early diagnosis • best treatment • better quality of life • finding a cure

PHA EUROPE

European pulmonary hypertension association

for the patients

Comments on the PH guidelines from the patients

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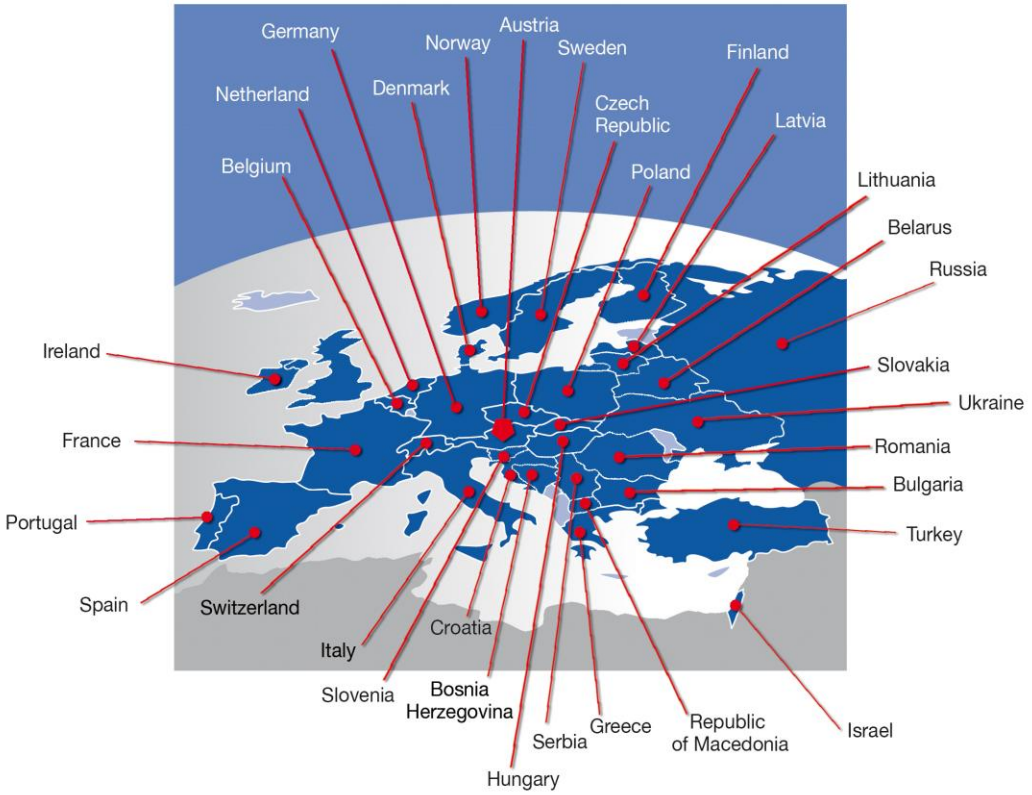
Challenging the 2015 PH Guidelines and annual G6 meeting

Sophia Antipolis 14-15 October 2016

About PHA Europe

- **2003:** Founded in Vienna by 8 national patient associations

- **Today:** A united, dynamic, expanding community comprising, 39 associations from 33 countries

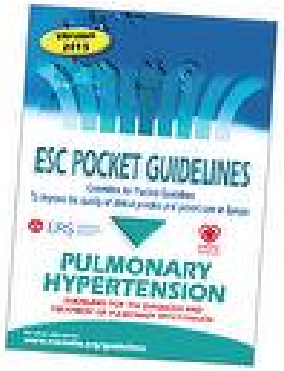


Our mission: Early diagnosis - Best treatment - Better QOL - Finding a cure

My presentation

- Positive aspects of PH guidelines for patients - in general and on specific issues
- Gaps in PH guidelines
- Challenges in application
- Conclusions

Positive aspects in general



- Progresses in past 20 years include:
  - New drugs
  - Improved surgery techniques
  - Better treatment management
- Guidelines are constantly updated to reflect the current state of the the art
- Guidelines ensure consistency of care and contribute to standardization at the highest possible level
- Clinical guidelines can help patients by influencing public policy on access/reimmbursement

Specific points of guidelines

1. Psycho-social support

- Greater recognition of the need for pyscho-social support (moved from IIc to Ic)
- “PH is a disease with a significant impact on the psychological, social (including financial), emotional and spiritual functioning of patients and their families” (6.3.1.5)
- Guidelines refer to an article in ERR co-authored by reps of PHA Europe, PHA US, PHA UK, physicians, nurses.
- This article follows up on findings of International Patient and Carer Survey conducted in 2011 in five European countries on 466 patients/carers.

2. Role of patient associations

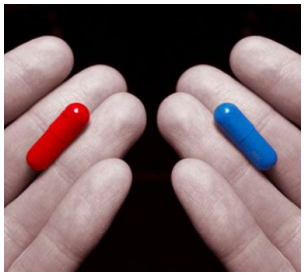
- Important role of PAs is now openly recognized:
  - "Encouraging patients and their family members to join patient support groups can have positive effects on coping, confidence and outlook"(6.3.1, General measures).
  - "Patient support groups may also play an important role and patients should be advised to join such groups (6.3.1.5, Psychosocial support).
  - "Referral centres should consider having a link to their national and/or European PH patients' associations" (12.1 Referral centres)

3. Patient risk profile

Determinants of prognosis[1] (estimated 1-year mortality)	Low risk <5%	Intermediate risk 5–10%	High risk >10%
Clinical signs of right heart failure	Absent	Absent	Present
Progression of symptoms	No	Slow	Rapid
Syncope	No	Occasional syncope[2]	Repeated syncope[2]
WHO functional class	I, II	III	IV
6MWD	>440 m	165–440 m	<165 m
Cardiopulmonary exercise testing	Peak VO_2 >15 ml/min/kg (>65% pred.) VE/VCO_2 slope <36	Peak VO_2 11–15 ml/min/kg (35–65% pred.) VE/VCO_2 slope 36–44.9	Peak VO_2 <11 ml/min/kg (<35% pred.) VE/VCO_2 slope ≥45
NT-proBNP plasma levels	BNP <50 ng/l NT-proBNP <300 ng/l	BNP 50–300 ng/l NT-proBNP 300–1400 ng/l	BNP >300 ng/l NT-proBNP >1400 ng/l
Imaging (echocardiography, CMR imaging)	RA area <18 cm^2 No pericardial effusion	RA area 18–26 cm^2 No or minimal pericardial effusion	RA area >26 cm^2 Pericardial effusion
Haemodynamics	RAP <8 mmHg CI ≥2.5 l/min/m^2 SvO_2 >65%	RAP 8–14 mmHg CI 2.0–2.4 l/min/m^2 SvO_2 60–65%	RAP >14 mmHg CI <2.0 l/min/m^2 SvO_2 <60%

- Until now FC was the criterion for treatment selection in related algorithm, patient risk profile has replaced it
- FC can be subjective, depends on patient's description of his/her symptoms and interobserver variability
- Risk assessment table is very comprehensive, includes many different variables, FC is one of them

4. Comb. therapy/transplant



- Combination therapy may now be applied sequentially or initially (upfront)
  - Recognition of the very aggressive nature of disease
  - Approach which has proven to be effective in other diseases eg. heart failure, HIV
- TX has been “moved up” in treatment algorithm
  - Delayed referral in combination with the length of the waiting time may increase the mortality of patients on the waiting list and their clinical severity at the time of transplantation.

5. Referral centres

- The need to refer to expert centres has been one of our key advocacy topics (Call to Action in EP in 2012).
- Most important for us is the recommendation in guidelines for multidisciplinary teams: “Referral centres are recommended to provide care by an interprofessional team” (12.1 Referral centres)
- List includes physicians, clinical nurse specialist, radiologist, experts in ECHO, RHC and VRT, access to psych/social support
- Guidelines should encourage regular contact between expert centre and local centres

Gaps in the PH guidelines

- Patients may have different expectations/priorities about their disease than what physicians think
- Patients may have preferences with regard to treatment (including refusal)
- There is only a very brief mention of consulting patients about treatment options (“as appropriate and/or necessary”) in the Preamble and nothing about feedback
- There is only one mention of “need for open and sensitive communication” in end of life section (6.3.11)

Challenges in application of guidelines

- Do all physicians know about the guidelines?
- Are these applied in daily practice?
- Are they maybe too complex and unrealistic for some countries?
- What happens if national health authorities draw up their own guidelines?

Conclusions

- The introduction of guidelines represents a very positive development for patients
- We welcome the increased recognition of psycho-social issues, of the importance of the role of PAs and new treatment strategies
- What is best for patients overall, as recommended in guidelines, may be inappropriate for individual patient needs
- More should be done to promote better communication between HCP and patients, shared decision making and patient self management
- Application of guidelines remains problematic
- The patient perspective should be included in future

Thank you for your kind attention

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