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Heart Transplantation in Adult patients with Congenital Heart Disease

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Since 85% of patients with Congenital Heart Disease (CHD) can currently survive and reach adulthood, Heart Transplantation (HTx) is becoming a novel challenge in adults with CHD (ACHD). The aim of this study was to assess results and outcomes of HTx in ACHD.
Material and methods: Clinical data, demographics and outcomes of patients with underlying diagnosis of CHD transplanted at ≥18years of age, were retrospectively reviewed.
Results: Among --- adult heart transplantations performed from 1991 to 2011, 20 recipients were ACHD (13 males). Age at transplant was 21.6 ± 4years (median 19.6). Sixteen of 20 (80%) were performed since 2000 and only 4 (20%) before. CHD included: double inlet single ventricle (10 cases), tricuspid atresia (2), transposition of great arteries (2), unbalanced atrioventricular septal defect (2à and miscellaneaous (4 cases: 1 Ebstein anomaly, 1 HLHS, 1 ALCAPA, 1 double outlet right ventricle). Seventeen had at least one previous cardiac surgery (85%), i.e: Fontan procedure in 5 (25%), palliative procedures in 10 (50%) and Mustard operation in 2 (10%). Three cases had no previous cardiac surgery (15%), i.e: 1 Ebstein, and 2 unbalanced AVSD.
Two early deaths occurred at day-1 and day-23 post-transplant, due to pulmonary hypertension and graft RV failure. One late death occurred at 9.6 year FU post-transplant, and one patient underwent re-HTx at 6-year FU, due to graft coronary artery disease. Follow-up post-transplant was 6.4 ± 5.3 years (1 to 20.9, median 5.3). Survival rates were 90% at 1 and 5-years post-transplant and 72% at 10 and 15 years. All survivors (17 cases= 85%) are doing well in NYHA class I. Overall < 1year mortality. Fontan procedure, gender, pre-transplant status did not impact significantly on survival.
Conclusion: The results of this small sample size study shows that HTx is an acceptable option for ACHD patients with end-stage failure of myocardial function and no other therapeutic alternative.