Antenatal and postnatal right isomerism of the atrial appendages in a 20-year single institution experience

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Background: Right isomerism of the atrial appendages (RAI), particularly if diagnosed prenatally is thought to be one of the worst forms of cardiac disease.

Methods: Single centre retrospective observational study of all cases with pre- or postnatal diagnosis of RAI during a 20-year period. All prenatal cases with visceroatrial situs ambiguous, severe congenital heart disease and the presence of an inferior vena cava were included. Postnatally RAI was confirmed by the presence of asplenia and/or bilateral right sided bronchial tree in chest X-ray or magnetic resonance.

Results: Between 1992 and 2012 RAI was diagnosed in 36 cases. Prenatal diagnosis was achieved in 18 fetuses. Pregnancy was terminated in 9 (50%), palliative care was chosen in 3 newborns and 6 patients were treated. In total only 4 of 18 (22%) prenatally diagnosed children were alive at end of follow up. 18 had a postnatal diagnosis with 6 choosing palliative care. Four of the remaining 12 died after surgery and 8 patients are alive. Of total 18 patients intended to treat 12 had an AVSD (unbalanced in 7, + pulmonary atresia (PA) in 4), single right ventricle in 3 and complex anatomies in 3 cases (+PA in 2). TAPVD to a systemic vein was diagnosed in 7/18 cases. Non confluent pulmonary arteries were present in 5/18 patients. The 12 survivors had a median follow up of 4.1(0.02-18) years. Fontan (n=13) or biventricular (n=1) repair were planned in 14/18 (77%). 2 patients were considered unsuitable for Fontan completion later and receive palliative care. 2 cases underwent successfully cavopulmonary connection and are waiting for Fontan completion. 9 underwent complete Fontan repair where 2 failed and died. One patient underwent successful biventricular repair. Successful univentricular palliation or biventricular repair were achieved in only 8/18(44%) patients. Fetal diagnosis or anatomical findings, such as TAPVD, non-confluent pulmonary arteries were not predictive for Fontan failure or outcome. (p=NS).

Conclusion: Our data confirm that RAI patients have a dismal prognosis with successful univentricular palliation or biventricular repair being feasible in less than 50% of all cases. Prenatal diagnosis and/or anatomical findings seem not to influence outcome.