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15 Year Experience in Pulmonary Hypertension Due to Congenital Heart Disease Before and After Targeted Therapies: The Durability of the Right Ventricle in this disease

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Introduction: Pulmonary arterial hypertension (PAH) associated with congenital heart disease (CHD) is characterized by severe increase in pulmonary pressure and resistance with relatively preserved RV function and significantly better prognosis compared to idiopathic PAH. The aim of this study was to examine the 15 year single-center experience in this population before and after use of PAH targeted therapies.

Methods: Between 1995 and 2011, 75 patients, aged at diagnosis 21±16 years, with PAH due to CHD were followed at our center. Diagnoses were: atrial septal defect 3 patients, ventricular/atrioventricular septal defect and/or arterial duct 30, complex CHD 37, Fontan 5. Five patients had Down syndrome while 20 had previous heart surgery. Patients were followed with clinical exam, EKG, echocardiogram, 6-min walk test and cardiorespiratory exercise test every 6 months with laboratory tests every 3 months.

Results: Over 15 years (mean follow-up 8±5 years) 96% of patients received PAH targeted therapy after 2002, initially monotherapy with second therapy added in 8 patients due to clinical worsening (3 patients improved, 5 progressed to death). Most (75%) patients remained stable clinically (WHO Class II) and in exercise capacity, while 14 (19%) deteriorated gradually to Class III. All stable patients had no clinical or echocardiographic signs of RV failure nor did they require treatment for it despite the long follow-up. There were 14 (18.6%) deaths (5 sudden, 9 due to worsening RV failure) at age 27±19 years, 2.9±2.3 years after initiation of therapy. PAH therapy was discontinued in 2 patients after surgical and interventional treatment respectively. All Fontan patients improved significantly with PAH therapy and are expected to discontinue it in the future.

Conclusions: Patients with PAH due to CHD, especially in the era of the new PAH targeted therapies, remain more stable with better prognosis than idiopathic PAH without significant RV failure over long follow-up periods, possibly because of RV adaptation since fetal life. Still, PAH due to CHD remains a complex disease with significant mortality and morbidity. Close follow-up and individualization of therapy seems critical for these patients' quality of life and long-term survival.