

Fetal laterality disturbance: Single centre experience of 227 cases spanning three decades

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Introduction: Disturbances of laterality in the fetus represent some of the most complex forms of congenital heart disease. This is a heterogenous group associated with a spectrum of cardiac and extracardiac abnormalities. In many cases single ventricle palliation is the only management option after birth. The cumulative effect of associated lesions means that many do not achieve Fontan completion long-term. We report a large prenatal series of right (RAI) and left atrial isomerism (LAI), describe their associated malformations and review outcome.

Methods: Retrospective search of our fetal cardiology database between 1980 and 2010 for cases of RAI and LAI. Fetal and postnatal management and long-term outcome were reviewed.

Results: A total of 230 cases of laterality disturbance were identified. Three cases were excluded as outcome data was not available. There were 145 cases of LAI of which 32 patients (22%) remain alive, 6 have a Fontan circulation and two have undergone cardiac transplantation. In 76 (52%) cases parents opted for termination of pregnancy (TOP), 13 pregnancies resulted in intra-uterine death (IUD), 18 patients died in the neonatal period (NND) and a further 6 died in childhood (INFD). Eighty-two patients had RAI of which 14 remain alive (17%) with 7 reaching Fontan circulation. In this group 43 (52%) opted for TOP, 5 resulted in IUD, there were 9 NND and 11 INFD. The number of parents opting for TOP has fallen from 66% in the first decade (1980-1990) to 60% in the second decade (1990-2000) and 42% in the recent cohort (2000-2010). Outcome by type and era is presented in Table 1. Examining the data on an 'intention to treat' basis reveals survival of 0%, 50% and 59% for LAI and 0%, 36% and 37% for RAI across the three decades respectively.

Conclusion: Survival for those with LAI has improved with many undergoing biventricular repair. Congenital heart block remains a concern in this group resulting in IUD and NND. Prognosis remains poor in those with RAI with high attrition rates in those with single ventricle physiology.