Isolated Left Subclavian Artery, Multiple Ventricular Septal Defects, and Pulmonary Hypertension in a Child: A Case Report

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The isolated left subclavian artery is a rare congenital aortic arch anomaly where the left subclavian artery connects to the pulmonary artery via a patent ductus arteriosus or its remnant, rather than the aorta. Typically associated with other congenital heart defects, it can lead to subclavian steal syndrome, pulmonary steal syndrome, and discrepancies in size, blood pressure, or O2 saturation between the 2 upper limbs. Our case report describes a 14-month-old infant with an isolated left subclavian artery, multiple ventricular septal defects, and pulmonary hypertension, a rare anomaly impacting surgical planning and outcomes.

Congenital anomalies of the aortic arch can significantly impact cardiovascular function and clinical outcomes. One such rare anomaly is the isolated left subclavian artery that anomalously connects to the pulmonary artery through a patent ductus arteriosus (PDA) or its remnant instead of the aorta. This condition often accompanies other congenital heart defects, such as atrial and ventricular septal defect, double-outlet right ventricle, and tetralogy of Fallot. Notably, it is frequently associated with a right-sided aortic arch.

Patients with an isolated left subclavian artery may develop subclavian steal syndrome, characterized by retrograde blood flow from the vertebral-basilar circulation, potentially causing vertebral-basilar insufficiency symptoms, especially during physical exertion. In addition, pulmonary steal syndrome can occur, where retrograde filling of the pulmonary artery is supplied by the vertebral artery via the PDA. These syndromes lead to significant clinical manifestations, including discrepancies in upper limb size, blood pressure, and oxygen saturation, necessitating careful diagnostic and therapeutic strategies.

Herein, we describe a 14-month-old infant presenting with an isolated left subclavian artery, multiple ventricular septal defects, and pulmonary hypertension. This case underscores the complexity of managing such anomalies and the importance of tailored surgical approaches to improve patient outcomes.

Case Presentation

A 14-month-old girl was referred to our paediatric cardiology clinic due to a murmur detected by her paediatrician. The patient exhibited no syndromic features and displayed typical neurodevelopment. Born at 2070 g, her current weight of 6500 g falls below 2 standard deviations for her age. During the examination, a grade 2/6 systolic murmur was noted at the left mid-sternal border, along with a loud P2 at the left upper sternal border. There was no limb atrophy observed in any of her 4 limbs, and capillary refill time in all limbs was normal. The distal left upper limb appeared cyanotic compared with the right upper limb. Therefore, O2 saturation was checked in both upper limbs, revealing an O2 saturation of 93% in the right upper limb and 75% in the left upper limb. Blood pressure readings were 100/65 mm Hg for the right upper limb and 87/60 mm Hg for the left upper limb, with symmetric bilateral radial pulses. Room air SpO2 measured 93% and 75% for the right and left upper limbs, respectively.

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The patient showed no signs of subclavian steal syndrome. Electrocardiography demonstrated right ventricular hypertrophy with a normal sinus rhythm and axis. Chest radiograph revealed cardiomegaly, a right aortic arch, and increased pulmonary vascular markings.

Transthoracic echocardiography, using a vivid 7 with a 3.5-MHz sector transducer, revealed multiple ventricular septal defects (perimembranous, muscular, and apical), a left-sided PDA, and a right-sided aortic arch.

In response to the observed O$_2$ saturation discrepancy in the 2 upper limbs, computed tomography angiography was performed, revealing the presence of an isolated left subclavian artery originating from the pulmonary artery (Fig. 1). Given the optimal diagnostic capabilities of angiography for evaluating an isolated subclavian artery, the patient underwent cardiac catheterization. Ascending aorta angiography revealed a right-sided aortic arch, encompassing both the ascending and descending aortas. The aortic arch branches were identified as the left common carotid, right common carotid, and right subclavian artery, respectively. Opacification of the left subclavian artery occurred in a retrograde fashion from the left vertebral artery, with a slight delay. Systolic blood pressure for both the subclavian and pulmonary arteries measured 80 mm Hg, mirroring the systolic systemic blood pressure. Oxygen saturation levels were 79% in the pulmonary artery and 99% in the aorta. Left ventricle angiography revealed multiple ventricular septal defects resembling a Swiss cheese ventricular septum, along with good left ventricular function and normal coronary arteries (Fig. 2).

On confirming the diagnosis, a brain computed tomography scan was conducted to assess the vertebrobasilar insufficiency, revealing no signs of brain ischemia. Subsequently, the patient underwent pulmonary artery banding surgery and ligation of the

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**Novel Teaching Points**

- An isolated left subclavian is a rare aortic arch anomaly that should be considered in right aortic arch patients experiencing pulse or blood pressure discrepancies between their 2 upper limbs.
- The isolated subclavian artery, like other aortic arch anomalies, may be associated with DiGeorge syndrome.

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*Figure 1.* Cardiac computed tomography angiography shows multiple ventricular septal defects (A,B) as well as the isolated left subclavian artery (ILSA) connecting the pulmonary artery (PA) (C,D).
left subclavian artery’s origin from the pulmonary artery, without reimplantation. Notably, during the procedure, the surgeon observed the absence of a thymus in the patient.

After the operation, the patient’s postoperative course proceeded without any signs or symptoms of vertebrobasilar insufficiency syndrome. In addition, blood pressure measurements indicated 89/52 mm Hg for the left upper limb and 92/57 mm Hg for the right upper limb. The patient progressed without complications in the postoperative period, and she was discharged after 1 week, with captopril and furosemide as oral medications. The patient received regular follow-up appointments for 3 months after the surgery. During the most recent echocardiogram (conducted 3 months after surgery), it was observed that the pulmonary artery band was effective, as indicated by a pressure gradient of 60 mm Hg between the right ventricle and the pulmonary artery. In addition, no significant size difference was noted between the upper limbs during the examination. Regrettably, the patient did not continue with follow-up appointments beyond the 3-month mark after the surgery.

Discussion

Isolated left subclavian artery occurs when the left subclavian artery is not connected to the aortic arch or the left common carotid artery; instead, it is attached to the left pulmonary artery via a left-sided ductus arteriosus. As a result, filling of the isolated subclavian artery occurs in a retrograde fashion from the vertebral artery. This rare aortic arch anomaly is typically associated with the right aortic arch, with only 0.8% of right aortic arch cases presenting an isolated subclavian artery. As anticipated, our patient also exhibited a right-sided aortic arch.

Typically asymptomatic, isolated subclavian artery does not commonly result in a vascular ring. Symptoms, if they occur, may manifest as a discrepancy between the 2 upper limbs in terms of size, pulse, or blood pressure. However, in our patient, systemic pulmonary hypertension prevented a noticeable difference in blood pressure or pulses between the upper left limbs, and the size of both upper limbs remained similar.

The isolated subclavian artery may lead to subclavian steal syndrome and pulmonary steal syndrome. Subclavian steal syndrome is a vascular condition characterized by the blockage or narrowing of the subclavian artery near the origin of the vertebral artery. This altered vascular flow leads to reversed blood circulation in the vertebral artery on the same side, flowing towards the upper arm beyond the blocked or narrowed section of the subclavian artery, where reduced blood pressure is present. In subclavian steal syndrome, retrograde flow from the vertebralbasilar circulation provides the blood flow needed by the subclavian artery, leading to symptoms of vertebrobasilar insufficiency, particularly during exercise. Our patients showed no signs or symptoms of vertebrobasilar insufficiency, and a computed tomography scan revealed a normal brain. Pulmonary steal syndrome typically arises with decreased pulmonary vascular resistance in early infancy, causing retrograde blood flow from the vertebral artery to the subclavian artery and then to the pulmonary artery. This scenario results in brain ischemia on one side and pulmonary congestion on the other. However, given our patient’s systemic pulmonary hypertension, the pulmonary steal phenomenon was not expected.

In up to 60% of isolated left subclavian cases, coexistent congenital heart defects are common, often involving conotruncal anomalies such as double-outlet right ventricle and tetralogy of Fallot, as well as atrial septal defects and tricuspid septal defects. Not surprisingly, our patient presented with multiple ventricular septal defects, with the largest being a perimembranous ventricular septal defect measuring 6 mm in diameter. Reiterating, extracardiac anomalies and genetic syndromes like 22q11 deletion are more prevalent in patients with aortic arch anomalies, particularly in right aortic arch cases. Despite the absence of a syndromic appearance, our patient, lacking a thymus, was still considered to have DiGeorge syndrome, prompting a genetic study.

Given our patient’s Swiss cheese ventricular septum with multiple ventricular septal defects, current guidelines favour pulmonary artery banding as the preferred surgery. The management of an isolated subclavian artery is predominantly influenced by the severity of concurrent congenital heart disease and growth of the subclavian artery. Although the literature supports acceptable outcomes for surgical reimplantation of the isolated left subclavian along with ductus arteriosus ligation, due to the limitations in our centre, we
opted for ligation of the isolated left subclavian origin from the pulmonary artery. Three months after surgery, our patient’s condition was deemed satisfactory. Pulmonary arterial pressure has decreased, and no size discrepancy was observed in the child’s upper limbs. Nevertheless, reimplantation surgery remains the primary option, particularly when combined with other complex congenital heart defects, for high-risk patients. PDA ligation may be the optimal management approach. Closing a PDA reduces steal from the circle of Willis and may enhance blood flow to the same-side upper limb. However, it does not stop subclavian stealing from the circle of Willis or guarantee ample limb blood supply. Long-term follow-up until adulthood is necessary to assess whether asymptomatic patients require reimplantation surgery for the isolated subclavian artery and to determine the optimal timing for such an operation.

**Ethics Statement**

All procedures performed in this study were per the ethical standards of the institutional review board (IRB) of Shahid Modarres Educational Hospital and with the 1964 Helsinki declaration and its later amendments.

**Patient Consent**

This is a retrospective case report using deidentified data; therefore, the IRB did not require consent from the patient.

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**Disclosures**

The authors have no conflicts of interest to disclose.

**References**

1. Pati PK, Verghese MJ, George PV. Isolation of left subclavian artery with reversal of neurological and hemodynamic abnormalities after percutaneous closure. *Indian Heart J.* 2014;66:477–480.

2. Chaturvedi K, Prasad D, Ashwath R, Strainic JP, Snyder CS. Isolated left subclavian artery, complete atrioventricular block, and tricuspid atresia in a neonate. *Tex Heart Inst J.* 2016;43:546–549.

3. Lee JS, Park JY, Ko SM, Seo DM. Isolation of the left subclavian artery with right aortic arch in association with bilateral ductus arteriosus and ventricular septal defect. *Korean J Thorac Cardiovasc Surg.* 2015;48:415–418.

4. Psillas G, Kekes G, Constantinidis J, Triaridis S, Vital V. Subclavian steal syndrome: neurotological manifestations. *Acta Otorhinolaryngol Ital.* 2007;27:33–37.

5. Chen MR, Yu CH. Subclavian and pulmonary steal phenomenon in isolated left subclavian artery with left lung agenesis. *Jpn Heart J.* 2002;43:429–432.

6. Priya S, Thomas R, Nagpal P, Sharma A, Steigner M. Congenital anomalies of the aortic arch. *Cardiovasc Diagn Ther.* 2018;8(suppl 1):S26–S44.

7. Zhu D, Zhou Y, Ji W, Gu X, Li F. Clinical and imaging characteristics of isolated subclavian artery in pediatric patients. *Clin Imaging.* 2021;77:224–229.