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Effect of pulmonary vasodilatators on clinical and Echocardiographic parameters in children with primitive pulmonary hypertension

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This study was to assess the influence of anti-PHT medications on clinical and echocardiographic parameters in children.
Methods: Patients <18years diagnosed with primitive PHT were retrospectively reviewed. Functional class, RV and LV diameters, tricuspid regurgitation grade, systolic and mean pulmonary pressure, TAPSE and TAC were recorded before and after anti-PHT treatment. Deceased or transplanted patients (group I) were compared to survivors (group II).
Results: 16 patients (11 males), were diagnosed with PHT (11 primitive and 5 illegitime association: closed VSD= 2, closed PDA= 2, TGA= 1), at mean age of 6.7years (0.7 to 16.2). All received anti-PHT treatment: monotherapy (IV prostacycline= 4, bosentan= 2), or bitherapy (IV prostacycline + bosentan= 2, bosentan + sildenafil= 7) or tritherapy (IV prostacycline + bosentan + sildenafil= 1). Mean FU was 4.3years (0.7 to 10.5). Five patients died, 1 underwent heart-lung transplantation (group I= 6) at mean FU 3.4y (1 to 4.1), and 10 survived (group II) at mean FU 4.7y (0.8 to 9.5).
NYHA class changed from 50% class II, 37.5% III and 12.5% IV before treatment, to 50% class I, 12.5% II and 37.5% IV after.
Overall mean RV/LV did not change (1 to 0.95). RV/LV decreased in group II: 1.14 to 0.51, and increased in group I: 0.87 to 1.57. The proportion of TR grade 0-I increased from 47% to 60%, and of TR grade II-III decreased from 53% to 40%. All patients with TR grade III at end FU ranged in group I. Systolic and mean pulmonary pressure did not change: respectively 92.1 to 99.9mmHg and 65.8 to 56.2mmHg, and were similar between groups I and II.
TAPSE and TAC were not available in group I, and increased in group II: respectively from 14 to 17.9mm and 76.8 to 101.6ms.
All patients in group I were either in NYHA class IV at first presentation (2cases), or received first therapy more than 6 months after diagnosis (4cases).
Conclusion: Anti-PHT therapy contributes to ameliorate functional class and RV function despite no significant effect on pulmonary pressure level. Persistent TR grade III and increasing RV/LV seem to be factors of bad prognosis.