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The Real Landscape of Pulmonary Artery Hypertension Associated with Congenital Heart Disease in Vietnam National Heart Institute – Bach Mai Hospital – Vietnam

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Abstract:

Aims: The purpose of this study is to present an overall informtion on the characteristics of patients living with pulmonary artery hypertension associated with congenital heart disease (PAH-CHD) and the current management of this disease in Vietnam.
Methos: This study inclued a cross- sectional restrospective study on 96 patients from April 2014 to July 2015 and a cross- sectional prospective study on 121 patients from January 2016 to July 2017 who were diagnosed PAH- CHD and treated in Vietnam National Heart Institute, the National Hospital Terminal of the North of Vietnam. Both incidence and prevalence cases were included. We not only reported the clinical and physiological chracteristics at the time of diagnosis for this cohort of patients but also described the management strategy of these patients in Vietnam.
Reults: A total of 217 patients were identified as having PAH – CHD with 36 mean age, 73% female. The ASD was the most common pathologic classification with 51%. At the time of enrollment, 19% of patients were diagnosed Eisenmenger. 6 MWD, proBNP, RA area, TAPSE usuaslly were used in assessments the severity and the risk of death of all PAH patients, there was 63% patients with low risk, 25% patients with intermediated risk and 11% patients with high risk, the death rate in hospital was 7.3% with the most common trigger was pregnancy. The high risks for pulmonary thromboembolism in ES group were described as hypervisocoity, NYHA IV, high CRP, large dilated pulmonary arteries, and dual ventricular dysfunction. In this cohort patients, we performed the corrective treatment for 28% patients, and the 72% remaining patients were treated with PAH specific therapy and would be reassessed after 3-6 months duration. PGEi5 was the most mommonly used target therapy (36%) followed by the combination of ERA and PGiE5 (14%).
Conclusion: Our patients are detected at the advanced stage of the disease, and thus the mortality is still unacceptably high. The rate of the PAH patient group could not be completely corrective treatment was still high and we lacked of special procedure to correct the defects beside special PAH therapy .

Keywords: Pulmonary artery hypertension, congenital heart disease, Eisenmenger syndrome.