Challenges of Prenatal Diagnosis of fetal Hypoplastic Aortic Arch and Predication of the Need for Intervention

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Abstract

Prenatal diagnosis of hypoplastic aortic arch and coarctation of aorta is still challenging and remains one of the most difficult cardiac defect to diagnose. The results reveal significant improvement of prenatal diagnosis of hypoplastic arch and coactation of aorta. The data also shows the significant overlapping of fetal aortic isthmus z score between the infants who need the arch procedure and those who do not.

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Abstract:

Prenatal diagnosis of hypoplastic aortic arch and coarctation of aorta is still challenging and remains one of the most difficult cardiac defect to diagnose. The results reveal significant improvement of prenatal diagnosis of hypoplastic arch and coactation of aorta. The data also shows the significant overlapping of fetal aortic isthmus z score between the infants who need the arch procedure and those who do not.

Invited Commentary:

Prenatal diagnosis of hypoplastic arch and coarctation of aorta improves survival and reduces morbidity after birth. (1, 2) Significant improvement in diagnosis of fetal congenital heart defect has been made with advance in technology and experience including fetal ultrasound and fetal echocardiography. However prenatal diagnosis of hypoplastic aortic arch and coarctation of aorta is still challenging and remains one of the most difficult cardiac defects to diagnose before birth with relatively high false positive and false negative rate compared to other major congenital heart defects. (3, 4)

The presence of hypoplasia of the isthmus and transverse arch is the most consistent and the definitive antenatal sign of postnatal coarctation. (4) In this study, Evans et al retrospectively reviewed a large number of cases with prenatal diagnosis of hypoplastic aortic arch and explored the challengers and complexities of the referral, indication, risk factor, diagnosis and the accuracy of prediction for the need of postnatal procedure. The results demonstrate the significant improvement of prenatal diagnosis of hypoplastic aortic arch and coarctation of aorta over the past five years. Since 2017 in their state-wide fetal cardiology program in Nevada, the detection rate of infants who need intervention of aortic arch for coarctation of aorta and hypoplastic arch after birth, improved from 38% to 82%, p=.04.

The results also show the significant overlapping of the fetal aortic isthmus z score between the infants who need the arch procedure and those who do not. Of the 34 that had a postnatal procedure 11 (32%) had a fetal aortic isthmus z-value with a median and range of -2.8 (-1 to -5.0), and of 51 that did not undergo postnatal procedure 20 (38%) had a z-value with a mean and range of -2.6 (-1.5 to -4.6). Of the 44 presumed ductus arteriosus dependent aortic arch obstruction by fetal echocardiogram 10 had subsequently prostaglandin E 1 discontinued.

Their findings further demonstrate the anatomic and pathophysiological complexity of ductus arteriosus and aortic isthmus region in patients with coarctation of aorta and dynamic change following the transition from fetal to neonatal circulation and the need to adopt multiparametric diagnostic model and multicriteria prediction model. (5, 6, 7, 8) Many investigators have been working to explore the application of 3D fetal echocardiography and CMR in fetal aortic arch anomalies to increase the accuracy of the diagnosis of coarctation of aorta and the prediction for postnatal intervention. (9, 10, 11)

1. Franklin O, Burch M, Manning N, Sleeman K, Gould S, Archer N. Prenatal diagnosis of coarctation of the aorta improves survival and reduces morbidity. Heart. 2002; 87 (1):67–9.

2. Słodki M, Rizzo G, Augustyniak A, Seligman NS, Zych-Krekora K, Respondek-Liberska M; International Prenatal Cardiology Collaboration Group. Retrospective cohort study of prenatally and postnatally diagnosed coarctation of the aorta (CoA): prenatal diagnosis improve neonatal outcome in severe CoA. J Matern Fetal Neonatal Med. 2020;33(6):947–951.
3. Tegnander E, Williams W, Johansen OJ, Blaas HG, Eik-Nes SH. Prenatal detection of heart defects in a non-selected population of 30,149 fetuses–detection rates and outcome. Ultrasound Obstet Gynecol. 2006; 27:252–265.
4. Hornberger LK, Sahn DJ, Kleinman CS, Copel J, Silverman NH. Antenatal diagnosis of coarctation of the aorta: a multicenter experience. J Am Coll Cardiol. 1994; 23:417–423.
5. Enery Gómez-Montes , Ignacio Herraiz García , David Escribano Abad , Jesús Rodríguez Calvo , Cecilia Villalaín González , Alberto Galindo Izquierdo [1]Application of a Global Multiparameter Scoring System for the Prenatal Prediction of Coarctation of the Aorta. Journal clinical medicine. J Clin Med. 2021; 20;10 (16):3690.
6. Meaghan Beattie, Shabnam Peyvandi , Suguna Ganesan , Anita Moon-Grady Toward Improving the Fetal Diagnosis of Coarctation of the Aorta. Pediatr Cardiol. 2017; 38 (2):344-352.
7. Wang H, Lei W, Liu J, Yang B, Li H, Huang D The Diastolic and Systolic Velocity-Time Integral Ratio of the Aortic Isthmus Is a Sensitive Indicator of Aortic Coarctation in Fetuses. J Am Soc Echocardiogr. 2019; 32(11):1470-1476.
8. DeVore GR, Haxel C, Satou G, Sklansky M, Pelka MJ, Jone PN, Cuneo BF. Improved detection of coarctation of the aorta using speckle-tracking analysis of fetal heart on last examination prior to delivery. Ultrasound Obstet Gynecol. 2021; 57(2):282-291
9. Zhuo Chen MD, PhD, Yue Zhou MS,Jingying Wang PhD,Xiaowei Liu MD, PhD, Shuping Ge MD,Yihua He MD, PhD Modeling of coarctation of aorta in human fetuses using 3D/4D fetal echocardiography and computational fluid dynamics Echocardiography. 2017; 34(12):1858-1866
10. David F A Lloyd, MPhil, Kuberan Pushparajah, MD, John M Simpson, Prof, MD, Joshua F P van Amerom, BASc, Milou P M van Poppel, MD, Alexander Schulz, Bernard Kainz, PhD,Maria Deprez, PhD,Maelene Lohezic, PhD, Joanna Allsop, Sujeev Mathur, MD, Hannah Bellsham-Revell, MD, Trisha Vigneswaran, MRCPCH, Marietta Charakida, PhD, Owen Miller, MD, Vita Zidere, MD, Gurleen Sharland, Prof, MD, Mary Rutherford, Prof, MD, Joseph V Hajnal, Prof, PhD, and Reza Razavi, Prof, MD Three-dimensional visualization of the fetal heart using prenatal MRI with motion-corrected slice-volume registration: a prospective, single-centre cohort study Lancet. 2019; 20393(10181): 1619–1627.
11. David F.A. Lloyd, Milou P.M. van Poppel, Kuberan Pushparajah, Trisha V. Vigneswaran, Vita Zidere, Johannes Steinweg, Joshua F.P. van Amerom, Thomas A. Roberts, Alexander Schulz, Marietta Charakida, Owen Miller, Gurleen Sharland, Mary Rutherford, Joseph V. Hajnal, John M. Simpson, Reza Razavi. Analysis of 3-Dimensional Arch Anatomy, Vascular Flow, and Postnatal Outcome in Cases of Suspected Coarctation of the Aorta Using Fetal Cardiac Magnetic Resonance Imaging. Circulation: Cardiovascular Imaging. 2021; 14 (7): 2021; P e012411