Fetal versus postnatal echocardiogram accuracy in the diagnosis of interrupted aortic arch

*Larios G., Jaeggi E., Yoo S-J., Dutil N., Thakur V.*
The Hospital for Sick Children, Division of Cardiology. University of Toronto

Introduction: Accurate prenatal diagnosis of interrupted aortic arch (IAA) and type allows for comprehensive fetal and genetic counselling and postnatal management. Our study aims to evaluate the rate and accuracy of fetal (FE) versus postnatal (PNE) echocardiogram for IAA presence and type in patients with usual associated lesions (UAL), such as ventricular septal defects, or complex associated lesions (CAL, e.g. truncus arteriosus, TA).

Methods: Retrospective review of the FE and PNE in patients with suspected IAA was analysed, from 2004 to 2016. Patients were grouped as FE or PNE based on timing of initial suspicion of IAA. Final diagnosis of IAA and type was confirmed in the operating room, on axial images or autopsy. The presence and impact of UAL or CAL in the diagnosis was analysed.

Results: Of 96 subjects conforming the study group, 44 IAA were suspected on FE and 52 on PNE. Of the 44 subjects with FE, 11 (25%) pregnancies were terminated, 4 (9%) resulted in stillbirth and 7 (16%) were misdiagnosed as IAA and were found to have coarctation of the aorta. Of the 52 found on PNE, 6 had a missed diagnosis on FE. All PNE patients had diagnosis confirmed. Of 73 confirmed IAA, there were 25 type A (34.2%), 47 type B (64.4%) and 1 type C (1.4%). FE detected 20/27 subjects and PNE 52/52 (Sensitivity 74.1% versus 100%, p= 0.0005). FE detected correctly the type of IAA in 11/20 (55%) and PNE in 48/52 (92.3%), p=0.001. A higher proportion of CAL was observed during FE, 18 (41%) subjects versus 8 (27%) patients in the only PNE group. The presence of CAL did not interfere with the performance of the tests (p=0.38).

Conclusions: Fetal diagnosis of IAA remains challenging with a lower detection rate and accuracy compared to PNE. Careful evaluation of 3 vessel tracheal and long axis views, especially when usual associated cardiac anomalies are found, is required to improve the accuracy of FE. Improved fetal diagnosis is important to allow women to have different options in pregnancy management and the option for genetic testing and counselling.
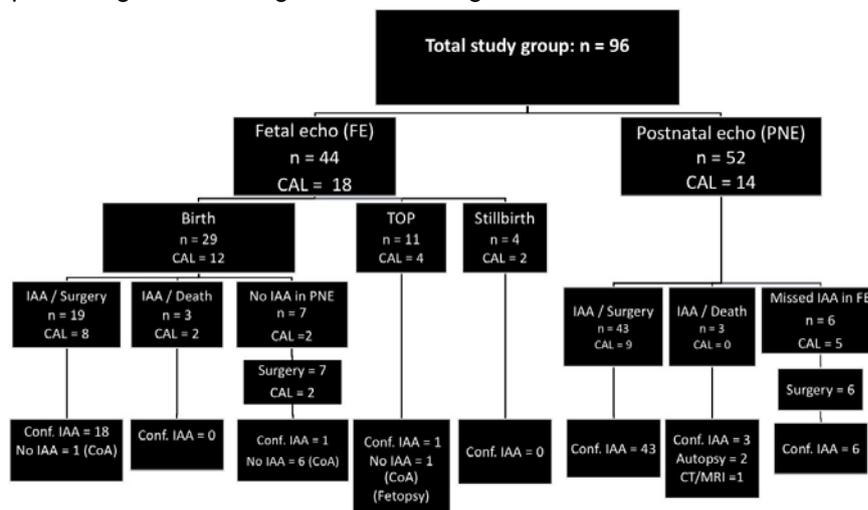
Figure 1: Distribution of the subjects according to type of study and outcome.CAL: Complex associated lesions; CoA: Coarctation of the aorta; Conf. IAA: Confirmed IAA (in the operating room, axial images or autopsy/fetopsy, or true positive); IAA: Interrupted aortic arch; FE: Fetal echocardiogram; PNE: Postnatal echocardiogram; NC: Not confirmed diagnosis; No IAA: Incorrectly diagnosed as IAA (false positive); TA: Truncus arteriosus; TOP: Termination of pregnancy; UAL: Usual associated lesions.