

# Anomalous left brachiocephalic vein: important vascular anomaly concomitant with congenital anomalies and heart diseases

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**Background:** Anomalous left brachiocephalic vein (ALBCV) is a rare and less known systemic venous anomaly. Infrequently, this vein takes an abnormal course and passes to the right behind or beneath the aortic arch to create the superior vena cava (SVC). Its incidence was reported much higher in patients with congenital heart disease especially in conotruncal and aortic arch anomalies. It could be misdiagnosed with normal or abnormal mediastinal structures. It also could make complication during surgeries or invasive strategies. Previously, this anatomical finding has been reported in case reports and there are just few studies evaluating these patients as a group to find other abnormalities

**Materials and methods:** We conducted a retrospective, multicentre study between 2008 and 2014 at three institutions. We reviewed thoracic computed tomography angiography of 1372 patients referred to these centres. The diagnosis of ALBCV was confirmed by an expert radiologist and the imagings were reassessed to identify new cases and concomitant anomalies. We analysed the imagings' details and measured the prevalence of each anomaly.

**Results:** Among the 22 cases of ALBCV, 12 (54.5%) and 10 (45.4%) patients were males and females, respectively, with median age of 12.5 years. Tetralogy of Fallot (TOF) was considered as a most concomitant anomaly with ALBCV (54.5%). Two patients had associated atrial septal defect (ASD) and defined as pentalogy of Fallot. Right-sided aortic arc (RSAA) was detected in 12 (54.5%) patients; mirror image was found in 5 of them. Pure ventricular septal defect or pulmonary stenosis without TOF was recognised in 4 patients. Three cases had isolated overriding aorta (13.6%). In 3 patients, we could find patent ductus arteriosus (13.6%). In 2 (9%) patients, abdominal haemangioma was incidentally diagnosed. Aberrant left retrotracheal subclavian artery was detected in 1 (4.5%) patient. One patient only had isolated ALBCV (4.5%).

*Conclusions:* In our study, ALBCV was frequently seen in association with other congenital anomalies, mostly TOF and RSAA. In patients with pulmonary hypoplasia or aplasia, some parts of lung's blood supply were provided by abnormal aorto-pulmonary connections. For a radiologist, it is important to differentiate this anomaly in cross-sectional imaging from persistent left SVC, partial anomalous pulmonary veins return and an enlarged lymph node. Detection of ALBCV could draw the attention to the more serious heart disease and in isolated forms prevented further evaluations. (Folia Morphol 2017; 76, 1: 51–57)

**Key words:** anomalous left brachiocephalic vein, computed tomography angiography, anomaly

## INTRODUCTION

The brachiocephalic (innominate) veins are the two large veins of the thorax which are formed by the fusion of each corresponding internal jugular and subclavian veins at the level of the sternoclavicular joint. The left brachiocephalic vein (LBCV) is usually longer than the right. This vein generally has a horizontal course. It crosses the anterior mediastinum from left to right to join the right brachiocephalic vein (RBCV) [5]. In its normal course, the LBCV descends obliquely towards the right and after passing over the aortic arch branches, it merges with superior vena cava (SVC) [8].

Thoracic venous anomalies can be categorised to pulmonary and systemic [1]. Anomalous left brachiocephalic vein (ALBCV) is a rare systemic venous anomaly. In this situation, LBCV takes an abnormal course and passes to the right behind or beneath the aortic arch and join the SVC below the orifice of azygos vein [4, 6–8, 16, 17, 19]. This was first described by Kershner [9] more than a hundred years ago. Figure 1 shows the course of normal and anomalous LBCV.

Isolated ALBCVs have no clinical significance and they are just normal variants but it is crucial to diagnose them prior invasive procedures. Unawareness of such anomalies before sternotomy or heart transplantation, could lead to vascular injury, disastrous surgical results and increase the morbidity and mortality [2, 7, 17]. Particularly in patients with right ventricular outflow tract obstruction, it should be considered in order to decide the plan during surgery [10]. Airway compression and obstruction symptoms are reported to be caused by this anomaly. It could make massive bleeding during tracheostomy and thyroidectomy and other neck surgeries [2]. Unawareness of such anomaly may cause technical difficulty for cardiologists while performing transvenous pacemaker inser-

tion. Anaesthesiologists may have problems during placing central venous pressure line and port catheter in a left arm approach [3].

Anomalous left brachiocephalic vein is generally associated with other congenital anomalies [2, 4, 6, 10–14, 18, 19]. Its incidence was reported much higher in patients with congenital heart disease (CHD) compared to the patients without CHD. Detection of ALBCV could draw the attention of the radiologists to the more serious heart disease [11].

Previously, this anatomical finding has been reported in case reports and there is just few studies evaluating these patients as a group to find other abnormalities.

We performed our study to find the new cases of ALBCV in order to evaluate associated and more complicated anomalies. By this way, the clinicians could be aware of it in order to avoid the complications, especially before surgical interventions.

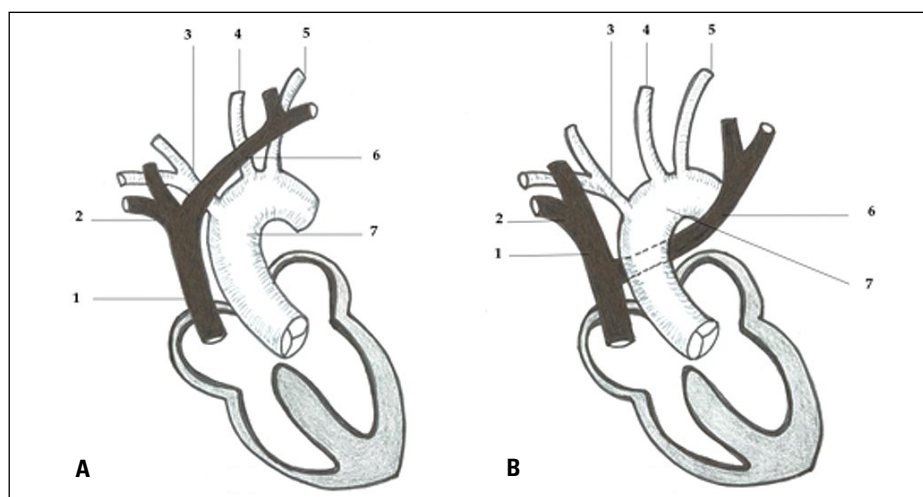
## MATERIALS AND METHODS

We conducted a retrospective, multicentre study between 2008 and 2014 at three institutions (Masih Daneshvari University Hospital, Shahid Rajaie University Hospital and one private referral imaging centre named Tooska). We reviewed thoracic computed tomography (CT) angiography (CTA) of 1372 patients referred to these centres.

All the CT angiographies were done for the diagnostic purposes by the clinicians' decisions and we didn't impose any extra radiation dose or cost or probable risk to the patients.

Each patient had unique code and their identities were preserved secretly.

The proposal of our study was approved by the Ethics Committee of Shahid Beheshti University of Medical Sciences.



**Figure 1.** Normal anatomy (A) and anomalous retro- and subaortic left brachiocephalic vein (B); 1 — superior vena cava; 2 — right brachiocephalic vein; 3 — brachiocephalic artery; 4 — left common carotid artery; 5 — left subclavian artery; 6 — left brachiocephalic vein; 7 — aortic arch.

Multi-section CT angiograms were obtained with a 4-row multi detector CT scanner (General Electric Medical Systems, LightSpeed Q/Xi). Typically, CTA was initiated 15 s to 20 s after the start of IV infusion of non-ionic iodinated contrast material. Contrast material (Iodixanol, [Visipaque 320 mg]) was injected (2 cc/kg) at a rate of 2–4 mL/s, for a total scanning time of 15 s to 20 s, with the use of a power injection and an 18- or 20-gauge needle inserted in the antecubital vein. The volume of iodinated contrast material in each study was typically 200 mL. The scanning parameters included 120 kV, 95 mA and section thickness of 1.3 mm to 2.5 mm. The scan revolution time was 0.5 s. Data for CTA were obtained in a caudocranial direction. Three dimensional reconstructions were performed GE workstation, LightSpeed Q/Xi.

The diagnosis of ALBCV was confirmed by an expert radiologist and the images were reassessed to identify new cases and concomitant anomalies.

We recorded patients' age, sex and then registered the radiologic findings such as overriding of aorta, mirror image, ventricular septal defect (VSD), atrial septal defect (ASD), patent ductus arteriosus (PDA), tetralogy of Fallot (TOF), aorto-pulmonary (AP) connections and main pulmonary artery diameter. We also evaluated the images for other probable anomalies.

## RESULTS

Among the 22 patients with ALBCV, 12 (54.5%) were males and 10 (45.4%) were females with median

age of 12.5 years (ranged 2 months to 40 years, Q1: 3 y, Q3: 20 y, IQR: 17 y).

Tetralogy of Fallot was the most concomitant anomaly with ALBCV. It was seen in 12 (54.5%) patients. Two of TOF patients had associated ASD and were diagnosed with pentalogy of Fallot (POF). Abnormal AP connection and overriding of aorta were seen in 15 (68.2%) cases. Right sided aortic arch was detected in 12 (54.5%) patients; mirror image was found in 5 of them. Pure VSD and pulmonary stenosis anomaly without TOF were considered in 4 patients. Three subjects had isolated overriding aorta (13.6%). In 3 patients, we could find PDA anomaly (13.6%). In 2 (9%) patients, abdominal haemangioma was incidentally diagnosed. Aberrant left retrotracheal subclavian artery was detected in 1 (4.5%) patient. One patient had isolated ALBCV (4.5%) without other cardiovascular abnormality and was investigated with vasculitis symptoms and incidentally this anomaly was found. This patient's age was 40 years. Table 1 shows the frequency of cardiac abnormalities. Detailed findings of each patient could be seen in Table 2. Figures 2 and 3 show CTA of ALBCV.

## DISCUSSION

Anomalous retro-aortic left brachiocephalic vein is an anomaly seen in about 0.5–1.7% of all children with CHD [3, 4, 6, 10, 14]. Herein, we reported the prevalence of cardiovascular anomalies in 22 young patients with ALBCV. Based on the findings of the present study, almost all of the young

**Table 1.** Number and frequency of cardiovascular anomalies in 22 patients with abnormal left brachiocephalic vein

Malformations	Number	Frequency (%)
TOF	12	54.5%
RSAA	12	54.5%
Pure VSD	4	18.1%
Pure PS	4	18.1%
Overriding aorta	3	13.6%
PDA	3	13.6%
ASD (POF)	2	9%
Aberrant RSA	1	4.5%
Isolated ALBCV	1	4.5%

ALBCV — abnormal left brachiocephalic vein; ASD — atrial septum defect; PDA — patent ductus arteriosus; POF — pentalogy of Fallot; PS — pulmonary stenosis; RSA — retrotracheal subclavian artery; RSAA — right sided aortic arc; TOF — tetralogy of Fallot; VSD — ventricular septum defect

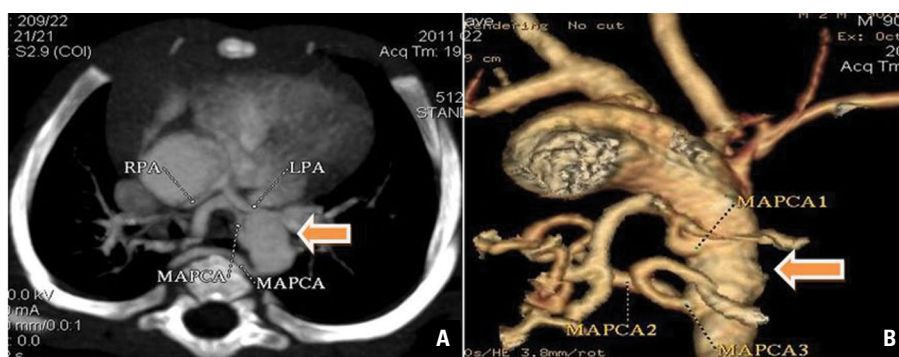
patients with ALBCV are expected to suffer from other concomitant congenital cardiovascular defects. High incidence of other cardiac abnormalities in these patients has been reported previously [4, 6, 10]. In a study published at 2010, Nagashima et al. [14] reviewed surgical records of 4805 patients and found 15 patients with ALBCV. CHD was seen in 14 and there was just 1 patient without other congenital heart anomalies.

The results of the present study showed that the most common anomaly seen in these patients is TOF (54%) and right-sided aortic arch (54%). Presence of TOF in the ALBCV patients is a common finding and varies between 40% and 42% [4, 14], 77% [3], 87.5% [10] and 93% [6] among studies. Although the frequency of TOF in these patients is extremely high, ALBCV is not a common finding in TOF patients

**Table 2.** Detail information of all 22 patients

	Age	Sex	Aortic arc	Mirror image	Over-riding aorta	VSD	RVH	PS	Abnormal AP connection	Other
1	2 m	M	L	-	+	+	-	+	+	-
2	5 m	M	R	-	+	+	+	+	+	-
3	5 m	M	R	+	+	+	-	+	+	Dilated PDA
4	8 m	F	L	-	-	+	-	-	-	2 liver haemangiomas
5	3 y	F	L	-	+	+	+	+	+	ASD, PDA
6	3 y	F	L	-	+	+	+	+	+	-
7	4 y	M	R	+	-	-	-	-	-	-
8	5 y	F	R	-	+	+	+	+	+	Aberrant retrotracheal LSCA
9	6 y	F	R	+	-	+	+	+	+	-
10	6 y	M	R	+	+	+	+	+	+	-
11	7 y	M	L	-	-	-	-	-	+	LVH, RSCA
12	10 y	F	L	-	+	+	+	+	+	Aortic arc
13	17 y	F	R	-	+	+	+	+	+	ASD, PDA
14	18 y	M	L	-	+	+	+	+	+	-
15	18 y	F	R	-	-	-	+	+	-	-
16	19 y	M	R	-	+	+	+	+	+	-
17	20 y	F	L	-	+	+	+	+	+	-
18	22 y	M	R	+	+	+	+	+	-	-
19	24 y	M	L	-	+	+	+	+	+	Liver haemangioma
20	27 y	M	R	-	-	-	-	-	-	-
21	31 y	M	R	-	+	+	-	+	+	-
22	40 y	F	L	-	-	-	-	-	-	Vasculitis (isolated ALBCV)

ALBCV — abnormal left brachiocephalic vein; ASD — atrial septal defect; AP — aorto-pulmonary; F — female; L — left; LSCA — left subclavian artery; LVH — left ventricular hypertrophy; M — male; PDA — patent ductus arteriosus; PS — pulmonary stenosis; R — right; RSCA — right subclavian artery; RVH — right ventricular hypertrophy; VSD — ventricular septal defect; y — year



**Figure 3.** Axial (A) and 3-dimensional (B) reconstructed computed tomography angiography in a 19-year-old man with anomalous left brachiocephalic vein shows multiple major aorto-pulmonary collateral arteries (MAPCA) joining to the descending aorta (arrows) due to right (RPA) and left pulmonary artery (LPA) stricture.



**Figure 2.** Coronal (A) and axial (B) views of a multisecton computed tomography angiography in an 18-year-old man with abnormal left brachiocephalic vein; **A.** The left brachiocephalic vein (LBCV) crosses obliquely to the right beneath the aortic arch (red arrow) and joins to right brachiocephalic vein (blue arrow); **B.** LBCV (curved arrow) running under the aortic arch (thin arrows) which is right sided.

and has been seen in 5% to 8.5% of them [4, 6, 10]. Right sided aortic arch is another abnormality seen with high frequency in ALBCV patients. This aortic abnormality has been reported in ALBCV patients as high as 46% [14], 70% [19], 77% [3] and 86% [10]. In a study by Choi et al. [4] it was shown that the presence of ALBCV in other congenital heart anomalies such as TOF, VSD or pulmonary atresia is associated with higher incidence of right sided aortic arch.

Pulmonary atresia and isolated VSD (not in the context of TOF) were seen in 22.8% of the patients. In previous studies, pulmonary atresia has been reported as 43% [3] and 20% [14].

We incidentally detected 2 new cases of abdominal haemangioma associated with ALBCV. This combination has not been reported in previous studies.

Isolated ALBCV without concomitant cardiac or arch anomalies is tremendously rare [14–17].

This anomaly arises from an abnormal embryological development of precardinal veins at 8<sup>th</sup> week of foetal development. It could be due to the persistence of terminal part of the anterior inferior left cardinal vein. Embryologically, anterior and posterior cardinal veins in each side connect to each other and make common cardinal vein. This vein opens to primitive sinus venosus. Most of the left anterior cardinal veins should disappear and new transverse anastomotic channels by capillary plexus should be made in order to drain the left side of head and neck and left arm. If the aortic arch does not shorten during embryological development, it will prevent the development of transverse venous plexus and leads to formation of an anomalous course of innominate vein [6, 12, 16, 17, 19].

There have been some concomitant abnormalities with ALBCV reported previously that we did not find

in our patients. Bartoli et al. [1] reported the pseudocoarctation of the aorta associated with ALBCV which has never been described before. Choi et al. [4] reviewed 2457 echocardiograms of CHD and found 24 cases of ALBCV. They discovered 1 patient with atrial inversion and a RBCV below the left aortic arch.

The isolated form of ALBCV is usually seen in adults. Srinivasan et al. [17] presented a 68-year-old man who was incidentally diagnosed to have this anomaly on thorax CT without other cardiac or aortic anomalies. Park et al. [16] reported a 63-year-old man of isolated ALBCV that was incidentally detected by performing transoesophageal echocardiography and CT. In isolated form, this abnormality can cause technical problems during interventions through left arm or neck surgery. Also in cardiac surgery, ALBCV needs different surgical strategies; in cardiopulmonary bypass, the SVC should be cannulated more caudally than usual in order to avoid obstruction of retro-aortic brachiocephalic vein or sometimes they need ductal ligation, subclavian to pulmonary artery shunting or operative management of staged single ventricle palliation [2, 10, 17].

On the other hand, ALBCV can obscure the surgical field during surgeries of congenital heart abnormalities. Airway compression and obstruction symptoms are also induced by this anomaly. Budhiraja et al. [2] reported that bleeding is a hazardous consequence of high brachiocephalic vein during tracheostomy and thyroidectomy and other anterior neck surgeries. The isolated form of the disease should also be differentiated from some structures. In non-contrast CT, this anomaly can be misdiagnosed as enlarged lymph nodes [17]. It should also be differentiated from other vascular structures such as left SVC and an atrophic right pulmonary artery [6, 13, 16]. The descending portion of the retro-aortic innominate vein may be mistaken for persistent left SVC or an ascending vertical vein in a total anomalous pulmonary venous connection on echocardiography [10]. The middle portion needs to be differentiated from the central pulmonary artery [2, 4, 13]. The retroaortic segment may be misinterpreted as right pulmonary artery in patients with hypoplasia or atresia in central pulmonary arteries, or an early branching of right upper lobe pulmonary artery on echocardiography [4, 6, 10, 17]. The retroaortic crossing segment of the anomalous brachiocephalic vein may be misinterpreted on unenhanced CT as an enlarged lymph node

[12], an elevated right pulmonary artery in patients with hypoplasia or atresia in central pulmonary arteries, or an early branching right upper lobe pulmonary artery on cross-sectional echocardiography [6, 17, 18]. Carefully tracing this vascular channel through sequential images is the key to differentiate these structures. The data presented in the present study were based on ultrafast CT findings. It is noteworthy that newly developed multiple detector CT, which is widely used in recently, is more capable to differentiate these structures [3].

## CONCLUSIONS

In conclusion, ALBCV is a rare congenital anomaly and is usually associated with other cardiovascular anomalies. This anomaly is due to wrong embryological development of vascular systems. Although the isolated ALBCV does not cause clinical significance, preoperative diagnosis of this anomaly is crucial especially in patients with conotruncal cardiac anomalies. Detection can be easily attained via echocardiography, Doppler sonography and CT.

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