Left Subclavian Artery Isolation with Right Aortic Arch and D-Transposition of the Great Arteries

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INTRODUCTION
Isolation of the subclavian artery is defined as an anomalous origin of the subclavian artery from the pulmonary artery. Isolation of the left subclavian artery (LSA) is rare and is exceedingly rare in association with d-transposition of the great arteries (d-TGA). On review of current literature, there have been 10 cases of d-TGA with systemic arterial supply from the pulmonary artery (Supplemental Table 1). Herein, we present a case with prenatally diagnosed d-TGA, postnatally found to have isolation of the LSA from the left pulmonary artery (LPA). We discuss the patient’s clinical course, classic examination findings, embryologic basis of isolated subclavian artery, imaging modalities used in diagnosis, and implications for the surgical repair.

CASE PRESENTATION
The patient was prenatally diagnosed at 29 weeks gestation with d-TGA with an intact ventricular septum. The patient was referred for delivery at our tertiary care center. Labor was induced at 39 weeks gestation, and delivery was uncomplicated. He weighed 3.6 kg, with Apgar scores of 7 and 8 at 1 and 5 min, respectively. Pre- and postductal saturations were obtained from the right upper extremity and the lower extremity, respectively. Preductal saturation remained at 40% despite 100% fraction of inspired oxygen and initiation of prostaglandin. Arterial blood gas demonstrated a partial pressure of oxygen of 24 mm Hg obtained from the indwelling umbilical arterial catheter. Initial focused postnatal echocardiography confirmed the diagnosis of d-TGA with an intact ventricular septum, a restrictive atrial septum, and a large patent ductus arteriosus. Bedside balloon atrial septostomy with ultrasound guidance was performed. After septostomy, ultrasound confirmed the widely patent, unrestrictive atrial communication with an immediate improvement in postductal saturation to 85%.

Complete postnatal echocardiography after septostomy again demonstrated the transposed great arteries (Figures 1 and 2, Videos 1 and 2), demonstrated a right aortic arch (Figure 3, Video 3) with suspicion for double aortic arch (Figure 4, Video 4), and also demonstrated a bicuspid pulmonary valve (Figure 5, Video 5). On review of echocardiographic images after cardiovascular magnetic resonance imaging (CMR) confirmed the diagnosis of LSA isolation, the LSA was determined to be the LSA arising from the LPA, not from the left aortic arch as initially suspected.

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To definitively characterize the anatomy, contrast-enhanced CMR using ferumoxytol and four-dimensional flow sequences was performed. CMR confirmed transposition of the great arteries (Figure 8A) and a right aortic arch (Figure 8B) with an abnormal transverse axis video demonstrates the LPA directed leftward with the LSA anterior to the bicuspid pulmonary valve. Video 5: 2D transthoracic echocardiography: parasternal short-axis video demonstrates the aortic valve rightward and anterior to the bicuspid pulmonary valve. Video 6: 2D transthoracic echocardiography: subcostal short-axis video demonstrates the LPA directed leftward with the LSA arising from the LPA proximally.

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head and neck branching pattern. The first branch was a common origin of the right and left internal carotid arteries, followed by the right subclavian artery (Figure 8C). The LSA originated from the LPA (Figure 8D). The LSA was demonstrated to have flow reversal in diastole, in contrast to the forward flow shown in the right

Figure 1 Echocardiography: subcostal long-axis image and video (Video 1) demonstrate transposition of the great arteries. The left ventricle (LV) gives rise to a great artery, which branches into left and right pulmonary arteries (PAs), as shown in the images on the left (A,C). The right ventricle (RV) gives rise to a great artery that does not branch, the aorta (Ao), as shown in the images on the right (B,D).

Figure 2 Echocardiography: parasternal long-axis image and video (Video 2) demonstrate the pulmonary artery (PA) arising from the left ventricle (LV) and the aorta (Ao) arising from the right ventricle (RV). LA, Left atrium.

Figure 3 Echocardiography: suprasternal short-axis sweep side-by-side images with color and video (Video 3) demonstrate the first head and neck vessel coursing to the left, establishing a right aortic arch (Ao). There appears to be a “starburst pattern” in which two vessels course to the left and two vessels course to the right. This initially raised the suspicion for a double aortic arch. On later review of these images after the diagnosis was made by CMR, it was determined that the LSA was arising from the LPA, not from the left aortic arch as initially suspected.
subclavian artery (Figure 9), consistent with an anomalous origin of
the LSA from the lower pressure pulmonary artery.

At three days of life, the patient underwent an arterial switch oper-
ation in standard fashion, as the bicuspid pulmonary valve was
adequate in size. The LSA was ligated and divided from its LPA origin
(Figure 10), a small amount of presumed ductal tissue was resected,
and the artery was anastomosed to the leftward aspect of the proximal
aortic arch. The intraoperative course was uncomplicated, with stable
hemodynamics and normal left upper extremity perfusion.

Postoperative epicardial echocardiography demonstrated mildly
depressed left ventricular systolic function, normally functioning atrio-
ventricular valves, normally functioning neopulmonic and bicuspid
neoaortic valves (Figures 11–13), and the LSA arising from the aortic
arch (Figure 14). The left arm was monitored closely postoperatively
and demonstrated normal perfusion and blood pressure.

DISCUSSION

Congenital aortic arch anomalies include a range of anomalies that
arise from abnormal persistence or regression of embryologic bran-
chial arches. Of aortic arch anomalies, an aberrant right subclavian ar-
tery from a left aortic arch is most common, whereas isolation of a
subclavian artery, in which the subclavian arises directly from a pulmo-
nary artery with no communication to the aortic arch, is the rarest. In a
review of 39 cases of isolation of the LSA in 1990, congenital heart
disease was present in 59%, and tetralogy of Fallot was the most
commonly associated lesion.1 Of all patients with right aortic arches,
0.8% are found to have isolation of the LSA.2

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Transposition of the great arteries and anomalous origins of the
head and neck vessels are explained embryologically by distinct
mechanisms: septation with failed spiraling and abnormal embryologic arch regression, respectively. In Edwards’ hypothetical double-arch model (Figure 15A), the right fourth and sixth aortic arches regress while the left arches persist, leading to a left aortic arch with a left ductus arteriosus. Failed regression of the right fourth and sixth arches can lead to a double aortic arch or, if accompanied by inappropriate regression of the left fourth arch, result in a right aortic arch. The seventh intersegmental artery migrates cranially and becomes the subclavian artery. While this migration is occurring, if there is persistence of the left sixth arch in the setting of a right aortic arch with right ductus, there is potential for the persistent left ductus to form continuity with the LSA, thus leading to an anomalous origin of the LSA from the LPA with potential for ductal tissue within the LSA (Figure 15B). Our patient’s anatomy is represented in Figure 15C.

Subclavian isolation from a pulmonary artery can lead to unique physical examination findings in which the corresponding arm receives deoxygenated blood. Reverse differential cyanosis is a well-described physical examination finding classic in neonates with d-TGA and is due to elevated pulmonary vascular resistance in early neonatal life, leading to flow through the patent ductus arteriosus from the pulmonary artery to the descending aorta, and thus, deoxygenated blood is sent to the lower body via the ductus. Therefore, when isolation of a subclavian artery is combined with d-TGA, there will be normal saturations in the limb supplied by the isolated vessel with an otherwise cyanotic head and contralateral upper extremity. The diagnostic examination of a patient with isolated right subclavian artery from the right pulmonary artery in the setting of d-TGA was reported by Arunamata et al.: the right upper extremity and lower extremities had normal saturations, whereas the face and left upper extremity were cyanotic, consistent with reverse differential cyanosis of d-TGA.

The extremity corresponding to the isolated subclavian artery will likely have dampened pulses and decreased blood pressure relative to the contralateral and normally perfused upper extremity. The subclavian artery is supplied by the lower pressure pulmonary artery, which explains the difference in pulsatility. In many cases, the isolated subclavian artery has ductal tissue, which can constrict and close, preventing antegrade blood flow into the corresponding upper extremity, and instead, the subclavian must be supplied by retrograde flow from the ipsilateral vertebral artery. This can result in subclavian steal, manifested by symptoms of vertebrobasilar insufficiency (vertigo, dizziness, syncope, confusion, etc.) as well as symptoms of ischemia of the corresponding upper extremity.

A common theme among case reports of subclavian isolation is the challenge of diagnosis by echocardiography. The diagnosis is most commonly made by cardiac computed tomography, CMR, or invasive angiography. However, it can be made by echocardiography and requires thorough interrogation of the branching pattern of the...
Figure 9 CMR: flow measurements in the left subclavian artery (LSCA) and the right subclavian artery (RSCA). There is reversal of flow in diastole in the left subclavian artery, an indicator that it has an anomalous origin.

Figure 10 Intraoperative photograph demonstrating the LSA originating from the LPA.

Figure 11 Postoperative echocardiographic image from the parasternal long-axis view demonstrating the outflow tracts after the arterial switch operation. LV, Left ventricle; RV, right ventricle.
head and neck vessels. Although arch sidedness and branching are classically defined in the suprasternal axial view with a sweep superiorly, locating the origin of an anomalous subclavian artery requires slow sweeps inferiorly to visualize the main and branch pulmonary arteries, as described by Makadia et al. Thus, when arch branching anatomy is unclear from typical suprasternal sweeps, further evaluation of each head and neck vessel’s origin is necessary to define the anatomy. In our case, on review of the postnatal echocardiogram after the diagnosis was made, there were supraclavicular and subcostal views in which the LSA could be seen originating from the LPA (Figures 4 and 5, Videos 4 and 5). Echocardiographic diagnosis is most often made when there is suspicion for abnormal branching on the basis of a neonate’s abnormal saturation pattern but can otherwise be easily missed. Although isolation of a LSA with a right aortic arch is rare, it is often associated with congenital heart disease that may require surgical intervention. Identification of an anomalous origin of the subclavian artery is important preoperatively to enable an opportunity during the intracardiac repair to reimplant the isolated subclavian artery onto the aorta, with possible resection of ductal tissue, and allow normal perfusion of the extremity.

CONCLUSION

Isolated LSA in a patient with d-TGA is a rare occurrence with unique findings on physical examination, characterized by reverse differential cyanosis with cyanosis of the left upper extremity, which is supplied by the LPA via the isolated LSA. Four extremity blood pressures and saturations are necessary to raise suspicion for the diagnosis before imaging confirmation. Diagnosis by echocardiography can be difficult and should include thorough sweeping views of aortic arch branching from suprasternal views. Even so, the diagnosis may not be made until the intraoperative period, unless suspicion is raised by examination or echocardiographic findings, in which case CMR, cardiac computed tomography, or invasive angiography can confirm the diagnosis preoperatively. Diagnosis is important to allow correction intraoperatively at the time of the arterial switch operation, thus preventing late sequelae of subclavian steal.

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SUPPLEMENTARY DATA

Supplementary data to this article can be found online at https://doi.org/10.1016/j.case.2021.09.010.
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