New developments in treatments and treatment strategies for pulmonary hypertension

The two largest congresses in the fields of cardiology and respiratory medicine have taken place recently and it was very encouraging to see that a number of sessions were dedicated to pulmonary arterial hypertension and to chronic thrombo-embolic pulmonary hypertension (CTEPH), featuring some of the world’s leading experts as speakers or chairpersons. Recent clinical trials were presented and it is likely that treatment strategies may change as new insights into the disease were revealed.

Annual congress of the European Society of Cardiology

Barcelona (Spain) was the venue for this year’s ESC annual congress, which took place from August 30 to September 3 and was attended by 30,000 participants from all over the world. PHA Europe was represented by Pisana Ferrari. As in past years, there was considerable interest on the part of the scientific community for the field of PH, as demonstrated by the six dedicated scientific sessions and 200 posters. The sessions were extremely well attended and many had an audience of over 500, with people standing at the back of the room for lack of seating space. Different aspects of the disease were analysed in the various sessions, including pathophysiology, genetics, screening of populations at risk (in particular scleroderma), diagnostic work up, new drugs and future treatment strategies. The clinical trials leading up to the recent approval of two new drugs were examined in great detail: Macitentan-Opsimut, a new endothelin receptor antagonist (“ERA”), and Riociguat-Adempas, a soluble guanylate cyclase stimulator (“sGC”) which acts on the nitric oxide pathway. There was a lot of interest for the GRIPHON clinical trial on selixipag, an oral prostacyclin receptor agonist which targets the prostacyclin pathway. The preliminary results of the GRIPHON study are extremely promising and may pave the way to a new era of oral prostanooids.

There was also much discussion of what the new ESC/ERS clinical practice guidelines for PH, which are due out next year, will look like, in particular with regard to the treatment algorithm. There is much expectation for this and it is likely that there will be quite a number of changes. Importantly for us psycho-social support for patients has been given a higher grade of recommendation in the “general measures” section. We have been advocating for the recognition of the need for psycho-social support for a long time, with our “Call to Action” (presented at the European Parliament in 2012) and the findings of the International Patient and Carer survey (presented at three key congresses in 2013-14 as a scientific poster). Lung transplant surgery has also been moved slightly higher up in the algorithm to avoid patients being listed too late, which can seriously compromise their capacity to recuperate post surgery. Other changes discussed related to the use of “combination therapy” (concomitant use of two or more PAH specific drugs, see below ERS report).

Annual congress of the European Respiratory Society

Pisana Ferrari, Juan Fuertes and Christian Richter, from phev, Germany, represented PHA Europe at the ERS annual congress, held in Munich from September 6 to 10. The congress was the largest to date, with 22,000 delegates. A number of scientific sessions were dedicated to pulmonary hypertension (these were also very well attended) and there were numerous poster presentations which also covered quality of life and emotional (eg. depression) issues.

The main highlight of the ERS congress for the PH community was the presentation of the results of AMBITION, a large, long term clinical trial investigating the benefits of “upfront” combination therapy with Ambrisentan-Vorlibris, an “ERA”, and Tadalafil-Adcirca, a “PDE-5 inhibitor” which acts on the nitric oxide pathway. Previous studies had analysed only “sequential” combination therapy, where patients start with one drug and a second one is added at a later stage if treatment goals are not met. Three of these studies have proven substantial benefits sustained over time. The AMBITION study shows even more dramatic improvements with a 50 percent reduction in risk of clinical failure in the group having received both drugs right from the start as opposed to the ones on monotherapy. The rationale of this new approach is that the severity of the disease and its progressive nature may justify a more aggressive “hit early, hit hard” strategy. We now look forward to seeing how the new and exciting findings from these congresses will translate into future treatment management. Upfront combination therapy could potentially become the new standard of care for PAH.

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