 years PAH has transitioned from being a fatal disease with a poor prognosis to a long-term condition that, with the right treatment and support, can be managed.¹

• Patient care delivered via specialist, multidisciplinary healthcare teams now has a greater focus on quality of life considerations alongside the clinical symptoms of the disease

Patient engagement and self-management should be key considerations in PAH care.
Patients that feel well informed cope better with their disease

- PAH patients who were satisfied with the received information found it easier to cope with their disease ($p = 0.0045$)
- Those who found the overall information useful found it easier to cope with their disease ($p = 0.003$)

54% of PAH patients do not feel adequately informed about their disease


<table>
<thead>
<tr>
<th>Information area</th>
<th>% of patients receiving info (n=68)</th>
</tr>
</thead>
<tbody>
<tr>
<td>Disease</td>
<td>58</td>
</tr>
<tr>
<td>Medical tests</td>
<td>67</td>
</tr>
<tr>
<td>Treatments</td>
<td>47</td>
</tr>
<tr>
<td>Other services</td>
<td>21</td>
</tr>
<tr>
<td>Different places of care</td>
<td>23</td>
</tr>
<tr>
<td>Ways to help yourself</td>
<td>33</td>
</tr>
</tbody>
</table>
Areas in which patients desired more information

<table>
<thead>
<tr>
<th>Topics in questionnaire</th>
<th>Other topics raised by patients</th>
</tr>
</thead>
<tbody>
<tr>
<td>Diagnosis</td>
<td>Survival estimates/prognosis</td>
</tr>
<tr>
<td>Whether disease is under control</td>
<td>Research / future treatments</td>
</tr>
<tr>
<td>The severity of the disease</td>
<td>Possibility to become cured</td>
</tr>
<tr>
<td>Things patients can do to get well</td>
<td>Future palliative care</td>
</tr>
<tr>
<td>Side effects of treatments</td>
<td>Own hospital records</td>
</tr>
<tr>
<td>Purpose and results of medical tests</td>
<td>Co-morbidity</td>
</tr>
<tr>
<td>Possible causes</td>
<td>Risks of surgery</td>
</tr>
<tr>
<td>Rehabilitation services</td>
<td>Appropriate form of exercise</td>
</tr>
<tr>
<td>Psychological support</td>
<td>Unstressed, honest, straight forward information without having to ask and easy to get</td>
</tr>
<tr>
<td>Individually written information</td>
<td></td>
</tr>
</tbody>
</table>

Based on questionnaire for PAH patients ($n = 68$) in Sweden

# The optimal patient journey: Effective communication throughout the care process

<table>
<thead>
<tr>
<th></th>
<th>Patient information</th>
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</thead>
</table>
| First clinic visit       | • Mechanisms of disease  
                          | • Investigations required  
                          | • Contact details of the PH Centre |
| Diagnosis                | • On disease, treatment options, specialists involved in PAH care  
                          | • Managing the emotional impact; support via patient associations  
                          | • Written materials for patient to take away |
| Appointment 1 after      | • Written care plan following shared decision-making  
                          | • Patient ‘passport’ with individualised content  
                          | • Further information from patient association |
| diagnosis                |                                                                                     |
| Appointment 2 after      | • Referral to specialists as needed e.g. psychiatry, welfare  
                          | • Patient-to-patient mentoring matched according to age/background etc. |
| diagnosis                |                                                                                     |
| Appointment 3 after      | • Self-management course to support rehabilitation  
                          | • Ongoing engagement with multidisciplinary healthcare team and patient associations |
| diagnosis                |                                                                                     |

Adapted from “A holistic approach to patient care in pulmonary arterial hypertension”, 2015.
The overall treatment goal in patients with PAH is achieving a low risk status.

### Table 13  Risk assessment in pulmonary arterial hypertension

<table>
<thead>
<tr>
<th>Determinants of prognosis* (estimated 1-year mortality)</th>
<th>Low risk &lt;5%</th>
<th>Intermediate risk 5–10%</th>
<th>High risk &gt;10%</th>
</tr>
</thead>
<tbody>
<tr>
<td>Clinical signs of right heart failure</td>
<td>Absent</td>
<td>Absent</td>
<td>Present</td>
</tr>
<tr>
<td>Progression of symptoms</td>
<td>No</td>
<td>Slow</td>
<td>Rapid</td>
</tr>
<tr>
<td>Syncope</td>
<td>No</td>
<td>Occasional syncope&lt;sup&gt;a&lt;/sup&gt;</td>
<td>Repeated syncope&lt;sup&gt;b&lt;/sup&gt;</td>
</tr>
<tr>
<td>WHO functional class</td>
<td>I, II</td>
<td>III</td>
<td>IV</td>
</tr>
<tr>
<td>6MWD</td>
<td>&gt;440 m</td>
<td>165–440 m</td>
<td>&lt;165 m</td>
</tr>
<tr>
<td>Cardiopulmonary exercise testing</td>
<td>Peak VO&lt;sub&gt;2&lt;/sub&gt; &gt;15 ml/min/kg (&gt;65% pred.) VE/VCO&lt;sub&gt;2&lt;/sub&gt; slope &lt;36</td>
<td>Peak VO&lt;sub&gt;2&lt;/sub&gt; 11–15 ml/min/kg (35–45% pred.) VE/VCO&lt;sub&gt;2&lt;/sub&gt; slope 36–44.9</td>
<td>Peak VO&lt;sub&gt;2&lt;/sub&gt; &lt;11 ml/min/kg (&lt;35% pred.) VE/VCO&lt;sub&gt;2&lt;/sub&gt; slope ≥45</td>
</tr>
<tr>
<td>NT-proBNP plasma levels</td>
<td>BNP &lt;50 ng/l</td>
<td>BNP 50–300 ng/l</td>
<td>BNP &gt;300 ng/l</td>
</tr>
<tr>
<td></td>
<td>NT-proBNP &lt;300 ng/l</td>
<td>NT-proBNP 300–1400 ng/l</td>
<td></td>
</tr>
<tr>
<td>Imaging (echocardiography, CMR imaging)</td>
<td>RA area &lt;18 cm&lt;sup&gt;2&lt;/sup&gt; No pericardial effusion</td>
<td>RA area 18–26 cm&lt;sup&gt;2&lt;/sup&gt; No or minimal, pericardial effusion</td>
<td>RA area &gt;26 cm&lt;sup&gt;2&lt;/sup&gt; Pericardial effusion</td>
</tr>
<tr>
<td>Haemodynamics</td>
<td>RAP &lt;8 mmHg CI ≥2.5 l/min/m&lt;sup&gt;2&lt;/sup&gt; SvO&lt;sub&gt;2&lt;/sub&gt; &gt;65%</td>
<td>RAP 8–14 mmHg CI 2.0–2.4 l/min/m&lt;sup&gt;2&lt;/sup&gt; SvO&lt;sub&gt;2&lt;/sub&gt; 60–65%</td>
<td>RAP &gt;14 mmHg CI &lt;2.0 l/min/m&lt;sup&gt;2&lt;/sup&gt; SvO&lt;sub&gt;2&lt;/sub&gt; &lt;60%</td>
</tr>
</tbody>
</table>

6MWD = 6-minute walking distance; BNP = brain natriuretic peptide; CI = cardiac index; CMR = cardiac magnetic resonance; NT-proBNP = N-terminal pro-brain natriuretic peptide; pred. = predicted; RA = right atrium; RAP = right atrial pressure; SvO<sub>2</sub> = mixed venous oxygen saturation; VE/VCO<sub>2</sub> = ventilatory equivalents for carbon dioxide; VO<sub>2</sub> = oxygen consumption; WHO = World Health Organization.
Treatment Goals

• Clear parameters - easy to assess
• BUT...
• Many patients remain in the intermediate zone despite increases in PH therapy
• There are many other factors which can influence parameters
• Many goals are not meaningful for patients
What do Patients Want?

- To get an accurate diagnosis and the best treatment without avoidable delay
- To be treated with humanity, dignity and respect
- Information and communication
- Psychological, social and spiritual support
- Optimal symptom control
- Rehabilitation
- Well coordinated care
- End of life care
- Support for carers and bereavement
Majority of patients think QoL is the most important factor when choosing treatment.
Impact of PAH on a patient’s life

• Overall QoL and concerns about life expectancy and the future have the biggest impact on the life of a patient with PAH.
Individualised goal setting

- Coaching patients to identify their own realistic, meaningful and achievable goals
- Develop a plan to achieve their goals
- Ongoing assessment, re-evaluation and setting of new goals

I want to live to see my granddaughter born
I want to go swimming with my children
I want to be pain free
I want to be able to work and support my family
6.3.1.5 Psychosocial support

PH is a disease with a significant impact on the psychological, social (including financial) emotional and spiritual functioning of patients and their families.

Teams managing these patients should have the skills and expertise to assess and manage issues in all of these domains, with close links to colleagues in relevant disciplines for those with severe problems, e.g. psychiatry, clinical psychology, welfare and social work.
Depression

Because of diminished quality of life, decreased exercise capacity and mobility, social isolation, financial burdens, unemployment, and an uncertain prognosis, patients with PAH may be more vulnerable to developing depression¹

Resulting in:

- Medical non-compliance¹
- Increased levels of IL6¹², CRP¹², endothelial dysfunction³ and BNP¹
- Decreased 6 minute walk test distance¹
- Decreased life expectancy⁴

4. Ross (2012) BMJ. 345
6.3.11 End of life care and ethical issues

The clinical course of PH is one of progressive deterioration interspersed with episodes of acute decompensation. It is difficult to predict when patients will die since death may come either suddenly or slowly because of progressive heart failure. It has been shown that physicians caring for patients tend to be overoptimistic in their prognostication and frequently misunderstand their patients’ wishes.

Open and sensitive communication with patients allows advanced planning and discussion of their fears, concerns and wishes, which is essential to good care.
Palliative care in PH

- Many patients have a significant symptom burden despite optimal therapy.
- Can be used in parallel with PH therapy to improve symptoms and provide support for patients and their carers.
- Should be introduced earlier in the course of the disease as a component of standard care.
- Has been shown to improve depression and increase survival in other diseases.¹

Conclusion

• Patients should be encouraged to accumulate knowledge and skills to become empowered to live well with PAH

• Successful chronic disease interventions usually involve a coordinated multidisciplinary care team ¹

• CNSs can provide high quality and cost effective care to patients and families through education, support, advocacy and coordination of care ²

Thank you