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Case report

Multi-modality imaging in the diagnosis of fetal vascular ring

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Abstract

Prenatal diagnosis of vascular ring can improve the time to intervention. Correlation of fetal diagnosis with postnatal imaging in the newborn period is needed for appropriate clinical management, surgical referral, and surgical approach if indicated. We present a case of a fetal diagnosis of double aortic arch with postnatal feed and swaddle cardiac MRI diagnosis of right aortic arch and aberrant left subclavian artery.

Key words: fetal echocardiogram, vascular ring, double aortic arch, feed and swaddle MRI.

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Case report

A 25-year-old G2P1 mother without significant past medical history was referred for fetal echocardiogram at 25 6/7 weeks of gestation. The mother was pregnant with dichorionic diamniotic twins, and 3 days earlier the anatomy scan was suspicious for vascular ring on an anatomy scan of Twin B. The pregnancy was uncomplicated with normal cell-free DNA and no family history of congenital heart disease. The fetal echocardiographic images showed a bifurcating aortic arch by 2-D imaging (Figure 1) as well as "O"-shaped vessel arrangement by color Doppler (Figure 2), consistent with double aortic arch, as well as a perimembranous ventricular septal defect (VSD). The other twin had bilateral superior venae cava without a bridging vein. The parents were counseled about vascular rings, the differential diagnosis of double arch, as well as the potential for postnatal surgery.

The babies were delivered by schedule repeat C-section at 37 weeks at a community hospital, and Twin B was transferred to our facility on DOL 7 for hemolytic anemia and respiratory distress. Echocardiography at our facility showed double aortic arch and perimembranous VSD as well as a small left superior vena cava to coronary sinus with a bridging vein. Our institution then proceeded with our standard protocol feed and swaddle cardiac magnetic resonance imaging (MRI), which was performed during admission. MRI showed a right aortic arch with aberrant left subclavian artery, noting that MRI is unable to rule out atretic vessels such as an atretic nondominant arch or a closed duct (Figures 3, 4). Imaging also demonstrated mild narrowing of the trachea at the level of the transverse arch that was not felt to be significant enough to warrant intervention. The child was discharged home on day of life 23 in good health. The patient is clinically well and asymp-

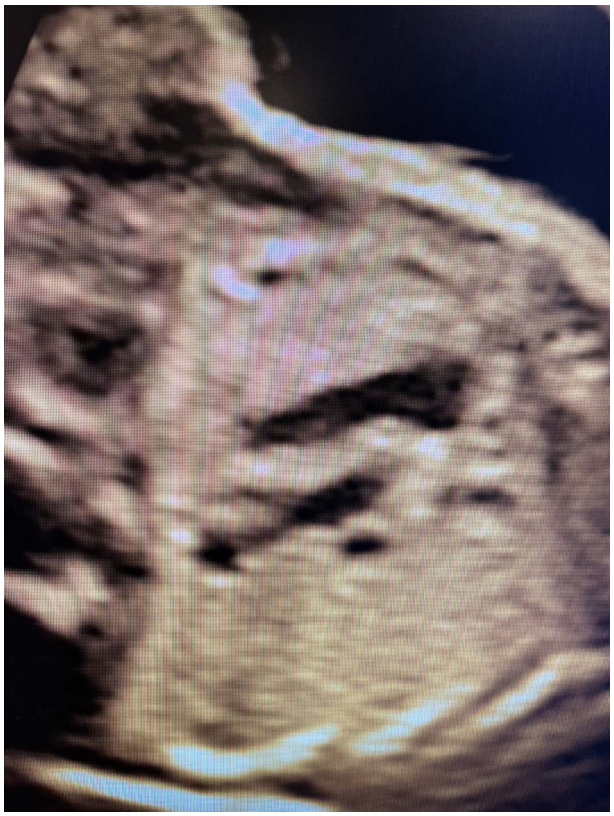

Figure 1. 2D fetal echocardiogram of the 3-vessel trachea view showing a left sided main pulmonary artery (top), a right-sided aortic arch with what appears to be a smaller left-sided transverse arch (asterisk), and a right sided superior vena cava

tomatic; his VSD is small and hemodynamically insignificant. Of note, genetic testing showed a duplication of 4q34.1q34.3 affecting the region containing HAND2.

Discussion

The embryologic aortic anatomy is comprised of 6 paired dorsal aortic arches, of which the third through sixth ultimately contribute to the formation of the final aortic architecture. The fifth arches typically regress, and the seventh intersegmental arteries complete the anatomy by forming the subclavian vessels. The final anatomy is created with strategic regression of most commonly the right-sided fourth aortic arch and various subsegments of the paired arches, to create a normal left aortic arch. Failure of regression of any variety of these segments can lead to a wide range of anatomical findings, many of which are clinically silent. Surgical intervention is needed when these anatomical variants impinge on other structures within the chest, namely the trachea (leading to respiratory difficulty) and esophagus (causing failure to thrive and/or feeding difficulty). The anatomical variations that most frequently require intervention are double aortic arch, with persistence of both fourth aortic arches leading to encircling and impingement of the airway and esophagus, and aberrant subclavian arteries forming a Kommerell's diverticulum with similar compression of thoracic structures [1].

As the requirements for obstetric assessment have changed, with incorporation of more cardiac views and assessment on

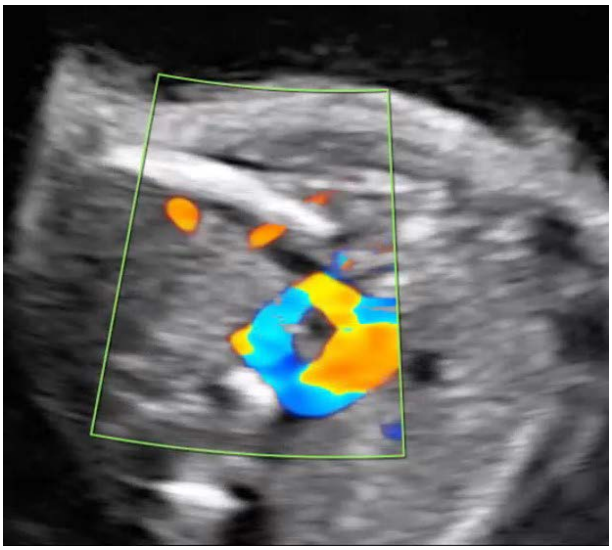

Figure 2. Color Doppler of at the same level showing O-shaped vessel arrangement consistent with double aortic arch

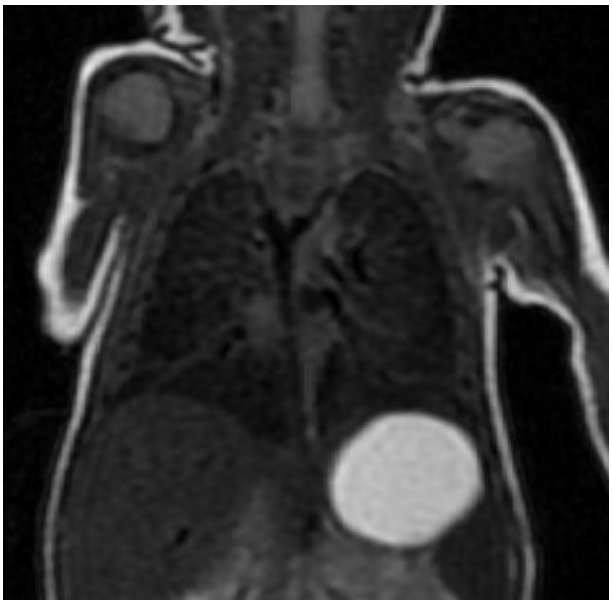

Figure 3. Feed and swallow MRI with coronal imaging of the descending aorta, demonstrating right-sided proximal descending aorta with Kommerell's diverticulum and aberrant left subclavian artery

routine high-level anatomy scan, so has the increased detection of these lesions [2]. Prenatal counseling, including a description of anatomy, comorbidities, and genetic conditions, improves parental awareness, improves detection of extracardiac abnormalities, and prepares the postnatal medical team to optimize delivery conditions and postnatal care [3]. Compared to postnatally diagnosed patients, prenatal diagnosis of vascular rings leads to earlier repair and fewer pre- and postoperative symptoms, with comparable surgical outcomes [4-6].

Assessment of the aortic arch can be performed by transthoracic echocardiography, computed tomography (CT), and MRI. While CT has shorter image acquisition time and higher spatial and temporal resolution, MRI has the advantage of imaging of other thoracic structures without the need for ionizing radiation or iodinated contrast [1]. Since the development

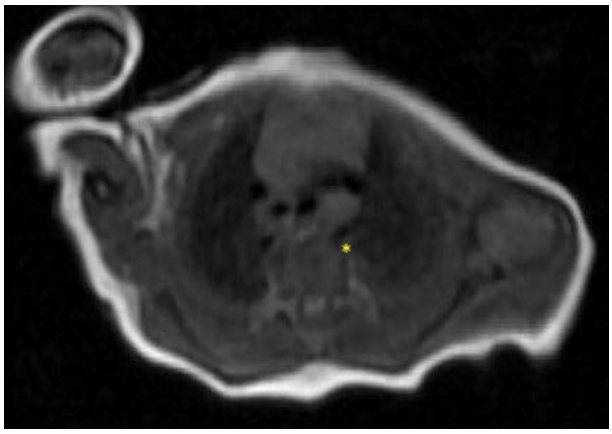

Figure 4. Axial view demonstrating asymmetry of the head and neck vessels, inconsistent with double aortic arch (aberrant left subclavian artery represented by asterisk)

of feed and swaddle MRI in infants under 6 months old, patients can be imaged with high diagnostic accuracy without sedation [7]. With advances in technology and the ability to minimize risk to patients, multimodality imaging of cardiac defects has become commonplace for guidance of medical and surgical management. Comparison of fetal echocardiography to postnatal MRI is a nascent area of imaging [8] with the potential to improve diagnostic accuracy even further.

Conclusions

As fetal detection of aortic arch anomalies has become commonplace, postnatal imaging determines clinical management and clarifies the diagnosis made on fetal echocardiographic images. Multimodality imaging of aortic arch abnormalities is essential for accurate diagnosis and informs medical decision-making.

Conflict of interest

The authors declare no conflict of interest.

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