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Case Report

Tracheal stenosis due to vascular rings: its possible prenatal diagnosis based on four cases of vascular rings with or without eventual tracheal stenosis

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Summary

Vascular rings encircle the trachea and esophagus, and sometimes compress these organs, causing trachea/esophagus-compression-related symptoms. Four fetuses were diagnosed with vascular rings and treated in this tertiary institute. One infant showed the manifestation of tracheal stenosis after birth, whereas the remaining three did not. The authors analyzed the relationship between pre- *vs*. post-natal images and postnatal tracheal stenosis. In the infant with tracheal stenosis, a three-vessel tracheal (3VT) view by fetal echocardiography clearly showed the tracheal wall; however, a tracheal lumen was not evident. In the other three infants without tracheal stenosis, the tracheal walls were clearly observable and an echolucent tracheal lumen was evident. Fetal MRI also showed the loss of the continuity of the tracheal lumen. These two findings, an invisible tracheal lumen on fetal echocardiography and loss of tracheal-lumen-continuity on fetal MRI, may indicate tracheal stenosis and accompanying air-way-obstruction symptoms.

Key words: Fetal echocardiography; Magnetic resonance imaging; Three-vessel tracheal view; Tracheal stenosis; Vascular ring.

Introduction

Vascular rings are rare aortic arch abnormalities. Vascular structures encircle the trachea and esophagus, and sometimes compress these organs, causing trachea/esophagus-compression-related symptoms. In a case with severe tracheal compression, respiratory symptoms appear soon after delivery, requiring emergency care and surgery. Fetal diagnosis of vascular rings, especially presence/absence of tracheal compression is of clinical importance.

Image analyses (ultrasound and MRI) have enabled us to prenatally diagnose vascular rings. Fetal echocardiography with a three-vessel tracheal (3VT) view has especially enabled detection of the topological relationship of great vessels, leading to fetal diagnosis of this disorder [1]. However, prenatal image findings indicative of tracheal compression are not yet determined. The authors here present four consecutive cases with vascular rings. One infant showed respiratory symptoms due to tracheal stenosis, whereas the remaining three did not. Based on this experience, the authors suggest some clinically useful prenatal image-analysis-signs for predicting presence/absence of tracheal compression and respiratory symptoms.

Case Report

During a two-year period (December 2014 – December 2016), four fetuses were diagnosed with vascular rings in this institute. In almost all primary or secondary institutes in this area, obstetricians routinely perform fetal echocardiography at 28-31 weeks, usually employing four chamber-, three vessel-, and 3VT views. When obstetricians suspect structural abnormality, they transfer the mothers to this institute. The four fetuses were transferred to the authors, which they here demonstrate.

Table 1 shows the diagnostic opportunity, prenatal findings of the tracheal structure (tracheal luminal continuity ± : described later), postnatal diagnosis, and presence/absence of tracheal stenosis-related symptoms after birth. All mothers vaginally gave birth to term infants weighing a median of 3,230 (2,272-3,906) grams. Vascular rings were prenatally diagnosed at a median of 31 (range: 31-35) weeks.

Of the four infants, one (case 1) showed the manifestation of tracheal stenosis after birth, whereas the remaining three (cases 2-4) did not. To highlight the relationship between the pre- *vs*. post-natal image and presence/absence of postnatal tracheal stenosis, cases 1 and 2 will be described in detail. In case 1, fetal ultrasound clearly showed the tracheal wall (Figure 1a: arrow); however, importantly, a tracheal "lumen" was not evident. Fetal MRI (1.5-tesla system) also showed the "loss of tracheal-lumen-continuity"; in other words, the tracheal lumen was not followed for its entire length (Figure 2d: arrow). In case 2, contrarily, tracheal walls (anterior and posterior) were clearly observable and among them, an echolucent tracheal lumen was evident (Figure 1b: asterisk). Fetal MRI showed the continuity of the tracheal lumen (Figures 2e-h: arrowhead). After birth, echocardiography and three-dimensional CT revealed that case 1 had a double aortic arch, the origin of great vessels from the right heart, and pulmonary artery atresia, and case 2 had a double aortic arch. The trachea was tightly compressed in case 1 but it was not in case 2 (Figures 2i and j, respectively). In cases 3 and 4, fetal echocardiography revealed a tracheal lumen (Figures 1c and d: as-

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Table 1. — *The diagnostic opportunity, prenatal findings of a tracheal structure (tracheal luminal continuity), postnatal diagnosis, and presence/absence of tracheal stenosis-related symptoms after birth.*

	Case 1	Case 2	Case 3	Case 4
Gestational week at prenatal diagnosis (weeks)	31	31	31	35
Diagnostic opportunity for fetal ultrasound	FGR	3VT view abnormality	3VT view abnormality	3VT view abnormality
Tracheal lumen on 3VT view in fetal ultrasound	(-)	(+)	(+)	(+)
Tracheal luminal continuity in fetal MRI	(-)	(+)	NE	(+)
Delivery week (weeks)	40	41	39	39
Birth weight (grams)	2272	3906	3114	3340
Postnatal diagnosis	Double aortic arch, DORV, PA, VACTER association	Double aortic arch	Right aortic arch, left ductus arte-, riosus, aberrant left subclavian artery	Right aortic arch, Kommerell diverticulum
Tracheal stenosis-related symptom	Present	Absent	Absent	Absent

Abbreviations: DORV: double outlet right ventricle; FGR: fetal growth restriction; NE: not examined; PA: pulmonary atresia; 3VT: three-vessel trachea.

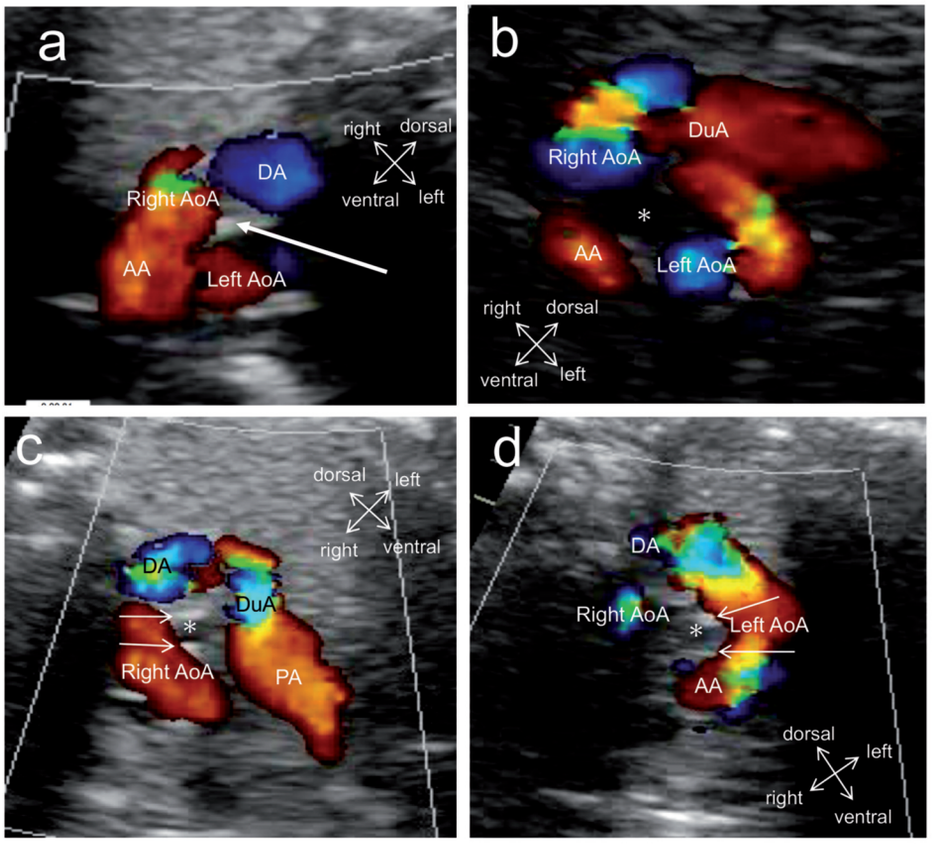

Figure 1. — Fetal ultrasound findings indicative of tracheal stenosis + *vs*. –.
Figures a, b, c, and d are from cases 1, 2, 3, and 4, respectively. Echogenic tracheal walls are evident (long arrow) but there is no echolucent area between them in case 1 (a), whereas an echolucent area (asterisk) is evident in cases 2-4 (b-d). The tracheal anterior wall (echogenic: small arrow), echolucent tracheal lumen (asterisk), and echogenic posterior tracheal wall (small arrow), the "three-layer pattern", can be clearly seen in Figures c and d.
AA: ascending aorta, AoA: aortic arch, DA: descending aorta, DuA: ductus arteriosus, PA: pulmonary artery.

terisk) and MRI also revealed a tracheal lumen along its entire length (figure not shown).

After birth, cases 2, 3, and 4 showed no dyspnea indicative of tracheal stenosis, whereas case 1 had severe dyspnea immediately after birth, requiring mechanical ventilation. On day 4, open resection of the right aortic arch was performed, relieving tracheal compression and allowing the discontinuation of mechanical ventilation. In addition to a double aortic arch, a double outlet right ventricle and pulmonary atresia were observed. Modified Blalock-Taussig shunt and ligation of the patent ductus arteriosus were performed at one month old.

Discussion

Here, the authors made two important observations. Fetal echocardiography and MRI facilitated an appropriate diagnosis of vascular rings, which enabled preparation for appropriate management after delivery. An "invisible" tracheal lumen based on fetal echocardiography and the loss of continuity of the tracheal lumen on fetal MRI may indicate tracheal stenosis and its accompanying air-way-obstruction symptoms.

Firstly, prenatal echocardiography and MRI enabled an ap-

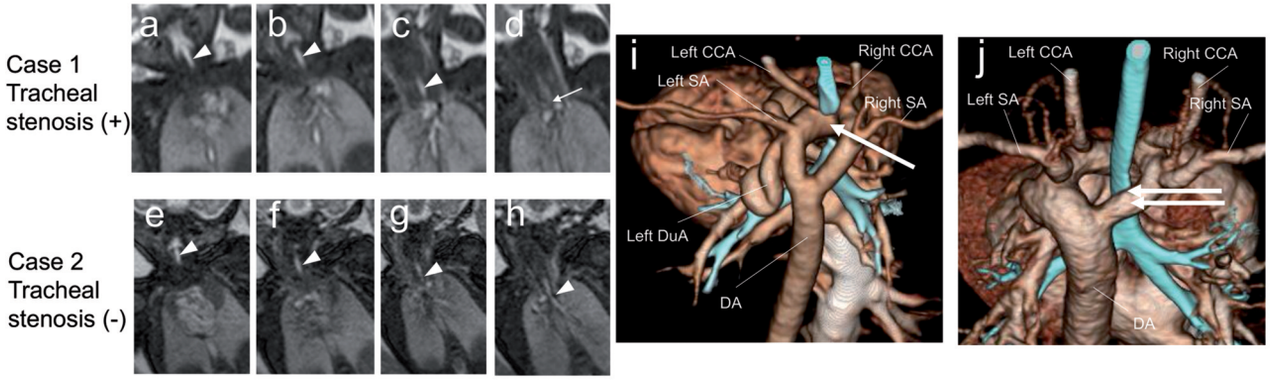

Figure 2. — Fetal coronal T2-weighted MRI (Figures a-h) and three-dimensional CT (Figures i and j) of cases 1 and 2. Figures a-d: MRI of case 1 indicates that the trachea is clearly visible in its upper portion (Figures a-c: arrowheads), but it cannot be distinguished in its lower part around the great vessel loop (possibly around the double aortic arch) (Figure d: arrow) and the trachea cannot be followed for its entire length. Figures e-h: In case 2, the entire length of the trachea can be visualized (Figures e-h: arrowheads) and there is no "loss of continuity" observed in case 1 (Figure d: arrow). Figures i, j: CT image after the births of cases 1 (Figure i) and 2 (Figure j), a view from the patients' dorsal side. A vascular ring due to a double aortic arch is evident (Figures i, j). Tracheal compression is evident in case 1 (Figure i: arrow), but not in case 2 (Figure j: double arrow). CCA: common carotid artery, DA: descending aorta, DuA: ductus arteriosus, SA: subclavian artery.

propriate diagnosis of vascular rings. Since the introduction of the 3VT view on fetal echocardiography, some reports described the prenatal diagnosis of vascular rings. It enables the preparation of treatment immediately after birth and facilitates the transfer of pregnant mothers to a well-equipped center [1]. Confining the topic to a double aortic arch, according to Trobo *et al.* [2], 13 articles on 35 cases of the prenatal echocardiographic diagnosis of a double aortic arch are reported up until 2015. Japanese obstetric guidelines [3] suggest the merit of performing fetal ultrasound morphological screening including the heart at 28-31 weeks, which was effective in this study to detect vascular rings. The present authors confirmed the efficacy of the 3VT view to detect vascular rings. Secondly, an "invisible" tracheal lumen on fetal echocardiography and the loss of continuity of the tracheal lumen on fetal MRI may indicate tracheal stenosis and accompanying air-way-obstruction symptoms. Some efforts have been made to associate tracheal compression and ultrasound findings; however, data are very limited. For example, in a case of severe tracheal compression, ultrasound findings mimicking congenital high airway obstruction syndrome were observed [4, 5]. The present study showed that in cases without postnatal tracheal stenosis-related symptoms, the tracheal lumen was seen between the anterior and posterior tracheal walls as an "echolucent area". The absence of this echolucent area/lumen indicated tracheal stenosis in case 1. Fetal MRI may also provide another additional finding: in cases without tracheal stenosis, an intact tracheal lumen could be easily followed for its entire tracheal length, whereas in a case with tracheal stenosis, its lumen was obscure, showing "loss of continuity". Although fetal MRI imaging may be influenced by the fetal movement, position, or level of equipment resolution, fetal MRI may also be a useful diagnostic tool for tracheal stenosis accompanied by vascular rings. These findings are based on only a four-case experience. Further study is needed to confirm the present observations.

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