The Lateral Chest Film as an Unreliable Indicator of Azygos Continuation of the Inferior Vena Cava

RonalD J. O'Reilly, M.D., and J. H. Grollman, Jr., M.D.

Summary Recognition of azygos continuation of the inferior vena cava (AC-IVC) is necessary to prevent unnecessary surgery, to modify the approach to right heart catheterization and to avoid potentially fatal azygos ligation at the time of thoracic surgery. Absence of the inferior vena cava (IVC) shadow on the lateral chest film has been claimed to be an important feature of AC-IVC. Our findings, however, indicate that AC-IVC is frequently associated with a normal appearing IVC shadow on lateral chest film study. When the findings on frontal chest film examination suggest AC-IVC, a caval shadow on the lateral view does not exclude the diagnosis. There persists a need for further radiographic evaluation and, if necessary, angiographic confirmation. The lateral chest film does not provide reliable evidence for or against the diagnosis of azygos continuation of the inferior vena cava.

Azygos Continuation of the inferior vena cava (AC-IVC) is a relatively infrequent cardiovascular developmental abnormality occurring both in association with congenital heart disease and as an isolated anomaly of no hemodynamic importance. Since AC-IVC presents as a right mediastinal mass density, recognition is critical to prevent unnecessary tumor work-up and thoracic exploration. Diagnosis is necessary prior to cardiac catheterization since this anomaly contraindicates the femoral route to the right heart. Preoperative recognition of AC-IVC is important before certain thoracic and abdominal procedures. Sacrifice of the azygos vein may prove fatal in AC-IVC. The approach to portocaval decompression surgery may have to be modified in these patients. Finally, the presence of AC-IVC may give a clue to possible underlying congenital heart disease and/or situs abnormalities. Heller and others state that absence of the IVC shadow on the lateral chest film is an important sign associated with azygos continuation. The purpose of this paper is to report six patients with proven AC-IVC who demonstrate an inferior vena caval shadow on lateral chest radiography. The unreliability of the lateral chest film to either confirm or exclude this venous abnormality will be stressed.

Methods and Results

Ten patients with angiographically proven AC-IVC were reviewed. Adequate lateral chest films were available in seven patients (table 1). The chest films of 100 consecutive patients with no congenital cardiovascular abnormality were also studied. The presence or absence of an IVC shadow on lateral chest film examination of both the AC-IVC group and the group without AC-IVC was recorded. In five of the seven cases of AC-IVC, an inferior vena caval shadow was clearly recognized on lateral chest film study. In one patient, a caval shadow was faintly visualized (fig. 1).

Conversely, no IVC shadow was identified on two of the normal chest films of the 100 patients without this venous anomaly. In seven patients without AC-IVC, the IVC line shadow was poorly seen or absent due to adjacent diaphragmatic pleural or parenchymal abnormality.

The vessel responsible for the IVC silhouette was surgically or angiographically demonstrated in four of the patients with AC-IVC. In Case 1, the surgeon described an inferior vena cava at operation for ASD repair. In Case 6, visualization was obtained via reflux from right atrial injection (fig. 2). In two cases (3 and 4) (fig. 3) the vessel was opacified by selective catheterization of the inferior vena cava.

Embryology

The anatomy and embryology of AC-IVC have been well described by Anderson, Chuang and Hollinshead. Based on embryologic development, the normal IVC can be classified into four segments: (hepatic, prerenal, renal and postrenal) (fig. 4). The hepatic segment can be further divided into its cranial and caudal extensions (table 2). It is the caudal extension which in normal development fuses with the prerenal segment to provide caval continuity. The cranial extension receives the hepatic veins and drains into the right atrium.

AC-IVC results from interruption or failure of fusion of the prerenal segment and the caudal extension of the hepatic segment. As a result, blood flow from the postrenal and renal segments of the IVC is diverted into the azygos system.

Unfortunately, AC-IVC has frequently been referred to as absence of the IVC or absence of the hepatic segment of the IVC. We concur with Anderson that this terminology is somewhat misleading. In both normal development and in AC-IVC, a vessel is present which receives the hepatic veins, courses through the caval hiatus and communicates with the right atrium. In previous descriptions of AC-IVC, this vascular structure has been called the hepatic vein, the confluence of the hepatic veins or the IVC (cranial component of the hepatic segment). The hepatic veins and the hepatic segment of the IVC have similar embryologic origin from the primitive omphalomesenteric system. Further, the hepatic segment of the IVC is derived from the right hepatic
vein. In this paper we use the term hepatic IVC segment to further emphasize that the basic embryologic defect represents abnormal fusion rather than a congenital absence of the entire hepatic segment of the IVC.

Discussion

The cranial component of the hepatic segment of the IVC is present in AC-IVC. Surgery and angiography have verified that it is this vascular structure which accounts for the IVC silhouette which may be present on lateral chest film studies of patients with this anomaly.

Surgical descriptions reflect the lack of uniformity in the vascular nomenclature at the right atrial-IVC junction in patients with AC-IVC. Berdon and Baker report a case of AC-IVC in which the surgeon initially thought he saw a normal IVC. “On closer inspection,” however, the vessel was described as a “confluent hepatic vein.” As in our Case 1, Stern and Bloomberg reported a patient with AC-IVC in whom an “inferior vena cava” was described entering the right atrium. Peterson noted that while an “IVC” was present, it was “quite small in comparison to its usual size.” Likewise, Chapman and Bogedain described vessels entering the right atrium at the usual location of the IVC which were “hypoplastic.” Since the hepatic segment in AC-IVC does not receive significant flow from the extra-hepatic IVC, it is not surprising that the vessel has frequently been described as “hypoplastic” or of small caliber.

Angiographic opacification of the hepatic segment was provided by right atrial reflux in Case 6 (fig. 2). An illustration in the paper by Vaughan et al. shows similar reflux opacification of the hepatic segment in a patient with AC-IVC. These cases clearly demonstrate the anatomic basis of the lateral chest IVC shadow in AC-IVC.

To our knowledge, the selective venograms performed in Cases 3 and 4 (fig. 3) represent the first reported antegrade hepatic segment visualization and the first angiographically documented communication between the hepatic and extrahepatic segments of the IVC in AC-IVC. Selective catheter position was critical since initial cavagrams obtained from iliac vein injection in these patients did not show caval communication with the hepatic segment. The fact that

<table>
<thead>
<tr>
<th>Patient</th>
<th>Age/Sex</th>
<th>Associated cardiovascular abnormality</th>
<th>IVC shadow present on lateral chest film</th>
</tr>
</thead>
<tbody>
<tr>
<td>1) S.C.</td>
<td>23/F</td>
<td>ASD and partial anomalous pulmonary venous return from left upper lobe.</td>
<td>Yes</td>
</tr>
<tr>
<td>2) D.F.</td>
<td>27/M</td>
<td>None</td>
<td>Yes</td>
</tr>
<tr>
<td>3) P.S.</td>
<td>20/F</td>
<td>None</td>
<td>Yes</td>
</tr>
<tr>
<td>4) E.S.</td>
<td>11/F</td>
<td>None</td>
<td>Yes</td>
</tr>
<tr>
<td>5) D.D.</td>
<td>30/M</td>
<td>No associated congenital abnormality: Iliac vein thrombosis.</td>
<td>Yes</td>
</tr>
<tr>
<td>6) D.G.</td>
<td>10/F</td>
<td>ASD</td>
<td>Probable</td>
</tr>
<tr>
<td>7) M.B.</td>
<td>5/F</td>
<td>Situs inversus. Pulmonic valvular stenosis.</td>
<td>No</td>
</tr>
</tbody>
</table>

Figure 1. Lateral chest projections of three representative patients (Cases 1, 3, 6) with proven AC-IVC in which an inferior vena caval shadow is seen (arrows).

Figure 2. Lateral angiocardiogram in Case 6 (fig. 1 right) showing reflux opacification of the hepatic IVC segment.
venography in previously reported cases of AC-IVC has not
followed selective catheterization may explain the failure to
recognize this caval communication angiographically. Thus,
more often than realized, there may be a small, extra-
hepatic contribution to hepatic segment flow in this condi-
tion. The fact that the hepatic-extra-hepatic communication
could only be demonstrated by selective caval catheteri-
zation would suggest minimal functional continuity between
the two systems. Our cases do show that a potential caval
collateral pathway through the hepatic segment is possible
in AC-IVC. Effler's description1 of a fatality following azygos
ligation in AC-IVC emphasizes, however, that the adequacy
of this potential collateral route can in no way be assumed.

A number of plain film signs of AC-IVC have been
described on the frontal chest study. The dilated azygos
presents as a convex density at the tracheal-right bronchus
angle (fig. 5). Dilation of the ascending azygos segment may
result in deviation of the right paravertebral pleura which
can be determined from Bucky technique films or laminog-
raphy.4 The azygos silhouette increases in size as the patient
assumes the supine from the upright position. Fluoroscopic
findings (i.e., decrease in size with Valsalva and increase
in size with Muller maneuvers) may further aid in evalua-

Fifteen cases of AC-IVC have been reported in which no

<table>
<thead>
<tr>
<th>IVC segment</th>
<th>Boundaries</th>
<th>Derivation</th>
</tr>
</thead>
<tbody>
<tr>
<td>Cranial Extension</td>
<td>From level of hepatic veins → through caval hiatus → into right atrium</td>
<td>Hepatic sinusoids → Hepatic vein → hepatic segment</td>
</tr>
<tr>
<td>Hepatic</td>
<td></td>
<td></td>
</tr>
<tr>
<td>Caudal Extension</td>
<td>From pre-renal segment → level of hepatic veins</td>
<td>Subcardinal veins → pre-renal segment</td>
</tr>
<tr>
<td>Pre-renal</td>
<td>From level of renal veins to the caudal extension of the hepatic segment</td>
<td></td>
</tr>
<tr>
<td>Renal</td>
<td>Level of renal veins</td>
<td>Subcardinal-Supracardinal venous anastomosis → renal segment</td>
</tr>
<tr>
<td>Post-renal</td>
<td>From level of iliac veins to level of renal veins</td>
<td>Supracardinal vein → post-renal segment</td>
</tr>
</tbody>
</table>
IVC shadow was evident on lateral chest film study (table 3; fig. 6). The "absent caval sign" has been claimed as an important feature of AC-IVC.

Our findings indicate that the presence of an IVC shadow on lateral chest study does not exclude the diagnosis of AC-IVC. Conversely, the absence of the IVC silhouette does not imply the diagnosis. As stressed by Hoffman and Rigler, the IVC line shadow may be obscured by abnormal adjacent diaphragmatic pleural or parenchymal densities and may not be visible in a small number of normal persons. Their observations are confirmed in our small random study of a patient population without AC-IVC. Diaphragmatic elevation, pleural effusion, pulmonary basal atelectasis, infiltrate or fibrosis often obscure the silhouette of the IVC. In 2% of our patients with normal chest films, the IVC could not be identified.

It is as important to be aware of the limitations of X-ray signs as it is to know the signs themselves. We agree with Heller and others that in AC-IVC there may be absence of the IVC silhouette on lateral chest film study (fig. 6). However, the usefulness of the absent caval sign in our experience has been limited. The hepatic segment of the IVC (cranial component) is present in AC-IVC. Our findings indicate that even a hypoplastic segment may be indistinguishable from a normal IVC on lateral chest film examination.

When the findings on frontal chest study suggest AC-IVC,
a caval shadow on the lateral chest view does not rule out the diagnosis. There persists the need for further radiographic evaluation and, if necessary, angiographic confirmation.

In Effler's description of a surgical fatality which resulted from failure to diagnose AC-IVC, he states, “inspection of the right diaphragm” was performed at surgery and “a normal vessel entering the right auricle” was observed. In retrospect, he concluded that the “presence of the hepatic vein prompted the erroneous diagnosis.” To avoid a similar erroneous diagnosis, an IVC shadow on lateral chest film study should not exclude consideration of azygos continuation of the inferior vena cava.

Acknowledgment

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References

2. Edwards EA: Clinical anatomy of lesser variations of the inferior vena cava, and a proposal for classifying the anomalies of this vessel. Angiology 2: 85, 1951
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