

Features and outcomes of fetuses with tricuspid atresia

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Background: Tricuspid atresia (TrA) has variable anatomic features and a rather severe prognosis, requiring a univentricular correction.

Objectives: Our aims were to analyse retrospectively the features and outcomes of cases diagnosed in our Center with this lesion.

Material and methods: Out of around 1250 fetal cases with CHD studied in our Centre between Jan.1998 and Dec. 2011, 55 (4.4%) were found to have TrA, at 16-34 weeks' gestation (w.g.) The characteristics and outcomes of 53 cases with known follow-up were retrospectively analysed.

Results: 1/ Characteristics: Four fetuses were twins; 23 fetuses (43.4%) had associated pulmonary stenosis (PS); 9 (17%) had pulmonary atresia (PAtr), 2 with sinusoids; 14 (26.4%) had associated one or more ventricular septal defects (VSD), one with coarctation of aorta; 7 (13.2%) had transposition of great arteries (TGA) with VSD, 1 with interrupted aortic arch. One fetus had a minor chromosomal anomaly - inversion (2) p11.2q13; 8 had associated extracardiac anomalies (ECA). Three fetuses had heart failure already at presentation, due to restrictive foramen ovale.

2/ Outcomes: Sixteen fetuses (30.2%) were terminated, 5 with ECA. Out of 37 fetuses that continued pregnancy, one died in utero at 24 w.g., 4 died shortly after birth (2 in heart failure, 1 in severe cyanosis and 1 fetus with PAtr and sinusoids had cardiac arrest). The remaining 32 cases were operated, all with palliative procedures (shunt or banding pulmonary artery), 11 underwent Glenn operation and 6 the Fontan procedure. A total of 5/32 cases (15.6%) died postoperatively, 3 after shunt operation (1 with PAtr and sinusoids), 1 (with interrupted aortic arch) after the Norwood 1st step and 1 after Glenn operation. Total intrauterine and postnatal mortality was 10/37 cases (27%). The remaining infants are alive, a half of them on medications, at 2-12 years.

Conclusions. Despite an improvement in surgical results and in perinatal management the outcomes of TrA remain still poor, almost one third dying postoperatively or spontaneously. Negative prognostic factors were restrictive foramen ovale, heart failure and PAtr with sinusoids.