The Global Alliance against Chronic Respiratory Diseases

Pulmonary hypertension

Dr Marc Humbert
What is the burden of pulmonary hypertension?

The true burden of pulmonary hypertension is currently unknown and largely underestimated.
Pulmonary Hypertension

It remains widely believed that pulmonary hypertension is a rare disease.

Although true for idiopathic pulmonary arterial hypertension (formerly known as primary pulmonary hypertension), the true burden of pulmonary hypertension is currently unknown and certainly underestimated.

As novel and simple oral therapies are now available, a programme to improve awareness of pulmonary hypertension should be promoted.
Pulmonary hypertension and cor pulmonale may complicate many advanced pulmonary conditions including COPD, bronchiectasis, cystic fibrosis, lung fibrosis, sarcoidosis and tuberculosis.

When present, pulmonary hypertension directly contributes to disability and early mortality.
Pulmonary Hypertension: who is at risk?

Pulmonary hypertension is a major cause of disability and mortality in patients with **sickle cell disease** and **thalassemia** (significant burden in Africa and in people of African origin worldwide, as well as in subjects from Mediterranean countries) as well as in patients with **schistosomiasis** (significant burden in Brazil, Egypt...).
Pulmonary Hypertension: who is at risk?

Pulmonary hypertension may affect a significant proportion of **highlanders** in many countries (significant burden in Bolivia, Peru, China and other parts of Asia, South America and Africa...)
Up to 3% of patients may develop chronic thromboembolic pulmonary disease after severe acute pulmonary emboli.

This may lead to severe chronic thromboembolic pulmonary hypertension, a condition that can be cured by means of surgical thromboendarterectomy.
WHO classification of functional status of patients with pulmonary hypertension

Class Description

I No limitation of usual physical activity; ordinary physical activity does not cause dyspnea, fatigue, chest pain, or presyncope

II Mild limitation of physical activity; no discomfort at rest; but normal activity causes increased dyspnea, fatigue, chest pain, or presyncope

III Marked limitation of activity; no discomfort at rest but less than normal physical activity causes increased dyspnea, fatigue, chest pain, or presyncope

IV Unable to perform physical activity at rest; may have signs of RV failure; symptoms increased by almost any physical activity
Without treatment: survival correlates with functional class

Median survival – in years

Class I & II: 6 years
Class III: 2.6 years
Class IV: 0.5 years

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A clinical classification and evidence-based guidelines for pulmonary arterial hypertension have been recently produced and are going to be revised in 2007-2008.

Pulmonary arterial hypertension may be idiopathic, familial or associated with various conditions such as HIV infection, liver cirrhosis, autoimmune diseases, or congenital heart diseases which are prevalent worldwide.

Recently published evidence-based guidelines for pulmonary arterial hypertension should be provided to the medical community, and affordable drugs offered to affected individuals.
Action plan

- In countries where there is an increased risk for pulmonary hypertension (altitude, schistosomiasis, sickle cell disease...) we propose to launch a program in order to improve
- Pulmonary hypertension detection
- Diagnosis of pulmonary hypertension
- Management: basic therapy (exercise limitation, diuretics, oxygen, anticoagulants) and evaluation of novel therapeutic strategies (with a focus on oral drugs: sildenafil, bosentan?)
Goals to be achieved in the next 5 years

- Inform the medical community worldwide of the burden
- Inform the medical community of country-specific risk factors
- Provide information on signs or symptoms
- Provide information on a step-by-step management of dyspnoea
- Provide educational tools on simple tests suggestive of pulmonary hypertension: chest X ray and EKG
- Develop reference centers for pulmonary hypertension in each country with access to screening procedure (echodoppler) and if possible confirmation right-heart catheterization
- Propose epidemiological studies to evaluate the burden of pulmonary hypertension
- Improve access to conventional therapy for patients (oxygen, diuretics, anticoagulant)
- Demonstrate efficacy of novel therapies in patients displaying pulmonary hypertension of various origins
What can GARD do?

Next steps:

- Improve awareness on pulmonary hypertension
- Support a network of experts through the country to estimate the true burden that is certainly many folds higher than currently believed
- Enhance early detection for and treatment of pulmonary hypertension
- Promote better management according to local guidelines based on available therapies

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