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

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**Case Report**

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# Abstract

solated left subclavian artery is a rare congenital aortic arch anomaly in which the left subclavian artery is connected to the pulmonary artery via a patent Ductus Arteriosus or a remnant of it, instead of the arota. Generally, it is associated with the right aortic arch and other congenital heart defects, mostly tetralogy of Fallot. Isolated left subclavian artery can cause subclavian steal syndrome, pulmonary steal syndrome and size or blood pressure discrepancy between the two upper limbs. We present a 14-months-old infant with isolated left subclavian artery, multiple ventricular septal defects and pulmonary hypertension. To our knowledge, it is a rare anomaly which can influence the surgical planning and outcomes.


# Introduction

Isolated left subclavian artery is a rare congenital aortic arch anomaly in which the left subclavian artery is connected to the pulmonary artery via patent ductus arteriosus or a remnant of it, instead of the arota (1). It is often associated with other congenital heart anomalies such as ventricular septal defect and tetralogy of Fallot. Additionally, in the isolated left subclavian artery, the aortic arch is usually right-sided (2). Isolated left subclavian can result in pulmonary steal syndrome, subclavian steal syndrome or difference in the size of the two upper limbs (3). In subclavian steal syndrome, the blood flow required by the subclavian artery is supplied retrogradely from the vertebro-basilar circulation, as a result of which is symptoms of vertebro-basilar insufficiency, especially during exercise (4). Similarly, pulmonary steal syndrome refers to retrograde filling of the pulmonary artery via patent ductus arteriosus from the left subclavian which in turn is supplied by the vertebral artery (5).

Herein, we present a 14-month-old infant with isolated left subclavian artery, multiple ventricular septal defects and pulmonary hypertension. To our knowledge, it is a rare anomaly which can influence the surgical planning and outcomes.

# Case Presentation

A 14-months-old girl was referred to our pediatric cardiology clinic for evaluation of a murmur heard by her pediatrician. The patient did not have syndromic face and was neurodevelopmentally normal. She had birth weight of 2070 gr and current weight of 6500 gr, which was lower than two standard deviations of normal weight for her age.

In her physical examination she had a grade 2/6 systolic murmur at the left mid sternal border and a loud P2 on the left upper sternal boarder. There was no limb atrophy and the capillary refilling time of all four limbs was normal. SPO2 at room air was 93% and 75% for the right and left upper limb, respectively. Blood pressure was 100/65 mmHg for the right upper limb and 87/60 mmHg for the left upper limb. Bilateral radial pulses were symmetric. She also had no sign or symptoms in favor of subclavian steal syndrome.

Electrocardiography (ECG) demonstrated right ventricular hypertrophy with normal sinus rhythm and normal axis.

Chest X-ray revealed cardiomegaly, right aortic arch, and increased pulmonary vascular markings (Fig. 1).

Trans-thoracic echocardiography using a vivid 7 with a 3.5-MHz sector transducer showed multiple ventricular septal defects (Perimembranous, muscular and apical), a left sided patent ductus arteriosus and right sided aortic arch (Fig. 2).

Due to the two upper limbs O2 saturation discrepancy, Computed tomographic angiography was also done for the patient, in which the isolated left subclavian artery off the pulmonary artery was discovered (Fig. 3).

Since angiography is the best tool for evaluation of isolated subclavian artery, the patient underwent cardiac catheterization. Ascending aorta angiography showed a right-sided aortic arch as well as descending aorta. Aortic arch branches were left common carotid, right common carotid and right subclavian artery respectively. Opacification of the left subclavian artery was retrogradely from the left vertebral artery with a few seconds delay. Systolic blood pressure for both of the Subclavian and Pulmonary arteries was 80 mmHg, which was identical to systolic systemic blood pressure. O2 saturation was 79% in the pulmonary artery and 99% in the aorta. Left ventricle angiography also showed multiple ventricular septal defects (swiss cheese ventricular septum), good left ventricular function and normal coronary arteries (Fig. 4).

Once the diagnosis was confirmed, a brain CT scan was done for evaluation of the vertebra-basilar insufficiency, which showed no brain ischemia.

Afterwards, the patient underwent pulmonary artery banding surgery as well as ligation of the left Subclavian origin from pulmonary artery without reimplantation of the left subclavian artery. During the procedure, the surgeon noticed that the patient did not have a thymus.

The patient’s post operation course was without any sign or symptoms of vertebra-basilar insufficiency syndrome. Besides, the blood pressure was 89/52 mmHg for the left upper limb and 92/57 mmHg for the right upper limb.

# Discussion And Conclusions

Isolated left subclavian artery occurs when the left subclavian artery is not connected to the arch or the left common carotid artery, but rather it is attached to the left pulmonary artery via a left sided ductus arteriosus; therefore, filling of the isolated subclavian artery is from the vertebral artery in a retrograde fashion(6). Isolated left subclavian artery is a rare aortic arch anomaly which is associated with the right aortic arch. However, only 0.8% of right aortic arch cases have isolated subclavian artery (7). As we expected, our patient also had a right sided aortic arch.

In most cases, the isolated left subclavian artery is asymptomatic and does not cause a vascular ring. Symptoms, if present, including discrepancy between the two upper limbs in terms of size, pulse or blood pressure (8). However, in our patient due to systemic pulmonary hypertension, neither blood pressure nor pulses was lower in the upper left limb and the size of both upper limbs was similar.

In isolated subclavian artery, subclavian steal syndrome and pulmonary steal syndrome are also expected. In subclavian steal syndrome, the blood flow required by the subclavian artery is supplied via retrograde flow from the vertebro-basilar circulation, which in turn causes symptoms of vertebro-basilar insufficiency, especially during exercise. Our patients did not have any sign or symptoms of vertebro-basilar insufficiency, and the brain was found to be normal on the patient's brain CT scan.

Pulmonary Steal Syndrome occurs when pulmonary vascular resistance decreased in early infancy. Therefore, retrograde blood flow from the vertebral artery to the subclavian artery, and then to the pulmonary artery. This situation results in ischemia of the brain on one side and pulmonary congestion on the other. (9) Although since our patient had systemic pulmonary hypertension, the Pulmonary Steal phenomenon was not expected.

In up to 60% of isolated left subclavian cases, there are co-existent congenital heart defects, mostly Conotruncul anomalies such as double outlet right ventricle and tetralogy of Fallot, atrial septal defects and ventricular septal defects (9). Not surprisingly, our patient has multiple ventricular septal defects, the largest of which was a Perimambranous VSD with 6 mm diameter. On the other hand, extra-cardiac anomalies and genetic syndromes such as 22q11 deletion are more common in patients with aortic arch anomalies, especially in right aortic arch cases(8) Our patient did not have a thymus, so even though without a syndromic appearance, she was considered Digeorge syndrome and consequently genetic study requested for her.

Since our patient had multiple ventricular septal defects (Swiss cheese ventricular septum), based on the current guidelines, pulmonary artery banding was the preferred surgery for her. Reviewing the literature demonstrated that surgical reimplantation of the isolated left subclavian along with ductus arteriosus ligation have had acceptable outcomes, however, we did not perform it for our patient. As a replacement, we did ligation of the isolated left subclavian origin from the pulmonary artery. Obviously, in the follow-up period, evaluation of the size of the upper limbs as well as development of sign or symptoms of vertebra-basilar insufficiency seems necessary.

# Conclusion

isolated left subclavian is a rare aortic arch anomaly which should be considered in right aortic arch patients who have pulse or blood pressure discrepancy between their two upper limbs as it has clinical consequences such as subclvian steal syndrome and resultant brain ischemia.

# Declarations

## Ethics approval and consent to participate :

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

*Informed consent:* informed consent was obtained by the participant's parents included in the study.

## Consent for publication:

Not applicable

## Availability of data and materials

Data sharing is not applicable to this article as no datasets were generated or analysed during the current study

## Competing interests

The authors declare that they have no competing interests


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None


## Authors' contributions

Design of the work: M.K., M.H.

Writing original draft: T.T., M.T., S.S

Writing-review & editing: M.k., T.T., M.T.

Data acquisition: T.T., M.T, M.K., M.H.

Supervision: M.K, M.H., S.S.


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# Figures

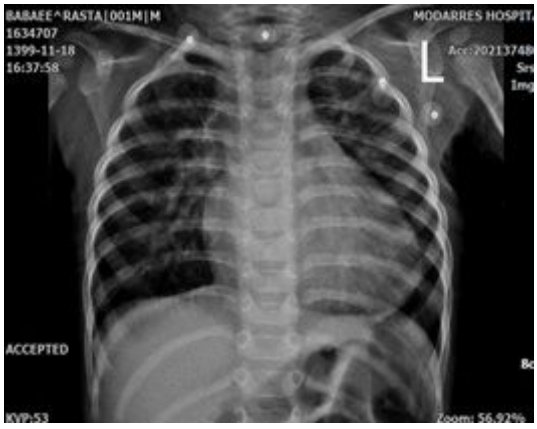


**Figure 1**

Frontal chest roentgenogram, demonstrating cardiomegaly, increased pulmonary vascular marking and right aortic arch.

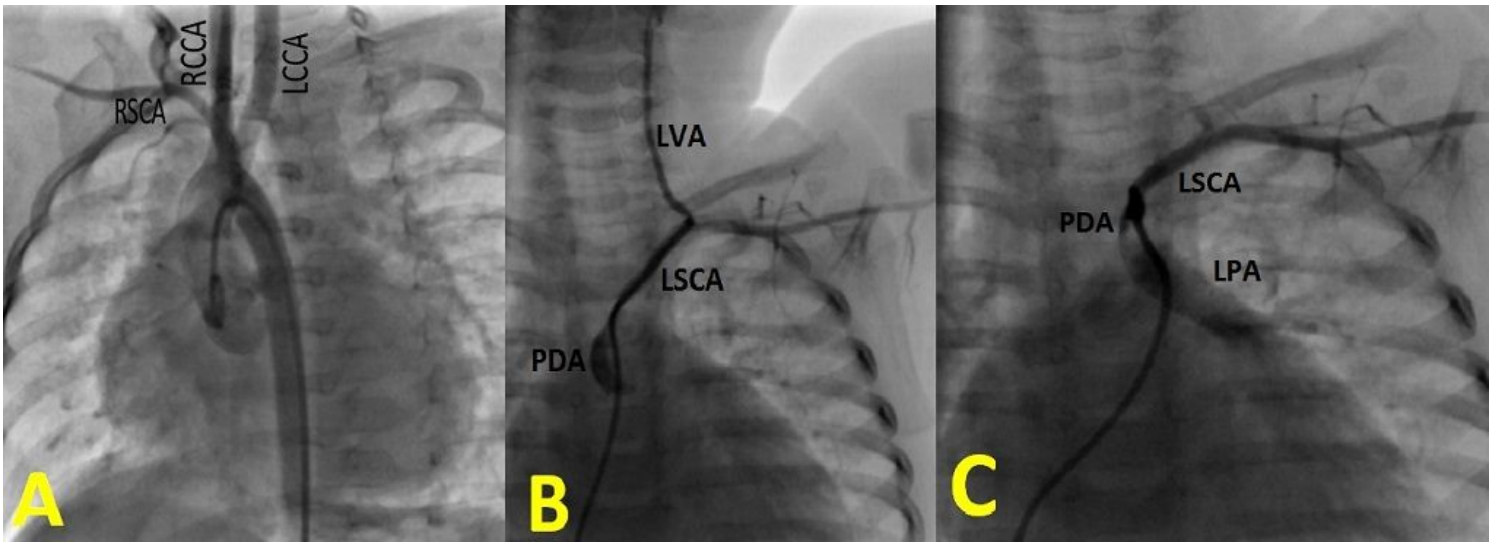


**Figure 2**

Trans-thoracic Echocardiography shows A, B: Muscular ventricular septal defects (asterisk) and large perimembranous ventricular septal defect (arrow) C: Dilated pulmonary artery (PA)

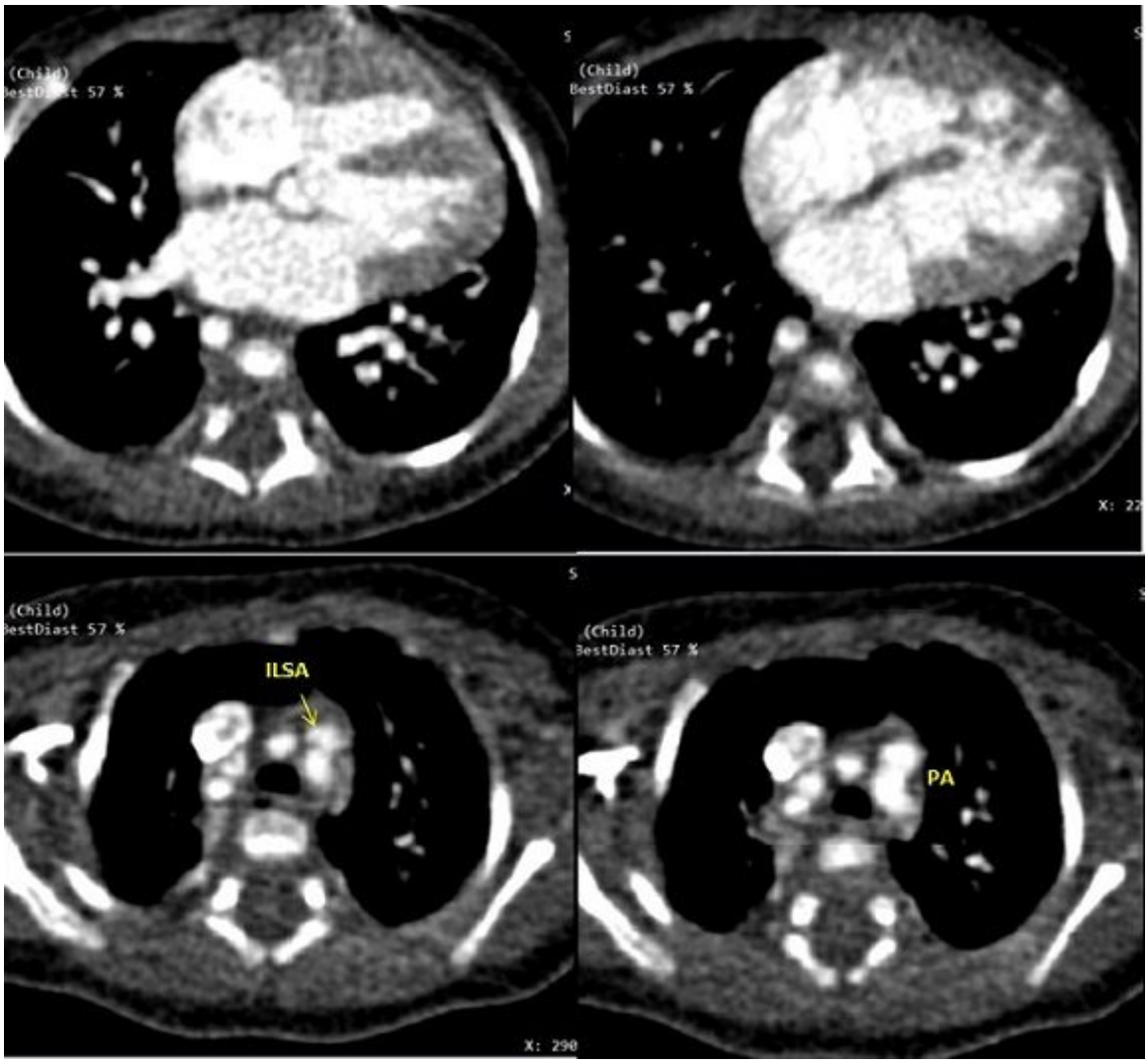


Figure 3

Cardiac CT angiography shows multiple ventricular septal defects as well as the isolated left subclavian artery (ILSA) connecting the pulmonary artery (PA)

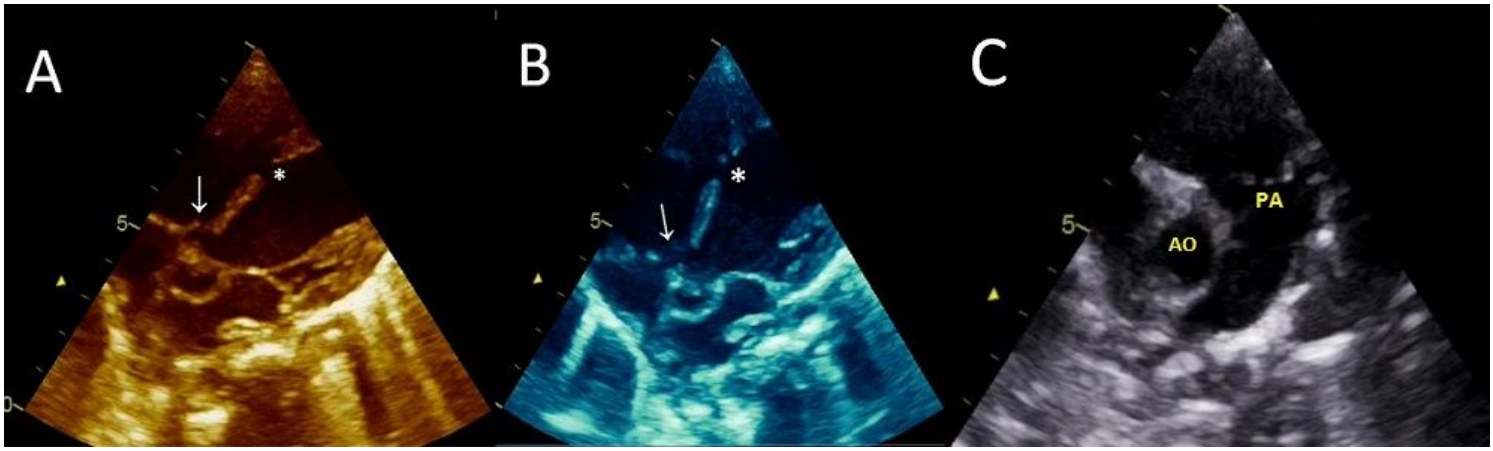


Figure 4

Ascending aorta angiography shows A: Right aortic arch as well as the arising of three vessels from the aortic arch: left common carotid, right common carotid and right subclavian artery in order. B and C: The left vertebral artery drains into the left subclavian artery, which is connected to the pulmonary artery via a patent ductus arteriosus. LCCA, left common carotid artery; RCCA, right common carotid artery; LSCA, left subclavian artery; RSCA, right subclavian artery; LVA, left vertebral artery; PDA, patent ductus arteriosu