New Options for Risk Assessment in Pulmonary Hypertension

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Pulmonary arterial hypertension (PAH) is a rare but not orphan disease with a prevalence of about 15 - 60 /Million in western industrialized nations. Whereas pulmonary hypertension (PH) other than Group 1 (WHO classification) is a more frequently observed condition in different clinical settings. Derived data form the United Kingdom estimate a prevalence of almost 100/Million. The most common cause of PH will be found in patients with left heart disease (Group 2). Due to the diversity in pathophysiological pathways of PH patients face a broad spectrum of risk to deteriorate in their clinical status.

A proper diagnosis of pulmonary hypertension can be only made with right heart catheterization in order to measure the magnitude of pulmonary pressure and just as important to define the vascular bed in which the high originate (pre or post - or combined pre and post capillary).

However, when pulmonary hypertension is diagnosed and the classification group defined, the focus will be put on risk classification taking in account diverse and sophisticated imaging techniques and clinically and laboratory assessed variables. For patient with PAH the ESC guidelines recommend a comprehensive risk using 13 variables an in the manner of a traffic signal divide the risk in low (green) intermediate (yellow) and high (red) estimating a one year risk of death of < 5%, 5 - 10% and > 10% respectively [1]. The risk groups were set up from result of different outcome studies. For this risk assessment different variables like NYHA FC, NT-pro-BNP, clinical signs of right heart failure, CPET, imaging or hemodynamic measures are used. No single variable of this panel is perfect in predicting the risk. This rise the question, which combination may be best using reliable but readily variables and when to start or modulate therapy? In the last years a huge input of research was set by searching for risk calculation and imaging modalities to optimize risk prediction and treatment strategies. At time the French, Swedish and COMPERA registries more or less validate the ESC/ERS (European Society of Cardiology/European Respiratory Society) PH guidelines risk assessment strategy but none of this risk calculations are using parameters deriving form imaging. The COMPERA registry, an ongoing web-based PH registry was launched in 2007. It collects baseline, follow-up and outcome data only in treatment naïve PAH patients. Data from this registry were used in an abbreviated version of the risk assessment strategy of the European PH guidelines [1]. Instead of 13 variables outlined in the European PH guidelines, COMPERA uses only 6 variables (NYHA-FC; 6-min walking test, BNP or NT-pro-BNP, right atrial pressure, cardiac index and mixed venous oxygen saturation). Data from 1588 patients were analyzed, much more than in the Swedish registry [2]. The reduced set of variables in the COMPERA registry turned out to accurately match with the ESC/ESR PH risk assessment strategy and effectively separated patients in low, intermediate and high risk of death with above the mentioned one year mortality rates. The risk stratification was valid for baseline and follow up assessment which would allow a dynamic approach in the treatment strategy. The main objective in the current PH guidelines is achieving or holding a low risk profile which presume timely follow up. In the French registry even less variables were used to serve in a simplified risk assessment (NYHA-FC; 6 min walking test, right atrial pressure and cardiac index) [3]. Thereby each of the low risk criteria predicted outcome after first re-evaluation within a median follow up time of 34 months. Again, no variables derived from the diverse available imaging modalities were used.

Unquestionable the right heart determines the prognosis of PAH patients and is an unfavorable prognostic marker in PH patients other than group 1 [4]. Echocardiography permits deep insight in the structures and function of the right ventricle. Many prognostic markers

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are extracted from echocardiographic examinations and some newer echo methods like Strain and 3-D echocardiography are likely to be useful in future. Right atrial pressure, stroke volume and right ventricle volumes sufficiently describe right ventricle function and echocardiography is capable to measure this functions. Right heart reverse remodeling seems to be a strong indicator of effective therapy. Recently right ventricle end-diastolic area, right atrial area and left ventricle systolic eccentricity index were found to give an excellent measure for reverse remodeling and this quality was strongly associated with survival in patients with pulmonary hypertension. Furthermore reverse remodeling was only achieved when pulmonary vascular resistance could substantially be reduced [5].

However cardiac magnetic resonance imaging (CMR) may overcome some problems in echocardiography like a better visibility of the whole right ventricle by measuring volumes and stroke volume and could even better indicate reverse remodeling of the right heart. Results from studies dealing with CMR in pulmonary hypertension promise this imaging as an important modality for risk assessment and treatment strategy.

In summary, ESC PH guidelines recommended risk stratification is well validated by diverse risk assessment strategies. To date probably at best with the COMPARA risk assessment. This assessment however, is dependent from invasive measures but does not use variables derived from imaging. Resent research demonstrate promising results from imaging surveys which excellently associate with clinical risk and right ventricle remodeling. Therefor future studies will combine simple imaging measures with an even more abbreviated version of the ESC/ERS PH guidelines risk stratification tool. The goal will be a sample of easy derived parameters which are valid for risk assessment but also timely detect functional worsening associated with risk for morbidity and mortality.

Conflict of Interest

I don't have any conflict of interest regarding this editorial.

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