Impact of pediatric heart transplant program on the outcome of dilated cardiomyopathy

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Introduction:
Pediatric dilated cardiomyopathy (DCM) is a multifactorial, progressive disease. Heart transplantation (Htx) is an acceptable treatment option for children with end-stage heart failure resulting from DCM. Impact of introduction of a pediatric Htx program (2007) was analyzed.

Patients and methods:
82 children (female/male:35/47, age at time of diagnosis:0-17 yrs, weight:2-105 kg) with DCM (LVDD>2SD and/or LV-FS<30%) were enrolled between 2000 and 2014 in our center. Tachycardia-induced (6), pacemaker-related (4), anthracycline-induced (2) and muscular dystrophy associated (1) cardiomyopathies were excluded. During follow-up medical treatment, time from diagnosis to end-stage heart failure (HF) need for VAD therapy and htx results were examined.

Results:
Myocarditis was confirmed in 10 cases (8 viral, 2 autoimmun, 10/69, 14.5%), 9 (90%), of them recovered, Htx was necessary in 1 case. Idiopathic DCM was diagnosed in 59 cases, 13 of them were familial (13/59, 22%). In the later group diagnosis was established at younger age (4.1 yrs versus 5.6 yrs). Medical treatment included ACEi (57/59, 97%), beta-blocker (44/59, 75%), aldosterone receptor antagonist (41/59, 69%), furosemide (50/59, 85%), digoxin (34/59, 58%) and anticoagulation (31/59, 52%). Antiarrhythmic drug and ICD implantation were rarely used (amiodarone 4/59, 7%, ICD 7/59, 12%). All pts with end-stage HF before the Htx program died. After 2007 27 pts required htx with/without VAD. Time from diagnosis to end-stage HF was 2.3 yrs (0-12 yrs) regarding all 27 idiopathic cases. Despite introducing Htx program, 4 pst died while waiting (4/27, 15%). 15 children were transplanted without VAD therapy (15/27, 55%), in 8 cases VAD therapy was necessary (8/27, 29%). 2 children died during VAD treatment (2/8), 6 were successfully transplanted (6/8). Among the total of 21 Htx patients 2 pts died in the post-transplant period.

Conclusions:
1. Before the introduction of Htx program all pts with end-stage HF died. 2. The progression of pediatric DCM is rapid. 3. Need for ICD implantation is very rare in childhood. 4. Since the introduction of Htx program almost all pts with end-stage HF were successfully transplanted. 5. VAD treatment is a necessary component of any pediatric Htx program. 6. Mortality after htx is acceptable.