Prenatal Diagnosis of Vascular Rings

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Objectives. The purpose of this series was to identify vascular rings prenatally on fetal echocardiograms.

Methods. We reviewed the fetal echocardiograms and clinical histories of 9 patients with a diagnosis of vascular rings at our institution from 2004 to 2009. Eight patients had a prenatal diagnosis by fetal echocardiography. One other patient who had undergone fetal echocardiography had a diagnosis of a vascular ring and a cervical arch only postnatally. Results. Among the 8 patients with a prenatal diagnosis (4 with a double aortic arch and 4 with a right aortic arch [RAA], an aberrant left subclavian artery, and a left ductus arteriosus [LDA]), the vascular ring was isolated in 4 and associated with other structural congenital heart disease in 4. In all 8 patients with a prenatal diagnosis, the vascular ring was identified by cephalad transducer sweeps from a 3-vessel view (3VV) with and without color Doppler imaging, which revealed vascular structures coursing around the trachea. Seven of these patients had postnatal confirmation of the abnormality (there was 1 pregnancy termination without autopsy). The only known vascular ring missed at fetal echocardiography but diagnosed after birth was that associated with a cervical RAA (which could not be visualized in cross-sectional sweeps) and an LDA evaluated only late in pregnancy. Only 3 of the 8 postnatally treated neonates had clinical symptoms of the vascular ring, and 5 underwent surgical division of the ring. Conclusions. Vascular rings can be largely identified before birth with cephalad sweeps from the 3VV with attention to the relationship of the aortic and ductal arches to the trachea on fetal echocardiograms. Key words: congenital heart disease; double aortic arch; fetal echocardiography; prenatal diagnosis; vascular ring.

Abbreviations
CHD, congenital heart disease; DAA, double aortic arch; DORV, double-outlet right ventricle; LAA, left aortic arch; LDA, left ductus arteriosus; LSCA, left subclavian artery; PS, pulmonary stenosis; RAA, right aortic arch; TOF, tetralogy of Fallot; 3VV, 3-vessel view

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Vascular rings are congenital abnormalities of the aortic arch and its branches that form a ring around and may compress the trachea and esophagus. The most common forms of vascular rings include a double aortic arch (DAA) and a right aortic arch (RAA) with a left ductus arteriosus (LDA) from an aberrant left subclavian artery (LSCA) (Figure 1). These either can occur in isolation or may be associated with other congenital heart defects. Affected patients typically present after birth with respiratory distress and feeding difficulties. In addition, some have an associated 22q11.2 deletion, which places them at risk for neurodevelopmental delays, speech difficulties, hypocalcemia, increased risk of infection, and other structural abnormalities, including cleft lip/palate. Others may only be identified in patients undergoing echocardiography for structural congenital heart disease (CHD). To date, only a few small case series in the obstetric literature have documented prenatal detection of vascular rings.1–3
With clinical application of the 3-vessel view (3VV) and attention to the aortic and ductal arch relationships to the fetal trachea, it is now possible to consistently recognize vascular rings before birth. In this series, we review our experience with the prenatal diagnosis of vascular rings, describing the prenatal features and clinical outcome of the affected pregnancies.

Materials and Methods

We identified all cases of prenatally diagnosed vascular rings between January 2004 and January 2009 through our Fetal Cardiovascular Program database. We also identified all infants and children with a diagnosis of vascular rings in our institution through the pediatric echocardiography database over the same period to identify fetuses prenatally examined by fetal echocardiography who only had a diagnosis of a vascular ring after birth. We reviewed the fetal echocardiograms to define the prenatal features and confirmed the diagnoses by postnatal echocardiography, computed tomography, and/or magnetic resonance images.

All fetal and neonatal echocardiographic examinations were performed on an Acuson Sequoia echocardiographic machine (Siemens Medical Solutions, Mountain View, CA) using 8- or 6-MHz curvilinear or 10-MHz phased array transducers. Routine fetal cardiac assessment included demonstration of segmental anatomy using gray scale imaging, including the atrial and visceral situs, systemic and pulmonary venous connections, symmetric 4-chamber view, patency and symmetry of the atrioventricular valves, pulmonary and systemic venous connections, both outflow tracts and great arteries, and the ductal and aortic arches, including their positions relative to the trachea shown in cephalad sweeps from the 3VV. Pulsed and color Doppler interrogation were also performed to confirm patency of the ventricular inflows, outflows, and arches and to assess the flow patterns in the pulmonary veins, umbilical artery, umbilical vein, and ductus venosus. The examination was performed by a sonographer or fetal/pediatric cardiologist and always reviewed by a fetal/pediatric cardiologist.

Prenatal and postnatal medical records, including surgical, sonographic, and genetic reports, were also reviewed to document the reason for referral, associated cardiac and extracardiac abnormalities, and clinical outcomes of affected fetuses. Approval was given for this research by the Institutional Review Board of the University of California, San Francisco, Medical Center.

Results

During the study period, 8 patients had a prenatal diagnosis of a vascular ring in our institution. One other patient was evaluated prenatally with fetal echocardiography but had a diagnosis of a vascular ring only after birth. Table 1 summarizes the clinical observations. There were 4 diagnoses of a DAA and 5 of an RAA/LDA. Of the 9 patients, 5 had isolated vascular rings (DAA in 2 and RAA/LDA in 3). Four patients had a vascular ring associated with more complex intracardiac abnormalities, including tetralogy of Fallot (TOF) in 1, a double-outlet right ventricle (DORV) with critical pulmonary stenosis (PS) in 2, and a DORV with subaortic obstruction and left juxtaposition of the atrial appendages in 1. Gestational ages at fetal diagnosis ranged from 19 to 34 weeks. A 22q11.2 deletion was not detected in the 8 patients tested.

Fetal Echocardiographic Features

In all 8 cases with a prenatal diagnosis, the vascular ring was initially recognized in cephalad cross-sectional sweeps from gray scale images beginning at the level of the 3VV (Figures 2–5 and Videos 1–5). From these sweeps, the relationship of the aortic arch and ductus arteriosus to the fetal trachea could be easily shown, and a vascular structure coursing around and behind the trachea. Center and right, Two of the most common forms of vascular rings: a DAA (left) and an RAA with aberrant LSCA and LDA (right). DA indicates ductus arteriosus; LCC, left common carotid artery; and MPA, main pulmonary artery.
Chea could be identified. Color flow mapping confirmed the presence of patent vascular structures coursing around the trachea and was particularly helpful in identifying a diminutive ductus arteriosus associated with pulmonary outflow tract obstruction. Coronal images through the fetal trachea obtained in 3 cases further confirmed the presence of the vascular structures on either side of the trachea just above the carina (Figure 3). In 3 fetuses with normally related great arteries, a gap between the main pulmonary artery and ascending aorta seen in the 3VV led to the referral for fetal echocardiography. In 8 of the 9 cases, the descending aorta coursed more centrally rather than to the left in front of the fetal spine and behind the left atrium and was less closely related to the left pulmonary veins than usual.

In the 4 cases with a DAA, there was a large left-sided ductus arteriosus associated with a dominant right arch and more diminutive left arch in 3 and nearly equally sized left and right arches in 1. In 2 of the 4 with a DAA (cases 2 and 7), mirror image brachiocephalic vessels could be visualized further suggesting the diagnosis of a DAA. In 1 with a DAA (case 7), severe tracheal obstruction resulted in hyperexpansion of the lungs and evolution of ascites: the so-called congenital high airway obstruction syndrome as previously reported.4

In 2 of 5 cases of an RAA/LDA (cases 4 and 5), intracardiac abnormalities with pulmonary outflow tract obstruction and a more diminutive ductus arteriosus from an aberrant LSCA were also documented, with retrograde ductal flow in 1 and antegrade ductal and proximal LSCA flow in the other. In 1 fetus (case 1), a vascular ring was clearly evident prenatally in the 3VV, but we were unable to differentiate between a DAA, an LDA with a very diminutive left aortic arch (LAA), and an RAA/LDA. In this case, the definitive diagnosis of an RAA/LDA was made postnatally.

One patient (case 8) with an RAA/LDA who had undergone fetal echocardiography at 33 and 36 weeks for suspected fetal coarctation in our program was only after birth found to have a high

### Table 1. Cases With a Diagnosis of a Vascular Ring

<table>
<thead>
<tr>
<th>Case</th>
<th>EGA at Diagnosis, wk</th>
<th>Reason for Fetal Echocardiography</th>
<th>Type of Associated Vascular Ring</th>
<th>Associated CHD</th>
<th>Extracardiac/Chromosome Abnormalities</th>
<th>Clinical Outcome</th>
</tr>
</thead>
<tbody>
<tr>
<td>1</td>
<td>20</td>
<td>Abnormal 3VV</td>
<td>DAA vs RAA/ LDA/LSCA, postnatally confirmed RAA/LSCA</td>
<td>NL</td>
<td>NT</td>
<td>Clinically asymptomatic neonate</td>
</tr>
<tr>
<td>2</td>
<td>34</td>
<td>Abnormal 3VV</td>
<td>DAA</td>
<td>NL</td>
<td>0</td>
<td>Clinically asymptomatic neonate</td>
</tr>
<tr>
<td>3</td>
<td>20</td>
<td>Maternal diabetes</td>
<td>RAA/LDA/LSCA</td>
<td>NL</td>
<td>0</td>
<td>Clinically asymptomatic at 3 y</td>
</tr>
<tr>
<td>4</td>
<td>25</td>
<td>Suspected CHD</td>
<td>RAA/LDA/LSCA</td>
<td>DORV, critical PS</td>
<td>Findings of CHARGE association Ehrler-Danlos syndrome</td>
<td>Clinically asymptomatic neonate (from respiratory standpoint)</td>
</tr>
<tr>
<td>5</td>
<td>19</td>
<td>Suspected CHD</td>
<td>RAA/LDA/LSCA</td>
<td>TOF</td>
<td>Surgical resection of vascular ring at time of elective TOF repair (5 mo)</td>
<td>Surgical resection of vascular ring</td>
</tr>
<tr>
<td>6</td>
<td>19</td>
<td>Suspected CHD</td>
<td>DAA</td>
<td>DORV, SubAS</td>
<td>0</td>
<td>Pregnancy termination at 20 wk without autopsy</td>
</tr>
<tr>
<td>7</td>
<td>24</td>
<td>Suspected CHAOS</td>
<td>DAA</td>
<td>NL</td>
<td>0</td>
<td>Surgical division of left arch at 1 d</td>
</tr>
<tr>
<td>8</td>
<td>33</td>
<td>Suspected CHD</td>
<td>Cervical RAA/LPDA, left descending aorta</td>
<td>Mild CoA (initially suspected prenatally)</td>
<td>DORV, critical PS</td>
<td>Vascular ring resection and aortopye at 2 y for respiratory symptoms</td>
</tr>
<tr>
<td>9</td>
<td>22</td>
<td>Suspected CHD</td>
<td>DAA</td>
<td></td>
<td>0</td>
<td>Neonate symptomatic with stridor, had surgical resection of vascular ring</td>
</tr>
</tbody>
</table>

AS indicates aortic stenosis; CHAOS, congenital high airway obstruction syndrome; CoA, coarctation of the aorta; EGA, estimated gestational age at diagnosis; NL, normal; NT, chromosome abnormality not tested; and 0 no chromosome abnormality, including 22q11.2 deletion.
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Figure 2. A, A cephalad cross-sectional sweep from the 3VV shows the normal leftward position of the aortic arch and ductus arteriosus relative to the trachea in a healthy fetus. B, Sweeping toward the fetal head from the 3VV, the most inferior arch, the ductus arteriosus, can be seen diving from the left side of the midline anteriorly straight back to the left of the midline and trachea (asterisk) posteriorly to join the descending aorta. C, Sweeping further toward the fetal head, the normal LAA can be shown coursing from the anterior mediastinum to the right of the midline, crossing the midline and coursing to the left of the trachea (asterisk) to join the descending aorta. In the normal arch arrangement, there is no vascular structure behind the trachea. See Video 1. Ao indicates ascending aorta; DA, ductus arteriosus; dAo, descending aorta; L, left; PA, pulmonary artery; R, right; and SVC, superior vena cava.

cervical RAA with mild distal arch obstruction, an LDA from an aberrant LSCA, and a left-sided descending aorta, resulting in mild, somewhat high tracheal and right main stem bronchus obstruction. On retrospective review of the fetal echocardiograms, a long-axis view of the aortic arch and a transverse arch view in a 3VV sweep could never be shown, and the descending aorta was entirely to the left behind the left atrium and in front of the spine (the usual position). In addition, no vascular structure could be visualized coursing behind the lower aspect of the trachea, making us less suspicious of a vascular ring.

Clinical Outcomes
Of the 9 pregnancies, 1 had termination of pregnancy without fetal autopsy, and 8 were continued, of which 7 delivered at term. One fetus with major polyhydramnios was delivered prematurely at 32 weeks as a consequence of preterm labor. The presence of severe tracheal compression associated with a DAA in this fetus (case 7) necessitated emergent surgical ring resection in the first day. Two neonates were clinically symptomatic with inspiratory stridor and had division of the vascular ring, resulting in resolution of the symptoms (cases 8 and 9). Two neonates who were clinically asymptomatic underwent elective vascular ring resection, 1 within the first week of life at an outside institution (case 2) and the second at the time of TOF repair (case 5). Three others, all with an RAA/LDA in utero, had not received any intervention for the vascular rings and remained asymptomatic at 3 months to 3 years.

Discussion
The literature regarding the prenatal diagnosis of vascular rings is extremely limited. In a pictorial essay, Yoo et al first illustrated the fetal sonographic features of aortic arch anomalies with emphasis on the utility of the 3VV and reviewed developmental mechanisms. Patel et al reported 6 cases of prenatally diagnosed vascular rings, including 2 with associated structural cardiac defects. Finally, in a retrospective study by Achiron et al, only 1 true vascular ring was described. In this series, we describe the prenatal findings of vascular rings in 9 patients, including
4 with associated intracardiac cardiac defects, show the utility of color Doppler imaging in confirming the diagnosis, and document the clinical outcomes, including associated symptoms and presence of extracardiac abnormalities.

We have confirmed that vascular rings can be diagnosed with fetal echocardiography using both 2-dimensional (gray scale sonography) and color flow imaging. Cross-sectional sweeps from a standard 3VV toward the fetal head, in particu-

**Figure 3.** Echocardiograms obtained at 34 weeks’ gestation in a fetus with a DAA (case 2). **A,** In the 3VV, a gap is identified between the main pulmonary artery and ascending aorta (arrow). The ductus arteriosus is of normal size and courses to the left of the trachea. See Video 2. **B,** Sweeping toward the fetal head, both the RAA and LAA, the latter of which is smaller, can be seen coursing on either side of the trachea (asterisk). **C,** Sweeping toward the fetal head, the mirror image brachiocephalic vessels (4 in total shown with arrows: the right and left carotid arteries, most anterior, and the right and left subclavian arteries, posterior) can be shown. **D,** Color flow mapping confirms the patency of the vascular ring coursing around the trachea (asterisk). See Video 3. **E,** In a coronal image, 2 vascular structures (the LAA and RAA) in a short-axis view on either side of the carina are shown. Abbreviations are as in Figure 2.
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lar, show the relationship of the arches to the trachea and the vascular structures coursing around and behind the trachea. Although the diagnosis was often initially suspected on the basis of the 2-dimensional images alone, color flow mapping provided confirmation that the structures were vascular in nature and was particularly useful in identifying the more diminutive ductus arteriosus associated with pulmonary outflow obstruction. Given the presence of fluid-filled lungs, fetal echocardiography provides more information with respect to the relationship of vascular and airway structures to each other that is more akin to postnatal magnetic resonance and computed tomographic images than postnatal echocardiography. Furthermore, after birth, the ductus arteriosus is often closed by the time of diagnosis, and its contribution to the ring is not so easily understood on echocardiography or even other imaging modalities. In the fetus, the ductus arteriosus can be readily shown even when more diminutive than normal in the context of pulmonary outflow tract obstruction, as we observed. In all 4 cases of a DAA, a large left-sided ductus arteriosus was found, another aspect of the abnormality not usually identified after birth.

Postnatally, many affected infants and children have a diagnosis of a vascular ring, particularly when in isolation, with the development of clinical symptoms, including large airway obstruction or dysphagia. In a 25-year experience with vascular rings, Woods et al. reported stridor and frequent upper respiratory tract infections as presenting symptoms for referral in 46% and 35% of patients, respectively. In a retrospective review, Shah et al. found that 91% of patients had airway symptoms, and 47% had esophageal symptoms. Patients with DAA had a significantly higher rate of respiratory distress and presented earlier compared with patients with RAA/LDA, who had a higher rate of dysphagia and presented later in childhood, often associated with onset of solid feeds. In our patients, only 3 of the 8 treated after birth were clinically symptomatic. However, most have had only short-interval follow-up thus far. It is also possible that prenatally diagnosed vascular rings represent a less severe spectrum of disease, perhaps the true spectrum.

Although not identified among the patients in this series, chromosomal abnormalities (particularly a 22q11.2 deletion) can be observed in patients with vascular rings, an association that is important for prenatal counseling. In a series of 66 cases of isolated anomalies of laterality or branching of the aortic arch, McElhinney et al. found a chromosome 22q11.2 deletion to be present in 24%, including 14% of patients with DAA and 32% with an RAA and LSCA. In addition to a 22q11.2 deletion, vascular rings have also been observed in oculo-auriculo-vertebral-Goldenhar complex, and in 1 patient from this series, Ehler-Danlos syndrome was diagnosed.
In conclusion, vascular rings can be readily detected at fetal echocardiography with attention to the relationship of the aortic and ductal arches to the trachea and the identification of a vascular structure coursing around and behind the trachea. Prenatal detection of these vascular congenital anomalies, whether in isolation or associated with other structural heart disease, should result in more appropriate counseling of affected pregnancies and additional testing including fluorescence in situ hybridization for a 22q11.2 deletion. Patients with prenatally diagnosed vascular rings may represent a less severe spectrum of the disease, with proportionately fewer infants manifesting symptoms after birth.

Figure 5. Vascular ring in a 25-week gestational age fetus with complex CHD including a DORV with a more rightward and anterior aorta, PS, and a ventricular septal defect (case 4). A, The 4-chamber view shows the relationship of the descending aorta (arrow) to the posterior wall of the left atrium and pulmonary veins. B, The 3VV shows the relationship of the great arteries to each other, and the position of the descending aorta suggests the possibility of an RAA. See Video 4. C, Sweeping toward the fetal head, the trachea is shown encircled by the rightward aortic arch and the smaller leftward ductus arteriosus. See Video 5. D, On color flow imaging, the vascular ring coursing around the trachea is confirmed, with retrograde flow shown in the ductus arteriosus, in keeping with critical pulmonary outflow obstruction. LPV indicates left pulmonary vein; LV, left ventricle; RPV, right pulmonary vein; and RV, right ventricle; other abbreviations are as in Figure 2.
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References


