Prenatal three-dimensional ultrasound detection of left pulmonary artery sling

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Summary
Left pulmonary artery (LPA) sling is a very rare congenital anomaly. This anomaly is often associated with tracheal narrowing caused by either intrinsic stenosis or malformation of tracheal rings. To the present authors’ knowledge, prenatal diagnosis of LPA sling has not been reported frequently, and three-dimensional ultrasound was not used in any of the previous cases. The aim of this article is to report one case of LPA sling, which was diagnosed in a fetus at 24 weeks gestation by routine echocardiography and four-dimensional spatio-temporal image correlation (STIC), with focus on the particularity of this rare anomaly prenatally diagnosed.

Key words: Left pulmonary artery sling; Prenatal ultrasound; Spatio-temporal image correlation (STIC).

Introduction
Left pulmonary artery (LPA) sling is a very rare congenital anomaly where the LPA arises distally, originates from the right pulmonary artery, turns sharply leftwards posterior to the trachea, and arrives to the left lung hilum through the space between the trachea and esophagus. This anomaly is often associated with tracheal narrowing, caused by either intrinsic stenosis or malformation of tracheal rings. To the present authors’ knowledge, prenatal diagnosis of LPA sling has not been reported frequently, and three-dimensional ultrasound was not used in any of the previous cases. The aim of this article is to report one case of LPA sling, which was diagnosed in a fetus at 24 weeks gestation by routine echocardiography and four-dimensional spatio-temporal image correlation (STIC), with focus on the particularity of this rare anomaly prenatally diagnosed.

Case Report
A 21-year-old gravida 2, para 0 woman was referred for a fetal echocardiography at 24 weeks gestation due to pulmonary artery bifurcation anomalies. Fetal echocardiography revealed LPA arising from the right pulmonary artery resulting in an LPA sling (Figure 1A). Likewise, combined with four-dimensional spatio-temporal image correlation (STIC) technology (Figure 1B) to better reflect spatial relationships between the LPA with the trachea and esophagus, LPA sling was confirmed in the fetal period. Ultrasonography once again did not show any other cardiac or body abnormality. At 37 weeks’ gestation, a female infant was delivered vaginally, she weighed 2,880 grams and Apgar scores were 9 and 10 at one and five minutes after birth, respectively. Due to the prenatal diagnosis of LPA sling causing high risk of postnatal respiratory failure, the baby developed tachypnea and circumoral cyanosis requiring supplemental oxygen on day of life 2. After delivery, the infant’s condition did not arouse sufficient attention. Meanwhile, the patient required discharge due to personal reasons, while the infant had respiratory complications as tachypnea and circumoral cyanosis at discharge. One year later, the infant was referred to Guangdong Provincial People’s Hospital for respiratory insufficiency and unexplainable recurrent pulmonary infections. Postnatal multislice spiral computed tomography (MSCT) (Figure 2) confirmed the diagnosis of the LPA sling. The baby therefore underwent correction via transection of the LPA and reimplantation anterior to the trachea and ligation of ductus arteriosus when she was one year and a month old.

Discussion
LPA sling is a rare congenital vascular anomaly where LPA abnormal originates from the right pulmonary artery and compresses the trachea. In the largest case series of 81 fetuses with prenatal diagnosis of vascular rings or slings, LPA slings only represented 0.02% [1], however, the mortality rate can be as high as 50%. The prevalence of LPA sling is purely conjectural, as there is no way to know the true number of asymptomatic cases that exist [2]. As far as the present authors know, this is the first definitive prenatal diagnosis of LPA sling with new technology, which has two-dimensional combined with four-dimensional spatio-temporal image correlation (STIC). In contrast, the accuracy of routine two-dimensional and color Doppler echocardiography to assess origin of LPA has been evaluated mostly in postnatal series of patients with LPA sling, and report on prenatal examinations are lacking [3, 4]. STIC is an automatic volume acquisition through the fetal heart, resulting in complete fetal cardiac cycle being dis
played in motion in a continuous three-dimensional cineloop sequence. STIC in offline settings could be used to confirm or exclude major congenital heart disease (CHD), and the anatomy of the fetal heart could be demonstrated effectively via means of STIC acquisition carried out by an operator unskilled in fetal cardiology [5]. With advances in both fetal routine echocardiography and STIC, abnormalities of the LPA is increasingly being recognized prenatally, allowing healthcare providers to anticipate management issues at the time of delivery or later in neonatal life, and assist parents to comprehend the prognosis. Hence, STIC can be used to improve and confirm in detail the diagnosis of LPA sling. LPA sling, which is not routinely investigated during fetal ultrasound examination, therefore, LPA sling can easily escape prenatal detection. The fetal three-vessel trachea (3VT) view is transverse plane of the upper mediastinum that evaluate the relationships of the main pulmonary artery, aortic arch, superior vena cava, ductus arteriosus and trachea [6]. The 3VT is particularly crucial in assessing a vascular anomaly [7]. 3VT includes another view which is three-vessel-pulmonary artery bifurcation, which detects the anomaly and defines the spatial relationship between the anomalous pulmonary artery and the trachea which are emphasized. So adding visualisation of three-vessel-pulmonary artery bifurcation view to routine echocardiography has been suggested, likely to increase the sensitivity of ultrasound screening for pulmonary artery sling and other rare congenital heart diseases. LPA sling is rare anomaly that can be clearly delineated by fetal echocardiography with STIC. Likewise, the fetal lung absence of gas interference in the fetal period, two-dimensional combined with STIC ultrasound can clearly show the left and right pulmonary artery branches. Prenatal diagnosis of an LPA sling can suggest better delivery planning and help parents comprehend the surgical repair to improve long-term outcomes.

References


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