Aberrant Left Brachiocephalic Vein versus Persistent Left Superior Vena Cava without Bridging Vein in Adults: Evaluation on Computed Tomography

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Objective: Persistent left superior vena cava without bridging vein (PLSVC w/o BV) is a common thoracic venous anomaly, while aberrant left brachiocephalic vein (ALBCV) is an uncommon condition. We compared the incidences of the two venous anomalies and assessed congenital cardiovascular diseases (CCDs) in adults using computed tomography (CT).

Materials and Methods: We reviewed the recorded reports or CT images of 49,494 adults for PLSVC w/o BV and ALBCV in two hospitals. We determined incidences of two venous anomalies and the rate of associated CCDs.

Results: 76 PLSVCs w/o BV and 27 ALBCVs were found. The incidence of PLSVC w/o BV was 0.15% and the incidence of ALBCV was 0.055%. PLSVC w/o BV had higher incidence than ALBCV (p<0.001). Four PLSVCs w/o BV and one ALBCV were associated with congenital heart diseases. Two PLSVCs w/o BV and four ALBCVs were associated with congenital aortic arch anomaly (CAA A). ALBCV had higher incidence of associated CAA A than PLSVC w/o BV (P=0.02).

Conclusion: The incidence of ALBCV was <50% that of PLSVC w/o BV. The two venous anomalies found on CT during adulthood were rarely associated with CCDs.

Keywords: persistent left superior vena cava, aberrant left brachiocephalic vein, computed tomography, transverse venous plexus

Introduction

Persistent left superior vena cava (PLSVC) is the most common venous anomaly of the thorax and occurs in approximately 0.3%–0.5% of the general population and in <10% of patients with congenital heart disease (CHD).1–5 There are three types of PLSVC; double superior vena cava (SVC) with and without bridging vein, and PLSVC with absent right SVC.1–5 More than 90% of PLSVCs are double SVCs, and PLSVC without bridging vein (w/o BV) accounts for 60%–70% of double SVCs.2,4 PLSVC is formed from remaining right and left anterior cardinal veins, and PLSVC w/o BV results from regression of both the superior and inferior transverse venous plexuses around the primitive aorta (Fig. 1).1–7

In the first 6 weeks of gestation, the superior and inferior transverse venous plexuses present around the primitive aorta. Then the 8th week of gestation, the inferior transverse venous plexus (TVP) regresses and the superior TVP remains, which finally forms a normal left brachiocephalic vein (BCV) (Fig. 1).8 When the superior TVP regresses and the inferior TVP remains, an aberrant left brachiocephalic vein (ALBCV) develops, and most are retroaortic courses.8–15 ALBCV is rare and its incidence in child patients with CHD has been reported to be 0.98%–1.7%.8,12 The incidence of ALBCV was higher in patients with conotruncal and congenital aortic arch anomaly (CAA A).8,10,11,13–15 There was no report of the incidence on adults including patients with CHD.

Since PLSVCs w/o BV and ALBCVs tend to be associated with congenital cardiovascular diseases (CCDs) including CHD and CAA A, in childhood associated CCDs are problem.

The isolated venous anomalies tend to be regarded sim-
ply as normal variants. However, identification of major thoracic venous anomalies on radiologic examination is important for placing central venous line (CVL) or intra-cardiac electrode/pacemaker.\textsuperscript{2,4,9,12} 

The incidence of ALBCV found in adulthood was not as rare as described in the previous literature.\textsuperscript{14} To the best of our knowledge, there are no reports that compare the incidences of ALBCV and PLSVC w/o BV. Clinical significance of the two venous anomalies in adulthood remains unclear. So, we determined the incidences of two venous anomalies and evaluated associated CCDs in adults on computed tomography (CT).

\begin{figure}[h]
\centering
\includegraphics[width=\textwidth]{figure1.png}
\caption{Schematic diagrams of venous development. \textbf{a}, Venous channels arise from 3 major cardinal veins (the anterior, posterior, and common cardinal veins). Superior and inferior transverse venous plexuses are present around the primitive aorta at the sixth week of gestation. \textbf{b}, Normal development of thoracic veins. Both anterior cardinal veins interconnect with the superior transverse venous plexus. The aortic arch occupies the embryonal space of the inferior transverse venous plexus and a normal left brachiocephalic vein passes anterior to the aortic arch. \textbf{c}, The persistent left superior vena cava occurs because the left anterior cardinal vein remains and there is regression of both the superior and inferior transverse venous plexuses. \textbf{d}, The aberrant left brachiocephalic vein occurs because the inferior transverse venous plexus remains and there is regression of the superior transverse venous plexus. RACV: right anterior cardinal vein; LACV: left anterior cardinal vein; STVP: superior transverse venous plexus; ITVP: inferior transverse venous plexus; RPCV: right posterior cardinal vein; LPCV: left posterior cardinal vein; RCCV: right common cardinal vein; LCCV: left common cardinal vein; RBCV: right brachiocephalic vein; LBCV: left brachiocephalic vein; RSVC: right superior vena cava; LSVC: left superior vena cava; AA: aortic arch; AZ: azygos vein; IVC: inferior vena cava.}
\end{figure}

\section*{Materials and Methods}
This retrospective study was approved by our institutional review board (16R-253).

Our study included 49,494 adults (27,282 males and 22,212 females; mean age: 62 ± 19.1 years) who underwent CT examinations in two university hospitals from January 2013 to July 2018. One was Tokai University Hospital and the other was Tokai University Tokyo Hospital.

In Tokai University Hospital, 42,791 chest CT examinations were performed during the study period. Among these reports made by board-certified radiologists with more than six years of experience each, the patients with ALBCV and PLSVC w/o BV were searched by using the
Congenital Venous Anomalies

We compared the incidences of the venous anomalies and associated CCD using Fisher’s exact test. P values less than 0.05 were defined to indicate statistical significance. Statistical analysis was performed using SPSS version 23 software (IBM Co., Armonk, NY, USA).

Results

A total of 49,494 adults (27,282 males and 22,221 females; mean age: 62.5 ± 16.2 years) underwent chest CT examinations during the study period (Tokai University Hospital: 23,880 males and 18,911 females; mean age: 64.6 ± 16.8 years, Tokai University Tokyo Hospital: 3,402 males and 3,301 females; mean age: 58 ± 16 years). Numbers and incidences of venous anomalies in two hospitals were shown in Table 1.

Seventy-six PLSVC w/o BV patients (45 males and 31 females; mean age: 60 ± 18 years) and 27 ALBCV patients (16 males and 11 females; mean age: 61 ± 15 years) were found. In the PLSVC w/o BV group, 56 patients could be evaluated on contrast-enhanced CT, including 20 CT angiographies. In the ALBCV group, 21 patients could be evaluated on contrast-enhanced CT including eight CT angiographies.

The incidence of PLSVC w/o BV was 0.15% and the incidence of ALBCV was 0.055%. The incidences of PLSVC w/o BV were higher than those of ALBCV (P = 0.001). Four patients with PLSVC w/o BV and one patient with ALBCV had CHDs (Table 2). The incidences of associated CHD were 5.3% in the PLSVC w/o BV group and 3.8% in the ALBCV group. There were no significant differences in the incidences of associated CHD between two groups (P = 0.62). Two patients with Tetralogy of Fallot (TOF) were associated with an ALBCV with right aortic arch with mirror image branching and a PLSVC w/o BV with polysplenia (No. 1 and 3 in Table 2). Five patients with ventricular septal defect (VSD) including the two patients with TOF had reserved operations (Table 2). One patient with ALBCV had CHD and CAAA (No. 1 in Table 2).

Two patients with PLSVC w/o BV and four patients with ALBCV had CAAA (Fig. 2). Two adults of PLSVC w/o BV were associated with incomplete double aortic arches and four adults of ALBCV were associated with

Table 1  Numbers and incidences of venous anomalies in two hospitals

| Patient numbers of two hospitals | Number and Incidence |
|----------------------------------|----------------------|
|                                  | PLSVC w/o BV | ALBCV     |
| Tokai University Hospital (42,791) | 63 (0.15%) | 21 (0.049%) |
| Tokai University Tokyo Hospital (6,703) | 13 (0.19%) | 6 (0.089%) |
| Total (49,494)                     | 76 (0.15%) | 27 (0.055%) |

ALBCV: aberrant left brachiocephalic vein; PLSVC w/o BV: persistent left superior vena cava without bridging vein.
CAA A, including an incomplete double aortic arch, a right aortic arch with mirror image branching, a right aortic arch with aberrant left subclavian artery, and a coarctation. The incidence of associated CAA A was 2.7% in the PLSVC w/o BV group and 15% in the ALBCV group. The ALBCV group had higher incidence of associated CAA A than the PLSVC w/o BV group (P = 0.02).

Fourteen patients with ALBCV had high aortic arches and two patients of PLSVC w/o BV were associated with high level incomplete double aortic arch.

All patients with CHD and CAA A underwent transthoracic echocardiographic examinations.

One patient with Marfan syndrome was found in the PLSVC w/o BV. She had no CHD but had a right tracheal bronchus, left bronchial diverticulum, scoliosis, and lens subluxation.

Seventy-four PLSVCs w/o BV drained into the right atrium via the coronary sinus. On two patients with PLSVC w/o BV including a patient of double outlet right ventricle, PLSVCs drained into the azygos vein via the hemiazygos vein, and they were associated with the left inferior vena cava (No. 4 in Table 2). In three patients with PLSVC w/o BV, pacemaker insertion from the left SVC was not performed (Fig. 3). One of the patients underwent myocardial infarction.

**Table 2** Associated congenital heart disease and others

| No | Venous anomaly       | Age | Sex | Findings                                                                 |
|----|----------------------|-----|-----|--------------------------------------------------------------------------|
| 1  | ALBCV                | 49  | M   | TOF (PS, VSD), patent foramen ovale, RAMI                                |
| 2  | PLSVC w/o BV         | 37  | M   | VSD                                                                      |
| 3  | PLSVC w/o BV         | 34  | M   | TOF (PS, VSD), Absent PV, Polysplenia, absent IVC with AZ continuation   |
| 4  | PLSVC w/o BV         | 27  | F   | Double outlet right ventricle (VSD), left IVC                             |
| 5  | PLSVC w/o BV         | 51  | M   | VSD                                                                      |

ALBCV: aberrant left brachiocephalic vein; PLSVC w/o BV: persistent left superior vena cava without bridging vein; TOF: Tetralogy of Fallot; PS: pulmonary stenosis; VSD: ventricular septal defect; RAMI: right aortic arch with mirror image branching; PV: portal vein; IVC: inferior vena cava; AZ: azygos vein; M: male; F: female

Fig. 2 A case of aberrant left brachiocephalic vein (ALBCV) coexisting with right aortic arch with aberrant left subclavian artery. Axial computed tomography images (a-e) show the right aortic arch with aberrant left subclavian artery and highly enhanced retroaortic left brachiocephalic vein by left hand injection of contrast material (white arrows). The asterisk indicates the Kommerell diverticulum (b). Chest radiograph (f) shows central venous line through the ALBCV (black arrows). The right aortic arch appears in the high position.
dial electrode implantation by the subxiphoid approach. In two patients with the PLSVC w/o BV, central venous catheters were inserted from the left jugular vein into the PLSVC due to malignant lymphoma.

Twenty-six patients with ALBCV had retroaortic left BCVs. In one patient with supraaortic left BCV, an ALBCV coursed between the right brachiocephalic and left common carotid arteries and was associated with PLSVC. Two patients had double left BCVs including normal position and retroaortic left BCVs. Two patients with ALBCV underwent central venous catheter insertions safely (Fig. 2).

**Discussion**

In this study, the incidence of PLSVCs w/o BV in adults determined by CT evaluation was 0.15%, which was similar to the incidence in general population on previous reports.2,3) A high incidence of CHD associated with PLSVC has been determined by angiography or echocardiography in children.3,16,17) Steckiewicz et al. reported that eight adults with PLSVC were found in 4,708 cardiac implantable electronic device recipients and no one was associated with CHD.18) In our study, only four adults with PLSVC w/o BV had CHD. The most common associated CHD is VSD, and others include TOF and pulmonary stenosis.3,6,7,16) In our study, five VSD cases, including two TOF cases, were found. However, PLSVCs detected during adulthood on less-invasive radiological examinations such as CT or MRI are typically asymptomatic. PLSVC without CHD in adults is diagnosed incidentally during CVL placement, and pacemaker and implantable cardioverter defibrillator implantation, and CT examinations.2,6,18)

ALBCV is very rare, and Chen et al. reported that the incidence of ALBCV in 1,812 patients with CHD was approximately 1.7%.8) Kahkouee et al. reported that among 22 patients with ALBCV with a mean age of 12.5 years, only one patient had isolated ALBCV (4.5%).15) Similar to PLSVC, ALBCV found during childhood has a tendency to be associated with CHD.8,12,15,17–19) Kim et al. reported that among 14 patients found in adulthood, only one ALBCV was associated with CHD, and this result was similar to ours.11) In Tokyo Hospital, 19 venous anomalies were not associated with CHD because of small subject size. Yamamura et al. reported that the incidence of ALBCV in adults without CHD found on CT was 0.03%–0.062%.14) In our study, the incidence of ALBCV in adults was 0.055%. The incidence of ALBCV was similar to previous reports and were also not associated with CHD.14)

Our study was the first one to compare the incidences of PLSVC w/o BV and ALBCV in adults using CT, and
results indicate that the incidence of ALBCV was less than half of PLSVC w/o BV. Since PLSVC without right SVC accounted for about 10% of all PLSVC cases, our study indicates that the ALBCV had higher incidence than PLSVC without right SVC.

Simple failure of obliteration of the left anterior cardinal vein results in PLSVC. Failing to obliterate the ligament of Marshall leads to the formation of a left SVC that drains into the coronary sinus. When the superior and inferior TVP around the primitive aorta regress, the PLSVC w/o BV is formed. The embryologic cause of PLSVC is unknown despite the tendency to be associated with CHD.

ALBCV is related to the development of the height of the aortic arch and the prevention of normal shortening of aortic arch during embryogenesis. In our study, 12 CAA As, including high aortic arch, were found in 27 cases of ALBCV. The high aortic arch might be formed from the third pharyngeal arch artery, which differs from the normal aortic arch formed from the fourth pharyngeal arch artery. ALBCV patients had higher incidence of having CAAA than PLSVC w/o BV patients. However, the exact embryogenesis of ALBCV remains unknown.

PLSVC is not associated with a retroaortic left BCV because the proximal part of the retroaortic left BCV, which is a part of the inferior TVP, derives from the distal portion of the left common cardinal vein. In our study, an ALBCV was associated with PLSVC because the supraaortic left BCV formed from the remaining anomalous superior TVP.

In previous studies, most PLSVCs drain into the right atrium via the coronary sinus as our cases did. On rare occasions, the coronary sinus is absent, and the PLSVC drains directly into the right atrium. Additionally, the PLSVC can connect directly or through the pulmonary veins to the left atrium, which causes a right to left shunt. Because an ALBCV that develops from the lower anastomosis is joined by the right BCV to the SVC, the SVC segment has to be shorter than normal, and the azygos vein has to be situated at the same level or higher than the root of the SVC.

An isolated PLSVC or ALBCV is a normal variant, and does not have any clinical significance. However, the exact anatomical diagnoses of PLSVC and ALBCV are important prior to interventional procedures. The technical difficulties associated with PLSVC may lead to the misplacement of a catheter and the vessel wall injury by the catheter. Serious complications, including angina, arrhythmia, cardiogenic shock, and even cardiac arrest have been reported when a guide wire or catheter is manipulated via the PLSVC. In our study, to avoid complications due to PLSVC, cardiac pacemaker placements from right subclavian vein through the right SVC were performed in three patients, and a myocardial electrode implantation was performed by the subxiphoid approach in one patient. Meanwhile, two central catheter insertions and one cardiac pacemaker placement through the ALBCV were performed on patients with ALBCV carefully because the clinicians were informed of the anomalies by CT reports previously. If incorrect insertion into the azygos vein at the same confluence level of the ALBCV can be prevented, CV catheter placement through ALBCV might be safe because download via the ALBCV is more gentle than that via the normal supraaortic left BCV.

Left partial anomalous pulmonary venous return (PAPVR) must be considered differential diagnosis of the PLSVC and ALBCV. Because PAPVR was found to occur in 0.04% of the population in a post-mortem series, more often on the right than on the left, the incidence of ALBCV may be higher than that of left PAPVR, which is frequently detected on CT.

Our study had some limitations. At first, search methods were different in two hospitals: one was key word search method and the other was direct CT image review method. When we used a keyword search of CT reports in Tokai University Hospital, in some cases, the desired venous anomalies might not have been detected. However, the incidences of both venous anomalies were similar to those of previous reports. Second, because not all patients underwent echocardiography, minor CHD, such as a tiny atrial septal defect, could not be detected. Third, not all patients underwent cardiac CT and therefore, we could not evaluate unroofed coronary sinus and coronary sinus atresia, so both coronary diseases coexisted with PLSVC. Finally, the number of patients examined by CT was too small to evaluate associated diseases and a larger study using the direct image review method is necessary to evaluate the incidence, exact anatomy of both venous anomalies and associated complications.

Conclusion

We assessed CT images of PLSVC w/o BV and ALBCV in adults. The incidence of ALBCV was less than half that of PLSVC w/o BV. The two venous anomalies were rarely associated with CHD and CAAA. Identification of those anomalies on CT, even if isolated venous anomalies, are important for correct planning of CVL and cardiac pacemaker placement.

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Disclosure Statement

There are no conflicts of interest to declare.

Author Contributions

Study conception: MK, TI
Data collection: MK, TI
Analysis: MK
Investigation: MK, TI, JK
Writing: MK
Funding acquisition: JK
Critical review and revision: all authors
Final approval of the article: all authors

Accountability for all aspects of the work: all authors

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