French monocentric experience with antenatal diagnosis of Hypoplastic Left Heart Syndrome

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The objective of this study was to assess the outcomes of fetus and decision making after prenatal diagnosis of hypoplastic left heart syndrome (HLHS).

Material and Methods: This study is a single-center retrospective analysis of all fetus diagnosed with HLHS syndrome from 2010 to 2016. Antenatal parameters included: mother age and gestational age of fetus at diagnosis, genetic testing, and prenatal outcomes. Postnatal parameters included: gestational age, birth weight, apgar score, echocardiographic measurements, and decision-making.

Results: Overall 70 foetus were included in the study, diagnosed at 26.4±5.2 weeks of gestation, with: typical HLHS (54), unbalanced DORV (2) or AVSD (1), borderline LV (4), severe aortic coarctation and LV hypoplasia (3), double inlet ventricle (3) and complex CHD (3). Four cases had a chromosome anomaly. Thirty-seven terminations of pregnancy (53%) at 25.2±3.1WG and 25 (36%) live births occurred; the remaining cases included: sudden in utero death (1), ongoing gestation (1), and lost of FU (6= 9%). Gestational age at birth was 38.8±1.9wg (34 to 42wg), birth weight was 3180±600g (1900 to 4300g); apgar score was ≤ 3 in 16% and > 7 in 80% of the cases. Postnatal echocardiographic analysis was concordant with prenatal assessment: HLHS (15), borderline LV (7), DORV (1), DIV (1). One patient died early after birth from uncontrolled cardiogenic shock and hypoxemia. Among the 24 remaining patients, 8 were un-operated and died (parents decision for compassionate care), and 16 underwent first-stage surgery (i.e. 23% of overall 70 fetus) at the age of 10.8±6.2 days (5 to 25d) from whom 7 had second-stage surgery at the age of 4.1±2.6 months (1 to 7mos).

Conclusion: Despite experience and improvement of surgical techniques in HLHS, decision making in a French tertiary-care center Pediatric cardiology department still prefer TOB or compassionate care to active surgical management.