Surgical Outcome in Paediatric Patients with Ebstein’s Anomaly: a Nation-wide Multicentre Study

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Objectives: To review outcome of a Dutch nation-wide cohort of paediatric patients with Ebstein’s anomaly (EA) treated with surgery in childhood (0-18 years). We focus on the intention to treat as biventricular, 1.5 ventricle or univentricular repair, the unplanned reoperations and survival rates in this distinct group of EA patients.

Methods: Records of all paediatric EA patients born between 1980 and 2013 were reviewed. Clinical data including demographics, intended type of operation, intraoperative procedures and postoperative outcomes were studied.

Results: Of the 176 included EA patients, 112 were not operated during childhood, whereas the other 64 patients underwent a total of 110 operations. Median time of follow-up after diagnosis of this surgical subgroup was 115 months (range 0 – 216 months). Thirty (47%) patients required surgery within the first year of life, yet 18/64 already within the neonatal period. Intention to treat was biventricular (n=37, 58%), 1.5 ventricle (n=5, 8%) or univentricular (n=22, 34%) repair. Of the 18 patients requiring surgery during the neonatal period, in 12 patients primarily a univentricular strategy was started, in the other six a biventricular repair was intended. 13/64 Patients required one or more unplanned reoperations (8, 2 and 3 patients in resp. the biventricular, 1.5 ventricle and univentricular group). Ten patients died of low cardiac output syndrome, hypoxemia with ventricular fibrillation, pulmonary hypertension or unknown cause, all within 33 days after surgery. Three patients died in the neonatal period, all but two patients died within the first year of life. The 1-, 5- and 10-years survival rates from time of diagnosis are 88%, 84% and 84% respectively. Univentricular repair was significantly associated with death (p=0.004), operation in the neonatal period was not (p=0.78).

Conclusions: While reporting on surgical interventions and their outcomes in patients with Ebstein’s anomaly it is essential to differentiate between patients under the age of 18 years and adult patients because they represent a different spectrum of the anomaly. In paediatric patients biventricular repair is often not feasible. Mortality is related to intentional univentricular repair.