

**Postnatal Outcome of Isolated AV-Block. A Dutch Retrospective Analysis.**

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**OBJECTIVE:**

Evaluation of the postnatal outcome in second and third degree congenital atrioventricular block (CAVB) in the Netherlands during the past decade.

**METHODS:**

A nationwide database of all Dutch paediatric cardiac centres was initiated to retrospectively analyse the postnatal outcome of neonates with a CAVB-II°/III° from 2003 to 2013. Exclusion criteria were LQTS, chromosome abnormalities or complex congenital heart disease in the absence of maternal autoantibodies.

**RESULTS:**

48 liveborn children (29 female) with CAVB were included with a mean follow-up time of 5.8 years. One child was first diagnosed with CAVB at birth. Four had CAVB-II°, 43 CAVB-III° and 1 progressed from II° to III°. 42 children were positive for anti-SSA/Ro, 29/42 for anti-SSB/La and 1 for only lupus-anticoagulant. The mean gestational age at birth was 38.0 weeks, with a mean heart-rate of 77 bpm (range 53-110) in CAVB- II° and 59 (40-98) in CAVB-III°. 6 patients had a hemodynamic relevant secundum type ASD during follow-up.

Concomitant clinical details were hydrops (3 newborns), ventilation requirement (19 neonates) and pacemaker implantation in 33 children (69%). 11 children required a pacemaker within 2 days (mean heart rate 50), 9 in the neonatal period and 13 beyond. Initial pacemaker implants were predominantly VVI systems (78.8%) and had epicardial leads (93.9%).

Two children with CAVB-III° died, one postnatally, having hydrops, a VSD, hypertrophic cardiomyopathy and was positive for anti-SSA/Ro. The other died at 3.3 years due to cardiac strangulation by the pacemaker lead. Cardiac function was echographically impaired in 11 of 46 children (23.9%) during follow-up. Because of severe dilated cardiomyopathy, two children (one autoantibody negative) required heart transplantation. At last follow-up, 36 patients were in NYHA-I, 8 were in NYHA-II. Two were not classified.

**CONCLUSION:**

The post-natal outcome of CAVB concerning functional class and the survival rate is satisfactory. 69% of the patients received a PM, for which a low heart-rate at birth was the main indication for early implantation. The prevalence of secundum type ASD was remarkably high in our study population. However, a national guideline for the follow-up of CAVB in the Netherlands is lacking, leaving room for improvement.