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Fetal coarctation of the aorta – can we predict who needs surgery?

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Introduction
Fetal coarctation of the aorta (CoA) has high false positive rates when assessed in tertiary centres. Retrospective studies indicate that quantification of arch measurements and visualisation of isthmal features such as coarctation shelf (S) and flow disturbance (F) allow separation of hypoplastic from normal arches but their ability to predict requirement for surgery in a prospective cohort remains unknown.
Aims
To prospectively assess the ability of isthmal Z-scores (I) and ratio of isthmus to duct (I:D) measured in three vessel and tracheal view and visualisation of S and F to identify fetuses requiring neonatal surgery for isolated coarctation.
Methods
We acquired Doppler ultrasound measurements on 42 consecutive fetuses we diagnosed with isolated CoA referred at median 22.5 (15 - 37) weeks' gestation to two fetal cardiology centres. We measured I, D and I:D ratio prospectively and recorded whether S or F were seen. We recorded at the first scan and again near term whether the baby was likely to require surgery.
Results
31/ 42 (74%) babies had CoA requiring perinatal surgery. None were diagnosed with CoA later in childhood. Both I:D ratio < 0.74 (25/31, 81%) and I Z- scores < -2 (28/31 90%) gave positive predictive value for surgery of 80% at initial scan. ROC curve analysis showed a modest effect of I:D ratio (AUC 0.66, 95%CI 0.44, 0.870, p=0.167) and isthmal z-score (AUC 0.69, 95%CI 0.51, 0.88, p=0.086) to correctly identify those requiring surgery. Serial scans were performed in 39/42 (median 3, range 1-5). Half those showing growth towards normal values (I Z-score (8) or I:D ratio (1))or deviation from normal (I Z-score (4) or I:D ratio (4) required CoA surgery. (figure) The variables were not powerful predictors in isolation but multivariable logistic regression combining all gave positive predictive value 80% and ROC AUC 0.753 (0.51, 0.99).
Conclusions
Incorporation of these measurements in clinical practice enabled prediction of antenatal diagnosis of true CoA at first scan in 74% with correction of false positive diagnosis in 3 following serial measurements. Serial targeted examination may reduce the false positive rate of fetal CoA in clinical practice.

Figure: Serial isthmal Z scores of fetuses diagnosed with suspected coarctation