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Outcome After Transplantation in Patients With Eisenmenger Syndrome

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Introduction:
Eisenmenger syndrome (ES) is the gravest expression of any unrepaired congenital cardiac defect with a systemic-to-pulmonary communication leading to pulmonary arterial hypertension (PAH), bidirectional or reversed flow, and cyanosis.
Novel therapy with pulmonary arterial hypertension (PAH)-targeted therapies offers long-term benefits for exercise capacity, clinical symptoms and possibly survival, however transplantation (Tx), either as combined heart-lung-transplantation (HLTx) and single/double-lung transplantations (LTx) with concomitant cardiac repair, is still the only curative treatment option available.
The aim of the present study was to evaluate Nordic patients with Eisenmenger syndrome who have undergone Tx from 1985-2012 and to describe morbidity-free survival along with overall survival.

Methods:
This was a retrospective, descriptive multicentre study on patients transplanted due to ES.

Results:
From 1985-2012 63 of 714 patients with ES (8.8%) in the Nordic region received a HLTx (90.5%) or LTx with concomitant repair of the cardiac defect (9.5%). Transplanted patients were 31.9 years (21.1-42.3 years) at the time of transplantation, ranging from 10 to 67 years. Paediatric transplantations accounted for 12.7%.
A total of 39 deaths (61.9%) occurred during the study of which 7 (11%) were peri-operative deaths within 30 days of transplantation. Overall median survival was 12.0 years (95%CI 7.6-16.4 years) and the overall 1-, 5-, 10-, and 15-year post-Tx survival rates were 84.1%, 69.7%, 55.8%, and 40.6 %, respectively (figure 1). Survival conditioned by surviving the first year after Tx was 13.9 years (95%CI 7.0-20.8 years) with 1-, 5-, 10-, and 15-year survival rates of 94.4%, 77.4%, 63.0%, and 47.3 %, respectively. There was no difference in median survival after HLTx and LTx (14.9 years vs. 10.6 years, p = 0.718). Median survival free of cardiac allograft vasculopathy, bronchiolitis obliterans syndrome (BOS), and dialysis/kidney transplant were 11.2 years (95%CI 7.8-14.6), 6.9 years (95%CI 2.6-11.1), 11.2 (95%CI 8.8-13.7), respectively.
The leading cause of death after the perioperative period was infections (36.7%), followed by BOS (23.3%) and heart failure (13.3%).

Conclusions:
This study shows that lengthy post-transplant survival without severe comorbidities is achievable in patients with Eisenmenger syndrome. Cause-specific mortality after the perioperative period is dominated by causes not directly related to transplantation.