

**Media release**  
**November 22, 2008**

## **Pulmonary Hypertension Patients Celebrate a Longer Life**

Australians with a severe form of a heart/ lung condition known as pulmonary hypertension are meeting today for the first ever patient seminar on their illness and to celebrate the treatment advances that have directly translated to longer lives.

“The meeting today is a significant milestone as in years gone by a diagnosis of pulmonary hypertension was a certain death sentence, though with the major advancement in medical treatments in the last ten years these patients can now live for many years in reasonable health and some reach a normal lifetime,” Professor Anne Keogh, Senior Heart Transplant Cardiologist, Professor of Medicine and Head of the Pulmonary Hypertension Clinic at St Vincent’s Hospital, Sydney said.

“It’s important to stop and acknowledge how life has changed for these people and also to gather hope and strength from the stories they can share with each other,” Professor Keogh said.

Pulmonary *Arterial* Hypertension, or PAH (a type of pulmonary hypertension), is a unique disease which causes a restriction of blood vessels from the heart to the lungs, leading to increased pressure in the pulmonary arteries. As a result the right side of the heart gradually enlarges, causing it to weaken and reduce its ability to pump enough blood for the body’s needs. The most common cause of death for PAH patients is heart failure.

“These advances mean a massive difference in survival rates as ten years ago someone diagnosed with PAH would only have a 30% chance of living for three years and we now have an 85% survival rate at three years,” Professor Keogh said.

Another interesting aspect of the change in approach to PAH is that there are less heart lung transplants as the medical treatment is better. “We used to do eight to ten transplants a year for PAH patients and today we do very few, which means there are savings in that those 30 organs can be used for other people,” Professor Keogh said.

Associate Professor Glenn Reeves, Staff Specialist in Immunology, Allergy, and Infectious Diseases at John Hunter Hospital in Newcastle said the estimated prevalence of PAH varies depending on the study, though there are between 500 – 1000 Australians diagnosed with the illness and many more are likely to go undiagnosed.<sup>1</sup>

“The way lives have changed for people with PAH is one of the really fantastic stories of modern medicine – we can now show hard and fast survival benefits and very strong symptomatic benefits where many people can return to their normal way of living and working,” Associate Professor Reeves said. “Getting an early diagnosis is vital”.

PAH can be associated with several different diseases, including:

- Scleroderma (also known as systemic sclerosis)
- Systemic Lupus Erythematosus
- HIV/AIDS
- Congenital heart defects
- Some liver disorders
- Use of certain drugs.<sup>2</sup>

These conditions represent risk factors for development of PAH, with up to 30% of lupus and scleroderma patients shown to have this condition.<sup>1</sup> Some of the early symptoms of PAH include shortness of breath during exertion or normal daily activities; fatigue or feeling tired all the time; dizziness, especially when climbing stairs or standing up and fluid build up in the limbs. In babies, the first signs may be difficulty with feeding, breathlessness, going blue and failure to gain weight. Often, adults do not take much notice of these early signs of PAH as they may think they are over-tired or lacking in fitness.

“Breathlessness is not OK,” Professor Keogh said. “It’s important that people with these symptoms see their doctor for a sound wave test on their heart and a set of lung function tests. PAH without treatment is more life threatening than breast cancer and more lethal than most tumours, yet it is easier and less painful to screen.”

November is PH Awareness Month in many countries and this year Australia is joining the fight to raise awareness of the condition and build funding to support finding a cure.

#### **Risk factors and treatment**

People need to be proactive and persistent in seeking an accurate diagnosis as breathlessness may be a symptom not only of PAH, but also other conditions requiring active treatment.

There are now three different classes of medication used to treat pulmonary hypertension which are effective in stabilising, and in many cases improving the disease.

Combining treatments is the way forward and new agents are also being investigated that may target cells in the blood vessel walls in the lungs that are responsible for the development of pulmonary hypertension.

Further information about pulmonary hypertension can be found at [www.phnsw.org.au](http://www.phnsw.org.au) and [www.phaaustralia.com.au](http://www.phaaustralia.com.au). To learn more about awareness week activities contact [enquiries@phnsw.org.au](mailto:enquiries@phnsw.org.au) or phone 02 8250 1150.

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**Issued on behalf of Pulmonary Hypertension NSW and supported by Actelion.**

#### **References:**

1. Collins N et al. European Journal of Echocardiology (2006) 7, 439e446
2. Australian Lung Foundation website, <http://www.lungnet.com.au/content/view/208/1/>, accessed 28 Oct 2008

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