Prenatal diagnosis of vascular rings and outcome

Shweta Bakhru¹, Nageswara Rao Koneti¹, Sujata Patil¹, Bhargavi Dhulipudi¹, Tapan Dash¹, Geeta Kolar², Suseela Vavilala²
¹Department of Pediatric Cardiology, Rainbow Children’s Heart Institute, Hyderabad, Telangana, India, ²Department of Fetal Medicine, Fernandez Hospital, Hyderabad, Telangana, India

ABSTRACT

Background: Vascular rings (VRs) present with varied symptoms and may result in significant morbidity before an accurate diagnosis is made. Prenatal diagnosis may be useful to plan surgery after birth.

Objectives: The purpose of the study was to see the feasibility of accurate diagnosis of VR during antenatal ultrasound examination and describe their outcome.

Methods: This is a retrospective observational study between January 2014 and December 2019. Vascular rings were diagnosed on the basis of three vessel tracheal view and neck vessels arrangements on fetal echocardiogram. Postnatal evaluation by transthoracic echocardiography and computerized tomography angiogram was performed. Surgical repair was done as per standard indications.

Results: A total of 35 cases of fetal VRs (median gestational age: 24 weeks [range: 19–35]) were diagnosed during the study period. There were four dichorionic diamniotic twin gestation pregnancies. The right aortic arch (RAA) with anomalous left subclavian artery (ALSA) was suspected in 31 fetuses, double aortic arch (DAA) in 3, and circumflex aorta in 1. Twenty-six (74%) patients had successful deliveries. One patient had a spontaneous miscarriage, 2 underwent termination, and 6 were lost to follow-up. Postnatal assessment showed RAA with ALSA in 18, DAA in 5, circumflex aorta in 2, and no abnormality in 1. Twenty-two (86%) were operated (RAA with ALSA: 17, DAA: 4, and circumflex aorta: 1) and four were waiting for surgery. Two patients died due to prematurity-related complications. All survivors are symptom free during follow-up (median: 2.24; range: 0.2–5.6 years).

Conclusions: Fetal echocardiography enables prenatal diagnosis and planning of postnatal repair of VRs.

Keywords: Double aortic arch, fetal echocardiogram, prenatal diagnosis, right aortic arch, vascular ring

INTRODUCTION

Isolated vascular rings (VRs) are commonly overlooked during routine antenatal ultrasonographic evaluation. The incidence of VR in the absence of other cardiac defects in a fetus is about 1 in 1000 pregnancies.[31] Systematic evaluation of the fetal heart, including three-vessel tracheal view, gives a vital clue to the diagnosis of VRs. Associated genetic abnormalities like 22q11.2 microdeletion are rare but essential in terms of...
prognostication.\cite{2,3} Clinical presentation of VR in the postnatal period may vary from asymptomatic to severe respiratory distress based on the severity of airway obstruction. Frequent aspirations, dysphasia, respiratory infections, and persistent stridor are some other manifestations of untreated VRs.\cite{4} Postnatally, VRs are often mistaken for upper airway pathology and missed unless specifically looked for. Delayed diagnosis can result in significant damage to the upper airways from persistent compression. It is, therefore, necessary to identify early and plan the treatment. This study describes a series of antenatally diagnosed VR with postnatal outcome.

**METHODS**

This is a retrospective observational study done from two centers: (i) tertiary cardiac center with pediatric and fetal cardiac program and (ii) tertiary fetal medicine unit. The data were collected from January 2014 to December 2019. All diagnosed and suspected cases of isolated VRs were included in the study.

**Fetal echocardiography**

Fetal echocardiogram was performed on Phillips IE 33 (Koninklijke Philips N. V., Amsterdam, The Netherlands) or GE Voluson sonography equipment (General Electric Company, Boston, MA 02210, United States). The study was done using fetal probes (Philips C5-2, GE Voluson C1-6). Color flow mapping and spectral Doppler were used whenever needed during the study. Three-dimensional (3D) fetal echocardiogram and volume rendering were available from 2019 at our center. A full-volume acquisition was done using eM6C (GE Voluson). The study was done using grayscale, color flow, and power Doppler. An offline analysis and 3D color flow images were rendered.

Inclusion criteria were as follows:

- All suspected cases of VR:
  1. Right aortic arch (RAA)
  2. Double aortic arch (DAA)
  3. Abnormal descending aorta.

Exclusion criteria were as follows:

- Associated intracardiac anomalies
- Visceral situs abnormalities, including situs inversus and ambiguous.

**Fetal echocardiogram and imaging protocol**

The following steps were used to diagnose fetal VR [Figures 1 and 2]:

i. Four-chamber view for the position of the descending aorta [Figure 1a]
ii. Three-vessel tracheal view is to identify the sidedness of the aortic arch and subclavian artery
iii. Identification of bifurcation of the ascending aorta
iv. Location of the arch in tracheal bifurcation view [Figure 1b].

The study was done after obtaining Pre-conception and Pre-natal diagnostics Techniques Act clearance form. Fetus cardiac situs was determined as an initial step in all cases. Mid-thoracic four-chamber view was obtained to see the position of the descending thoracic aorta. It is more toward midline than normal, in case of RAA. After assessing intracardiac anatomy, three-vessel tracheal view was obtained. The sidedness of the aortic arch can be identified in this view. The left aortic arch travels left of the trachea and joins left-sided ductus arteriosus and forms “V.” The right-sided aortic arch travels right and posterior of the trachea and joins left-sided ductus arteriosus to form “U” [Video 1]. Cranial vessels should be identified when VR is suspected. Most of the time, anomalous left subclavian artery (ALSA) is aberrant in the isolated RAA. The first vessel in that case (left carotid artery) travels anterior to the trachea and does not bifurcate (unlike the innominate artery). ALSA can be evaluated in a three-vessel tracheal view or aortic
arch long axis [Figure 2]. A study of the ascending aorta is essential in all cases of RAA in three-vessel view. Early bifurcation of the ascending aorta [Figure 3c] can be seen in case of DAA. Furthermore, three-vessel view shows both left and right arches encircling the trachea. Identification of cranial vessel pattern is important to have the diagnosis of DAA [Video 3]. Finally, the coronal view of the thorax should be obtained to see the longitudinal section of the trachea and its relationship with aortic arch.

**Imaging analysis**

An off-line analysis of suspected cases was done. (i) ALSA was demonstrated in all RAA cases to diagnose VR. ALSA (fourth branch) arises from the descending aorta travels left side crossing posterior to trachea. Kommerell diverticulum was seen in some of the cases. However, it was not mandatory for the diagnosis [Figure 2]. (ii) DAA was diagnosed based on the formation of “O” around the trachea due to the right and left aortic arches [Videos 2, 3 and 4]. An incomplete DAA is suspected when the right arch is present without ALSA. This implies that a segment distal to left innominate artery is atretic [Figure 3]. (iii) Unusual arch pattern from right to left, posterior to the trachea is considered as circumflex aorta [Figure 4].

Amniocentesis was done to look for the 22q11.2 microdeletion in selected cases based on the family consent.

**Postnatal evaluation**

All neonates with antenatal diagnosis of VR had undergone transthoracic echocardiogram after birth. The sidedness of the arch and cranial vessels was identified. Computerized tomography (CT) angiogram was done in the cases of symptomatic neonates or electively after 6 months of age in all cases [Figure 5]. A barium swallow was done whenever required.

**Management strategy**

Symptomatic neonates due to upper respiratory compression were advised surgery for the VR division. In an asymptomatic VR, surgery was delayed for 6 months to 1 year of age. Lateral thoracotomy without cardiopulmonary bypass was the usual approach for the division of VR [Figure 6]. Outcomes were analyzed.

**RESULTS**

A total of 35 fetuses with VR were identified during the study period. Figure 7 provides overall information about the total cases. The median gestational and maternal age at the time of diagnosis was 24 weeks (range: 19–35) and 26 years (range: 22–32), respectively. There were four twin pregnancies of diamniotic dichorionic type. Amniocentesis was done in 13 (37%) cases, and no genetic abnormality was found.

**Prenatal diagnosis**

RAA forming the VR was the most common diagnosis in our study population constituting 88% (n = 31). DAA [Figure 3a-c] was suspected in 8% (n = 3) and circumflex aorta [Figure 4a] in 2.8% (n = 1).

![Figure 3: (a) The complete double aortic arch encircling the trachea. (b) Three-dimensional volume-rendered color flow showing the double aortic arch. (c) Early bifurcation of ascending aorta in outflow tract view. (d) Computerized tomography angiogram demonstrating the double aortic arch in a neonate with prenatal diagnosis](image1)

![Figure 4: A case of circumflex aorta (a) three-dimensional volume-rendered imaging with color flow showing the ascending aorta and ductus arteriosus. (b) Computerized tomography angiogram confirms the posterior compression of the trachea and esophagus by the aorta. (c) Coronal plane showing the circumflex vessel. (d) Volume-rendered image showing the arch of the aorta and its branches. AA: Ascending aorta, DA: Ductus arteriosus](image2)
Fetal outcome

Twenty-six (74%) had a successful pregnancy and institutional delivery. One had a spontaneous miscarriage at 26 weeks of gestation. Two pregnancies were terminated due to family decisions. Six patients did not respond to our communications, and no further follow-up was available.

Postnatal outcome

Postnatal data were available for 26 (74%) patients. The diagnosis of VR was confirmed in 25/26 (96%) cases. Eighteen (69%) patients had RAA with ALSA forming a VR. One of the RAAs had an aberrant innominate artery instead of ALSA. DAA was found in total 5 (19%) cases, of which 3 were prenatally diagnosed and 2 were thought as RAA during fetal ultrasound, but CT scan confirmed double arch. Both the cases were found to have incomplete DAA. One (3.8%) patients had a circumflex aorta causing esophageal compression. One case in a twin pregnancy was found to have a normal left aortic arch instead of RAA.

Clinical presentation

Three patients delivered prematurely (of which two patients died due to sepsis and pulmonary problems. Two patients had severe respiratory distress and four presented with dysphagia. Eighteen (69%) patients were asymptomatic at birth and during follow-up. Extracardiac, skeletal abnormalities were seen in two cases.

Surgical management

Twenty-two (86%) patients with VR were operated during this period. Four patients with DAA were operated within 2 months after birth. One patient with DAA was born at 27 weeks and succumbed due to sepsis after the VR division. Seventeen patients with RAA with ALSA were operated from left thoracotomy where significant esophageal compression was demonstrated [Figure 6]. There were no postoperative complications in these patients. One patient with the right circumferential aorta [Figure 7] became severe dysphagia during neonatal period due to esophageal compression. Aortic relocation, the VR surgery was not done as it appeared high risk in this case and therefore baby had gastrostomy. Four patients were waiting for the surgery.

The survivors were followed up for a median period of 2.24 (range: 0.2–5.6) years. All patients were asymptomatic during the follow-up period.

DISCUSSION

VR is an anomaly where the trachea and esophagus are encircled either partially or entirely by the vascular structure, which can be patent or atretic.\(^5,6\) Prenatal diagnosis of VR may be beneficial to plan surgical repair at an appropriate time.\(^6\) A routine fetal echocardiogram may often overlook VR, however, meticulous, systematic evaluation gives a clue to the diagnosis in the fetus.\(^7\) In our cohort of 35 patients of fetal VRs, 26 patients were confirmed by postnatal assessment. The remaining 9 (25%) patients were either terminated the pregnancy (n = 3) or could not be communicated (n = 6). This suggests that social stigma related to congenital heart disease plays a significant role in the society and influences the fetal outcome.

The diagnosis of VR was confirmed in 96% of our postnatal cases. Prenatal diagnosis is possible if the fetal echocardiogram is performed meticulously by an expert. Four-chamber and outflow tract views will not provide adequate information to diagnose VR. A systematic evaluation of three-vessel view and a high thoracic section of the fetus will give essential clues to the diagnosis. Three-vessel tracheal view is mandatory to see the sidedness of the aortic arch. Mid- and high thoracic sections of both coronal and axial views are of utmost importance to identify the neck vessels to decide the type of VR.\(^8,9\) However, the specific subtype of VR was not possible in all cases. Two of our incomplete DAA were diagnosed as RAA with ALSA during prenatal evaluation despite meticulous evaluation. This was probably because of the misinterpretation of the left subclavian artery.

Figure 5: Computerized tomography angiogram of axial (a) and coronal (b) planes showing Right aortic arch forming the vascular ring. Diverticulum of Kommerell (b) is seen giving origin to the left subclavian artery

Figure 6: (a) Three-vessel tracheal view of fetal echocardiogram showing RAA. White arrow showing trachea. (b) Intraoperative picture showing RAA with ligamentum (white arrow) causing vascular ring. RAA: Right aortic arch, SVC: Superior vena cava, PA: Pulmonary artery, Ao: Aorta

Figure 7: Computerized tomography angiogram of axial (a) and coronal (b) planes showing Right aortic arch forming the vascular ring. Diverticulum of Kommerell (b) is seen giving origin to the left subclavian artery.
aortic arch as the left brachiocephalic trunk or overlap
of the left ductus arteriosus. Moreover, the atretic distal
left aortic arch in one case and less dominant left aortic
arch in another gave inadequate information for the
diagnosis. However, both the cases underwent surgical
repair.

Wang et al. accurately diagnosed VRs in fetuses
with aortic arch abnormalities using 2D and 3D
sonography. In their series of 30 fetuses, 4 were DAA,
21 were RAA, and 4 were left arch with aberrant right
subclavian artery (ARSA). They had better sensitivity
for the detection of brachiocephalic ($P < 0.1$) and arch
anomalies.\cite{10} We performed 3D volume rendering in
the last few cases and found useful to correlate with 2D
modality [Figure 1c]. Our series has predominantly RAA
followed by DAA. We did not include the left aortic arch
with ARSA as a VR due to its incomplete form.

Most of our cases were having the RAA with
ALSA (69% [18/26]) forming a VR. These cases
were all confirmed by the postnatal transthoracic
echocardiogram and subsequently by CT angiogram
before surgical repair [Figure 5]. The RAA, left ductus
arteriosus, and ALSA form the complete VR and appear
as “U” shape in three-vessel tracheal view [Figure 2a].
The ALSA can be assessed in most of the cases by
color Doppler. DAA was found in 19% (5/26) of our
cases. Two fetuses were diagnosed as RAA with ALSA
instead of incomplete DAA. We found that this type
is a difficult subset to diagnose by sonography, since
complete encircling and formation of “O” are not
possible.

The large series ($n = 81$) published by Li et al. showed
RAA with ALSA forming a VR in 56.8% and DAA in
3.7%.\cite{11} They had also included a left aortic arch with
ARSA an incomplete VR, and hence, our series could not
be compared in terms of incidence.

Undiagnosed cases may present with varied manifestations
from asymptomatic status to severe respiratory

Figure 7: The flowchart of all cases with postnatal outcome
distress. Symptomatic patients may have significant morbidity due to airway compression. All our patients had institutional delivery and immediate postnatal evaluation. Patients with DAA and circumflex aorta had undergone early surgery due to airway and esophageal compression. Only one patient died after surgery with associated comorbidities. Therefore, it is necessary to have a prenatal diagnosis of VRs to plan early surgery in severe types such as DAA and circumflex aorta. Most of our patients with RAA and ALSA (17/22) underwent elective surgical repair of VR during infancy to prevent the complications.

The data published by Tuo et al. considered surgical repair only in symptomatic patients (n = 4/19), mostly DAA causing airway compression. The remaining asymptomatic cases were kept on medical follow-up. This suggests that DAA causes significant obstruction of the upper airway and esophagus and hence necessitates early surgical repair. Symptomatic delayed cases with respiratory symptoms, especially DAA, carries significant morbidity and mortality due to upper airway compression and persistent bronchomalacia even after surgical division. Therefore, fetal diagnosis is crucial and does allow for a prospective plan.

Asymptomatic VR patients had an elective CT angiogram after 6 months of age for a future surgical repair if a radiological obstruction was seen. All our asymptomatic patients were studied by barium swallow to demonstrate esophageal compression (posterior indentation) before the surgical decision. Moreover, surgical management of VR is safe and definitive. Most of the time, surgery can be accomplished by thoracotomy incision. Naimo et al. described 36-year follow-up data of the surgical division of VR with excellent outcomes and improvement in tracheomalacia. Muscle-sparing thoracotomy reduces mean hospital stay, the need for chest tube drainage, and sternal scar.

VR is known to be associated with 22q11.2 deletion. This needs prenatal genetic diagnosis so that counseling can be offered to the family in positive cases. Our experience is somewhat different in our small series. We found normal karyotyping and fluorescence in situ hybridization in 13 cases who underwent amniocentesis.

**CONCLUSIONS**

The diagnosis of VRs during prenatal ultrasound examination is feasible. Careful cranial sweep from three-vessel view to tracheal view appears to provide more information for the diagnosis of a VR in the fetus. The prenatal diagnosis appears to be useful in the proper planning of surgical management before they become symptomatic.

**Limitations**

This is a small cohort of isolated diagnosis of fetal VRs. A comparative study between prenatal diagnosis and undiagnosed historical cases may be useful to see the actual outcome and usefulness of fetal diagnosis.

**Financial support and sponsorship**

Nil.

**Conflicts of interest**

There are no conflicts of interest.

**REFERENCES**

1. Bronshtein M, Lorber A, Berant M, Auslander R, Zimmer EZ. Sonographic diagnosis of fetal vascular rings in early pregnancy. Am J Cardiol 1998;81:101-3.

2. Patel CR, Lane JR, Spector ML, Smith PC. Fetal echocardiographic diagnosis of vascular rings. J Ultrasound Med 2006;25:251-7.

3. Jain S, Kleiner B, Moon-Grady A, Hornberger LK. Prenatal diagnosis of vascular rings. J Ultrasound Med 2010;29:287-94.

4. Suh YJ, Kim GB, Kwon BS, Bae EJ, Noh CI, Lim HG, et al. Clinical course of vascular rings and risk factors associated with mortality. Korean Circ J 2012;42:252-8.

5. Yoo SJ, Min JY, Lee YH, Roman K, Jaeggi E, Smallhorn J. Fetal sonographic diagnosis of aortic arch anomalies. Ultrasound Obstet Gynecol 2003;22:535-46.

6. Tuo G, Volpe P, Bava GL, Bondanza S, De Robertis V, Pongiglione G, et al. Prenatal diagnosis and outcome of isolated vascular rings. Am J Cardiol 2009;103:416-9.

7. Achiron R, Rotstein Z, Lipitz S, Mashiah S, Hegesh J. First-trimester diagnosis of fetal congenital heart disease by transvaginal ultrasonography. Obstet Gynecol 1994;84:69-72.

8. Yoo SJ, Lee YH, Kim ES, Ryu HM, Kim MY, Choi HK, et al. Three-vessel view of the fetal upper mediastinum: An easy means of detecting abnormalities of the ventricular outflow tracts and great arteries during obstetric screening. Ultrasound Obstet Gynecol 1997;9:173-82.

9. Yagel S, Arbel R, Antebiy EY, Raveh D, Achiron R. The three vessels and trachea view (3VT) in fetal cardiac scanning. Ultrasound Obstet Gynecol 2002;20:340-5.

10. Wang Y, Fan M, Siddiqui FA, Wang M, Sun W, Sun X, et al. Strategies for accurate diagnosis of fetal aortic arch anomalies: Benefits of three-dimensional sonography with spatiotemporal image correction and a novel algorithm for volume analysis. J Am Soc Echocardiogr 2018;31:1238-51.

11. Li S, Luo G, Norwitz ER, Wang C, Ouyang S, Yao Y, et al. Prenatal diagnosis of congenital vascular rings and slings: Sonographic features and perinatal outcome in 81 consecutive cases. Prenat Diagn 2011;31:334-46.

12. Evans WN, Acherman RJ, Ciccolo ML, Carrillo SA, Mayman GA, Luna CF, et al. Vascular ring.
13. Naimo PS, Fricke TA, Donald JS, Sawan E, D’Udekem Y, Brizard CP, et al. Long-term outcomes of complete vascular ring division in children: A 36-year experience from a single institution. Interact Cardiovasc Thorac Surg 2017;24:234-9.

14. D’Antonio F, Khalil A, Zidere V, Carvalho JS. Fetuses with right aortic arch: A multicenter cohort study and meta-analysis. Ultrasound Obstet Gynecol 2016;47:423-32.