Persistence of Left Superior Vena Cava*

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Persistence of a left superior vena cava is a congenital anomaly of considerable interest but by itself exerts no apparent adverse affect on cardiac function. It may occur as an isolated lesion but more often it is associated with many congenital cardiac anomalies. Heretofore it could only be discovered at autopsy or cadaver dissection, but angiocardiography and cardiac catheterization have made possible its recognition during life. This report is a summary of the findings of 11 cases detected by these methods at this center.

Embryology

In early development, the cephalic portion of the embryo is drained by bilateral symmetrical right and left anterior cardinal veins. These join the posterior cardinal veins near the heart and form the right and left ducts of Cuvier which enter the sinus venosus (Figure 1A). During the eighth week of fetal life, an oblique communication forms between the anterior cardinal veins and shunts blood from the left vein to the right. This vessel becomes the left innominate vein. The development of the left innominate vein is followed by atrophy of the left anterior cardinal vein. It normally persists as the first left intercostal vein. The left duct of Cuvier persists as the oblique vein of the left atrium which enters the coronary sinus. The latter structure is derived from the left half of the sinus venosus. The right duct of Cuvier normally becomes the superior vena cava (Figure 1B). The fully developed superior vena caval system is demonstrated in Figure 2.

If the left innominate vein fails to develop, the left anterior cardinal vein persists and continues to drain the left brachiocephalic veins. In this situation it becomes the left superior vena cava. Since the left duct of Cuvier empties into the left half of the sinus venosus which ultimately becomes the coronary sinus, a persistent left superior vena cava drains into this structure (Figure 1B).1-3

Review of Literature

Maude Abbott4 records nine cases of persistent left superior vena cava among 1,000 cases of congenital heart disease. McCotter in 19165 reported three additional cases and upon review of the anatomic literature collected a total of 120 cases. In 1939 Chouke6 thoroughly reviewed the literature and recorded a total of 205 cases. In 1946 Sanders7 added a case and reported the total number to be 215 and estimated the incidence to be 1 in 348 cadavers.

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Aided by grants from the Mallinckrodt Chemical Works, the New York Heart Association and the New York Hospital—Cornell Medical Center Research Fund.
In the angiocardiographic literature, Sussman and Brahms\(^8\) illustrated a case of double superior vena cava and in another publication\(^9\) recorded two instances of coronary sinus visualization with probable reflux filling of the terminal end of a persistent left superior vena cava. Cooley and co-workers\(^10\) reported two cases of persistent left superior vena cava associated with congenital heart disease. A case diagnosed by angiocardiography and cardiac catheterization was also reported from this center.\(^11\) Other cases diagnosed by angiocardiography appear in a recent publication.\(^12\) Cournand and co-workers demonstrated a left superior vena cava in a patient with bilateral superior venae cavae and dextrocardia during cardiac catheterization.\(^13\)

**Classification and Case Reports**

Persistence of the left superior vena cava may be classified into three groups: (1) bilateral superior venae cavae without other congenital cardiac anomalies; (2) bilateral superior venae cavae with associated congenital cardiac anomalies; and (3) absence of the right superior vena cava (Table

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**FIGURE 1A**

*Figure 1*: Embryological development of the vena caval systems.  
A. Early and B. Late. (Modified from Gray's Anatomy\(^1\)).

**FIGURE 2A**

*Figure 2*: A. Angiocardiogram demonstrating normal superior vena cava and tributaries (simultaneous bilateral arm injection).  B. Tracing.
FIGURE 3 (Case 1): Bilateral superior vena cavae and mitral stenosis.
A. Angiocardiogram with simultaneous bilateral arm injection demonstrating bilateral superior vena cavae.—B. Tracing of A. Cardiac catheterization revealed blood oxygen contents in volumes per cent at position 1-13.1; at position 2- 9.6 (46 per cent saturation); at position 3- 15.1.—C. Angiocardiogram, left oblique projection, showing bilateral superior vena cavae.—D. Tracing of C.—E. Angiocardiogram showing the enlarged left atrium of mitral stenosis.—F. Tracing of E.
I). In group I, three cases were encountered and an illustrative case (Case 1) is reported. In group II, seven cases were studied and the wide variety of associated lesions can be noted in the table. The one case in group III is unique in that a right superior vena cava was absent.

Several variations of bilateral superior venae cavae were encountered. The commonest variety consisted of separate bilateral systems with no intercommunication (Figures 3, 5A, 5B). Another variation consisted of a rudimentary intercommunicating innominate vein which allowed opacification during angiocardiography of both superior venae cavae (Figure 4C). Another type showed only a rudimentary left superior vena cava, and most of the blood from the left side passed to the right superior vena cava via a large left innominate vein (Figure 5C). One case showed double azygos veins as well as double superior venae cavae (Figure 5D). This anomaly has been described six times in the anatomical literature.7 A left superior vena cava which drains the entire venous brachiocephalic system

![Figure 4](http://journal.publications.chestnet.org/pdfaccess.ashx?url=/data/journals/chest/21240/)

**FIGURE 4 (Case 5): Complex cyanotic congenital heart disease.**

A. Course of catheter (retouched) through left superior vena cava into right atrium and out into right superior vena cava. Oxygen contents of both right and left superior vena cava blood were 19.3 volumes per cent.—B. Catheter (retouched) passed through interatrial defect into left atrium and a right pulmonary vein.—C. Angiocardiogram showing bilateral superior venae cavae. The left is filled by an intercommunicating rudimentary innominate vein.—D. Later phase demonstrating complex cardiac anomalies with overriding aorta, pulmonary stenosis and intracardiac shunts.
without a corresponding right is seen in Figure 6. Twenty cases of this rare variation have been reported previously.\textsuperscript{14,15}

Illustrative Case (Case 1, Table I):

A 33 year old woman with well documented severe mitral stenosis was admitted for mitral valvulotomy. Cardiac catheterization was performed for preoperative evaluation. The catheter was introduced into the left median basilic vein. In the thorax it was observed to pursue an anomalous course along the left cardiac border from whence it could be directed into the right atrium. Blood samples

\begin{table}
\centering
\caption{Classification of Persistent Left Superior Vena Cava}
\begin{tabular}{|l|l|l|l|}
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\textbf{GROUP I: Bilateral superior vena cavae without other congenital cardiac lesions:} & \\
\hline
Case Number & Age-Sex & Diagnosis & How Demonstrated \\
\hline
(1) 26643 & 33 F & Rheumatic heart disease Mitral stenosis & Cardiac catheterization Angiocardiography (Figure 3) \\
(2) 306716 & 43 F & Syphilitic aortitis & Angiocardiography \\
(3) 552360 & 49 F & Thymoma & Angiocardiography \\
\hline
\textbf{GROUP II: Bilateral superior vena cavae associated with other congenital heart lesions:} & \\
\hline
(4) 561178 & 4 M & Situs inversus Dextracardia Tricuspid atresia Common atrium & Cardiac catheterization \\
(5) 590919 & 4 M & Situs inversus with levocardia Interatrial and interventricular septal defects Infundibular pulmonary stenosis Transposition of great vessels & Cardiac catheterization Angiocardiography (Figure 4) \\
(6) 560278 & 21 F & Single atrium Single ventricle Dextraposition of aorta Congenital mitral stenosis & Angiocardiography (Figure 5D) Autopsy \\
(7) 517038 & 4 M & Tricuspid atresia Interatrial septal defect & Cardiac catheterization (11) \\
(8) 574367 & 3 F & Tetralagy of Fallot & Angiocardiography (Figure 5B) \\
(9) 492621 & 6 F & Tetralagy of Fallot Interatrial septal defect & Angiocardiography (Figure 5C) \\
(10) 498509 & 30 F & Infundibular pulmonic stenosis Post-stenotic dilatation of pulmonic arteries and branches & Angiocardiography (Figure 5A) \\
\hline
\textbf{GROUP III: Persistent left superior vena cava without a right superior vena cava:} & \\
\hline
(11) 619508 & 21 M & No other cardiovascular abnormalities & Angiocardiography (Figure 6) \\
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\end{tabular}
\end{table}
FIGURE 5A. Four angiocardiograms showing persistent left superior vena cava.


B. (Case 8): Left superior vena cava. Tetralogy of Fallot.
C. (Case 9): Complex cyanotic congenital heart disease. Bilateral aygous veins, bilateral superior vena cavae and cor boculare. The right superior vena cava is not seen.
taken from various sites along this course (Figure 3B) revealed oxygen contents compatible with superior vena caval blood (position 1), coronary sinus blood (position 2), and right atrial blood (position 3). These findings established the presence of a left superior vena cava, that terminated in the coronary sinus. No other congenital defects were identified and a hemodynamic pattern typical of mitral stenosis with marked pulmonary vascular sclerosis was observed. Angiocardiography made by simultaneous injection of both arms with contrast materials confirmed the presence of bilateral superior vena cavae (Figures 3A, B, C, D). The typical enlarged left atrium of rheumatic heart disease was also demonstrated (Figures 3E, F). At operation (Dr. Frank Glenn) the anomalous left superior vena cava was observed and, because of its location along the posterior surface of the left atrium, did not interfere with the procedure.

**Comment**

Unlike many congenital cardiac lesions that defy understanding, persistence of a left superior vena cava is a relatively simple development defect. Fortunately, its occurrence results in no complication aside from the fact that it may occasionally simulate widening of the aorta during roentgenography of the heart. The association of persistence of left superior
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vena cava with complex cyanotic types of congenital cardiovascular disease is, however, noteworthy (Figures 4, 5).

Although a left superior vena cava may be suspected on fluoroscopy it can be positively identified in the living patient only by angiocardiography or cardiac catheterization. A left superior vena cava may be missed during angiocardiography unless the left arm veins are used for the injection of contrast substances. Even though 11 cases were found at this center, many more were probably missed because injection of the right arm for angiocardiography is preferred since this simplifies positioning of the patient for left oblique and lateral studies of the heart.17

Catheterization of the heart via the left arm veins, routinely used at this center, affords the opportunity of entering a left superior vena cava. In some of our cases, however, demonstration of the left superior vena cava could be accomplished by catheterization of the right superior vena cava. In these instances the catheter passed through the right superior vena cava, the right atrium, into the coronary sinus, and up the left superior vena cava.

SUMMARY AND CONCLUSION

Eleven cases of persistent left superior vena cava discovered during life by cardiac catheterization and angiocardiography are described and illustrated. By itself, a persistent left superior vena cava produces no detectable effects on the function of the heart but it is often associated with other congenital cardiac anomalies often of a multiple complex type.

RESUMEN

Se describen y se ilustran once casos de vena cava superior izquierda persistente, descubiertos durante la vida por la cateterización cardiaca y la angiocardiografía. Por sí misma la vena izquierda persistente, no produce efecto apreciable en la función del corazón, pero se asocia a menudo con otras anomalías congénitas frecuentemente de un tipo múltiple y complejo.

RESUME

Les auteurs décrivent 11 cas de persistance d'une veine-cave supérieure gauche découverte pendant la vie, par cathétérisme cardiaque. A elle-seule, la persistance de la veine-cave supérieure gauche ne produit aucun trouble perceptible de la fonction cardiaque mais elle est souvent associée avec d'autres anomalies cardiaques congénitales, habituellement complexes.

REFERENCES