Early timing of surgical intervention in patients with Ebstein's anomaly predicts superior long-term outcome.

OBJECTIVE: Various surgical valve repair and replacement techniques have been developed over the past decades for patients with Ebstein's anomaly. Determination of the appropriate moment for surgery, however, has not been elucidated clearly enough. METHODS: From 1976 to 2007, 130 patients (mean age 23.8 +/- 17.8 years, range: 1 month to 73.6 years) underwent surgery for Ebstein's anomaly at our centre. Four patients (3.0%), who underwent univentricular palliation, and four (3.0%), who only had an atrial septal defect closure, were excluded. In 110/122 (90.2%) patients, a primary tricuspid valve repair was feasible. Valve replacement was necessary in 12 (9.8%). Mean follow-up time was 10.5 +/- 9.1 years (94.3% complete, 1284 patient years). RESULTS: There were two (1.5%) hospital deaths. Overall survival was 87.2% +/- 3.6%, 85.1% +/- 4.1% and 81.2% +/- 5.4% at 10, 20 and 25 years, respectively, without significant difference between the repair and replacement group (p=0.31). The New York Heart Association functional class>II (p=0.01) and cardiothoracic ratio>0.6 (p=0.02) were significant risk factors for mortality. Overall freedom from re-operation was 79.9 +/- 4.6%, 61.9 +/- 6.8% and 58.0 +/- 7.4% at 10, 20 and 25 years, respectively. Age<0.6 (p=0.009) were significant risk factors for the need of a re-operation. CONCLUSIONS: Repair, as opposed to replacement, is feasible in the vast...
majority of patients presenting with Ebstein's anomaly with a low early mortality rate. Outcome, in
terms of survival and freedom from re-operation in the long term is determined by the clinical state at
the time of surgery. Therefore, timely operation is warranted before significant cardiomegaly develops
and functional status deteriorates.