P-24

Management of neonates with congenital complete heart block – a tertiary paediatric cardiothoracic unit experience

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Background: Reported risk factors for poor outcome in neonates with congenital complete heart block (CCHB) include prematurity, low birthweight, hydrops, low ventricular rates, and structural heart disease.

Objective: By reflecting on experience in managing neonates with CCHB admitted to a tertiary paediatric cardiothoracic intensive care unit, to identify prognostic factors for outcome.

Methods: Retrospective study (January 2003 - May 2011). Data harvested from clinical records and electronic databases.

Results: Sixteen neonates included (11 male (69%)). Median (range) gestational age was 35 (30 - 40) weeks; 69% were preterm (<37 weeks). Mean (range) birthweight was 2.5 (1.5 – 3.2) kg.

CCHB was diagnosed antenatally in 13 (81%), with median (range) ventricular rate *in utero* of 58 (40 - 85) bpm. In 9 cases, maternal anti-Ro antibodies were present: 6 received antenatal steroids, 2 of whom were antenatally hydropic. Seven (44%) had underlying structural heart disease. Ventricular function (via 2D Echocardiography) on admission was good in 11 (69%), moderately impaired in 4 (25%), and very poor in 1 patient.

Nine (56%) patients were admitted within 24 hours (75% within 72 hours) of birth. Prior to transfer, one patient required temporary pacing; 7 (44%) required isoprenaline. Four (25%) patients required emergency cardiac surgery (all non-bypass). All 16 patients ultimately required permanent pacemaker (PPM) implantation; 6 (31%) were temporarily paced beforehand.

Two early deaths occurred: one arrested during PPM implantation, the second, 3 days post-PPM implantation with progressive LV dysfunction and multiorgan failure. Two patients died later (aged 4 – 5 months): one with rhinovirus infection and ventricular decompensation, the other, following progressive ventricular dysfunction. All four deaths occurred in children born at ≤33 weeks gestation with birthweight <2kg. Structural heart disease coexisted in 3 who died (all 3 had good ventricular function on admission).

Conclusions: In this cohort, prematurity and low birthweight in association with CCHB were significant risk factors for mortality; underlying structural heart disease, hydrops, and antenatal low ventricular rates were not. Good ventricular function at birth did not predict favourable outcome, especially when structural heart disease coexisted. Early and late mortality remains high in CCHB, making close surveillance essential following PPM implantation.