Termination of Left Superior Vena Cava in Left Atrium, Atrial Septal Defect, and Absence of Coronary Sinus

A Developmental Complex

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EXCLUSIVE of cases of congenital cardiac disease with asplenia we have observed eight cases in which a persistent left superior vena cava terminated in the left atrium. Pathologic study in each of the five cases in which death occurred suggests that when a left superior vena cava joins the left atrium, this vascular anomaly is part of a developmental complex in which absence of the coronary sinus and a defect in the postero-inferior angle of the atrial septum are also a part.

The primary purpose of this communication is to define the individual cardiac anomalies that form this developmental syndrome. Reference is also made to the clinical, physiologic, radiologic, and developmental features.

Anatomic Characteristics

Study of the five cases in which specimens were available form the basis for the anatomic definitions to be given.

Persistent Left Superior Vena Cava Terminating in the Left Atrium

The left jugular venous system and left subclavian vein joined normally to form a large vein, the left superior vena cava. This large vein descended vertically, passing anterior to the aortic arch and to the left pulmonary hilus (fig. 1a). The vein terminated in the left atrium, between the left pulmonary veins, posteriorly, and the base of the left atrial appendage, anteriorly. The hemiazygos vein joined the left superior vena cava after arching over the left main bronchus in a mirror-image fashion to the junction of the azygos vein with the superior vena cava on the right side. In four cases, an innominate bridge between the two superior venae cavae was absent. In the fifth case (case 5), wherein the right superior vena cava was absent, the right innominate vein crossed the midline to terminate in the left superior vena cava.

Absent Coronary Sinus

In each of the cases studied, the coronary sinus was absent. The ostium of the left superior vena cava was in the left superior aspect of the left atrium between the base of the atrial appendage and the left superior pulmonary vein. Anteriorly and to the left of this ostium a valve-like structure was seen in three cases and a ridge in a fourth case. The cardiac veins drained individually into the corresponding atria.

Atrial Septal Defect

In three cases there were no malformations of the atrioventricular valves, while in two cases the atrioventricular valves showed clefts characteristic of those in persistent common atrioventricular canal.

In three cases without valvular deformities, the atrial septal defect exhibited certain characteristics, to be described. From the right atrial side (fig. 2b and d) the atrial septal defect was in the position normally occupied by the right atrial ostium of the coronary
From this view the size and position of the defect might simply suggest an enlarged ostium of a coronary sinus. The defect was located postero-inferiorly to the fossa ovalis, inferior and medial to the ostium of the inferior vena cava, and above the septal leaflet of the tricuspid valve. Some septal tissue lay between the atrial septal defect and the tricuspid valve. The inferior limb of the fossa ovalis separated the fossa ovalis from the atrial septal defect.

When viewed from the left atrial cavity (figs. 1b, 2a and c), the atrial septal defect was above the postero-medial commissure of the mitral valve, and septal tissue separated the defect from the mitral valve. The defect was bounded anteriorly, superiorly, and inferiorly by atrial septal tissue. Posteriorly, it was bounded by the posterior atrial wall.

The defect, as described, possesses three features that distinguish it from the atrial septal defect of persistent common atrioventricular canal. In the latter condition (fig. 3a), (1) the atrial septal defect is centered over the central portion of the cleft anterior mitral leaflet, (2) no septal tissue separates the atrial septal defect from the atrioventricular valves, and (3) septal tissue is present between the posterior atrial wall and the defect.

In contrast, the defect of the condition described (1) is centered over the posteromedial commissure of the mitral valve, (2) is separated from the atrioventricular valves by septal tissue, but (3) is not separated from the posterior atrial wall by septal tissue.

In two cases the AV valves showed clefts characteristic of those in persistent common atrioventricular canal. In these, the atrial septal defect was larger and had features different from the three cases in which no valvular malformations were present (fig. 3b and c). The atrial septal defect appeared to
occupy in continuity the positions of both the defect considered specific for the entity being described and the atrial septal defect of classical persistent common atroventricular canal.

**Clinical Observations**

The essential clinical findings in the five patients who died and in the three living patients are given in tabular form in table I. Initially, when the five patients on whom necropsy was eventually done were studied clinically, no suspicion of connection of a vena cava with the left atrium was entertained. In one patient (case 3), however, cardiac catheterization initially had been attempted through a left antecubital vein. The cardiac catheter was advanced into the

![Figure 2](image)

**Figure 2**

a and b. Case 3. a. Left side of the heart exposing the interior of the left atrium. Posterior and superior to the mitral valve (arrow) is the atrial septal defect (D) characteristic of the malformation herein described. The left superior vena cava (LSVC) joins the left atrium. The left and right pulmonary veins (LPV; RPV) join the left atrium normally. The interatrial ostium secundum (II) is normal. b. Right side of heart. The atrial septal defect (D) lies anterior and inferior to the entrance of the inferior vena cava (IVC) into the right atrium and postero-inferior to the fossa ovalis (FO). RSVC, right superior vena cava joining right atrium. c and d. Case 1. c. Interior of left atrium and left ventricle. The characteristic atrial septal defect (D). d. Right atrium and ventricle. The probe lies in the entrance of the superior vena cava. Other abbreviations as in b.
left superior vena cava, left atrium, and left ventricle. This was subsequently substantiated by angiocardiography. In the remaining patients the clinical findings had been considered as explained by the septal defects present (atrial septal defect in each case; ventricular septal defect in two).

In retrospect, it is interesting to note that, in all but one (case 4) of the five cases, duskeness of the skin was apparent, even though an atrial septal defect was the only intracardiac defect in two of these four patients (cases 2 and 3). In the latter two cases the levels of oxygen saturation were 89 per cent in the left atrium in one (case 2) and 92 per cent in a femoral artery, in the other (case 3).

Similarly, in the three living patients, the initial clinical impression did not include a diagnosis of a systemic venous anomaly, although mild cyanosis was recorded in two cases (cases 6 and 7). Levels of oxygen saturation of the blood, either in a left-sided chamber or in a femoral artery, were 86, 92, and 93 per cent, respectively.

In two (cases 7 and 8) of the three living patients, the clinical diagnosis of termination of the left superior vena cava in the left atrium was made by cardiac catheterization when the heart was explored by a catheter through one of the left brachial veins. In each, suspicion of such a termination was substantiated by angiocardiography. In the third patient (case 6) the initial cardiac catheterization was done through one of the right-sided veins. In this case, however, the diagnosis of left superior vena cava terminating in the left atrium was made after advancing the tip of the catheter through the left atrial cavity, and then into the left superior vena cava. The venous anomaly was substantiated later by angiocardiography performed through a left
### Table 1

<table>
<thead>
<tr>
<th>Case no.</th>
<th>Age (yr.)</th>
<th>Diagnoses</th>
<th>Clinical observations</th>
<th>Arterial oxygen saturation (per cent)</th>
<th>Special studies (vein used)</th>
<th>Angiocardiogram</th>
</tr>
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<tbody>
<tr>
<td>1</td>
<td>4/12</td>
<td>ASD Absent coronary sinus LSVC to LA VSD</td>
<td>Poor weight gain Dusky Grade I-II systolic murmur Died of pul. infection</td>
<td>—</td>
<td>—</td>
<td>—</td>
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<tr>
<td>2</td>
<td>2</td>
<td>ASD Absent coronary sinus LSVC to LA Urogenital anomalies</td>
<td>Failure to grow Dusky Grade III systolic murmur ASD surgically closed Died of cerebral embolism 8 mos. after operation</td>
<td>LA 89 Right saphenous</td>
<td>Not diagnostic</td>
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<td>3</td>
<td>5</td>
<td>ASD Absent coronary sinus LSVC to LA</td>
<td>Small for age Minimal dishkiness Grade III systolic murmur Died soon after surgical closure of ASD</td>
<td>FA 92 Left basilic catheter in LSVC to LA</td>
<td>Left basilic</td>
<td>Diagnostic</td>
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<td>4</td>
<td>8</td>
<td>Persistent CAV canal Absent coronary sinus LSVC to LA Rt. pul. veins to RA Persistent CAV canal</td>
<td>Poor growth and development Grade III systolic and Grade II diastolic murmurs. Died at conclusion of operation for PC “AV” canal</td>
<td>FA 96 Right saphenous</td>
<td></td>
<td>—</td>
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<td>5</td>
<td>9</td>
<td>Persistent CAV canal Absent coronary sinus LSVC to LA Double outlet RV Absent RVSC Rt. pul. veins to RA Levocardia Persistent CAV canal LSVC to LA Levocardia</td>
<td>Cyanosis Grade III systolic murmur Died at conclusion of operation for PC “AV” canal</td>
<td>FA 79 Left basilic catheter in LSVC to LA</td>
<td>Left basilic</td>
<td>Diagnostic</td>
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<td>Condition</td>
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<td>9</td>
<td>Living</td>
<td>Single atrium, L SVC to LA, Mitral insufficiency, Respiratory distress, Dusky, Grade II systolic and diastolic murmurs, Surgical closure of single atrium. Postoperative murmur of mitral insufficiency, Preop. BA 82, Postop. LA 93, Rt. basilic catheter in RA to LA to L SVC.</td>
<td>Living, ASD? L SVC to LA, Pul. hypert, Mild cyanosis, Grade III systolic murmur, Surgical closure of VSD, Preop. BA 88, Left basilic catheter in L SVC to LA.</td>
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<tr>
<td>7</td>
<td>10</td>
<td>Living</td>
<td>Signs and symptoms of brain abscess, Soft systolic murmur, Surgical closure of ASD, Preop. LA 93, Postop. BA 93, Left basilic catheter in L SVC to LA.</td>
<td></td>
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</table>

ASD, atrial septal defect; L SVC, left superior vena cava; LA, left atrium; VSD, ventricular septal defect; CAV, common atrioventricular canal; RA, right atrium; FA, femoral artery; BA, brachial artery; LV, left ventricle; —, test not done.

Electrocardiographic Observations:

In each of the remaining two patients (cases 1 and 7) with associated atrial septal defects, the electrocardiograms showed right axis deviation and right ventricular hypertrophy. In the last three cases, electrocardiograms were available for study. In the first case, the atrial septal defect was the initial abnormality leading ultimately to a diagnosis of congenital cardiac disease.
Physiologic Observations

Cardiac catheterization was performed in seven of the eight cases. A vein of the right arm was used in three cases, in each of which a transatrial left-to-right shunt was identified. In two of these cases (cases 2 and 4), no unusual course was displayed by the catheter, while in the third case (case 6), the catheter passed from the right atrium through the atrial septal defect into the left atrium and finally into the left superior vena cava.

In each of the four remaining patients (cases 3, 5, 7, and 8), in whom cardiac catheterization was done, the initial catheterization was attempted through a left antecubital vein. In each, the catheter followed a usual course in the thoracic cavity to the level of the left superior mediastinum. At this point, the catheter, instead of crossing to the right side of the superior mediastinum, took an unusual turn and descended in line with the left sternal border to enter the left atrium. From the latter chamber the tip of the catheter was advanced into the left ventricular cavity. In three cases (cases 3, 7, and 8), angiocardiography was performed while the catheter was in the left superior vena cava, and the diagnosis of anomalous termination of this vein in the left atrium was thus confirmed.

Near-simultaneous values for oxygen saturation in both atria were obtained in three subjects (cases 2, 6, and 8). In each of two cases the levels were almost identical. In one case (case 2) the values were 90 per cent in the right atrium and 89 in the left, while in case 6 the values were 85 and 87 per cent, respectively. In the third case the level of oxygen saturation in the right atrium was 89 per cent; in the left atrium, 93 per cent. In each of these cases the left atrial pressure was slightly higher than the right atrial pressure (greatest mean pressure difference being 3 mm. Hg).

In those cases with an intact ventricular septum, the pulmonary arterial pressure was in a normal range. When a ventricular septal defect was also present, the pulmonary pressure was elevated.

The levels of oxygen saturation of blood as measured from a left-sided cardiac chamber or a systemic artery varied from minor degrees of desaturation to values that bordered on the normal (table 1).

Radiologic Observations

Thoracic roentgenograms in each of the eight cases revealed signs consistent with those of a left-to-right shunt, in the form of cardiomegaly and prominent pulmonary vasculature. In those patients with additional cardiac malformations (cases 1, 4, 5, and 7) the signs were more prominent than in the others. In four patients (cases 5, 6, 7, and 8) there was an unusual shadow along the left superior mediastinum corresponding to the left superior vena cava as demonstrated by angio­graphy. The left atrium was not significantly enlarged in any of the cases.

Angiocardiography (fig. 4a and b) was done through a left antecubital vein in four patients (cases 3, 6, 7, and 8). In each, the unusual shadow noted in plain films became opacified and merged with the left atrial cavity above the level of the left hilum.

At the junction between the left superior vena cava and the left atrium, the cava was quite large. After the opacification of the left atrium, the left ventricle and aorta were opacified normally. In one patient (case 7) the right atrium also opacified, indicating a left-to-right shunt at the atrial level. In the lateral views the dilated left superior vena cava seemed to join the left atrium superiorly.

This appearance is quite different from the more common termination of the left superior vena cava in the coronary sinus, in which the junction with the right atrium is much lower and to the right side of the spine.

Developmental Considerations

The developmental complex described in this report appears to result from faulty development in the sinoatrial region of the heart.

Before attempting an explanation for this
DEVELOPMENTAL COMPLEX

Figure 4

Case 7. Forward angiocardiogram made by injection into a left brachial vein in frontal view, a, and in lateral view, b. In addition to opacification of the left superior vena cava (LSVC) there is dense opacification of the left atrium (LA), the left ventricle (LV), and the aorta (A). In this patient the diagnosis of atrial septal defect could not be conclusively proved, although there is suggestion of faint opacification of the right cardiac chambers and the right pulmonary artery, as might occur if an atrial septal defect were present.

condition, it is appropriate first to review the features of normal development of the sinoatrial region of the heart and of related structures.

At the 10-11 somite stage, the central cardiovascular system is represented by paired tubes which, in part, ultimately fuse. The fusion begins at the cephalad end and continues toward the caudal end. Thus, a single tubular heart is formed, retaining its paired identity at the caudal end.

The cephalad end of the single tubular heart is the arterial end, which gives rise to the aortic root. Caudal to the aortic root, in turn, are the conus and the ventricle of the tubular heart. More caudal to the ventricle is the venous end, which is composed of paired atria and paired great veins. The paired venous end completes its fusion in the early phase of 11-20 paired-somite stage to form the single atrial cavity and the sinus venosus.

Tandler\textsuperscript{2} observed that the sinus venosus never loses its paired condition completely. Thus, he identified three parts of the sinus venosus: (1) the unpaired central part or transverse part (between the orifices of the vitelline veins), and (2) right and (3) left horns which, in turn, receive the respective common cardinal and umbilical veins (fig. 5, I).

Davis\textsuperscript{3} observations were that the cephalic migration and growth of the atrium, which starts when the bulboventricular U loop is formed, becomes significant at the 11-20 paired-somite stage.

As the rapid cephalad growth of the common atrium continues, a sulcus begins to develop (fig. 5, II) at the left side of the junction between the sinus venosus and the common atrium. From within the heart, the site of the sulcus is represented by a fold\textsuperscript{4} between the sinus venosus and the left side of the common atrium. With the deepening of the left atrioventricular sulcus, this fold progresses from the left side toward the midline of the common atrium. By the completion of 20 paired-somite stage the atriovenous fold

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separates most of the left side of the common atrium from the sinus venosus. Some communication between the left side of the common atrium and the sinus venosus, however, is still present at this stage (fig. 5, II and III).

On the right side, an atriovenous sulcus also develops. This separates the right side of the common atrium from the sinus venosus. In spite of this, the sinus venosus remains connected with the dorsal part of the entire width of the right side of the common atrium.

The left horn of the sinus venosus diminishes in size and, in concert with this, growth of the transverse portion of the sinus venosus becomes retarded. The diminution in size of the left horn results chiefly from the beginning of obliteration of the left umbilical vein. The right horn begins to receive the hepatic veins and thus enlarges.

Along with the changes at the region of the sinus venosus, the common atrium enlarges significantly and it seems that the right half surpasses the left part in volume. By the abutment of the distal portion of the bulbus cordis against the upper and anterior wall of the common atrium, a blunt prominence is formed, which projects into the atrial cavity. Toward the end of the 20-somite to 6-mm. stage, a sickle-shaped protrusion is formed on the aforementioned intraatrial prominence, the protrusion to be known as the interatrial septum primum.

In embryos between 6 and 9 mm., the sinus venosus is completely separated from the left side of the common atrium by progression of the left atriovenous fold toward the right where, in the vicinity of the midline of the common atrium, the fold fuses with the septum primum.

In separating the left side of the common atrium from the sinus venosus, the left atriovenous fold provides part of the posterior wall of the left atrium. It also acts as the anterior wall of the left side of the sinus venosus, thereby creating the coronary sinus. As the anterior wall of the newly formed coronary sinus is the left atriovenous fold, the posterior wall of the coronary sinus is the tissue of the transverse part and left horn of the sinus venosus (fig. 5, IV).

Following development of the coronary sinus by the process described, the left atrium enlarges. This results in compression.

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**Figure 5**

Diagrammatic portrayal of various stages in the development of the sinus venosus and associated structures. I. At an early stage there is a common atrium (A) which communicates freely with the receiving chamber known as the sinus venosus (S.V.). The latter receives, through its left and right horns, respectively (L.H.; R.H.), the common cardinal vein of the corresponding side (L.C.C.V.; R.C.C.V.). Each common cardinal vein, in turn, receives its corresponding anterior cardinal and posterior cardinal veins (L.A.C.; L.P.C.; R.A.C.; R.P.C.). II. An early indication of partitioning of this portion of the heart into two sides is derived from the appearance of septum primum (S.I.) and the development of a fold (L.A.V. Fold) along the left side of the junction of the common atrium and sinus venosus. III. In the lowermost portion of the atrium, and with further development, the septum primum is approximated by the enlarging left atriovenous fold. At this stage, there is still interatrial communication in the most posterior-inferior angle of the common atrium. IV. With further development of the left atriovenous fold, union is made between the fold and the septum primum, thereby obliterating the communication between the sinus venosus and the left atrium. The posterior aspect of the left atriovenous fold contributes to the anterior wall of the newly developed coronary sinus, while the posterior wall of the left horn of the sinus venosus forms the posterior wall of the coronary sinus. Because of the obliteration of the communication between the sinus venosus and the left atrium, the newly formed structure, namely, the coronary sinus, communicates with the right atrium.
and stretching of the left horn of the sinus venosus and of the left anterior cardinal vein. This obstruction in the anterior cardinal system stimulates development of collateral channels between the left and right anterior cardinal veins, ultimately leading to establishment of the bridge between the two anterior cardinal veins, known as the left innominate vein. The resulting increased blood flow through the right anterior cardinal vein causes the right horn of the sinus venosus to enlarge and to appear to ascend against the posterior wall of the right atrium. Gradually, the cavities of the right horn of the sinus venosus and the right side of the atrium merge to form a common cavity known as the right atrium.

Accounts on development of the atrial septum do not indicate that the left atriovenous fold plays a role in formation of the final atrial septum. From the nature of the anomalies in the entity being considered, it seems probable that the left atriovenous fold, either by contributing to the atrial septum or by supplying a support to the septum primum at its posterior aspect, participates in closing the postero-inferior angle of the interatrial ostium primum.

More anteriorly, the interatrial ostium primum is closed by fusion of the septum primum with the atrioventricular endocardial cushion, a process that is generally recognized.

From the foregoing, it seems that the fundamental developmental abnormality in the condition reported is failure of completion of the left atriovenous fold (fig. 6). With this deficiency, the left side of the sinus venosus would maintain continuity with the left atrium and would be represented grossly as termination of the left superior vena cava in the left atrium. Failure of the full growth of the left atriovenous fold would deny the septum primum an anchor, posteriorly, and account for the characteristic defect of this condition.

Moreover, as the coronary sinus is derived, in part, from the left atriovenous fold, a deficiency of the fold would result in absence of the anterior wall of the coronary sinus.

That part of the posterior wall of the definitive left atrium would be derived from that part of the sinus venosus that normally contributes the posterior wall of the coronary sinus. Furthermore, failure of full growth of the left atriovenous fold may be associated with failure of full development of atrioventricular endocardial cushion tissue, the latter accounting for persistence of the common atrioventricular canal. The association of these two developmental anomalies will result in a common atrial septal defect, part of which is in the position of the atrial septal defect in persistent common atrioventricular canal, and part in the position of the atrial septal defect specific for the anomaly being discussed.

Discussion

Among reported instances of termination of the left superior vena cava in the left

Figure 6

Digrammatic portrayal of the authors' concept as to the basis for the developmental complex here reported, of which the three elements are (1) termination of the left superior vena cava in the left atrium, (2) an atrial septal defect in the postero-inferior angle of the atrial septum, and (3) absence of the coronary sinus. It is envisioned that with incomplete development of the left atriovenous fold, there is failure of union of the fold with the septum primum. Under these circumstances the sinus venosus continues to communicate with the left atrium. As the left superior vena cava has a ready avenue of flow, it remains patent and, in the adult stage, is represented by termination of the left superior vena cava in the left atrium. Failure of union of the left atriovenous fold with the septum primum would explain the presence of an atrial septal defect in the location peculiar for this developmental complex. As the coronary sinus is derived from the normal development of the left atriovenous fold and its junction with the septum primum, failure of the latter process would result in absence of the coronary sinus.
atrium in which necropsy had been done, atrial septal defect is commonly, though not universally, mentioned. When an atrial septal defect is identified, its position is usually not clearly defined. In a case studied clinically by Mankin and associates, an atrial septal defect was considered in association with termination of the left superior vena cava in the left atrium. In that case, the pulmonary veins joined the left superior vena cava.

An atrial septal defect was identified in seven of our eight cases. In the remaining case, an atrial septal defect, although not specifically identified, cannot be excluded.

It is still uncertain whether termination of the left superior vena cava in the left atrium is always associated with an atrial septal defect. Nonetheless, it seems appropriate to consider that a specific developmental complex exists in which the venous anomaly mentioned is associated with a defect in the postero-inferior angle of the atrial septum. This complex appears to be derived from incomplete separation of the sinus venosus from the left atrium.

One of us (J.E.E.) earlier had considered the atrial septal defect of this condition not a true defect of the atrial septum. Rather, it was considered that the anterior wall of the coronary sinus was deficient and therefore allowed an interatrial communication. We are now of the opinion that the interatrial communication in the condition under discussion is to be considered a true atrial septal defect of a specific type and associated with absence of the coronary sinus.

From our material, it is apparent that termination of the left superior vena cava in the left atrium may be associated with persistent common atroioventricular canal (two of eight cases). In such instances, the extensive distribution of the atrial septal defect suggests that the defect represents confluence of the atrial septal defect which is part of persistent common atroioventricular canal and the defect which is specific for the developmental complex here described.

Usually, when the left superior vena cava terminates in the left atrium, the left innominate venous bridge between the two superior vena cavae is absent. This probably reflects absence of obstruction to flow in each superior vena cava. Support for this thesis comes from two cases of termination of the left superior vena cava in the left atrium, in each of which an innominate venous bridge was present. In each of these cases the right superior vena cava was narrow. Obstruction in the latter vessel probably favored the development of collateral flow to the left superior vena cava, the major collateral channel being a developed innominate venous bridge.

Calculations of the venous return to the heart show that under normal conditions about one third of total venous return is through the superior vena cava.

In cases as those herein reported, in which the two superior vena cavae are of about equal size, one may assume that about one sixth of the systemic venous return is to the left atrium. The latter results in desaturation of the left atrial and systemic arterial blood. This may occur to such a degree as to cause cyanosis or duskinsness. Such was observed in three of the four patients (cases 2, 3, and 8) in this series in whom there were no additional intracardiac abnormalities.

In our series, the diagnosis of termination of the left superior vena cava in the left atrium was made only when the heart was explored with a catheter through a left antecubital vein; this was substantiated by angiocardiology. In one case, however, during cardiac catheterization, the catheter was easily advanced from the right atrium to the left and into the left superior vena cava. Later studies through a left antecubital vein substantiated the termination of the left superior vena cava in the left atrium. Angiocardiography was the most reliable procedure with which to establish whether a persistent left superior vena cava terminates in the coronary sinus or in the left atrium. In the present series the atrial septal defect has been successfully surgically closed in three instances. One of these patients (case 2) died 8 months after operation, of central nervous
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system complications; the left superior vena cava had not been interrupted surgically. The other two patients (cases 6 and 8) are living. In each of these cases the arterial oxygen saturation, determined postoperatively, is below normal levels, thus further confirming the drainage of venous blood through the left superior vena cava into the left atrium, even after closure of the atrial septal defect.

In none of the patients in this series in whom surgical closure either of an atrial septal or ventricular septal defect was accomplished had the left superior vena cava been ligated or interrupted.

There are two cases in the literature in which the left superior vena cava was ligated successfully\textsuperscript{11, 12} after confirming the presence of the right superior vena cava and an anastomosing vein between the two superior vena cavae.

The occurrence of complications in the central nervous system in two of our patients, which resulted in death of one, suggests that interruption of the left superior vena cava or anastomosis of this vein with the right superior vena cava should be done to avoid complications of a right-to-left shunt.

Summary

Eight cases of termination of the left superior vena cava in the left atrium are reviewed pathologically and clinically.

In three of the five patients who died, pathologic examination revealed three anomalies which, together, are considered to form a developmental complex. The anomalies are (1) termination of the left superior vena cava in the left atrium, (2) absence of the coronary sinus, and (3) an atrial septal defect lying in the postero-inferior angle of the atrial septum. In the two remaining fatal cases, the aforementioned anomalies were associated with persistent common atrophicventricular canal. In this situation, the atrial septal defect of the latter malformation was confluent with the atrial septal defect of the anomalous complex described.

The coexistence of three anomalies—the left superior vena cava terminating in the left atrium, absent coronary sinus, and atrial septal defect—is considered to result from a single developmental abnormality. This takes the form of failure of complete formation of the left atriovenous fold, that fold which normally develops along the left side of the junction of the sinus venosus and the atrial portion of the heart.

Clinically, features of increased pulmonary blood flow, coupled with dusky skin, were the significant abnormalities.

When cardiac catheterization was attempted through a right-sided vein, the data revealed a left-to-right shunt at atrial level, while levels of arterial oxygen desaturation were present.

Except in cases with coexistent ventricular septal defect, pulmonary hypertension was absent.

In each of the four patients in whom cardiac catheterization was performed through a left antecubital vein, the catheter was advanced into the left superior vena cava, left atrium, and left ventricle. This was substantiated by angiography in three of the four patients. In our experience, angiography was the most reliable procedure to substantiate the termination of the left superior vena cava in the left atrium.

The present study suggests that, in the absence of pulmonary hypertension, a left-to-right transatrial shunt associated with systemic arterial oxygen desaturation may indicate the presence of a persistent left superior vena cava terminating in the left atrium. Moreover, an atrial septal defect identified surgically as in the postero-inferior angle of the atrial septum should suggest that an additional anomaly may be present in the form of termination of the left superior vena cava in the left atrium.

References

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Famous General Practitioners

One reason why the famous achievements of general practitioners have been unrecognized or overlooked comes from the very success they achieved, for the fame resulting from an important discovery has often compelled a transfer from general to consultant work. It tends to be forgotten that the essential research which brought fame was carried out while the man was still in general practice.—Zachary Cope, Kt. Some Famous General Practitioners and other Medical Historical Essays. London, Pitman Medical Publishing Co., Ltd., 1961, p. 1.
Termination of Left Superior Vena Cava in Left Atrium, Atrial Septal Defect, and Absence of Coronary Sinus: A Developmental Complex
GUNAY RAGHIB, HERBERT D. RUTTENBERG, RAY C. ANDERSON, KURT AMPLATZ, PAUL ADAMS, JR. and JESSE E. EDWARDS

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