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Opinion

Mind the gap! What we don't know about right aortic arches and aberrant branches

This edition of the Journal contains two reports on the detection and examination of right-sided aortic arches diagnosed prenatally using ultrasound[1,2]. Achiron *et al.* first described the importance of the three vessels and trachea view in determining the prevalence of aortic arch abnormalities in an unselected screened population in this Journal in 2002[3]. They reported that right aortic arch occurred in 0.1% of the unselected screened fetal population and, in common with the Edward's classification, subdivided the cases into those with mirror-image arrangement of the vessels, with which congenital heart disease was almost universally associated, and those with an aberrant left subclavian artery, in which a loose vascular ring was likely, but congenital heart disease was not, being present in about 10%. The two papers in this issue, by Zidere *et al.*[1] and Berg *et al.*[2], contribute to this subject by documenting the frequency of right aortic arch and its more common branching patterns in selected populations, and by highlighting the importance of using screening protocols, providing adequate training and supervision and obtaining postnatal confirmation of antenatal findings.

Many of us work with highly selected populations that present with interesting anomalies, but we often lack the data with which to describe the prevalence of these malformations or 'normal variants' in unselected populations. In some European countries, almost the whole pregnant population is offered anomaly screening of the fetus, and with improved training in echocardiography, an increasing number of 'normal variants' has been described. These include the presence of a right-sided aortic arch and the persistence of the left superior caval vein, both of which may coexist with an otherwise structurally normal heart.

The study of Zidere *et al.*[1] was motivated by the recognition of an increase in the number of fetuses referred with congenital heart disease that had a right aortic arch along with an increase in cases of isolated right aortic arch referred from screening programs. They felt that the most plausible explanation was that the condition had simply not been recognized in past years. This suspicion was borne out by a retrospective review of recorded videotapes, which demonstrated that right aortic arch had been missed almost as often as it was detected in cases of tetralogy of Fallot, despite the well-known coexistence of the two in 20–25% of cases[1]. This highlights an important issue of interpretation, in that while the diagnostic view may be imaged correctly, the observer may not necessarily know how to interpret the component parts accurately.

Berg *et al.*[2] examined the branching patterns of the fetal right arch using ultrasound, illustrating how new modes of investigation sometimes describe anew associations that have been long forgotten. Long before sonography became the established mode of investigation in the child and fetus with suspected congenital heart disease, barium swallow and angiography provided us with excellent information on aberrant vessels and their clinical associations. Technical improvements in ultrasound, including the use of color Doppler, energy and three-dimensional, in parallel with improved training, now allow us to achieve non-invasively the quality of extracardiac vascular imaging in the fetal population that was routine in children 40 years ago using angiography. Not surprisingly, we are describing similar associations. Hastreiter *et al.*[4] made us aware of the important association of aberrant subclavian arteries with conotruncal cardiac malformations in 1966 and Wilson *et al.*[5] the association with 22q11 deletion in the early 1990s. Goldstein[6] described the association of aberrancy of the right subclavian artery with trisomy 21 in 1965. This was supported in 1989 by the findings of an angiographic study of children with congenital heart disease by Rathore and Sreenivasan[7], who found aberrant arch vessels (left vertebral and right subclavian artery) in 53% of trisomy 21 cases compared with only 1.7% of controls. There is no doubt that improved sonographic training, particularly emphasizing the three vessels and trachea view, has resulted in increased recognition of arch position and unusual branching patterns, but it remains to be seen whether the detection of an aberrant right subclavian artery is a practical additional sonographic marker for trisomy 21 in antenatal screening programs[8].

We already know the natural history of small ventricular septal defects and echogenic foci *in utero* and recognize the potential of the progression of aortic stenosis to hypoplastic left heart syndrome, but new information produces new challenges. We will need to decide how to manage information about arch anomalies and provide appropriate counseling for families. For example, what surveillance is warranted for fetuses and children with normal variants, such as isolated right aortic arch or persistent left superior caval vein? Should the fetus with an isolated right-sided aortic arch be karyotyped to exclude 22q11 deletion? Zidere *et al.*[1] suggest not, unless extracardiac malformations, such as thymic hypoplasia, are identified. Currently, we counsel parents based on outcomes from published reports – usually from selected populations of children who have undergone cardiac investigations. However, we still do not know the true association of aberrant right subclavian artery with trisomy 21[8] and persistence of the left superior caval vein

with congenital heart disease, particularly coarctation of the aorta[9], from large antenatal population-based studies.

One feature common to many antenatal studies is their retrospective design and lack of a 'joined-up' approach: essential data that should be available from postnatal examinations or a more meticulous antenatal approach are often missing. We are in danger of compiling an ever-expanding library that advances our knowledge in a piecemeal manner and makes the overall picture more difficult to interpret. This in part reflects the difficulties we experience in reaching the top of the learning curve in the detailed anatomical examination of the fetus. With frequent technical improvements, the goalposts continually shift as our eyes are opened to ever-finer detail. To benefit from this fully as a scientific community, we need to combine the skills of the cardiologist and the specialized obstetrician working in fetal medicine units in order to obtain the detailed knowledge required to counsel families appropriately before delivery, and to combine this with careful postnatal audit in order to evaluate outcomes as fully as possible. One way forward would be to use the joint expertise available in the cardiological and obstetric bodies (ISUOG and the AEPC (Association for European Pediatric Cardiology), for example) to collect longitudinal population-based data on the outcome of fetuses with aberrant vessels, right aortic arch and persistent left superior caval vein. Since analysis of these structures relies on the three vessels and trachea view we also need to ensure that this view is employed routinely at screening and that it is interpreted correctly. This would enable us to define the prevalence of these structures in the unselected antenatal population and increase our knowledge of important associations to provide optimal counseling and to guide management of these cases in the future.

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