REVIEW ARTICLE

Isolation of the left brachiocephalic artery revisited: A 52-year literature review and introduction of a novel anatomic-clinical-prognostic classification

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ABSTRACT

Isolation of the left brachiocephalic artery (ILBA) is an extremely rare anomaly of aortic arch with diverse manifestations in the neurologic system, heart, and left upper arm. This anomaly is defined as the absence of connection of the left brachiocephalic artery (LBA) to aortic arch and connection of LBA to pulmonary artery (PA) through a patent arterial duct (PAD). However, this definition is not inclusive of all cases. Not only are there inconsistencies in the definition and terminology of this aortic arch anomaly but also there is no classification for this anomaly despite its heterogeneous nature in terms of anatomy, clinical presentation and prognosis. We performed a 52-year comprehensive literature review in the period between 1966 and 2018. Our inclusion criteria were any manuscript that included a case report or case series, with confirmed diagnosis of ILBA. All quantitative data were analyzed using descriptive analysis by SPSS version 21 (IBM SPSS Statistics, USA). Results were presented as mean ± standard deviation and median. Based on the presence or absence of connection of LBA to PA and the number of sources of steal from the LBA, we classified ILBA into three types: single-steal type with no connection of LBA to PA and single source of blood flow steal from LBA through the left subclavian artery (LSCA), double-steal type with connection of LBA to PA through PAD and two sources of steal through LSCA and arterial duct (AD), and triple-steal type with bilateral PADs and therefore, three sources of blood flow steal from LBA including the LSCA and the double ADs. Patients with single-steal type have the best prognosis and present latest with symptoms of cerebrovascular insufficiency or left arm claudication. The oldest reported patient was 69 years of age with symptoms of dizziness and near syncope. No death was reported in these patients. Double-steal type is the most common type and is often associated with genetic syndromes and/or extracardiac anomalies. Triple-steal type is the rarest type with the earliest presentation and worst prognosis. The oldest reported patient was 60 days of age. All reported cases had cardiac symptoms, pulmonary overcirculation, pulmonary hypertension, and fatal outcome.

Keywords: Anatomic-clinical-prognostic classifi cation, isolation of left brachiocephalic artery, left subclavian steal syndrome

INTRODUCTION

Isolation of the left brachiocephalic artery (ILBA) is an extremely rare anomaly of aortic arch with diverse manifestations in the neurologic system, heart, and left upper arm. This anomaly is defined as the absence of connection of the left brachiocephalic artery (LBA) to aortic arch and connection of LBA to pulmonary

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artery (PA) through an arterial duct (AD).[1-6] However, this definition is not inclusive of all cases, including our present case. We present an asymptomatic child with ILBA and without connection of LBA to PA. Since 1966 when Levine *et al.* reported the first case of ILBA as "atresia of left common carotid and left subclavian arteries," there are inconsistencies in the definition and terminology of this aortic arch anomaly.[1] There is no classification for this anomaly despite its heterogeneous nature in terms of anatomy, clinical presentation, and prognosis.

We performed a comprehensive literature review in the period between 1966 and 2018. Based on the anatomy of our case and the 52-year literature review of ILBA, from 1966 to April 2018, we propose a comprehensive, novel, and practical anatomic-clinical-prognostic classification of ILBA.

SHORTCOMINGS OF THE CURRENT TERMINOLOGY AND DEFINITION AND THE NEED OF A CLASSIFICATION FOR ISOLATION OF THE LEFT BRACHIOCEPHALIC ARTERY

There are confusing inconsistencies not only in terminology but also in the definition of isolation of LBA in the literature. Different terminologies have been used for this anomaly including "anomalous origin of LBA from pulmonary artery," "aplasia of left brachiocephalic trunk," "isolation," "anomalous origin of LBA from left pulmonary artery," and "origin of LBA from right pulmonary artery."[2-6] The term "origin" from PA is not justified because the direction of blood flow is not from PA into the LBA. This hemodynamic point differentiates ILBA from interrupted aortic arch Type C [Figure 1 and Table 1].

The original case reported by Levine *et al.* was a 33-year-old woman with symptoms of dizziness and poor perfusion of the left arm without any connection of LBA to PA through a patent AD (PAD).[1] However, later, some authors defined ILBA as origin of LBA from left PA through an AD.[5] Review of literature and study of our case, who had no connection of LBA to PA, indicate the shortcoming of the current definition in inclusion of all cases of ILBA. Obviously, there is significant hemodynamic difference between patients with PAD and those with obliterated AD. Left to right shunt across the AD occurs only in the former.

Patients with ILBA have widely heterogeneous characteristics in terms of anatomy, clinical presentation, and outcome. These diversities indicate the heterogeneous nature of these patients and the need for an appropriate anatomic-clinical-prognostic classification system.

CLINICAL IMPORTANCE OF ISOLATION OF THE LEFT BRACHIOCEPHALIC ARTERY

ILBA results in a heterogeneous spectrum of clinical manifestations expanding over three organ systems, which can be acronymed as three "H's." Three H's include head (i.e., cerebral symptoms due to vertebral-basilar insufficiency), heart (cardiac manifestations secondary to pulmonary overcirculation and congestive heart

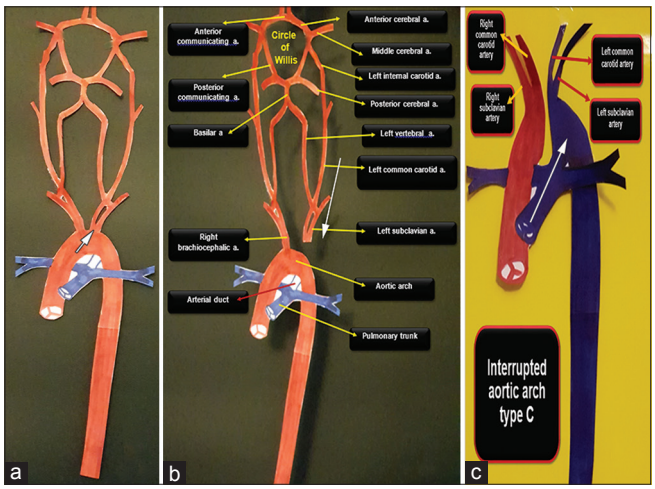

Figure 1: Aortic arch and cerebrovascular circulation in three different conditions: (b) normal, (b) isolation of the left brachiocephalic artery without connection to pulmonary artery, and (c) Type C interruption of aortic arch. For the sake of simplicity, all annotations are shown only in Figure 1b and are not repeated in Figure 1a. The white arrow in each of the figures shows the direction of blood flow into the left brachiocephalic artery

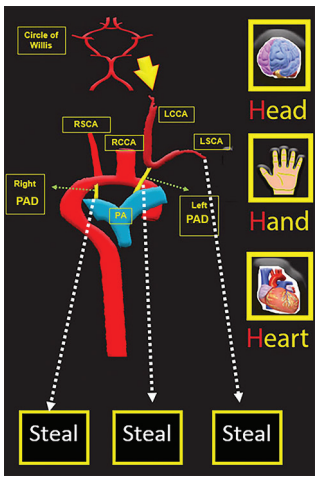

Figure 2: Schematic diagram showing "the anatomy and the number of steal phenomenon" and "the source of symptoms" in patients with isolation of the left brachiocephalic artery. Symptoms are related to head (manifested as dizziness and transient ischemic attacks due to cerebrovascular insufficiency), heart (manifested as congestive heart failure due to left to right shunt from left brachiocephalic artery to pulmonary artery), and hand (manifested as left arm claudication due to decreased flow to left subclavian artery)

Table 1: Clarifying the confusion on the source of perfusion of the left brachiocephalic artery and direction of blood flow between left brachiocephalic artery and pulmonary artery in four conditions: Normal, isolation of the left brachiocephalic artery (without and with connection to pulmonary artery), and type C interruption of aortic arch

The four conditions	Direction of perfusion of the left brachiocephalic artery	Source of perfusion of the left brachiocephalic artery
Normal	Antegrade perfusion	Ascending aorta > aortic arch > left brachiocephalic artery
ILBA without connection to PA	Retrograde perfusion	Ascending aorta > right brachiocephalic arteries > circle of Willis > left brachiocephalic artery
ILBA with connection to PA	Retrograde perfusion	Ascending aorta > right brachiocephalic arteries > circle of Willis > left brachiocephalic artery>Patent arterial duct>pulmonary artery
Type C interruption of aortic arch	Antegrade perfusion	Pulmonary trunk > patent arterial duct > aorta > left brachiocephalic artery

ILBA: Isolated left brachiocephalic artery, PA: Pulmonary artery

failure), and hand (claudication and numbness of the left arm due to decreased flow to the left subclavian artery [LSCA]) [Figure 2]. The cerebral and upper extremity symptoms are due to "under-perfusion" of these organs whereas the cardiac symptoms are secondary to pulmonary "over-circulation" resulting from the left to right shunt that occurs from LBA into PA through the PAD or ducts.

Patients with ILBA are also at risk of subclavian artery steal phenomenon. Connection of ILBA to PA provides the setting for an additional left to right shunt by PA steal (double steal) with consequent cardiac manifestations secondary to pulmonary overcirculation. In patients with bilateral ADs, there is a potential for triple steal to occur from LBA, including LSCA steal and pulmonary arterial steal through the left and the right ADs.

52-YEAR LITERATURE REVIEW ON ISOLATION OF THE LEFT BRACHIOCEPHALIC ARTERY: 1966–2018

We performed a 52-year literature review of ILBA from 1966 to April 2018.

Our inclusion criteria were any manuscript that included a case report or case series, with confirmed diagnosis of ILBA. The diagnostic criteria were lack of connection of LBA to aorta, Retrograde late filling of LBA through the collateral arteries from circle of Willis with or without connection to PA through unilateral or bilateral patent arterial duct or ducts.

The electronic database of PubMed, Embase, Cochrane, Web of Science, Scopus, Ovid MedLine, Clinical Key, and Essential Science Indicators and electronic journals of ScienceDirect, Wiley Online Library, Springer, Proquest, BMJ, and SAGE were searched respectively. We also searched Google Scholar and Google. We used the search terms of "isolation," "isolated," "anomalous origin," and "atresia" in combination with LBA or brachiocephalic artery, brachiocephalic artery and LBA without any limitation for age or language. We searched the references of each paper and included the relevant articles. We excluded cases with isolation of only left common carotid artery (LCCA) or only LSCA. We extracted the following twelve information about each case: age at presentation, main clinical presentation, presence of other associated congenital heart disease, presence of pulmonary hypertension, presence of extracardiac anomaly or syndromes, presence of weak or absent pulse of LCCA, smaller size, lower blood pressure (BP) and lower oxygen saturation of the left hand, smaller left cerebral hemisphere, type of treatment, and final outcome of the patient. All quantitative data were analyzed using descriptive analysis by SPSS version 21 (IBM SPSS Statistics, New york, USA). Results were presented as mean ± standard deviation and median. We found 30 cases that had the diagnostic criteria for inclusion in our study. After studying the full text of all papers, we found that case number 15 was a case of left aortic arch with isolated right brachiocephalic artery. Neither the title of this paper nor the full text clearly stated the sidedness of the brachiocephalic artery. Sidedness was shown in the figure of this case report.[6]

A separate literature search performed to find further cases of the left aortic arch and isolated right brachiocephalic artery proved the uniqueness of this case.

We also included the information of an asymptomatic 3.5-year-old child with ventricular septal defect (VSD) and ILBA. She was referred to our clinic with a wrong diagnosis of VSD and interrupted right aortic arch. On physical examination, she had normal growth and development. First and second heart sounds were normal and a grade 4/6 holosystolic murmur was heard at the left sternal border. A bruit was heard on the cranium. The left carotid, left brachial, and left radial pulses were weaker than the right ones. The BP in the right and left arms were 111/77 mmHg (mean = 90 mmHg) and

87/60 mmHg (mean = 70 mmHg), respectively. Oxygen saturation was 98% in both hands. Both hands were of the same size. On cardiac catheterization and angiography, there was a right aortic arch with two arch branches including right common carotid artery and right subclavian artery with ILBA as seen by late and retrograde filling of the LBA with no connection to PA [Figure 3 and Video 1]. The pulmonary arterial pressure was normal. Imaging of the brain and the heart by 384-slice computed tomographic angiography revealed normal size of both cerebral hemispheres, normal cerebral arteries and a long distance (21 mm) between the isolated LBA and the aortic arch [Figure 4]. The child was scheduled for surgical VSD closure. Considering the long distance between the LBA and the aortic arch, absence of symptoms of subclavian artery steal at this stage of life, and lack of consent of the parents, reimplantation of the isolated LBA was not planned at this age.

The results of the 52-year literature review on cases with ILBA, including our case, are tabulated in Table 2.

NOVEL ANATOMIC-CLINICAL-PROGNOSTIC CLASSIFICATION OF ISOLATION OF THE LEFT BRACHIOCEPHALIC ARTERY

Based on this literature review, we classified all the 31 cases based on the number of sources of steal from the brachiocephalic artery into three anatomic-clinical-prognostic types [Figure 5]. The basis for classification was the presence or absence of connection of LBA to PA and the number of sources of steal of flow from LBA. Case number 15 in Table 2 was a unique case of 2-month-old infant with left aortic arch and isolated right brachiocephalic artery.[6]

1. Single-steal type with single source of steal from LBA through the LSCA and with no connection to PA
2. Double-steal type with double sources of steal from LBA through LSCA and PAD
3. Triple-steal type with triple sources of steal from LBA through LSCA and bilateral PAD.

Clinical and prognostic characteristics of the three types of isolation of the left brachiocephalic artery

Comparison of clinical and prognostic characteristics of these three types is summarized in Table 3.

Single-steal type

In these patients with no connection of LBA to PA, the only source of steal from LBA is subclavian artery steal. Therefore, the least amount of steal occurs from the cerebral circulation. During infancy and early childhood when both the physical activities and the linguistic ability to express symptoms are limited, we do not expect overt complaints or symptoms of cerebrovascular insufficiency. The clinical and prognostic characteristics of this type are higher age at presentation, dominance of symptoms of cerebrovascular and left arm perfusion insufficiency, lack of association with genetic syndromes and extracardiac anomalies and universally good outcomes. Although atrial septal defect (ASD), VSD, pulmonary stenosis, and double-chamber right ventricle have been reported in these patients, the vast majority of these patients do not have congenital heart disease. Smaller left cerebral hemisphere, reported in other types, has not been reported in this type. There is no report of death in this type.[1,4,7,8,11,17,19,21]

Double-steal type

LBA is connected to PA in these patients, either directly or through a left-sided PAD. This type is the most common type of ILBA. These patients present much earlier than patients with single-steal type. More than half of these patients have associated genetic syndromes, including Down syndrome and particularly DiGeorge syndrome, or other extracardiac anomalies. CHARGE association and polysplenia syndrome have been reported. Cardiac manifestations are the main clinical presentation and 76.4% of these patients have at least one congenital heart disease other than

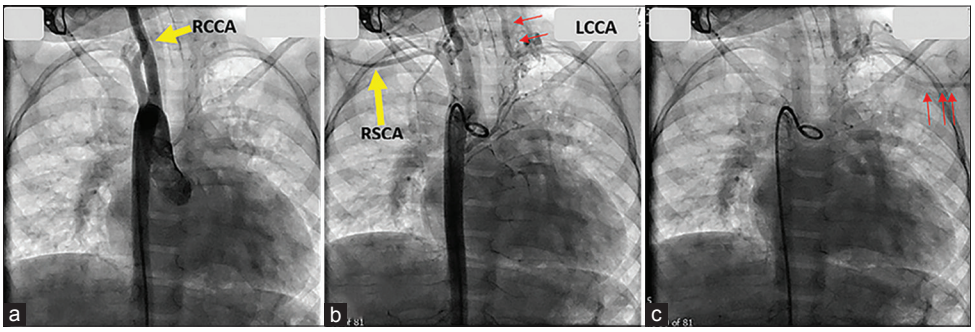

Figure 3: (a) Aortogram in anteroposterior view indicates right aortic arch with two branches of RCCA (yellow arrow) and RSCA. (b) After several beats, LCCA is visualized by retrograde flow from the cerebral circulation from above (red arrows). RSCA is shown by yellow arrow. LCCA is smaller than RCCA. (c) The left subclavian artery appears late through retrograde flow from above which is shown by the red arrows. RCCA: Right common carotid artery, LCCA: Left common carotid artery, RSCA: Right subclavian artery

Table 2: A 52-year literature review of complete isolation of brachiocephalic artery* (in order of the date from April 2018 back to 1966)

Number	Authors	Year	Age	Sex	Connection to PA (directly or through patent arterial duct)	Extracardiac anomaly	Associated CHD and/or PH	Other findings	Smaller size of the left hand	W/A pulse of LCCA	W/A pulse of the left upper limb	Lower oxygen saturation of the left hand	Lower blood pressure of the left hand	Smaller left cerebral hemisphere	Bilateral patent arterial ducts	Outcome	Type of ILBA
1	Our case	2017	3.5 years	Female	No	No	VSD	Bruit on the cranium	No	+	+	-	+	No	No	Scheduled for VSD closure	SS
2	Dubey *et al.*[5]	2017	9 years	Male	+	No	PDA + PH	Bruit in the neck and cranium	Atrophy of left upper limb	+	+	92%	Dampened waveforms	NS	No	SILBA	DS
3*	Gesuete *et al.*[7]	2016	1 day	Female	+	Prader-Willi syndrome	Multiple VSD + PDA + PH	Preterm (35 W)	NS	NS	NS	NS	NS	NS	No	Died at the age of 9 months She was not operated because of PH	DS
4	Yamasaki *et al.*[8]	2015	50 years	Male	No	No	No	Ruptured brain aneurysm and agenesis of the left internal carotid + untreated systemic hypertension	NS	NS	NS	NS	NS	NS	No	The brain aneurysm was successfully clipped	SS
5	Joseph *et al.*[9]	2016	56 years	Female	No	No	No	Headache + right-sided numbness of face and extremity	NS	NS	No difference	NS	No difference	NS	No	Intervention was considered	SS
6	Gowda *et al.*[10]	2014	5 years	Male	+	No	No	Referred from school Weakness of the left hand with playing	NS	NS	+	Left hand was 5% lower	Less than half of right hand BP	NS	No	SILBA	DS
7	Mangukia *et al.*[11]	2014	10 years	Male	No	No	VSD + Double-chamber RV	Presented with "Shortness of breath"	NS	NS	+	NS	Lower BP	No	No	Cardiac surgery for CHD was done. LBAILBA was not corrected	SS
8	Parody *et al.*[3]	2012	9 months		+	Down syndrome	VSD + PDA + PH	NS	NS	NS	NS	+	NS	NS	NS	SILBA + surgical repair of cardiac lesion	DS
9	Kreeger *et al.*[12]	2011	3 months	NS	+	DiGeorge syndrome	VSD	Incidental finding during an admission for an aspiration event	NS	NS	NS	NS	NS	NS	No	SILBA	DS
10	Gil-Jaurena *et al.*[13]	2011	6 months	NS	+	No	ASD PDA	Cardiac murmur and heart failure	NS	NS	NS	NS	NS	NS	No	SILBA	DS

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Table 2: Contd...

Number	Authors	Year	Age	Sex	Connection to PA (directly or through patent arterial duct)	Extracardiac anomaly	Associated CHD and/or PH	Other findings	Smaller size of the left hand	W/A pulse of LCCA	W/A pulse of the left upper limb	Lower oxygen saturation of the left hand	Lower blood pressure of the left hand	Smaller left cerebral hemisphere	Bilateral patent arterial ducts	Outcome	Type of ILBA
11	Reeves *et al.*[14]	2010	3 months	Female	+	"mild dysmorphic features" (Subclavian steal?)	Primum and secundum ASDs Cleft mitral valve LPA stenosis PPH + eft PDA and ductal stenosis + three branches of aortic arch	Developmental delay + Bulbar dysfunction	NS	NS	NS	NS	NS	diminished volume of the left cerebral hemisphere	No	NS	DS
12	Le Bret *et al.*[15]	2009	4 years	Female	+	22q11 deletion	PDA + PH	-	NS	NS	NS	NS	NS	NS	No	Excision of PDA and SILBA	DS
13	Martin *et al.*[16]	2006	23 days	Female	+	CHARGE association	PFO + PDA + PH + subaortic stenosis	-	NS	NS	NS	equal	+	Larger lateral ventricle on the left	No	SILBA + PDA ligation	DS
14	Pauliukas[4]	2005	35 years	Female	No	No	No	Symptoms of cerebrovascular and left arm hypoperfusion	No	+	NS	+	NS	NS	No	SILBA . using a 12-mm vascular graft	SS
15	Miyaji *et al.*[6]	2001	2 months	Female	+	DiGeorge syndrome	ASD + VSD + PDA + massive enlargement of pulmonary trunk producing a kink on the proximal left pulmonary artery + left aortic arch**	heart failure	NS	NS	NS	Right: 90% Left: 96%	NS	NS	No	Closure of ASD and VSD and reimplantation of brachiocephalic artery to the aorta	DS
16	Mart *et al.*[17]	2001	Neonate (precise age is not stated)	Male	+	No	TF	Cardiac murmur	NS	NS	NS	NS	Lower BP of the left arm	NS	No	Implantation of ILBA to aorta (with postoperative stenosis between aorta and LBA)	DS
17	Singh *et al.*[18]	2001	36 years	Female	No	No	No	Presented with: "dizziness vertigo claudication of left upper limb"	NS	+	+	NS	+	NS	No	Successful anastomosis of RSCA to LCC-LSCA confluence*	SS

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Table 2: Contd...

Number	Authors	Year	Age	Sex	Connection to PA (directly or through patent arterial duct)	Extracardiac anomaly	Associated CHD and/or PH	Other findings	Smaller size of the left hand	W/A pulse of LCCA	W/A pulse of the left upper limb	Lower oxygen saturation of the left hand	Lower blood pressure of the left hand	Smaller left cerebral hemisphere	Bilateral patent arterial ducts	Outcome	Type of ILBA
18*	Duke and Chan[19]	2001	1 day	Male	+	22q11 microdeletion	Acyanotic TF + Pulmonary overcirculation + PH + cervical aortic arch	Neonatal hypocalcemia + preterm (gestational age of 35 W)	NS	NS	NS	NS	NS	NS	No	Cardiac surgery at 13 months Isolated LBA was not corrected. The patient died of viral infection later	DS
19	Boren *et al.*[20]	2000	69 years	Male	No***	No	No	Symptoms of cerebrovascular insufficiency (dizziness and near syncope)	NS	NS	NS	NS	NS	NS	NS	Bypass graft of axillo-Axillary type	SS
20	Gamillscheg *et al.*[21]	2000	6 months	Male	+	Down syndrome	ASD + VSD + PDA + PH	No	NS	NS	NS	NS	Equal BP	NS	NS	SILBA using a 8-mm PTFE vascular graft	DS
21	Delgado and Barturen[22]	1998	5 years	Female	No	No	Long-segment coarctation	The aortic arch had three branches (RCC, RVA, RSCA)	NS	+	NS	NS	Left arm was slightly more because the RSCA was at the site of the coarctation	NS	No	Not operated because of the high risk of postoperative paralysis	SS
22	Kaku *et al.*[23]	1996	13 months	Female	+	Down syndrome	VSD + LPA stenosis + PDA + PFO + PH	Abnormal silhouette of heart on chest X-ray	NS	+	+	NS	Lower BP	NS	No	Awaiting operation	DS
23*	Bornemeier *et al.*[24]	1996	1 day	Male	+	Rib anomaly	Severe AS + coarctation + VSD + d-TGA + LJAA + PH	-	NS	NS	equal pulses	NS	Almost equal	NS	+	Died the day after operation	TS
24*	Papagiannis *et al.*[25]	1996	1 day	Male	+	Asplenia syndrome	Dextrocardia + DORV + situs inversus of atria + L-loop ventricles + common atrium + unbalanced AVSD + aortic atresia	NS	NS		+	NS	Lower BP	NS	+	Died in the operation room	TS
25*	Fong and Venables[26]	1987	6 weeks	Female	+	CHARGE association	Initially was operated for ligation of right-sided PDA	NS	NS	NS	NS	NS	NS	No	+	Sudden unexplained death before the second surgery	TS

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Table 2: Contd...

Number	Authors	Year	Age	Sex	Connection to PA (directly or through patent arterial duct)	Extracardiac anomaly	Associated CHD and/or PH	Other findings	Smaller size of the left hand	W/A pulse of LCCA	W/A pulse of the left upper limb	Lower oxygen saturation of the left hand	Lower blood pressure of the left hand	Smaller left cerebral hemisphere	Bilateral patent arterial ducts	Outcome	Type of ILBA
26*	Crump *et al.*[2]	1981	2 days	Female	+	Polysplenia syndrome	CAVSD + Bilateral SVC	NS	-	NS	+	NS	NS	No	No (oblite rated ductus)	Died shortly after cardiac surgery	DS
27	Harrington *et al.*[27]	1981	4 years	M	No	No	ASD + VSD + PS	Congestive heart failure in neonatal period	NS	+	+	NS	+	NS	No	Surgical correction of cardiac lesion (not clearly stated)	SS
28	Martin *et al.*[28]	1979	?	?	+	?	Left PDA	Access to full text of this paper was not possible.									DS
29	Park[29]	1979	3 days	M	+	No	CAVSD PDA+PH	NS	NS	+	+	NS	NS	NS	No	NS	DS
30*	Shaher *et al.*[30]	1972	2 months	NS	+	Multiple congenital anomalies	PDA+mild PH	NS	NS	+	+	NS	NS	Smaller left cerebral hemisphere +	+	Died at the end of the operation	TS
31	Levine *et al.*[1]	1966	33 years	Female	No (obliterated ductus)	No	No	Numbness of the left hand and "light-headedness"	+	+	NS	+	+	Normal radiomercury brain scan	No	SILBA	SS

AS: Aortic stenosis, B. PAD: Bilateral patent arterial duct, CAVSD: Complete atrioventricular septal defect, CHD: Congenital heart disease, ILBA: Isolated left brachiocephalic artery, LBA: Left brachiocephalic artery, LJAA: Left juxtaposition of atrial appendages, NS: Not stated (or not reported), PA: Pulmonary artery, PAD: Patent arterial duct, PH: Pulmonary hypertension, PPH: Persistent pulmonary hypertension, PS: Pulmonary stenosis, PTFE: Polytetrafluoroethylene, RCC: Right common carotid artery, RVA: Right vertebral artery, RSCA: Right subclavian artery, SVC: Superior vena cava, SILBA: Successful implantation of left brachiocephalic artery to aorta, VSD: Ventricular septal defect, W/A: Weak or absent; Color code of the table: White numbers, SS: Gray numbers, DS: Black numbers, TS, LCCA: Left common carotid artery

*Case numbers with an asterisk (i.e. 3, 18, 23, 24, 25, 26 and 30) are died. All cases are isolation of the left brachiocephalic artery except case number 15 that is isolation of the right brachiocephalic artery and left aortic arch. Using an autologous saphenous venous graft, **As to the best of our knowledge, this is the only reported case of isolation of right brachiocephalic artery with left aortic arch, ***Boren *et al.* (authors) state that: "flow toward" pulmonary artery (but not in the pulmonary artery) was seen through a vestigial patent arterial duct. The pulmonary artery is not opacified in the figure of their paper

PAD. Primum and secundum ASD, cleft mitral valve VSD, complete atrio ventricular septal defect (VSD), cervical aortic arch, subaortic stenosis, coarctation, and bilateral superior caval vein have been reported. More than 90% of cases had symptoms of pulmonary overcirculation. The majority of reported patients had pulmonary hypertension. Two patients died in this group, one died at the age of 9 months and the other before 2 years of age, had Prader–Willi syndrome and DiGeorge syndrome, respectively. Evaluation for DiGeorge syndrome and other genetic syndromes is recommended in this group.[3,5-7,10,12-16,18,20,22,26,27,30]

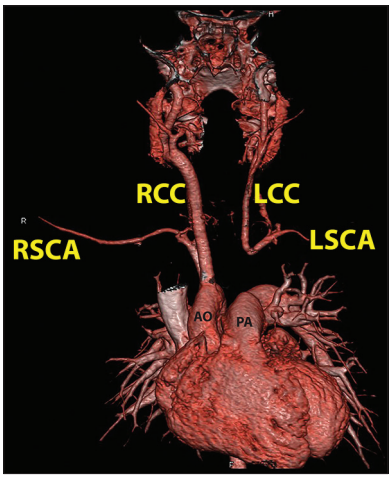

Figure 4: Three-dimensional, volume -rendered image of computed tomographic angiography shows the right aortic arch with isolation of the left brachiocephalic artery. LCC artery is smaller than RCC artery. The distance between the isolated left brachiocephalic artery with the aortic arch is significant. RCC: Right common carotid; LCC: Left common carotid, RSCA: Right subclavian artery, LSCA: Left subclavian artery, AO: Aorta, PA: Pulmonary artery

Triple-steal type

These patients have bilateral PADs which set the stage for triple steal to occur from the LBA. This is the rarest type with the earliest presentation, the worst prognosis, and 100% mortality. Despite the three sources of steal from LBA, the cause of death is not attributable to the steal phenomenon. The main culprit for mortality in these patients was the associated complex congenital heart disease. All reported cases had pulmonary overcirculation and pulmonary hypertension. Left to right shunt across the two PADs contributed to pulmonary congestion and heart failure. However, the smaller left cerebral hemisphere, reported in one-fourth of these patients on autopsy, can be explained by the triple-steal phenomenon. All reported cases had pulmonary overcirculation and pulmonary hypertension. A more detailed description of the cardiac and extracardiac anomalies of the four patients with this type is indicated in Table 4.[23-25,30]

Advantages of the novel classification

This classification system is comprehensive and includes all the reported cases of ILBA. It provides practical

Table 3: Clinical and prognostic characteristics of the three types of isolated left brachiocephalic artery*

	Single-steal type (*n*=10), *n* (%)	Double-steal type (*n*=17), *n* (%)	Triple-steal type (*n*=4), *n* (%)
Prevalence	10 of 30 (30)	17 of 30 (56.66)	4 of 30 (13.33)
Age at presentation (mean±SD)	30.15±23.73 years	1.35±2.52 years	26±29 days
Age at presentation (median)	34 years	3 months	21.5 days
Age at presentation (minimum-maximum)	3.5 years-69 years	1 day-9 years	1 day-60 days
Association with genetic syndromes and/or dysmorphic features or other extracardiac anomalies	0	10 of 17 (59)	4 of 4 (100)
Associated syndromes	-	Prader-Willi syndrome, DiGeorge syndrome, down syndrome and polysplenia syndrome, dysmorphic features, rib anomaly	Multiple congenital anomalies, asplenia syndrome, and charge association
Association with DiGeorge syndrome	0	4 of 17 (23.5)	0
Congenital heart disease other than a left-sided PDA	3 of 10 (30)	13 of 17 (76.4)	3 of 4 (75)
Symptoms of left to right shunt and pulmonary overcirculation	0	16 of 17 (94.1)	4 of 4 (100)
Symptoms of vertebral-basilar insufficiency	6 of 10 (60)	0	0
Left arm claudication	4 of 10 (40)	1 of 17 (0.06)	0
Pulmonary hypertension	0	At least 13 of 17 (76.4)	4 of 4 (100)
Weak or absent left radial pulse	5 of 10 (50)	6 of 17 (29.4)	2 of 4 (50)
Pulses of left and right hands were not reported	4 of 10 (40)	11 of 17 (65)	1 of 4 (25)
Equal pulses of left and right hands	1 of 10 (1)	0	1 of 4 (25)
Diminished volume of left cerebral hemisphere or Larger lateral ventricle on the left side	0	1 of 17 (0.06)	1 of 4 (25)
Death	0	3 of 17 (17.6)	4 of 4 (100)

*Case number 15 in Table 2, which was an isolated right brachiocephalic artery and left aortic arch is not included. SD: Standard deviation, PDA: Patent arterial duct

Table 4: Details of cardiac and extracardiac anomalies and other important characteristics of the four cases with triple-steal type of isolated left brachiocephalic artery

Author, year, country	Age	Cardiac anomalies	Extracardiac anomalies	Presence of PH	Possible etiology of pulmonary hypertension	Cardiac surgery
Bornemeir *et al*. (1996, USA)	1-day-old male newborn	Mesocardia, D-transposition of great arteries, hypoplastic aortic arch, tricuspid valve and right ventricle, severe coarctation with right arterial duct-dependent systemic circulation, valvar aortic stenosis, left juxtaposition of atrial appendages, isolation of left brachiocephalic artery with connection to pulmonary trunk through the left patent arterial duct	Rib and vertebral anomaly	+	Ductal-dependent systemic circulation	Aortic arch reconstruction using homograft Ligation of the right arterial duct Gore-Tex shunt between ascending aorta and right pulmonary artery (the newborn died the day after the operation)
Fong *et al*. (1987, Australia)	6-week-old female infant	Bilateral patent arterial duct with left to right shunt across the ducts and continuous murmur on cardiac auscultation	Charge association	Not explicitly stated (the continuous murmur denotes absence of significant PH)	-	First cardiac surgery included ligation of the right arterial duct. Sudden death occurred at 6 months of age at home before the second planned surgery (re-implantation of the left brachiocephalic artery). Diagnosis was confirmed by necroscopy
Shaher *et al*. (1972, USA)	2-day-old newborn	Continuous murmur, mild elevation of pulmonary artery pressure	Unilateral choanal atresia, low-set ears, unilateral facial peripheral paralysis, small mandible, abnormal digits, dysphagia.	Present (PA pressure=35/10 and aortic pressure=40/35 mmHg)	Left to right shunt	Division of bilateral patent arterial ducts without reimplantation of the left common carotid artery and left subclavian artery The infant died at the end of operation
Papagiannis *et al*. (1996, USA)	1-day-old boy	Autopsy-confirmed diagnosis: Asplenia syndrome, dextrocardia, situs inversus, l-loop ventricles, d-malposition of great arteries, common atrium, unbalanced complete atrioventricular septal defect and aortic atresia and bilateral patent arterial duct	Asplenia syndrome	+	Ductal-dependent systemic circulation	Detachment of both pulmonary artery branches from the pulmonary trunk with the mistaken diagnosis of truncus arteriosus, central shunt from the pulmonary trunk to left pulmonary artery and reimplantation of the isolated left brachiocephalic artery (the patient died in the operation room)

information on anatomy, expected clinical manifestations and outcome. It also provides hints on the essential diagnostic workup.

THREE SIMPLE AND HELPFUL DIAGNOSTIC CLUES FOR EARLY RECOGNITION OF THE TRICKY DIAGNOSIS OF ISOLATION OF THE LEFT BRACHIOCEPHALIC ARTERY

Look for the triad of "right aortic arch," "two aortic branches," and "weak pulse of the left arm"

The triad of "right aortic arch, two aortic arch branches and weak pulse of left upper arm" was present in almost 90% of all cases [Table 5].

All reviewed cases had right aortic arch. All patients except two cases had two aortic arch branches. The case reported by Pauliukas *et al.* had right aortic arch with mirror-imaging branches (case number 14 with single-steal type in Table 1).[4] Case number 11, a double-steal type, reported by Reeves *et al.* also had three aortic arch branches.[14] It is noteworthy that there is only once case with left aortic arch and isolation of the right brachiocephalic artery.[6] All other patients had right aortic arch with two aortic arch branches.

Absent or weak pulses of the left carotid, left brachial, and left radial artery are present in the vast majority of all types when pulses were examined. However, in about half of cases, bilateral examination of pulses was either not performed or not reported.

Table 5: The frequency of the triad of right aortic arch, two aortic arch branches, and weak or absent pulse of left upper limb in isolation of left brachiocephalic artery (total number of 30 cases*)

	n	Frequency (%)
Right aortic arch	30 of 30	100
Right aortic arch with two aortic arch branches	27 of 30	90
Right aortic arch with one aortic arch branch	1 of 30[1]	0.03
Right aortic arch with three aortic arch branches	Case number 31 in Table 2 1 of 30[14]	0.03
Right aortic arch with mirror-image branching	Case number 11 in Table 2 1 of 30[4]	0.03
Weak or absent left upper limb pulse in the 14 cases in whom pulses of the hands were examined	Case number 14 in Table 2 12 (of 14)	86
Pulses of the left upper limb were not reported**	16 of 30	53.3

*Case number 15 in Table 2, reported by Miyagi *et al*., had left aortic arch and isolation of right brachiocephalic artery and was not included in this table, **Access to the full text of case number 28 on Table 2, the case reported by Martin *et al*. in 1979, was not possible

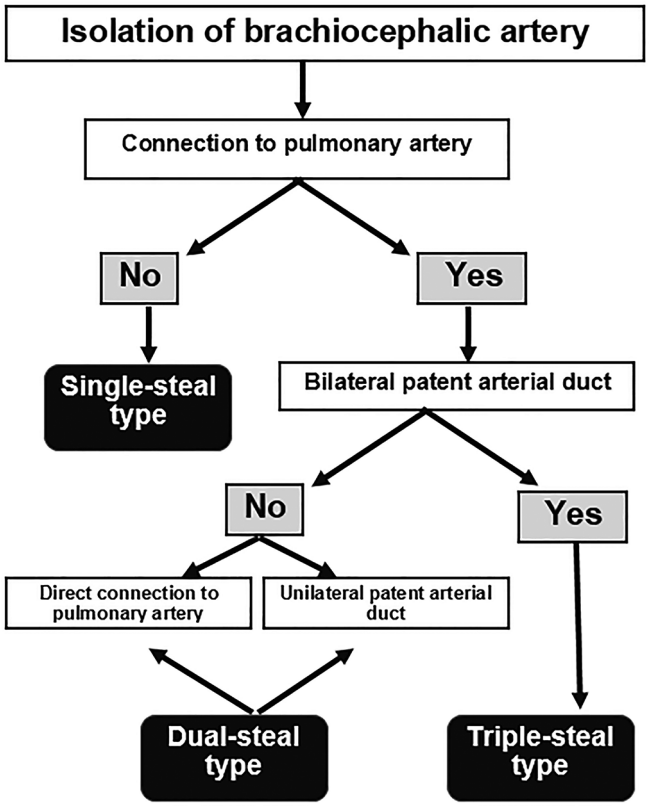

Figure 5: Classification of isolated left brachiocephalic artery

In all reported cases, except two, left hand pulse was either absent or lower than the right side. Of those 2 cases with equal pulses, one was due to origin of the right subclavian artery in the segment with severe coarctation.[8] The pathophysiology of equal pulses in the other case who was a 56-year-old woman with systemic hypertension and history of multiple transient ischemic attacks affecting the right side of the body is not clear.[9]

Therefore, simple palpation of the pulse of the left hand in a child with right aortic arch with two-aortic arch branches can be extremely helpful in early detection of this tricky and extremely rare anomaly.

Compare the blood pressure and the oxygen saturation of the left hand with the right hand

The BP in the left arm was lower than the right in patients with reported BP of both hands. In the 5-year-old girl reported by Delgado and Barturen, the slightly higher BP in the left arm was because of the long-segment coarctation involving the right subclavian artery.[21]

Mild decreased oxygen saturation of the left arm in comparison to the right arm may occur. Oxygen saturation was not reported in 73.3% of cases. In 7 cases with measured oxygen saturation, decreased saturation was seen in 4. The higher left arm oxygen saturation, reported in the case of Miyaji *et al*., represents the unique case of isolation of the right brachiocephalic artery and left aortic arch.[6] Thus, higher BP of the left arm in a patient with ILBA should raise the suspicion to associated lesions. As seen in Table 1, differential size of the left upper arm is rare and is not expected to be present at a very early age.

Auscultate the cranium for cranial bruits in isolation of the left brachiocephalic artery

Bruits in the cranium and in the neck can be detected in all types. However, as in our case, auscultation of bruits does not necessarily indicate a serious abnormality in intracranial arteries and may simply reflect the collateral flow that supplies the left vertebral artery in retrograde.

In summary, a weak radial pulse in a child with right aortic arch and two arch branches, without any history that indicates other etiologies, should very strongly raise the suspicion to the presence of ILBA. Bilateral palpation of radial pulses is an often forgotten and extremely simple physical examination that directs us to the early diagnosis of this extremely rare and tricky diagnosis, with potentially diverse neurologic, cardiac, and left upper extremity manifestations. We propose a simple, novel, and practical anatomic-clinical-prognostic classification that categorizes these patients into three types. As we go from single-steal type to triple-steal type, the prognosis

worsens and the age at presentation decreases. Complex congenital heart disease or mortality is not reported in patients with single-steal type. These patients often present in adulthood with symptoms of vertebral-basilar artery insufficiency or left arm claudication. About one-fourth of patients with the double-steal type have DiGeorge syndrome. Evaluation for microdeletion of 22q11 is recommended in these patients. Triple-steal type is the rarest type and has the earliest presentation with worst prognosis and 100% mortality. Of note, in this subset of patients, the mortality does not result from the steal phenomenon but from the associated complex congenital heart disease. All patients with triple-steal had extracardiac anomaly.

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Conflicts of interest

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