Breathlessness or dyspnoea is a common symptom of a variety of conditions, such as asthma, pneumonia and emphysema. However, one of the most serious causes of breathlessness, which is difficult to diagnose, is the rare lung disease known as pulmonary hypertension (PHT). Basli (2015) reminds us of the importance of remaining vigilant for less common causes of unexplained breathlessness, so diseases such as PHT are not missed.

PHT is a rare but serious multifactorial group of disorders with no real cure and can affect people from any race, gender or age at any time. Simply, PHT is raised blood pressure in the pulmonary circuit, clinically defined as a resting elevation of the mean pulmonary artery pressure (mPAP) greater than 25 mmHg as assessed by a right heart catheter (Simonneau et al., 2009).

In its early stages, the symptoms can be subtle and non-specific. Breathlessness and fatigue may only be problematic with exertion, such as when walking up steep inclines, climbing stairs or even the simple task of wheeling out the refuse bin. Often this is not enough of a discomfort to prompt a patient to seek medical attention. Patients may interpret this breathlessness and fatigue as “normal” (Basli, 2012).

What is enough to prompt a patient to seek a medical assessment is an escalation of symptoms, often accompanied by other symptoms such as ankle oedema, dry cough, and syncopal and presyncopal episodes (Doughty and Mainwood, 2001).

With disease progression, pressure in pulmonary arteries slowly increases, ultimately leading to the development of right heart failure. Respiratory tests investigating causes of a patient’s breathlessness often include spirometry, arterial blood gas and chest x-rays, which are often normal and will miss PHT.

Patients may be prescribed inhaled medications commonly referred to as “puffers” erroneously. It is not until patients have had an echocardiogram and conformational right heart catheter that a diagnosis of PHT is made. Unless suspected, this can often have significant delays in diagnosis. Data from one study reported inappropriate or delayed therapy can be up to two years after onset of first symptoms (Rich et al., 1997, cited Strange, 2014, p.117). Early diagnosis and treatment is vital in managing this disease known for its rapidly progressive nature if left untreated.

**HISTORY OF PHT**

Prior to the development of heart catheterisation in the 1930s, there were only occasional observational accounts that describe PHT, as we know it to be today.

The first was in 1891 when Ernst von Röntgen noticed at autopsy, abnormal structural changes in the pulmonary vessels (Anderson et al., 2016). The second was in 1901 when Abel Ayera coined the term “cardio negro” (black heart) for a syndrome characterised with breathlessness, coughing, cyanosis and abnormalities in the pulmonary vessels. Thirdly, David Dresdale in the 1950s, with the application of heart catherisation, found increased pulmonary artery pressure in patients that had neither lung nor heart problems (Fosket and Boroumand, 2016). PHT was seeing us but we were only just starting to see it.

The catalyst that would finally catapult PHT onto the global arena was in 1955 when there was a surge in the number of diagnosed cases of PHT-pulmonary arterial hypertension (PAH) caused by the appetite suppressant, Aminox. This epidemic prompted the World Health Organisation (WHO) to hold its first PHT symposium in 1973, a year after Aminox was withdrawn (Anderson et al., 2016). The purpose of this meeting was to gather the experts with knowledge and experience related to PHT.

25 years after the first meeting, the 1998 PHT symposium proposed the PHT classification system referred to as the Evián Classification, named after the location of the symposium. The Evián Classification established individual categories of PHT, where within groups there was shared pathological and clinical characteristics and therapeutic options. PHT being a single disease, five groups of disorders were identified as causing PHT (Simonneau et al., 2004).

The PHT symposium continues every five years and the classification is reassessed each time. Refinements are made reflecting the knowledge that has accumulated. The current classification is the Nice Classification that was updated in 2013 and will be reassessed in 2018 (see the Nice Classification here: http://www.pah-info.com/Classification_of_PHT).
PULMONARY ARTERIAL HYPERTENSION GROUP ONE

One of the most severe forms of PHT is Pulmonary Arterial Hypertension (PAH) Group One in the Nice Classification. PAH is a rare but devastating progressive disease that ultimately leads to right heart failure and death. The incidence of PAH is approximately 2.4 cases per million annually and it is more common in women than in men (Noel et al., 2017). One of the five classifications of PHT, it is this group that has received the most research attention.

Knowledge of the exact trigger for PAH is still unknown but Matura (2011) suggests that there is the existence of a common pathway whereby structural and remodelling changes occur in the pulmonary vasculature. These changes are thought to be a result of sustained increase in pulmonary pressures and pulmonary vascular resistance (PVR) from a combination of in situ thrombosis, pulmonary vasoconstruction and the remodelling of the vascular wall (Matura, 2011, p.269).

PAH SPECIFIC THERAPY

The main treatment goal for PAH is to slow the progression of the disease (Doyle-Cox et al., 2016). Current PAH therapies have been developed to target the three main pathways involved in the pathogenesis of PAH. These pathways and the therapies available include:

- Endothelin receptor antagonists: Ambrodan, Bosentan, and Macitentan
- Prostacyclin: Epoprostenol, Iloprost
- Phosphodiesterase type 5 inhibitor: Sildenafil and Tadalafil

The number of medications available continues to grow at a rapid pace. The first medication was approved in Australia in 2004 and since that time, a further seven have been approved, with the recent approval of Remicigut, a novel therapeutic class of therapy.

Prior to the development of PAH-specific targeted therapies the average life expectancy was less than three years. The life expectancy today is around seven years (Gin-Sing, 2010).

The future looks positive because with greater research focus and interest, more medications are being developed. Combination therapy, involving one medication from each of the three pathways, is proving to be more effective in achieving treatment goals than single use therapy (Strange et al., 2013).

In Australia, patients can only receive one medication funded by the Pharmaceutical Benefit Scheme (PBS) at this current time. The PBS mandates the prescribing of PAH therapies to designated specialist centres only and there are strict patient criteria and conditions of eligibility. Only patients diagnosed with Group One PAH are eligible to receive therapy. Currently there are 60 specialised designated centres throughout Australia.

THE PHT NURSE

We have seen the development of other types of specialist nursing roles in the care of patients with breast cancer, diabetes and asthma but only recently the PHT nurse. The majority of PHT nurses are attached to specialist teams at designated PHT centres in both public and private locations. Most of PHT care provided at these centres is outpatient based.

Essential to the role is an advanced nursing knowledge of PHT and the skill to make complex decisions, crucial in supporting, teaching and caring for both patients and their families.

Time is spent with patients and families helping them to understand the PHT disease process, recognising signs and symptoms of worsening, self-monitoring and discussing the various treatment options.

PAH-specific therapy requires patients to have ongoing monitoring mainly though outpatient appointments with telephone support. The more complex PAH therapies, like intravenous or inhaled therapies, require substantial time teaching patients and carers about how to self-deliver these medications using aerosol or intravenous delivery.

PHT nurses educate other nursing colleagues who are usually unfamiliar with this rare disease and its complex therapy. Patients with PAH from time to time will require hospital admission for acute care and for many nurses this may be their first time caring for a patient with PHT. This can be daunting and challenging experience. The PHT nurse is a point of reference and a vital link for frontline nurses working in partnership with collaborative patient care.

With the complexity of this disease coupled with the changing regulatory PBS requirements, the PHT nurse is proving to be invaluable and essential in the delivery of high quality nursing care. This new role will continue to expand and develop keeping abreast with medical advances in PHT.

CONCLUSION

PHT is a rare, complex and multifactorial disorder, which has a devastating impact on a person's quality of life. PHT has witnessed major advances since the turn of the century and now commands global attention, and significant interest and awareness continues too.

Continually being challenged by a prognosis that remains poor, the future still looks promising, and further treatments and drug pathways are on the horizon.

The PHT nurse has a key role in supporting and caring for patients with PHT and their families, and this role will continue to expand and be recognised as a new nursing specialist role.

Unexplained breathlessness must always be investigated and the possible diagnosis of the rare lung disease of PHT must always be considered.

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AUTHOR

TARA HANNOH MACN