Prenatal diagnosis of vascular rings and outcome

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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.[1] 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

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prognostication.[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.[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:

1. Associated intracardiac anomalies
2. 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

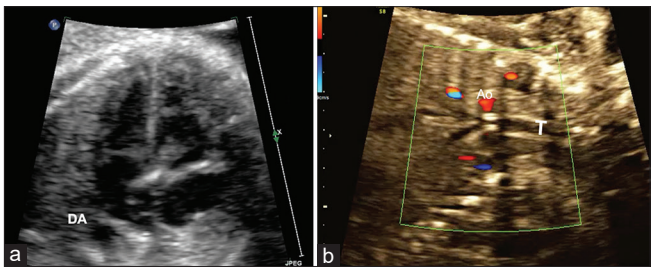

Figure 1: (a) Four-chamber view for the location of descending aorta. Descending aorta is medially shifted close to the spine than usual position. (b) Coronal section of the thorax at a tracheal bifurcation. This suggests the arch of the aorta on the right side of the trachea

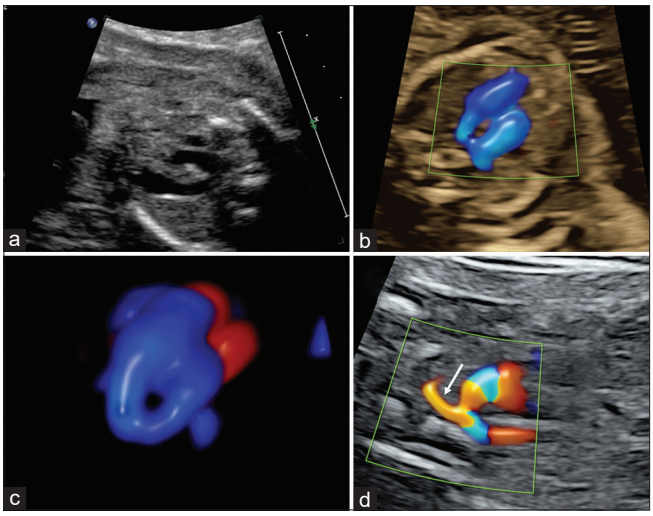

Figure 2: (a) Three-vessel tracheal view showing the formation "U" by the right aortic arch and left ductus arteriosus. (b) Color flow mapping showing the right aortic arch. (c) Three-dimensional volume-rendered image showing right aortic arch with left ductus arteriosus. (d) High thoracic transverse view showing aberrant left subclavian artery (white arrow). Right aortic arch: Right aortic arch

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).

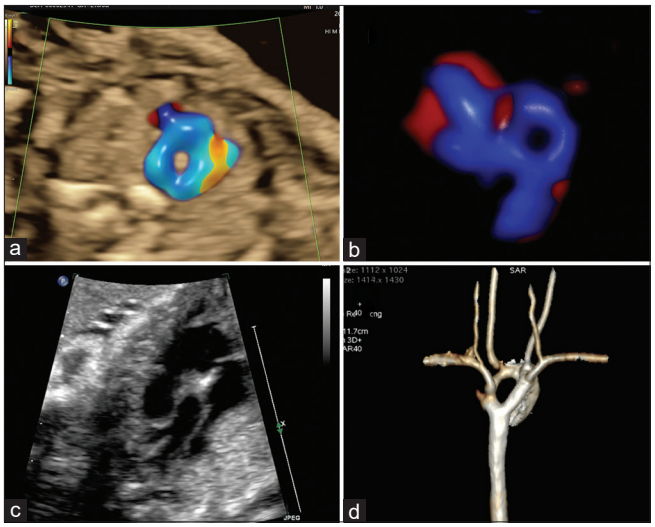

Figure 3: (a) The complete double aortic arch encircling the trachea. (b) Three-dimensional volume-rendered color flow showing the double aortic arch. (c) Early bifurcation of ascending aorta in outflow tract view. (d) Computerized tomography angiogram demonstrating the double aortic arch in a neonate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

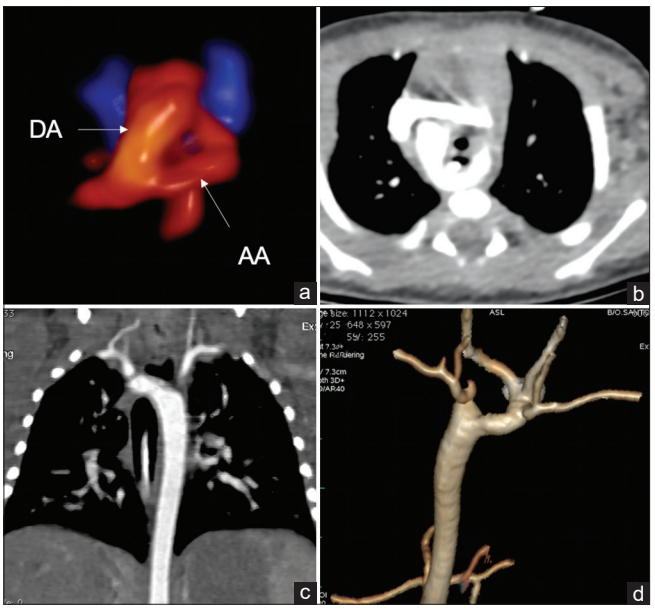

Figure 4: A case of circumflex aorta (a) three-dimensional volume-rendered imaging with color flow showing the ascending aorta and ductus arteriosus. (b) Computerized tomography angiogram confirms the posterior compression of the trachea and esophagus by the aorta. (c) Coronal plane showing the circumflex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

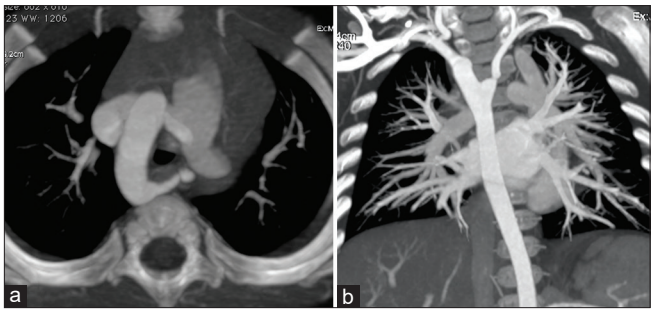

Figure 5: Computerized tomography angiogram of axial (a) and coronal (b) planes showing Right aortic arch forming the vascular ring. Diverticulum of Kommerell (b) is seen giving origin to the left subclavian artery

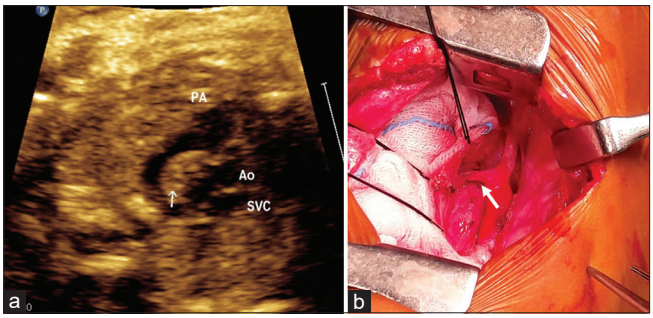

Figure 6: (a) Three-vessel tracheal view of fetal echocardiogram showing RAA. White arrow showing trachea. (b) Intraoperative picture showing RAA with ligamentum (white arrow) causing vascular ring. RAA: Right aortic arch, SVC: Superior vena cava, PA: Pulmonary artery, Ao: Aorta

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

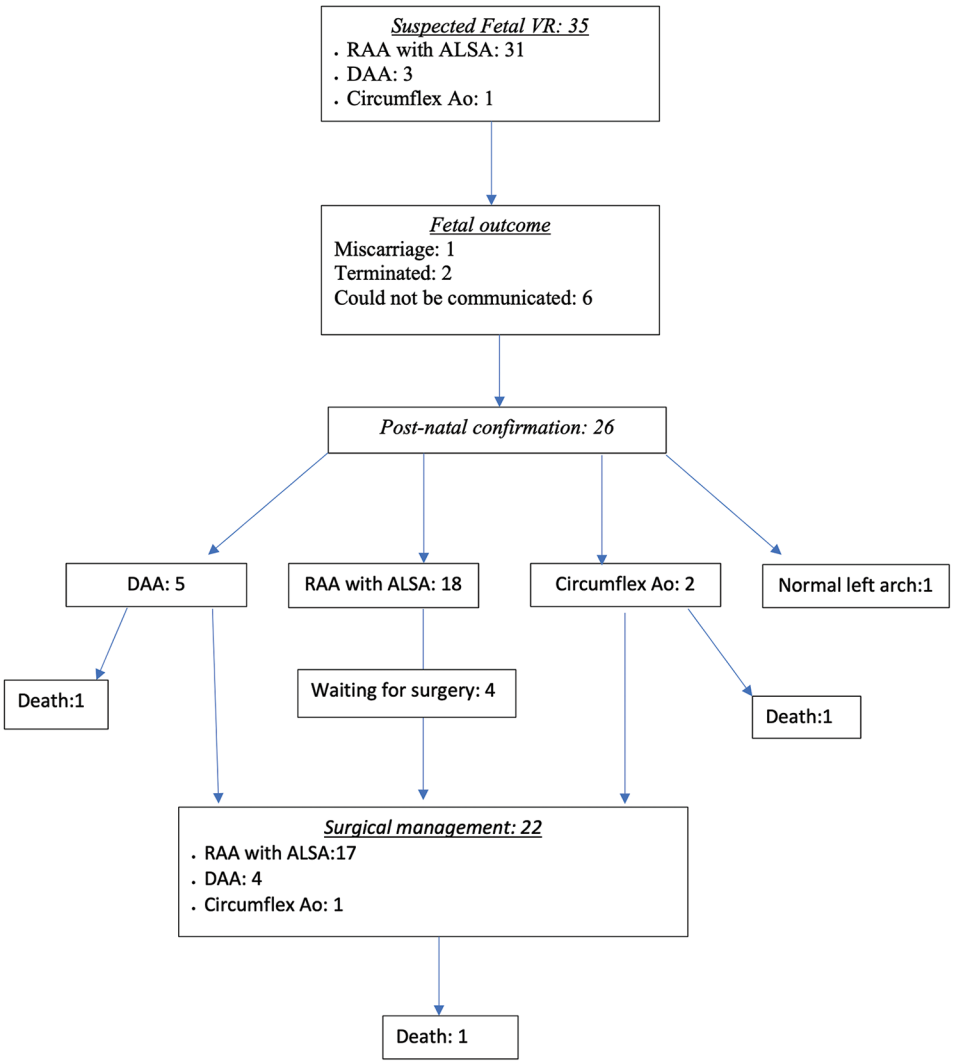

Figure 7: The flowchart of all cases with postnatal outcome

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.[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%.[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

distress. Symptomatic patients may have significant morbidity due to airway compression.[4] 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.[6] 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.[12]

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 esophagus 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.[13]

VR is known to be associated with 22q11.2 deletion.[14] 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.

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Conflicts of interest

There are no conflicts of interest.

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