MP3-8

Short-Term and long-Term Outcomes of Dilated and Non-compaction Cardiomyopathy Presenting during Childhood in West Sweden 1990-2015

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Objectives: To study the incidence, mortality and morbidity of dilated cardiomyopathy (DCM) and non-compaction of the left ventricle (LVNC) in children in West Sweden.
Materials and Method: Hospital records of children and adolescents, 0 – 18 years, diagnosed with DCM and/or LVNC over a 25-year period in Gothenburg region, were reviewed. Inclusion criteria for DCM were LV shortening fraction (LVFS) < 27% and symptoms of congestive heart failure. In LVNC inclusion criteria were according to international definition of the disease. Clinical data including echocardiography findings were registered. Survival was cross-checked against the Swedish Population Registry in October 2015. Standard statistical measures determined survival, normalization of cardiac function and morbidity in those with and without transplant or death.
Results: Sixty-nine patients were identified; 42 (61%) males and 27 (39%) females. The combined incidence of DCM and LVNC in children and young adults during this time period was 0.55 per 100 000 per year. Mean age at diagnosis was 6.7 years (1 day – 17.9 years). Eleven children (16%) presented before 4 weeks of age, 7 children (10%) between 1 month and 1 year, 21 children (30%) between 1 and 10 years and 30 children (43%) presented between 10 and 18 years. In the latter group most children also had a neuromuscular disease. Most patients had reduced ventricular function with a left ventricular shortening fraction (LVFS) < 27% at diagnosis (n= 43; 62%). The remaining patients with a LVFS >= 27% (n=26; 38%), mainly had LVNC (n=14) or were patients with left ventricular dilatation detected during screening for familial DCM. Transplant-free survival was 88.5%, 83,6% and 80,2% at one, two and five years after diagnosis, respectively. Combined mortality and transplant rate was 23.1% over the studied period.

Conclusion: The incidence of DCM and LVNC (0.55/100 000/year) in West Sweden was similar to reports from other countries. The majority of children with idiopathic DCM presented during the first year of life. Mortality was highest during the first year after diagnosis. The majority of children with DCM associated with other diseases were diagnosed beyond 10 years of age.