Impact of triple vasodilator therapy on prognosis stratification of pulmonary arterial hypertension associated with congenital heart disease

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Background: Pulmonary arterial hypertension (PAH) affects 5-10% of congenital heart disease (CHD) patients but there is limited information on the effect of combination therapy in these patients. The aim of this study was to assess the effect of triple therapy (TT) with prostacyclins on the risk status of patients with PAH-CHD.

Methods: A retrospective, longitudinal, cohort study of patients with PAH-CHD under active follow-up in our center. All patients were receiving baseline dual therapy at maximum doses. A simplified global risk assessment according to the 2022 PAH guidelines was performed prior to initiation of treatment, and at subsequent annual follow-up for 2 years, including functional class, 6-minute walk test and NT proBNP.

Results: 60 patients were included (median age 41, 31 women 61%). 32 had Eisenmenger syndrome, 9 had coincidental shunts, 18 had postoperative PAH, and 1 had a significant L-R shunt. TT was initiated based on the clinical situation, existence of shunt and ease of administration (epoprostenol 20%, treprostinil 41%, selexipag 32%, iloprost 7%). The use of triple therapy in patients with PAH-CHD demonstrated a significant overall improvement in risk status one year after initiation of treatment (p<0.05), which was maintained two years after initiation (Figure 1). Only four deaths and one lung transplantation occurred within this intermediate-high baseline risk PAH group. The estimated 2-year survival among those who completed treatment was 90%.

Conclusions: The use of triple therapy vasodilator treatment in patients with PAH-CHD demonstrates an overall improvement in risk status maintained over 2 years in line with that observed in other types of PAH.

Evolution of the risk situation