Wendy Gin-Sing

Personal statement regarding the use of pulmonary hypertension disease targeted therapies

I have been employed for the past seven years as a Pulmonary Hypertension Clinical Nurse Specialist at the Hammersmith Hospitals Trust, prior to this I worked for seventeen years within the cardiology setting, mainly on Coronary Care.

The Hammersmith Pulmonary Hypertension Service was designated by NSCAG in 2001, and currently consists of a Respiratory and Cardiology Consultant, two clinical nurse specialists, with a third joining the team next month, a full time Health Social Worker, three administrative staff and a part time hospital Chaplain. We also have two pulmonary hypertension (PH) research nurses co-ordinating clinical trials. The Hammersmith PH service has grown rapidly since designation when we had a patient population of 65, to seeing over 500 patients in the last financial year, of which 220 were new referrals. The majority of our patients are referred in WHO functional class III or IV being breathless on minimal exertion and sometimes at rest, with PH significantly affecting all aspects of their daily life. It is therefore essential that we diagnose and treat these patients without delay as the mean life expectancy from time of diagnosis without treatment is less than 3 years.

We regularly use all the PH technologies that are being assessed and Treprostinil in subcutaneous and intravenous infusions. The clinical decision as to which drug therapy is used depends on the individual patient – the severity of the disease, contraindications, published treatment algorithms, the ability to manage infusion therapy, and patient choice.

I feel strongly that patients should be educated about PH and well informed about treatment options to enable them to make decisions regarding their health and wellbeing. The majority of our patients will start therapy with oral disease targeted therapy, currently the first line treatment is Bosentan, unless there are contraindications, in which case we use Sildenafil. A number of patients who have experienced a good response to Bosentan but have developed a transaminitis, have been commenced on Sitaxentan.

It is essential to closely monitor patients on disease targeted therapies as these drugs do not cure the disease, but can improve symptoms and limit time to clinical worsening. Patients that are deteriorating need to be assessed and their treatment regime adjusted to ensure the best possible outcome, this often results in the use of combination therapy with several different agents. Nebulised Iloprost is most commonly used in combination with an oral therapy, rather than as a monotherapy as the patient is not treated overnight.

Patients in WHO functional class IV are ideally commenced on a continuous prostacyclin infusion, however some patients are unable to manage this independently, or refuse to undertake such invasive therapy this is not possible. When the patient needs emergency treatment we use intravenous

Epoprostenol because of the short half life of 3-4 minutes, and the ability to increase/decrease this rapidly according to the patient's condition. Once they have been stabilised this is then usually transitioned to either intravenous or subcutaneous Treprostinil as this is easier for the patients to manage at home as it has a longer half life of 3-4 hours and needs changing every 48 hours, instead of every 12 hours with Epoprostenol.

The PH population is heterogeneous, with no two patients, in the 900 seen here since designation, following exactly the same disease course. It is therefore essential to have a variety of disease targeted therapies available as it is difficult to predict how well a patient will respond to a particular therapy and for how long it will remain effective at improving symptoms.