

Long-term experience with Heart Transplantation in children and patients with congenital heart disease

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This study is to assess the long-term outcome of heart (HTx) and heart-lung transplantation (HLTx) in patients with congenital heart disease (CHD) and children with non-congenital cardiac or pulmonary disease.

Methods : the study is a retrospective single-centre analysis of long-term outcome after heart transplantation of children and adults with CHD, and children with non-congenital cardiac disease.

Results : From 1984 to 2013, 111 first-HTx, 5 HLTx and 6 second-HTx were performed (62males), in patients aged $11.7\pm 8.2y$ (med12y): 96(79%) were aged <18y at transplant. Cardiac diseases included 61 cardiomyopathies(50.8%), 50 CHD(41.7%), 6 retransplants(5%). HLTx included 1 Eisenmenger syndrome, 1 primitive pulmonary hypertension, and 2 pulmonary diseases. Patients with cardiomyopathy were younger than those with CHD (8.7y vs 14.9y). Seventeen (14%) patients were under circulatory mechanical support as a bridge to transplant. Immunosuppression was achieved by tritherapy in the majority of cases. Acute rejection occurred more frequently within the first year post-transplant (> 5th year) or late after transplant in teenagers because of non-compliance. Overall mortality was 27% : 33patients died, $3.5\pm 4.6y$ post-HTx (1 day to 16.4y, med 1.5months), due to : early multivisceral failure in 6 (18%), Pulmonary hypertension in 3 (9%), acute rejection in 7 (21%), graft coronary disease in 6 (18%), sepsis in 5 (15%) and miscellaneous in 6. Graft coronary disease occurred in 15cases (12.4%), of them 4 had 2nd HTx, 6 died and 5 are alive. Five posttransplant lymphoma occurred, 4months to 14y after HTx and were successfully cured in 4 (1died). Patients survival was 85% at 1y, 81% at 5y, 70% at 10y and 61% at 20y post-transplant. Graft survival rates were respectively 82%, 68% and 52% at 5y, 10y and 20y post-transplant. Survival did not differ with pretransplant disease, age, gender, pretransplant mechanical support. Mortality was higher in patients with coronary disease (40%) than those free from (25%). The majority of survivors (97%) are in NYHA class I.

Conclusion : Our experience with HTx and HLTx is favourable with acceptable long-term prognosis in patients with end-stage disease. Graft coronary disease is the main cause of failure, less frequent than in the adult non-CHD heart-transplanted population.