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Experience with restrictive cardiomyopathy in childhood age

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The objective of this study was to assess the clinical outcomes of children diagnosed with restrictive cardiomyopathy (RCM).
Material and methods: Retrospective data analysis of patients diagnosed with RCM at age less than 18y. Age at diagnosis, clinical oucome, therapeutics and survival were collected. Prognosis factors were assessed.
Results: From 1991 to 2016, 24 patients (15 males= 62.5%) were diagnosed with RCM, at the age of 5.5±4.8y (median 4.2y, range 1week to 14.8y). All patients had normal LV and RV systolic function. Events occurring over time included : supraventricular tachycardia in 5 cases (20%), atrioventricular block in 1 (4%), right heart failure in 10 (40%), stroke in 1 (4%) and syncope in 3 (12%). Pulmonary hypertension occurred in 4 cases (16%). Follow-up was uneventful in 4 patients at the time of analysis (16%). Treatment included : diuretics in 13 cases (52%), beta-receptor antagonists in 11 (44%), amiodarone in 4 (16%), and 2 patients had no treatment (8%). NYHA class I, II, III and IV range was respectively : 2 cases (8%), 11 (44%), 4 (16%) and 7 (28%). Death occurred in 5 cases (20%), at the age of 2±1.7y (median 1.5y, range 0.5 to 4.8y) and 0.6±0.7y after diagnosis (median 0.5y, 1week to 2y), from HF in 1 and sudden death in 4. Ten patients (40%) underwent heart transplantation, at the age of 12.5±6.7y (median 14.8y, range 3.7 to 22.1y) and 5±7y after diagnosis (median 2y, 0.6 to 19.4y). Overall death or transplantation occurred in 60% of the cases. Age at diagnosis was a predictive factor for death (1.4y in deceased patients versus 6.6y in alive cases, p= 0.03).
Conclusion : RCM prognosis is poor in children with high rates of death or transplantation. Infant age at diagnosis is a predictive factor of bad outcome.