

Subclavian and Pulmonary Steal Phenomenon in Isolated Left Subclavian Artery with Left Lung Agenesis

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SUMMARY

Isolated left subclavian artery has been documented to be associated with several congenital cardiac defects, including tetralogy of Fallot and transposition of the great arteries. In some cases subclavian or pulmonary artery steal phenomenon may occur when the isolated left subclavian artery connects to the main pulmonary artery via a ductus arteriosus. The isolated left subclavian artery may be part of the cardiac manifestations in multiple congenital anomalies, such as VACTERL association. We describe subclavian and pulmonary steal phenomenon in a neonate with complex congenital heart disease, including an isolated left subclavian artery, and left lung agenesis. An isolated left subclavian artery is rare and its association with agenesis of the left lung has not been reported previously. (Jpn Heart J 2002; 43: 429-432)

Key words: Isolated left subclavian artery, Subclavian steal, Agenesis of left lung

CASE REPORT

A male Taiwanese infant weighing 2760 grams was born at term to a G2P2 mother by vaginal delivery. The Apgar scores were 3 and 7 at one and five minutes, with a delay of initial crying for one minute. There was no family history of cardiac or skeletal anomalies. Physical examination at birth showed a slightly hypoplastic left hemithorax. The heart rate was 136 beats per minute, the respiratory rate 50 per minute, and blood pressure 45/25 mmHg and 62/34 mmHg in the left and right upper limbs, respectively. Systemic oxygen saturation was 80% on room air. The left brachial pulse was weak with a normal right brachial and femoral pulse. There was no cranial bruit. A grade 2/6 systolic ejection murmur was heard at the left midsternal border. Breath sounds were absent over the left chest. A chest x-ray showed almost complete opacification of the left hemithorax with a shift of the heart to the left, hemivertebrae of T4 to T7, scoliosis (Figure 1), and absence of the left radius. The baby's karyotype was 46XY. An electrocardiogram showed right axis deviation, right atrial enlargement, and right ventricular hyper-

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Received for publication January 18, 2002.

Revised and accepted March 25, 2002.



Figure 1. Chest radiograph (AP) shows opacification of the left hemithorax with shifting of the heart to the left, hemivertebrae of T4 to T7, and scoliosis.

trophy. Two-dimensional echocardiography demonstrated atrial situs solitus, a large primum type atrial septal defect, and a common atrioventricular valve with an indeterminate common ventricle. The ascending aorta was located to the right of the main pulmonary artery with a side-by-side arrangement. The aortic arch was right-sided. The first cephalic branch of the arch was the left common carotid artery. The pulmonary valve was mildly stenotic (pressure gradient 18 mmHg). The left pulmonary artery was not visible. There was a large patent ductus arteriosus, which connected to the left subclavian artery (LSA) distally and main pulmonary artery proximally with bi-directional shunting. Cardiac catheterization and angiography confirmed the aforementioned anatomy, including absence of the left pulmonary artery, the origin of the left subclavian artery from the main pulmonary artery via the ductus arteriosus, and retrograde flow in the main pulmonary artery (Figure 2). Other physical findings, computerized tomography and sonograms confirmed several other anomalies, including absence of the left radius and thumb, agenesis of the left lung, and agenesis of the left kidney. These extracardiac anomalies are characteristic of the VACTERL association.

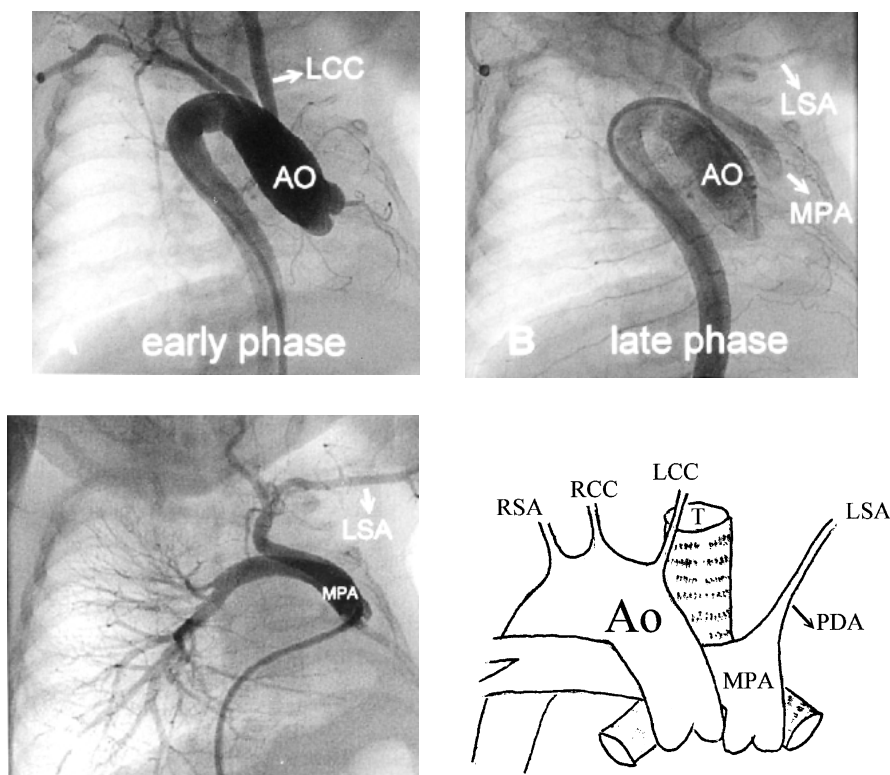


Figure 2. A: Early phase of aortogram demonstrates a right-sided aortic arch, with the first cephalic branch being the left common carotid artery (LCC). B: The main pulmonary artery (MPA) and left subclavian artery (LSA) are opacified by retrograde filling of contrast medium from the left vertebral artery. C: Pulmonary artery angiogram demonstrates the isolated left subclavian artery (LSA) connecting to the main pulmonary artery (MPA) via a patent ductus arteriosus (PDA). The left pulmonary artery is absent. D: Diagram of right aortic arch, isolated left subclavian artery (LSA), absent left pulmonary artery, and left patent ductus arteriosus (PDA) in this patient.

DISCUSSION

From the time of Shuford's description¹⁾ in 1970 up to 1990, only 39 cases of a right aortic arch with an isolated left subclavian artery (LSA) have been reported, diagnosed either by angiography or at postmortem examination.²⁾ This anomaly may derive from dual ipsilateral breaks in Edward's hypothetical arch development system.³⁾ One break is due to involution of the left dorsal aorta with migration of the left seventh intersegmental artery (subclavian) artery to the level of the left sixth (ductal) arch. Another break is caused by involution of the left fourth arch. These dual involutions leave the LSA isolated from the aortic arch but connected to the pulmonary artery system by a patent arterial duct or ligamentum arteriosum.

In isolated LSA, diminished pulses and a lower blood pressure (relative to the right arm) may be found in the left arm. Because the LSA is connected to the

main pulmonary artery via the arterial duct and to the left vertebral artery cephalically, both a "pulmonary steal phenomenon" from retrograde filling of the pulmonary arterial trunk and a "subclavian steal phenomenon" from retrograde circulation to the vertebrobasilar system may be noted.⁴⁻⁶⁾ These findings were present in our patient.

Various intracardiac anomalies may be associated with isolated LSA, the most common being tetralogy of Fallot.^{2,7)} Isolated cases with a double-outlet right ventricle,⁸⁾ complete transposition of the great arteries,⁵⁾ and interrupted aortic arch⁹⁾ have also been reported. Our patient's complex intracardiac malformations and absence of the left pulmonary artery have not previously been reported.

In addition to the cardiovascular malformations, our patient also had several extracardiac anomalies typical of the VACTERL association. Total agenesis of the left lung associated with an isolated LSA has not previously been reported, although there are reports of left lung hypoplasia in many cases of isolated LSA. While the mechanism is unknown, we hypothesize that our infant has an extreme form of this anomaly.

The management of isolated LSA consists of repair of the associated cardiac malformations and ligation of the PDA to prevent the pulmonary steal phenomenon.¹⁰⁾ However, patients with cerebral insufficiency or claudication of the left arm may require reimplantation of the LSA into the aorta. In our patient, a Fontan type operation or total cavopulmonary connection at a later period may be the best option, along with reimplantation of the LSA into the aorta.

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