

# Fetal Diagnosis of Dextroposition, Left Pulmonary Artery Sling, Partial Anomalous Left Pulmonary Artery, and Aortic Coarctation



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## INTRODUCTION

Left pulmonary artery (LPA) sling is a rare congenital anomaly accounting for less than 1% of congenital cardiac defects.<sup>1</sup> Left pulmonary artery sling is often associated with high respiratory morbidity and mortality due to accompanying tracheal stenosis.<sup>1-3</sup> The complex vasculature of the upper chest makes identification of a vascular anomaly challenging on transthoracic echocardiography due to air-filled trachea and lungs, but these structures may be defined by fetal echocardiography, when the trachea and lungs are fluid filled.<sup>4-6</sup> We present the unique case of a patient diagnosed prenatally with dextroposition, partial anomalous LPA sling, and aortic coarctation diagnosed by fetal echocardiography and highlight the use of abnormal cardiac position as an indicator for the presence of an atypical case of partial anomalous LPA sling.

## CASE PRESENTATION

A 27-year-old gravida 1, para 0, now 1, woman was referred for a fetal echocardiogram (ECHO) at 22 weeks' gestation due to maternal type 2 diabetes mellitus with elevated HgbA1C of 12.5%. The initial fetal ECHO (Figure 1A, Video 1) revealed a subtle ventricular size discrepancy (right ventricle larger than left) and mild aortic isthmus hypoplasia. Noninvasive prenatal testing was negative, and the parents declined additional genetic testing. Follow-up fetal ECHO at 27 weeks' gestation demonstrated the worsening finding of cardiac dextroposition, aortic isthmus hypoplasia, and a possible vessel posterior to the trachea coursing from right to left and suspicious for LPA sling versus double aortic arch (Figure 1B, Video 2). The normal origin of the LPA was seen, but imaging was suboptimal due to maternal body habitus (elevated body mass index). Fetal ECHO at 31 weeks demonstrated more significant cardiac dextroposition (Figure 1C, Video 3), ventricular size discrepancy with the right ventricle larger than the left, moderate transverse aortic arch and isthmus hypoplasia, and redemonstration of an abnormal vessel coursing posterior to the

trachea. Due to the progression in cardiac dextroposition, a fetal magnetic resonance imaging (MRI) was obtained to evaluate the lung parenchyma and rule out intrathoracic anomaly. Fetal MRI at 32 weeks demonstrated rightward deviation of the heart with hypoplastic appearance of the right lung (Figure 2); differential diagnosis at this stage included left congenital lobar hyperinflation, right pulmonary hypoplasia, or pulmonary artery sling.

Due to the concern for aortic coarctation and possible vascular sling (with associated respiratory failure after birth), delivery was arranged at a center adjacent to the children's hospital with pediatric cardiology and cardiovascular surgery services. The mother presented in preterm labor at 34 weeks and delivered a newborn female via normal spontaneous vaginal delivery. The infant's initial APGAR scores were 8 and 8 at 1 and 5 minutes, respectively. Within minutes of delivery, the infant became more cyanotic and developed irregular respiratory effort and intermittent apnea. She was admitted to the neonatal intensive care unit and started on prostaglandin E1 due to prenatal concerns for aortic coarctation. She was noted to have decreasing aeration and worsening hypercarbia; respiratory support was escalated to continuous positive pressure ventilation and subsequently to intubation. Chest X-ray showed the cardiac silhouette in the right chest, with a hyperexpanded left lung. Once stabilized, she was transferred to the cardiothoracic intensive care unit.

Postnatal ECHO showed the main pulmonary artery branched in typical position to the right pulmonary artery (RPA) and a hypoplastic LPA, with an additional branch pulmonary artery arising from the distal RPA heading leftward and posterior as is typical of a pulmonary artery sling (Figure 3, Video 4), in addition to aortic isthmus hypoplasia. Due to the atypical pulmonary artery anatomy, a cardiac computed tomography (CT) was obtained, which demonstrated a partial anomalous LPA sling with the LPA originating from the main pulmonary artery and a separate partial anomalous pulmonary artery branch originating from the RPA coursing posterior to the trachea (Figure 4). Due to the abnormal contours of the trachea and bronchi on the CT, a bronchoscopy was performed that showed tracheal rings with severe tracheal stenosis. A cardiac catheterization was performed to further characterize the flow distribution to the various lung segments given the complexity of the pulmonary arterial anatomy. Angiography confirmed that an anomalous left lower pulmonary artery branch coursed posterior to the trachea, forming the partial anomalous LPA sling, and that another left upper pulmonary artery branch arose from the main pulmonary artery, crossing anterior to the trachea and ultimately supplying the left anterior upper and lingual lobes (Figure 5). Bronchograms demonstrated an abnormal bronchial tree, with the trachea giving off a separate small anomalous left bronchial segment proximally that led to a hyperinflated left upper lobe and a significantly narrowed distal segment with tracheal rings (Figure 6). She underwent repair of the vascular sling, which involved

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## VIDEO HIGHLIGHTS

**Video 1:** Fetal ECHO four-chamber view at 22 weeks' gestation demonstrating normal cardiac axis. In retrospect, while the fetal cardiac axis is normal, there is mesoposition/mild dextroposition with subtle ventricular size discrepancy (right ventricle larger in size than left ventricle, with a right ventricle:left ventricle width ratio > 1.4).

**Video 2:** Fetal ECHO at 31 weeks showing the interval worsening of dextroposition, utilizing the situs sweep. The situs sweep begins in the abdomen and demonstrates a left-sided stomach (the superior aspect of this image is the left side of the fetus). Sweeping superiorly, the cardiac four-chamber view demonstrates the abnormal rightward cardiac position (dextroposition) with leftward apex (levocardia).

**Video 3:** Fetal ECHO three-vessel trachea view at 27 weeks' gestation demonstrating a vascular sling. In the three-vessel trachea view, a vessel courses posterior to the trachea consistent with a vascular sling.

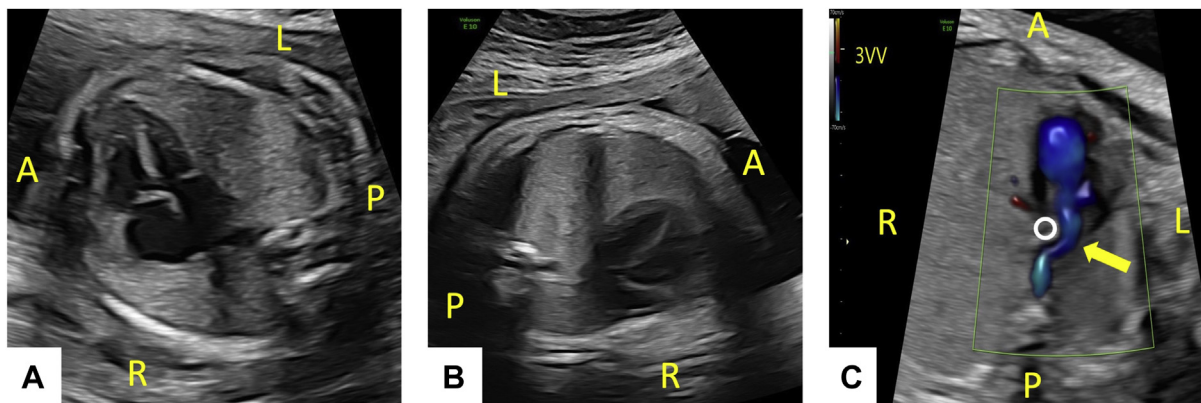
**Video 4:** Postnatal transthoracic ECHO parasternal short-axis view showing partial anomalous LPA sling. In the parasternal short-axis view (with two-dimensional and color-flow imaging), the native, hypoplastic LPA runs anterior to the trachea, while an additional anomalous LPA branch arises from the RPA and courses posterior to the trachea.

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prolonged intubation following tracheal reconstruction. She was subsequently discharged after an 88-day hospital convalescence, with close follow-up with cardiology, otolaryngology, and pulmonary specialists. Surveillance cardiac CT 3 months after surgical repair demonstrates that the heart is now normally positioned in the chest, with a patent reimplemented partial anomalous LPA without obstruction or stenosis and a patent aortic arch without coarctation (Figure 7). Follow-up ECHO at 6 months postdischarge shows normally sized left and right pulmonary arteries without obstruction or stenosis, an unobstructed aortic arch without coarctation, and normal biventricular function. She is currently doing well at home.

## DISCUSSION

The true incidence of vascular sling and ring anomalies is unknown, but it is believed that vascular rings account for less than 1% of congenital cardiac defects, with LPA sling representing a very rare subtype.<sup>1,3</sup> Normally, the main pulmonary artery bifurcates into the RPA and LPA anterior to the trachea at the level of the carina.<sup>7</sup> The RPA then traverses to the right lung and courses anterior to the right main bronchus while the LPA courses over and posterior to the left mainstream bronchus, below the aortic arch.<sup>7</sup> A pulmonary arterial sling is a malformation where a branch pulmonary artery courses aberrantly in between the trachea and esophagus, often associated with tracheal stenosis or resulting in tracheobronchial compression. There are right and left sling phenotypes, with LPA sling being the more common.<sup>8</sup> A partial anomalous LPA, however, is a rare branching pattern where, in addition to the typical bifurcation of the RPA and LPA from the main pulmonary artery, a pulmonary artery branch arises from the RPA and supplies one or more lobes of the left lung.<sup>9,10</sup> Left pulmonary artery slings may be further classified into type I or type II, depending on

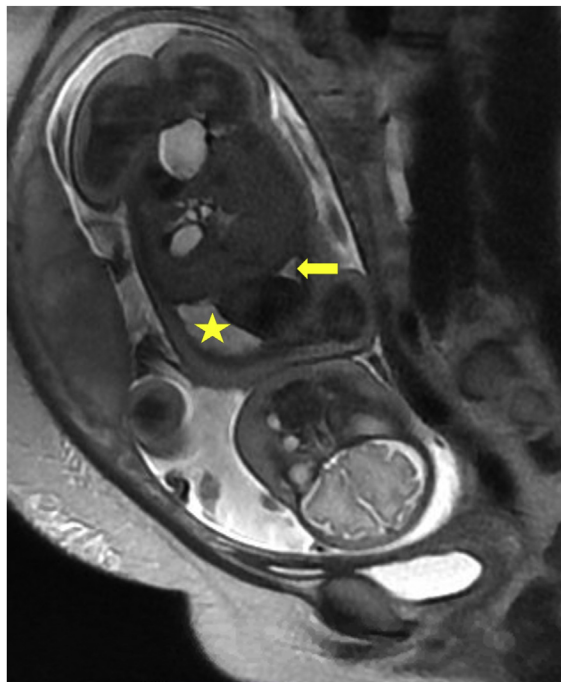


**Figure 1** Fetal ECHO (A) at 22 weeks' gestation, four-chamber view with mesoposition/mild dextroposition, and (B) at 31 weeks' gestation with worsening dextroposition. (C) Three vessel-trachea view at 27 weeks' gestation with a vessel (arrow) coursing posterior to the trachea (circle). A, Anterior; L, left; P, posterior; R, right.

reimplantation of the partial anomalous LPA onto the distal main pulmonary artery, segmental resection of the complete rings with end-to-end reconstruction of the trachea, repair of the aortic arch, and left upper lobe lobectomy on day of life 10. She had a prolonged postoperative course with, as expected, slow wean from the ventilator; a tracheostomy was placed on day of life 50, due to anticipated

the associated airway and lung anomalies. The type I phenotype typically has a normal tracheal bronchial tree, while type II has a more inferior origin of the LPA sling, inferior T-shaped carina, and association with tracheal stenosis from complete tracheal rings.<sup>11</sup>

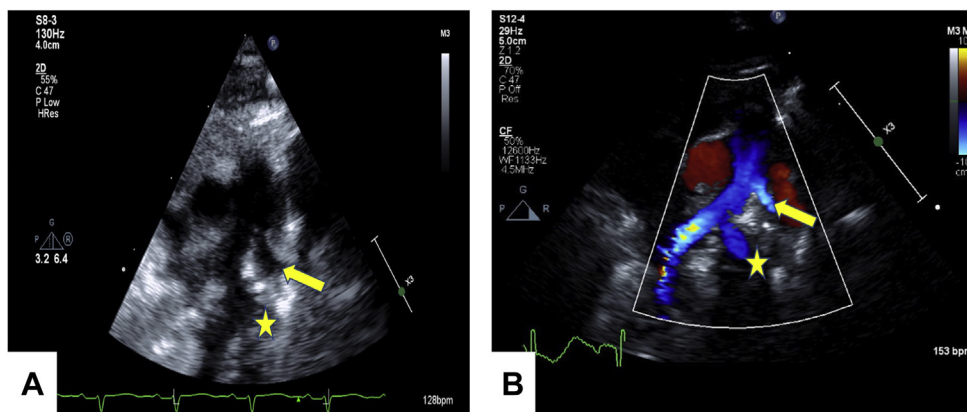
Historically, pulmonary vascular slings were discovered postnatally using echocardiography and high clinical suspicion. Patients with an



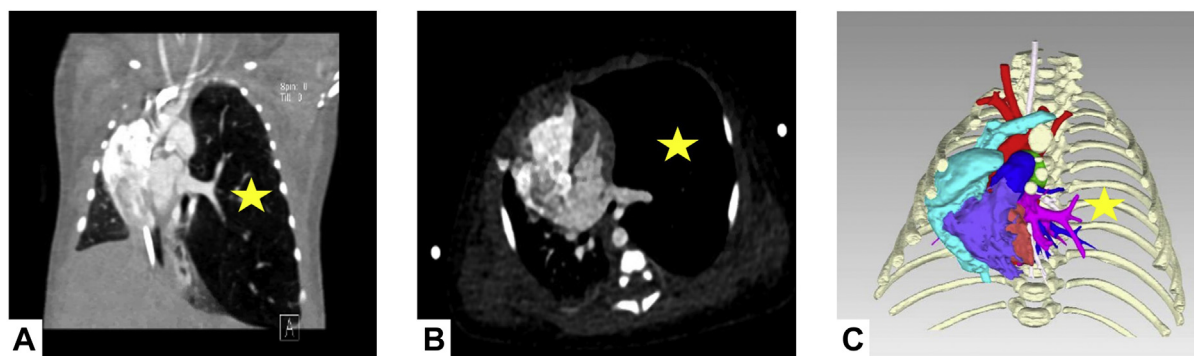
**Figure 2** Fetal MRI at 30 weeks' gestation with rightward deviation of the heart due to hyperinflation of the left lung (*star*). The right lung is severely hypoplastic (*arrow*).

LPA sling frequently present with respiratory distress, often due to tracheal stenosis and recurrent respiratory tract infections. The disease burden may be severe, and patients may present in the neonatal period depending on the degree of tracheal stenosis.<sup>3,5,8</sup> Given the complex pulmonary and cardiac components, echocardiography and advanced imaging techniques (such as CT or MRI) are used to fully delineate the anatomy and guide surgical repair.<sup>1,12</sup> Surgical repair involves mobilization and translocation of the aberrant pulmonary artery to a typical origin and course from the main pulmonary artery and tracheal reconstruction, if tracheal stenosis is present.<sup>3</sup>

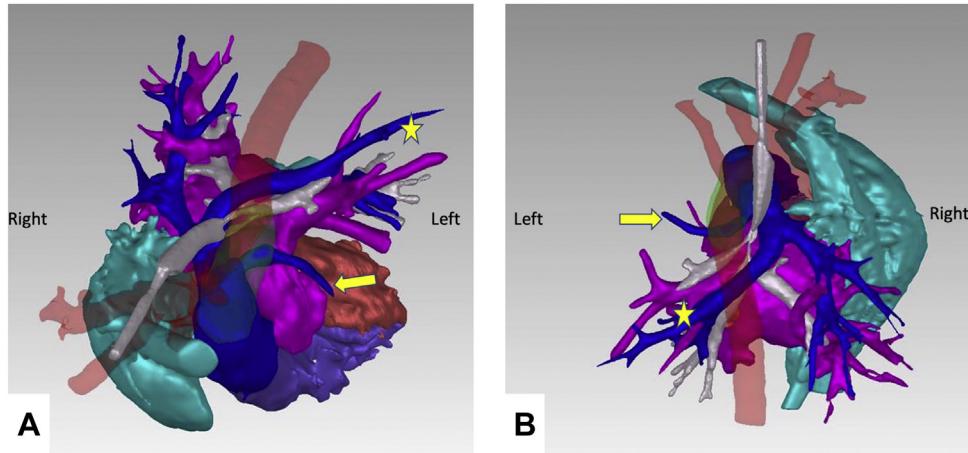
Due to the high association of LPA sling with respiratory morbidity and mortality, prenatal diagnosis should prompt planning for post-natal management at a pediatric tertiary care center.<sup>1</sup> Although challenging to diagnose, LPA sling is definable by fetal echocardiography and may be diagnosed prenatally with careful assessment of the branch pulmonary arteries and their relationship to the echogenic midline trachea, which is fluid filled. Postnatal transthoracic ECHO has limitations due to an air-filled trachea, which generates artifact and obscures visualization of the posterior vessels. Fetal ECHO uses the three-vessel tracheal view to evaluate the relationships among the main pulmonary artery, ductus arteriosus, aortic arch, superior vena cava, and trachea.<sup>6,13</sup> The ductus arteriosus and the aortic arch should form a V shape leftward of the trachea.<sup>6</sup> The main pulmonary artery should bifurcate into the RPA and LPA anterior to the trachea; care should be taken to distinguish the LPA (heading into the lung) from the adjacent ductus arteriosus (which converges with the descending aorta posteriorly). No vascular structures should course



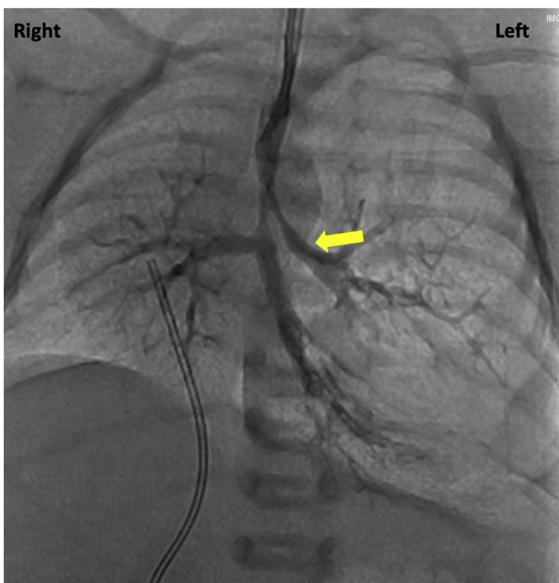
**Figure 3** Parasternal short-axis view on transthoracic ECHO in (A) two-dimensional imaging and (B) color flow showing native, hypoplastic LPA (*arrow*) anterior to the trachea with a partial anomalous LPA (*star*) coursing posterior, forming a vascular sling.



**Figure 4** Cardiac CT, (A) coronal and (B) axial plane with (C) three-dimensional reconstruction demonstrating dextroposition secondary to hyperinflated left lung (*star*).



**Figure 5** Three-dimensional reconstruction of contrast-enhanced cardiac CT scan demonstrates the pulmonary arterial tree (*blue*) and tracheobronchial anatomy (*gray*) from the **(A)** superior and **(B)** posterior views. The native, hypoplastic LPA (*arrow*) and a larger partial anomalous LPA form a vascular sling around the trachea (*star*).



**Figure 6** Bronchogram of the tracheobronchial tree. The trachea gives off a small anomalous bronchus (*arrow*) that travels leftward and anterior to the large, hyperinflated left upper lobe, which receives supply from the partial anomalous LPA.

posterior to the trachea. If the LPA takeoff appears more distal than normal and from the RPA, its relationship to the trachea should be closely evaluated for a posterior course, which would confirm the presence of an LPA sling.<sup>14</sup>

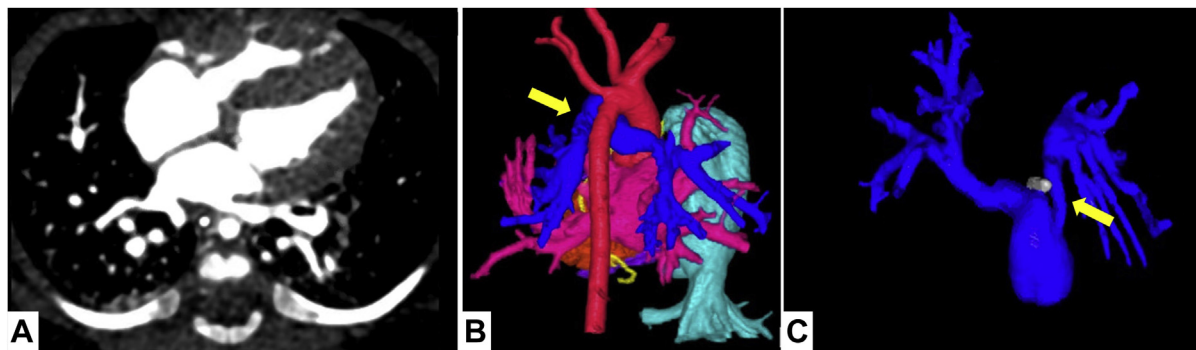
In the fetus, abnormal cardiac position has been reported as an indicator for the presence of a pulmonary sling.<sup>4</sup> Normally, only a small portion the right atrium and right ventricle resides in the right thorax, with the rest of the heart in the left thorax.<sup>15</sup> Dextroposition is an abnormal cardiac position, where the heart is shifted into the right thorax, with the apex remaining leftward. It is typically secondary to extracardiac lesions such as congenital diaphragmatic hernia, congenital pulmonary airway malformations, or pulmonary hyperinflation but

has also been associated with the presence of severe congenital heart disease.<sup>4,15</sup>

We report a unique case of a patient diagnosed in fetal life with cardiac dextroposition and a partial anomalous LPA sling in addition to aortic coarctation. We postulate that the evolution of cardiac dextroposition in utero was secondary to progressive hyperinflation of the left lung due to the abnormal tracheobronchial tree. To our knowledge, there has been only one case report that suggested cardiac positioning as a potential marker for LPA sling.<sup>4</sup> There have only been a handful of postnatal diagnoses of partial anomalous LPA, and of those reported, even fewer formed a sling resulting in airway or esophageal compression.<sup>9</sup> In addition, they were not associated with other cardiac malformations.<sup>9</sup> Furthermore, there is only one previous prenatal report of a partial anomalous LPA sling, and that patient had no other cardiac malformations.<sup>10</sup> This case is unique given the timing of fetal diagnosis, dextroposition used as an indicator of an LPA sling, and the presence of a partial anomalous LPA sling. The diagnosis of this partial anomalous LPA sling was confounded by the presence of an LPA branch anterior to the trachea and highlights the role of multimodal imaging in the diagnosis of complex cardiac anomalies. Once an LPA sling is identified by fetal echocardiography, postnatal ECHO with complementary imaging modalities, such as cardiac CT, is critical to comprehensive surgical planning to optimize the patient's long-term outcome.

## CONCLUSION

The diagnosis of a vascular sling by fetal echocardiography requires a high index of suspicion. In a fetus with cardiac dextroposition, LPA sling should be considered, in addition to evaluation of extracardiac pathology. Dextroposition in this setting is also indicative of more significant tracheobronchial compression and a higher risk of respiratory compromise in the neonatal period. Delivery planning of a fetus with an LPA sling should consider its association with tracheal stenosis and risk of respiratory failure after birth. Postnatal confirmation with ECHO and adjunctive imaging with cardiac CT or MRI provide comprehensive delineation of the vascular sling and tracheobronchial tree for comprehensive presurgical evaluation.



**Figure 7** Cardiac CT (**A**) and three-dimensional reconstruction (**B, C**) 3 months after surgical repair. The heart is now normally positioned in the chest, and the left lung is normally inflated. The reimplanted partial anomalous LPA (*arrow*) is normal in caliber without stenosis. The repaired aortic arch is unobstructed.

### SUPPLEMENTARY DATA

Supplementary data to this article can be found online <https://doi.org/10.1016/j.case.2022.01.007>.

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