Pulmonary Vasodilators Treatment in Failing Fontan: Data from the Spanish Registry for Pediatric Pulmonary Hypertension (REHIPED).


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INTRODUCTION: Protein losing enteropathy (PLE) and Plastic Bronchitis (PB) are a growing cause of morbidity, as survivors of the Fontan operation continue to age. Both disorders can lead to death, describing a poor prognosis, with a reported 50% mortality at 5-years after diagnosis. Limited data suggest the successful use of pulmonary vasodilators (PV) as a treatment strategy. Our purpose was to study the clinical/hemodynamic findings and survival of patients who had received PV for PLE/PB.

METHODS: From 2009 to 2014, 26 Fontan patients receiving PV following diagnosis PLE/PB (84% PLE) were identified from clinical database at the Spanish Pediatric Pulmonary Hypertension Registry. Data were collected retrospectively.

RESULTS: Mean age at PLE diagnosis was 10.4 (5.1-20) years. Fontan operation was performed at 5.9 (2.4–8.7) years of age. Fontan operation to PLE/PB diagnosis was 4.5 (0.2–13.2) years. Diagnosis of PLE/PB to PV started was 8.4 months. Transplant-free-survival or death was 88.5% at 5-years. Treatment achieved complete resolution of clinical symptoms and laboratory values in 6 patients (23%). Symptomatic and functional class improvement was achieved in 16 patients (61%) at 6-months follow-up, but this symptomatic improvement lasted only in 7 patients (27%) at 12-months follow-up, and 6 patients (23%) at 24-months follow-up. Patients who died or received a heart transplantation (HT), had lower cardiac index (2.36+/-.3 vs. 3.47+/-.1 l/min/m2;p=0.002), lower mixed venous saturation (Sv02) (67.5% vs. 73%;p=0.005), and NYHA functional class >2 at diagnosis, and later they began therapy with PV (2.8 vs. 0.6 years post-diagnosis,p=0.042). In 12 patients, in which clinical outcome was not satisfactory, an hemodynamic assessment was repeated. Only significant change was observed in the Sv02 (Pre-treatment:67% vs. Post-treatment:72%;p=0.03). However, a trend towards decreased central venous pressure and decreased cardiac index, as a result of intensive management after PLE/PB diagnosis, was observed.

CONCLUSIONS: PV have potential to improve symptoms and delay need for HT in this patient population. Although, the success rate of PV (associated to conventional therapy) was not superior to the previous reported series. As resolution always happened in the first 6-12 months after treatment, HT should be considered in any PLE/PB when there hasn’t been response after that period.