Pulmonary Arterial Hypertension: A New Challenge

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Pulmonary hypertension represents a new challenge for the current clinical practice, as recent advances in the field of diagnostic technologies led a to significant increase in the number of patients diagnosed with this disease and in the same time new therapeutic classes proved to have a significant role in improving survival of these patients.

The main types of pulmonary hypertension are the idiopathic form, called primary pulmonary arterial hypertension (PAH) and the secondary one, developed in the evolution of a cardiac disease, in many cases consequence of a congenital heart disease. Once occurred, PAH severely limits the life expectancy. Despite the recent introduction of many new therapeutic agents which are expected to have a revolutionary role in the improvement of life expectancy and quality of life of these patients, PAH continues to be associated with a mortality as high as 35% at 3 years after the diagnosis. The new therapies are mainly represented by vasodilators that target the nitric oxide, endothelin and prostacyclin pathways.

The diagnosis of PAH is usually established when a typical pattern of pre-capillary pulmonary hypertension is encountered in the hemodynamic profile, in the absence of any lung disease.

PAH is associated with a process called pulmonary vascular remodelling, represented by inflammation and angioproliferation, process which is difficult to assess in living patients. The process of vascular remodelling leads to a significant obliteration of the pulmonary vascular bed and therefore an important progress in the field of PAH diagnosis would be represented by the ability to detect the presence of obliteration of the pulmonary vascular bed and their extent.

A correct assessment of pulmonary hypertension severity should be based on histological examination of the pulmonary tissue and different techniques have been proposed to obtain samples of pulmonary tissue, such as transbronchial or surgical biopsies, which carry a high risk of procedural complication and therefore are rarely used. Cryobiopsy is a recently developed minimally invasive method to obtain lung tissue, still not being used on a large scale in the clinical practice.

However, it should be emphasized that monitoring PAH patients is extremely important for the correct management of these patients and should be based on the assessment of the primary disease process in the PAH lung vasculature. As a consequence of the difficulties encountered in obtaining samples of pulmonary tissue, this monitorization in clinical practice is based rather on indirect estimates of the effect of treatment on the clinical status of the patients, using pressure measurements in the right heart, evaluation of right heart function and evaluation of exercise capacity. Right heart catheterisation remains the golden standard for diagnosis of PAH and for its monitorization. Recently, in order to optimize the methods to monitor patients, cardiac magnetic resonance has been introduced as a non-invasive tool, with still inconclusive results.

In cases of pulmonary hypertension associated with congenital heart diseases, the reversibility degree is extremely important for establishing the indication for surgery, as surgical correction of the underlying disease could be performed only in early stages, while alterations of the pulmonary vascular bed are still reversible.

An alternative route to obtain sample of pulmonary tissue for histological analysis could be represented by the transvascular pulmonary puncture biopsy, using device patented in 1991. The method implies right heart catheterisation and advancement of the interventional device distally into the branches of the pulmonary artery, followed by vascular puncture and collection of pulmonary tissue samples [1]. The advantage of the method is that can be performed during the cardiac catheterisation, which is anyway performed in these patients, for both diagnosis and monitoring.

A study published in this number reveals the role of non-invasive diagnostic test for monitoring patients with PAH, including exercise capacity measured by the 6-minute walk test, biomarkers such as Brain Natriuretic peptide, electrocardiography, spirometry, and echocardiography. The authors showed that all these methods and especially echocardiographic evaluation of right heart hemodynamics based on the assessment of tricuspid regurgitation velocity, could be useful in the clinical practice for follow-up of the patients with PAH [2].

The authors underline the role of non-invasive tests for an appropriate management of patients with PAH and taking into consideration the need for identification of non-invasive, repeatable methods to be used in the follow-up of tese patients, the study is extremely valuable. However, it should not be forgotten that an acurate assesment of the pulmonary hypertension reversibility degree can be based only on direct histologic assesment of the samples

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of pulmonary tissue. The above mentioned transvasculary pulmonary puncture-biopsy could present in these circumstances an alternative and challenging method, carrying a significant added value for a correct estimation of the severity degree of PAH. In the hands of an experienced interventionist operator, the method is safe and effective for obtaining adequate samples of pulmonary tissue and could be indicated for monitorization of PAH reversibility degree in PAH, along with right heart catheterisation.

References

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