

Postnatal outcome in antenatally diagnosed atrioventricular and ventriculoarterial discordance

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Introduction:

There are few published data describing the outcome of fetuses with discordant atrioventricular and ventriculoarterial connections (DAVVAC). We aimed to describe survival and postnatal interventions in a large cohort of antenatally diagnosed DAVVAC, and identify antenatal predictors of survival.

Methods:

Fetuses with at least one discordant atrioventricular and one discordant ventriculoarterial connection were identified from the databases of two centres of fetal cardiology for the period 01/01/1989 – 01/06/2018, and antenatal and postnatal data were collected. Data were analysed using Kaplan Meier analysis with the Mantel-Cox test.

Results:

98 fetuses were identified. 39 pregnancies were terminated, there were no intrauterine deaths, and 16 were lost to follow up. Postnatal data were available for 43 patients, with a median follow-up of 9.5 years and maximum of 23 years. There were 8 deaths, 5 during first year of life, and all before the age of 4 years. The cohort was divided into 5 groups: 15 patients with no additional structural cardiac lesions (isolated), 17 with a ventricular septal defect (VSD) only, 21 with pulmonary stenosis +/- VSD, 8 with Ebstein's anomaly of the tricuspid valve +/- VSD, and 37 patients with other cardiac abnormalities. The best survival was seen in the pulmonary stenosis group, with no deaths. One death was seen in the isolated group, due to complete heart block that developed postnatally. The worst survival was seen in the Ebstein's group which was significantly worse than the rest of the cohort as a whole ($p=0.02$). 6 patients underwent permanent pacemaker implantation, and 23 underwent at least one other cardiac surgical procedure. The presence of antenatal tricuspid valve regurgitation was associated with worse postnatal survival ($p=0.002$). Antenatal complete heart block was seen in 4 patients, and the survival of this group was worse (although this did not reach statistical significance).

Conclusions:

This is the largest cohort described of antenatally diagnosed DAVVAC. The medium-term outcome of isolated DAVVAC and DAVVAC with pulmonary stenosis is very good, and the outcome of DAVVAC with Ebstein's anomaly is poor. Antenatal tricuspid valve regurgitation is associated with a worse postnatal outcome.