

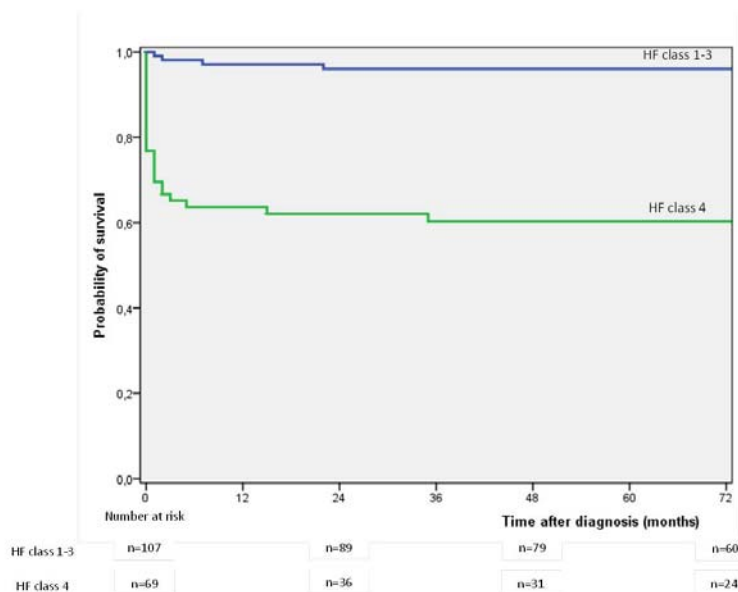
Ebstein's Anomaly in Childhood: Factors at Time of Diagnosis associated with Death

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Objectives: To analyse long-term survival in paediatric patients (0-18 years) with Ebstein's anomaly and to assess the factors at time of diagnosis that are associated with death.

Methods: Records of all Dutch consecutive live-born patients diagnosed with Ebstein's anomaly between 1980 and 2013 were reviewed. Survival curves of the time between diagnosis and death were obtained using the Kaplan-Meier method. By using the Cox proportional hazard model we analysed the factors (at diagnosis) that are associated with death.

Results: We included 176 paediatric patients (89 males). The majority (112/176, 64%) was diagnosed in the neonatal period. The median time of follow-up after diagnosis was 70 months (range 0-216). Thirty-one patients (18%) died. The 1-year survival was 84%, from 35 months after diagnosis and onwards the survival rate remained stable at 82%. The 1-year survival for patients diagnosed in the neonatal period was 78% compared to 95% for patients diagnosed after the neonatal period. Nineteen patients (11%) with severe rhythm disturbances underwent catheter ablation of an accessory pathway, none of them died. Sixty-four patients (36%) underwent cardiac surgery; 10 of these patients (16%) died (all within 33 days after the surgery) compared to 21 patients (19%) in the non-surgical group. The median age of death was less than one month (range 0-49 months). The modified Ross Heart Failure class 4 at time of diagnosis was the most important factor associated with death (Hazard ratio, HR, 12.5; 95% Confidence Interval, 95% CI, 4.4-35.9). Furthermore, diagnosis in the neonatal period (HR 4.2; 95% CI 1.5-12.0), severe tricuspid valve regurgitation (HR 2.4, 95% CI 1.2-5.0), severe pulmonary valve stenosis or (functional) pulmonary valve atresia (HR 3.7, 95% CI 1.8-7.7) and a patent ductus arteriosus (HR 2.8, 1.3-6.0) at time of diagnosis were univariately associated with death.



Conclusions: Heart Failure class 4 at time of diagnosis is the most important factor associated with death in paediatric patients with Ebstein's anomaly.