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CONGENITAL HEART DISEASE

NEVER TOO YOUNG OR TOO OLD TO BE DIAGNOSED WITH CONGENITAL HEART DISEASE

Breaking the Rules: Left Common Carotid Artery from the Main Pulmonary Artery in an Infant with a Vascular Ring



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INTRODUCTION

Aortic arch anomalies are common in isolation or as part of other forms of congenital heart disease. Echocardiography is often diagnostic of these abnormalities, whether pre- or postnatally; however, some patients will require additional cross-sectional imaging. The vast majority of aortic arch anomalies can be explained by the theoretical totipotent aortic arch, caused by abnormal embryonic regression of the pharyngeal arches.¹

One particularly unique and rare anomaly is an anomalous left common carotid artery (LCCA) arising from the main pulmonary artery (MPA). While very uncommon, this anomaly can also be explained by the theoretical totipotent arch, with the LCCA arising from the patent ductus arteriosus (PDA). Even more rarely, the anomalous LCCA can arise directly from the pulmonary artery independent of a PDA. Prior to this case, this exceedingly rare anomaly has only been reported twice in the literature.

CASE PRESENTATION

A full-term male neonate was urgently delivered at 37 weeks' gestation due to nonreassuring fetal heart rate tracing. His mother had been referred for fetal echocardiogram at 20 3/7 weeks' gestation due to personal history of secundum atrial septal defect that required patch closure, as well as viral myocarditis requiring left ventricular assist device for convalescence. Fetal echocardiogra-

phy demonstrated normal fetal intracardiac anatomy with bilateral superior venae cavae, right circumflex aortic arch with aberrant left subclavian artery, and left ductus arteriosus, consistent with a vascular ring (Figure 1, Video 1). The remainder of the prenatal course and care was uneventful.

At delivery, the patient emerged limp and apneic with no detectable heart rate. He had a prolonged resuscitation at delivery requiring intubation, multiple code doses of epinephrine and atropine, and chest compressions, eventually with detection of a heart rate of 107 beats per minute at 17 minutes of life. Blood pressure in the right arm was 56/30 mm Hg, and oxygen saturation in the right arm and right foot was 100% on 21% FiO₂. Therapeutic cooling for hypoxic ischemic encephalopathy was initiated. Brain magnetic resonance imaging was obtained for assessment after hypoxic ischemic encephalopathy therapy and incidentally noted an incomplete circle of Willis, largely isolating the left-sided cerebral circulation.

Postnatal transthoracic echocardiogram after perinatal stabilization confirmed the prenatal findings, along with stenosis of the aberrant left subclavian artery at its origin. On high parasternal view at the level of the bifurcation of the MPA, a previously unidentified vessel was visualized arising from the MPA, independent of the ductus arteriosus (Figure 2, Video 2).

Subcostal coronal views demonstrated the vessel coursing superiorly from the MPA (Figure 3, Video 3), with pulsatile antegrade flow and diastolic flow reversal. The course of the abnormal vessel was confirmed as the LCCA in additional suprasternal sagittal views (Figure 4). A computed tomography angiography was obtained for three-dimensional rendering of the arterial anatomy (Figure 5).

Once recovered from the perinatal insult, the patient underwent surgical reimplantation of the LCCA to the distal aortic arch, ligation and division of the left ligamentum arteriosum, and side-by-side anastomosis of the subclavian artery to the carotid artery. Intraoperative cerebral near-infrared spectroscopy monitoring demonstrated a marked increase in estimated cerebral oxygenation after the anomalous vessel was reimplanted (Figure 6). Surgical pathology confirmed that the resected ligamentum was indeed ductal tissue.

DISCUSSION

We present a case of an isolated LCCA arising from the MPA, discovered in a neonate with a right circumflex aortic arch, aberrant left subclavian artery, and independent left ductus arteriosus. This anomaly cannot be explained by the traditionally understood means of pharyngeal arch regression.

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VIDEO HIGHLIGHTS

Video 1: Fetal echocardiography, upper thoracic axial view, demonstrating a right aortic arch with left ductus arteriosus, creating a vascular ring. Corresponds to [Figure 1](#).

Video 2: Transthoracic echocardiography, high parasternal view sweeping superiorly, demonstrating the anomalous vessel arising from the MPA, independent of the ductus arteriosus. Corresponds to [Figure 2](#).

Video 3: Transthoracic echocardiography, subcostal coronal view, demonstrating the vessel arising from the MPA and coursing superiorly, with antegrade systolic and retrograde diastolic flow. Corresponds to [Figure 3](#).

[View the video content online at www.cvcasejournal.com.](http://www.cvcasejournal.com)

The vast majority of aortic arch anomalies can be explained by the theoretical totipotential aortic arch, caused by abnormal embryonic regression of the pharyngeal arches, as originally described by Knight and Edwards.¹ Numerous right aortic arch anomalies can form from traditional regression, ranging from the common vascular ring—right aortic arch with aberrant left subclavian artery and left ductus arteriosus ([Figure 7A](#) and [7B](#))—to rare anomalies such as anomalous left subclavian artery from the left pulmonary artery by way of a left ductus arteriosus.²

An anomalous LCCA arising from the MPA is a particularly unique and rare anomaly. Several cases of right aortic arch with the LCCA arising from the MPA have been reported; however, those aberrant vessels have generally originated from the left ductus arteriosus, a finding that can be consistent with the theoretical totipotential arch ([Figure 7C](#)).³⁻⁸ The common carotid arteries are derived from the third pharyngeal arches, both right and left segments normally emerging from the aortic arch via tissue derived from the aortic sac. The MPA and proximal ascending aorta are derived from septation of the truncus arteriosus by the development of the aortopulmonary septum.^{9,10}

Even more rarely, the anomalous LCCA can arise directly from the pulmonary artery independent of a PDA, as we present here. Such an anomaly cannot be explained by the traditionally understood means of pharyngeal arch regression. Prior to this case, this anomaly has been reported only twice in the literature.^{11,12}

Fouilloux *et al*¹¹ described a 3-year-old patient initially referred for a presumed PDA; however, preprocedural imaging demonstrated right aortic arch, anomalous left subclavian artery, and LCCA arising from the MPA. Intraoperatively, a fibrous strand was seen in the position of where a left ligamentum arteriosum would exist; however, this structure was not sent for pathology confirmation.

Kaushik *et al*¹² reported the case of a 10-month-old referred for murmur evaluation and left hemifacial microsomia who had a right aortic arch, aberrant left subclavian artery, a vessel later confirmed as the LCCA arising from the MPA, and a left superior vena cava. At surgical repair, a left ligamentum arteriosum was identified and ligated as well.

The anomaly we demonstrate here may be explained by different embryologic means, namely, malseptation of the aortic sac.^{10,13,14} This method was theorized by Männer *et al*,¹³ who were able to demon-

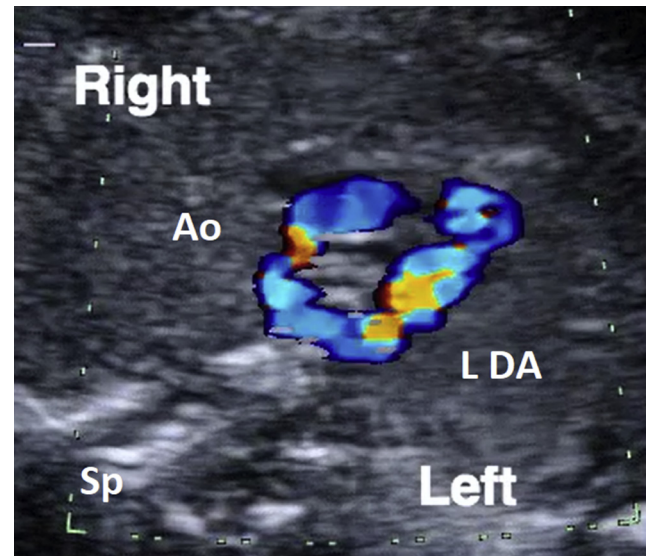


Figure 1 Fetal vascular ring. Axial image on fetal echocardiogram with color Doppler demonstrating the right-sided aortic arch (Ao) and left-sided ductus arteriosus (L DA), completing the vascular ring. Spine (Sp) marked for reference along with right and left orientation.

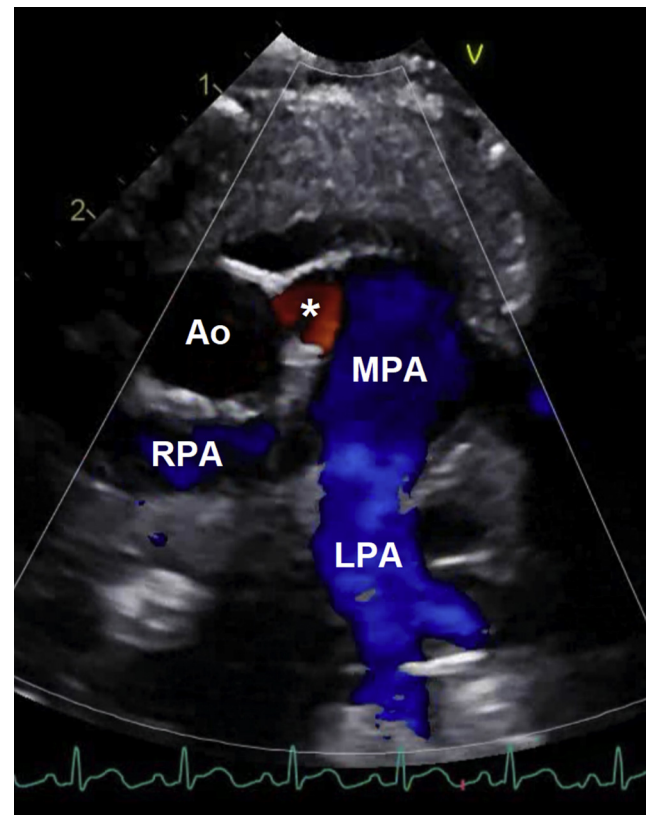


Figure 2 Short-axis origin of the anomalous vessel. High parasternal view on echocardiogram with color Doppler, demonstrating flow in an anomalous vessel originating from the MPA (*starred*). Ao, Aorta; LPA, left pulmonary artery; RPA, right pulmonary artery.

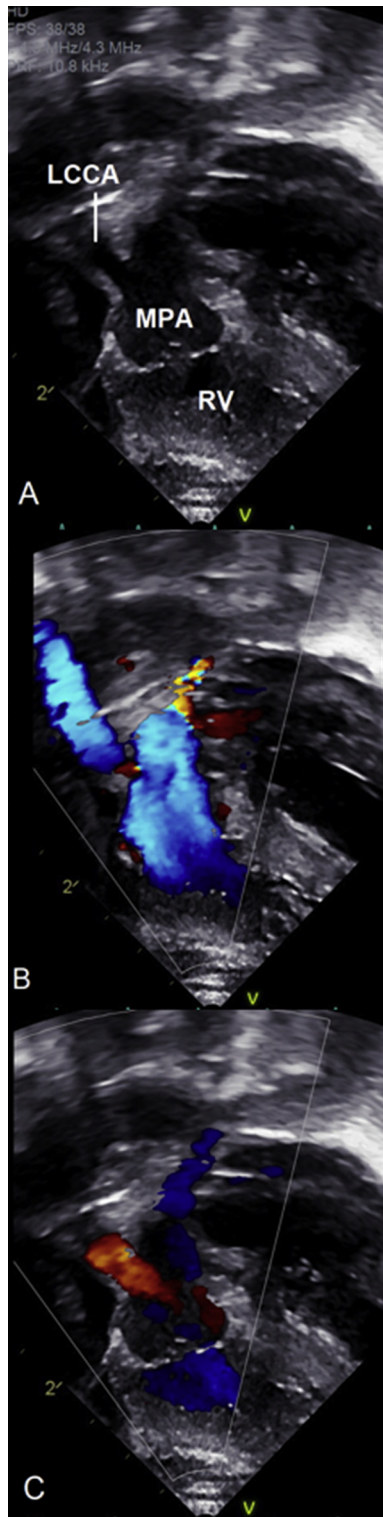


Figure 3 Subcostal coronal view on echocardiogram of the MPA. **(A)** Two-dimensional imaging and color Doppler in systole **(B)** and diastole **(C)**, demonstrating antegrade systolic flow and diastolic flow reversal in the anomalous LCCA. *RV*, Right ventricle.

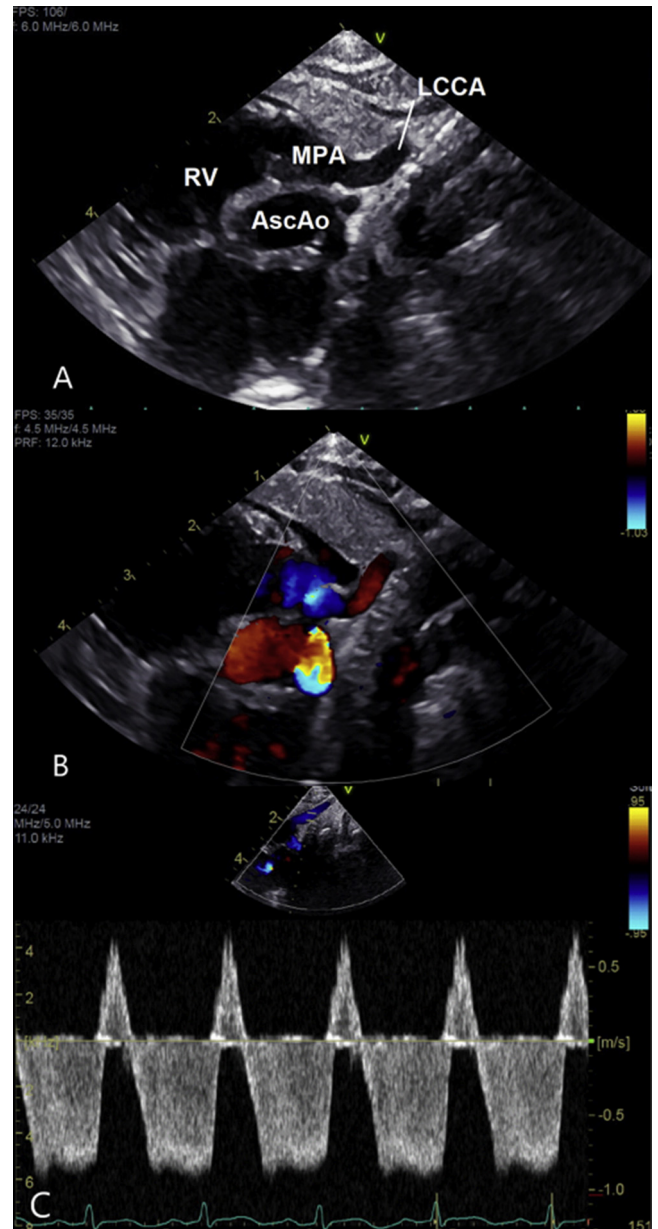


Figure 4 Suprasternal notch view on echocardiogram showing the anomalous LCCA arising from the MPA by two-dimensional echocardiography **(A)** and with color Doppler showing antegrade systolic flow **(B)**. Pulse-wave Doppler demonstrating antegrade systolic flow and holodiastolic flow reversal **(C)**. *AscAo*, Ascending aorta; *RV*, right ventricle.

strate this as a possible mechanism in vivo after ablating chick cardiac neural crest cells at Hamburger Hamilton stage 9; several embryos then developed isolated preductal brachiocephalic arteries.

While extremely rare, early identification of this anomaly was integral in avoiding potentially significant clinical sequelae, owing to the incomplete circle of Willis isolating the left circulation and the anticipated postnatal drop in pulmonary vascular resistance.

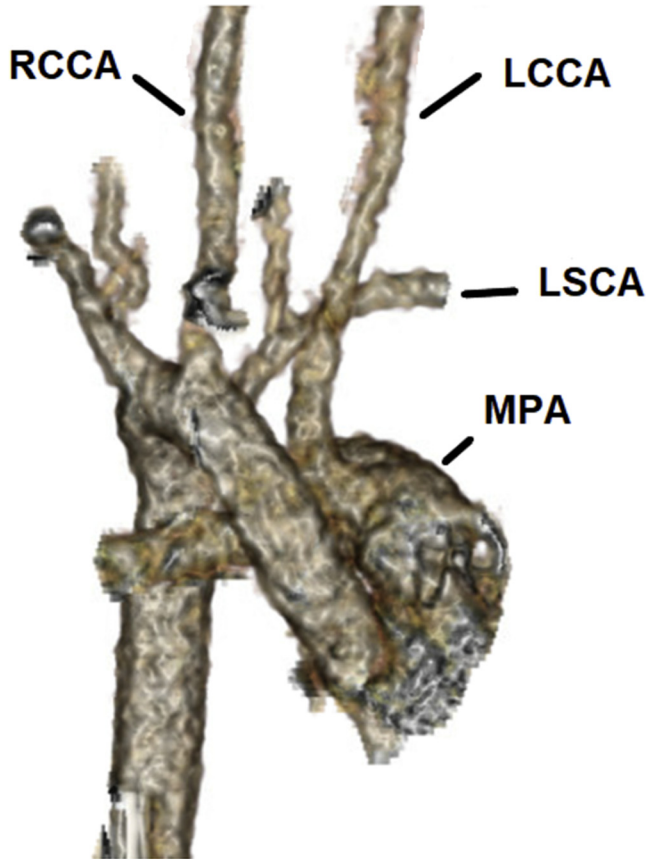


Figure 5 Three-dimensional reconstruction. Computed tomography with angiography three-dimensional rendering demonstrating the LCCA arising from the MPA right aortic arch with aberrant left subclavian artery. LSCA, left subclavian artery; RCCA, right common carotid artery.

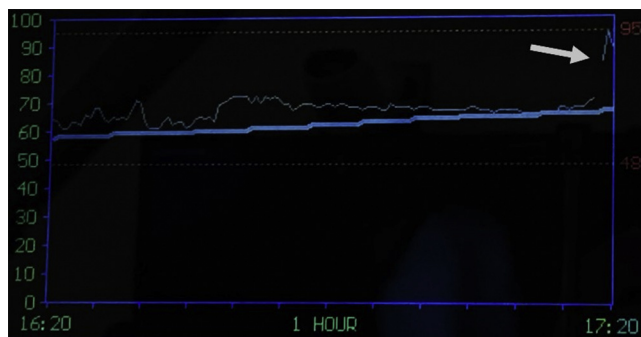


Figure 6 Intraoperative monitoring. Cerebral near-infrared spectroscopy monitoring data prior to and after (*white arrow*) re-implantation of the anomalous LCCA from the pulmonary artery to the aortic arch, documenting a significant increase in estimated cerebral oxygenation.

Echocardiography was essential in making the diagnosis, with three-dimensional rendering of the anatomy by cross-sectional imaging to assist in surgical planning.

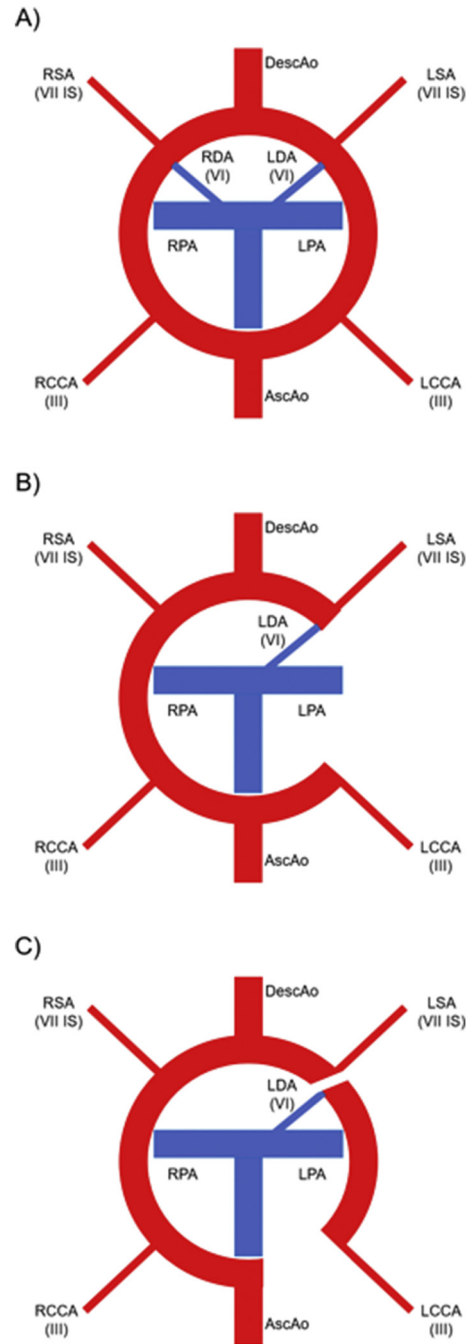


Figure 7 Demonstration of the totipotential aortic arch theory. Totipotential aortic arch prior to pharyngeal arch regression with corresponding arch segments in parentheses **(A)**. Regression patterns leading to a right aortic arch with aberrant left subclavian artery and left ductus arteriosus creating a vascular ring **(B)** and isolated LCCA from the pulmonary artery by way of a left ductus arteriosus **(C)**. AscAo, Ascending aorta; DescAo, descending aorta; LDA, left ductus arteriosus; LPA, left pulmonary artery; LSA, left subclavian artery (VII intersegmental artery); RCCA, right common carotid artery; RDA, right ductus arteriosus; RPA, right pulmonary artery; RSA, right subclavian artery (VII intersegmental artery).

CONCLUSION

The LCCA originating from the MPA in the absence of a connection to the left ductus arteriosus is an extremely rare aortic arch anomaly that defies traditional understanding of aortic arch development and regression. In this case, discovery of this lesion and surgical intervention early in infancy likely prevented development of significant cerebral steal. Transthoracic echocardiography was diagnostic, and multimodal imaging was helpful in confirming the anatomy when the clinical and echocardiographic suspicion was high.

SUPPLEMENTARY DATA

Supplementary data related to this article can be found at <https://doi.org/10.1016/j.case.2021.07.011>.

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