Clinical Case Reports

Open Access

CASE REPORT

Double trouble: fetal diagnosis of a pulmonary artery sling and vascular ring

Jesse W. Lee, Beth F. Printz, Sanjeet R. Hegde, Lisa A. Vargas & Heather Y. Sun

Division of Pediatric Cardiology, Rady Children's Hospital, University of California, San Diego, San Diego, California, USA

Correspondence
Heather Y. Sun, Division of Pediatric Cardiology, Rady Children's Hospital, University of California, San Diego, 3020 Children's Way, MC 5004, San Diego, CA 92123, USA. Tel: (858) 966-5855; Fax: (858) 966-7903; E-mail: hsun@rchsd.org

Funding Information
No sources of funding were declared for this study.

Received: 14 July 2016; Accepted: 4 October 2016

Clinical Case Reports **2016; 4(12): 1187–1190**

doi: 10.1002/ccr3.733

Key Clinical Message

Left pulmonary artery slings and vascular rings are rare congenital anomalies definable by fetal echocardiography. Left pulmonary artery slings are associated with high respiratory morbidity and mortality. Prenatal diagnosis of a left pulmonary artery sling should prompt delivery planning for postnatal management at a pediatric tertiary care center.

Keywords

Fetal echocardiography, pulmonary artery sling, vascular ring.

A 42-year-old gravida 4, para 3 woman was referred for a fetal echocardiogram (ECHO) at 26 weeks gestation due to concern for an abnormal aorta. The fetal ECHO revealed the left pulmonary artery (LPA) arising from the right pulmonary artery resulting in an LPA sling (Fig. 1A) as well as a right aortic arch with a left ductus arteriosus resulting in a vascular ring (Fig. 1B). Prenatal laboratories were unremarkable including a normal amniotic fluid genetic microarray. Due to the prenatal diagnosis of a vascular ring and an LPA sling causing high risk of postnatal respiratory failure, the fetus was delivered at a tertiary care center via repeat Cesarean section at 38 weeks gestation.

Postnatal ECHO and computed tomography (Figs 2 and 3) confirmed the diagnosis of the LPA sling and right aortic arch with diverticulum of Kommerell without additional intracardiac anomalies. The baby was initially asymptomatic, but developed stridor and respiratory distress requiring supplemental oxygen on day of life 7. A bronchoscopy confirmed tracheal stenosis with complete tracheal rings. The baby therefore underwent sliding tracheoplasty, LPA reimplantation, and division of the ligamentum arteriosum on day of life 11. After a prolonged recovery, the baby was discharged at 3 months old and remains clinically well with mild residual tracheal stenosis and mild LPA stenosis. Fetal ECHO diagnosis of both the LPA sling and vascular ring in this patient directed appropriate delivery at a pediatric tertiary care center and led to prompt postnatal care, confirmatory imaging, and surgical management prior to clinical deterioration.

To our knowledge, this is the first report describing the prenatal diagnosis of both an LPA sling and vascular ring in a single fetal patient. Vascular rings account for <1% of congenital cardiac defects; LPA slings are an even rarer anomaly. The actual incidence of vascular rings is difficult to determine as there is a wide spectrum of clinical presentations, including those who are asymptomatic into adulthood. In the largest case series of 81 fetuses with prenatal diagnosis of vascular rings or slings, only two fetuses had LPA slings [1].

Fetal ECHO is superior to postnatal transthoracic ECHO at diagnosing vascular thoracic anomalies, as the relationship of vascular structures surrounding a fluid-filled trachea in utero can be easily visualized. In contrast, the postnatal air-filled trachea obscures visualization of posterior vessels. Once the ductus arteriosus closes

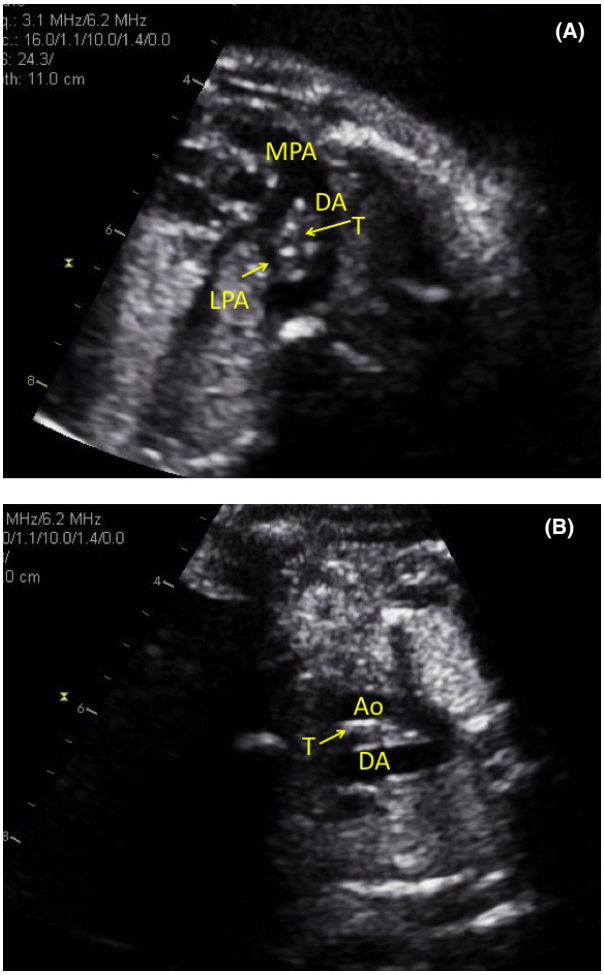

Figure 1. (A) Fetal ECHO axial view showing the left pulmonary artery (LPA) arising from the right pulmonary artery instead of the main pulmonary artery (MPA). The LPA courses posterior to the trachea (T) resulting in an LPA sling. (B) Fetal three-vessel view with trachea (T) demonstrating a right aortic arch (Ao) with a left ductus arteriosus (DA) forming a vascular ring around the trachea.

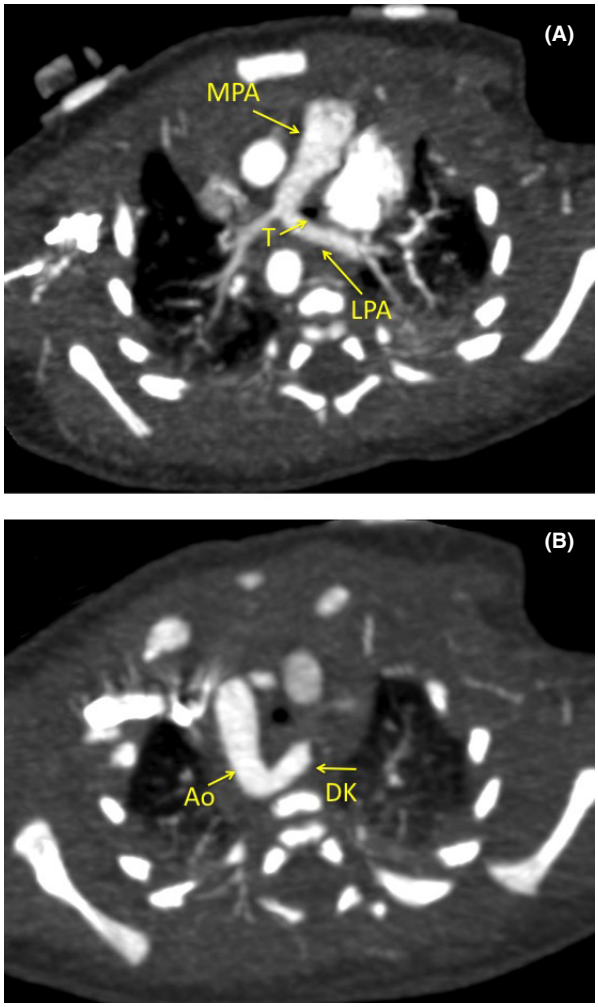

Figure 2. (A) Postnatal CT axial view showing the left pulmonary artery (LPA) sling coursing posterior to the trachea (T) and (B) right aortic arch (Ao) with diverticulum of Kommerell (DK).

becoming the ligamentum arteriosum after birth, it cannot be seen by any imaging modality. The fetal three-vessel view [2] and the three-vessel tracheal (3VT) view [3] are transverse planes of the upper mediastinum that evaluate the relationships of the main pulmonary artery, ductus arteriosus, aortic arch, superior vena cava, and trachea. The 3VT is particularly crucial in assessing a vascular anomaly. In the normal fetus, the pulmonary trunk, ductus arteriosus, and aortic arch form a V-shape appearance leftward of the trachea as they converge toward the descending aorta [4]. No vascular structures should pass posterior to or surround the trachea. In contrast, a vascular ring should be suspected if a vessel courses behind the trachea forming a U-shape appearance (Fig. 1B). If the LPA take off appears more distal than usual, its relationship to the trachea should be critically assessed to determine whether it courses posteriorly and creates an LPA sling (Fig. 1A).

To our knowledge, very few reports of prenatally diagnosed LPA slings exist [1, 5, 6]. The distal branch pulmonary arteries are normally formed arising from the lung buds and joining separately into the main pulmonary artery from the truncoaortic sac. When the left distal pulmonary artery instead courses behind the trachea and joins the right pulmonary artery instead of the truncoaortic sac, an LPA sling occurs [7]. The anomalous course of the LPA compresses the trachea and is frequently associated with intrinsic tracheobronchial abnormalities.

An LPA sling carries a poor prognosis. LPA slings are usually diagnosed in infancy due to their high association with respiratory insufficiency. Complete cartilaginous tracheal rings are found in more than 70% of patients with LPA slings leading to significant tracheal stenosis [8].

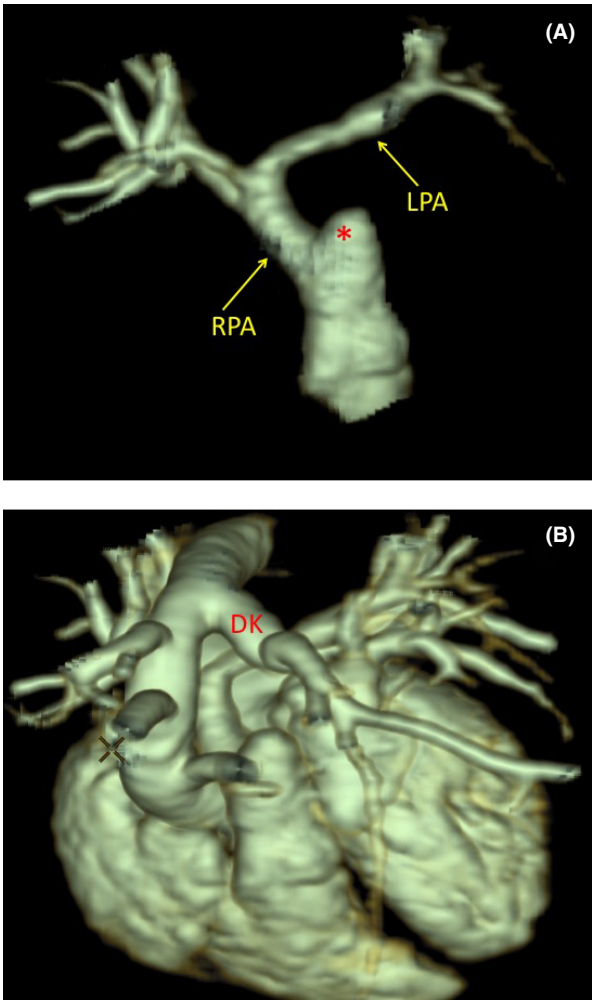

Figure 3. (A) Postnatal CT 3D reconstruction showing the stump of the normal origin of the LPA (*), which here arises from the right pulmonary artery (RPA). (B) Postnatal CT 3D reconstruction showing the diverticulum of Kommerell (DK) arising from the right aortic arch forming the vascular ring.

Morbidity and mortality are often dependent on the presence and extent of tracheal stenosis, with mortality rates as high as 50% [9]. Symptoms can include stridor, wheezing, recurrent respiratory infections, respiratory distress, and respiratory failure. Intubation and mechanical ventilation may be difficult depending on the severity of airway narrowing. Surgical repair frequently involves tracheoplasty, in addition to reimplantation of the LPA anterior to the trachea [8]. Postoperative and long-term respiratory symptoms and airway reinterventions are common and depend on the degree of residual airway stenosis and malacia.

The incidence of prenatally diagnosed vascular rings is <1% [4]. The most common cause of a prenatal vascular ring is a right aortic arch with left ductus arteriosus, which in postnatal life results in a diverticulum of Kommerell. As opposed to the usual dissolution of the embryonic right 4th arch, the embryonic left 4th arch disappears with persistence of the embryonic right 4th arch (which becomes the right aortic arch) and embryonic left 6th arch (which becomes the ductus arteriosus) between the truncoaortic sac and the left dorsal aorta. The ductus arteriosus receives the majority of the right ventricular cardiac output in the fetal circulation. The left dorsal aorta, in this instance, would then receive this output and become significantly larger than the left subclavian artery, forming the diverticulum of Kommerell [7]. The diverticulum of Kommerell gives rise to the aberrant subclavian artery and courses posteriorly to the esophagus and trachea. This particular vascular ring is thus comprised of the right aortic arch, left ductus arteriosus (or ligamentum), and the diverticulum of Kommerell with aberrant subclavian artery posteriorly.

Most vascular rings are isolated anomalies. However, intracardiac congenital heart defects and associated chromosomal abnormalities (such as 22q11.2 deletion) should be ruled out. Vascular rings can be associated with airway and feeding difficulties due to tracheal and/or esophageal compression from the surrounding vessels. If symptoms are mild and/or mistaken for pulmonary diseases such as bronchiolitis or asthma, diagnosis may be delayed until later in life. Dysphagia may also occur, especially with solid foods. In the absence of other congenital heart defects, surgical repair involves division of the ligamentum arteriosum, and long-term sequelae are rare.

Vascular rings and LPA slings are rare anomalies that can be clearly delineated by fetal echocardiography. Fetal diagnosis of an LPA sling should modify delivery planning to include early postnatal access to appropriate pediatric subspecialty care for surgical repair and improved long-term outcomes.

Conflict of Interest

The authors have no conflict of interests to disclose.

References

1. Li, S., G. Luo, E. Norwitz, C. Wang, S. Ouyang, Y. Yao, et al. 2011. Prenatal diagnosis of congenital vascular rings and slings: sonographic features and perinatal outcome in 81 consecutive cases. Prenat. Diagn. 31:334–346.
2. Yoo, S. J., Y. H. Lee, E. S. Kim, H. M. Ryu, M. Y. Kim, H. K Choi, et al. 1997. 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. 9:173–182.
3. Yagel, S., R. Arbel, E. Anteby, D. Raveh, and R. Achiron. 2002. The three vessels and trachea view (3VT) in

fetal cardiac scanning. Ultrasound Obstet. Gynecol. 20:340–345.
4. Patel, C. R., J. R. Lane, M. L. Spector, and P. C. Smith. 2006. Fetal echocardiographic diagnosis of vascular rings. J. Ultrasound Med. 25:251–257.
5. Yorioka, H., A. Kasamatsu, H. Kanzaki, M. Kawataki, S. J. Yoo. 2011. Prenatal diagnosis of fetal left pulmonary artery sling. Ultrasound Obstet. Gynecol. 37:245–246.
6. Ishii, Y., T. Miyamoto, K. Nakajima, K. Tanaka, K. Ikeda, N. Inamura, et al. 2016. Abnormal cardiac axis as a prenatal marker of left pulmonary artery sling. Pediatr. Int. 58:158–161.
7. Weinberg, P. 2013. Chapter 33: aortic arch and vascular anomalies. Pp. 758–798 in H. Allen, D. Driscoll, R. Shaddy and T. Feltes, eds. Heart disease in infants, children, and adolescents, 8th ed. Lippincott Williams & Wilkins, Philadelphia, PA.
8. Backer, C. L., H. M. Russell, S. Kaushal, J. C. Rastatter, C. K. Rigsby, and L. D. Holinger. 2012. Pulmonary artery sling: current results with cardiopulmonary bypass. J. Thorac. Cardiovasc. Surg. 143:144–151.
9. Berdon, W. E., D. H. Baker, J. Wung, A. Chrispin, K. Kozlowski, M. deSilva, et al. 1984. Complete cartilage-ring tracheal stenosis associated with anomalous left pulmonary artery: the ring-sling complex. Radiology 152:57–64.

Supporting Information

Additional Supporting Information may be found online in the supporting information tab for this article:

Video S1. Fetal echocardiogram and postnatal CT clips demonstrating left pulmonary artery sling and vascular ring.