Visceroatrial Situs Abnormalities: Sonographic and Computed Tomographic Appearance

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Twenty patients with visceroatrial situs abnormalities were examined by sonography or computed tomography (CT). Eighteen patients underwent cardiac catheterization. Left isomerism (polysplenia syndrome) was found in seven patients, right isomerism (asplenia syndrome) in eight, and total situs inversus in five. The abdominal features of polysplenia include interruption of the inferior vena cava with azygous/hemiazygous continuation (100%) and multiple splenules. The diagnostic signs of asplenia include an inferior vena cava and aorta that course together on the same side of the spine with inferior vena cava–atrial communication (100%). In each case, the key differential feature relates to the major abdominal vessels. Sonography or CT examination in cases of situs ambiguus may reveal diagnostic features that can be used with radiographic signs to accurately diagnose the visceroatrial situs.

The lungs, liver, spleen, stomach, and atria are usually asymmetric organs and the term *situs* is used to define the position of the atria and viscera relative to the midline or sagital plane. *Situs solitus* indicates the normal position of the atria and viscera and *situs inversus* the mirror image location. *Situs ambiguus* is a third and abnormal type of situs in which the relationship of the atria and viscera is inconsistent. In this type of situs abnormality, structures that are usually asymmetric can be symmetric (i.e., both sides of the viscera, lungs, and atria are nearly identical to one another). *Isomerism* is a term used to describe symmetric morphology. *Asplenia* is a syndrome characterized by situs ambiguus with bilateral right-sidedness (right isomerism). *Polysplenia syndrome* is a second situs ambiguus characterized by bilateral left-sidedness (left isomerism) [1–6].

Abdominal sonography and computed tomography (CT) can be useful in the diagnosis of visceroatrial situs abnormalities. In some instances, sonography and/or CT may be the initial diagnostic procedure. These heterotaxic patterns, that is, anomalous placement of organs and major blood vessels within the abdomen, should be recognized as indicating an abnormal situs and not be confused with other pathology.

Subjects and Methods

During a 2 year period, 20 patients with visceroatrial situs abnormalities were studied by sonography or CT in correlation with chest and abdominal radiographs. In 18 of the 20 patients, findings at diagnostic cardic catheterization [7] were also correlated (table 1). All patients were examined by sonography using a Picker 80L B-mode scanner with real-time capabilities, and a few patients were examined with two-dimensional sonography using a phased-array Varian V-3400 sector scanner with a 3.5 MHz transducer. Several patients were examined by CT using a GE 8800 CT/T scanner. The patients were 1 day to 26 years old. Sedation with rectal Pentothal (sodium thiopental, rectal suspension) (25 mg/kg) was used occasionally for CT and sonography of children under age 3 years [8]. In six patients, radionuclide liver-spleen scans were obtained.

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TABLE 1: **Clinical and Anatomic Features of Visceroatrial Situs Abnormalities**

Abnormality: Case No. (age, gender)	Cardiovascular Anomalies	Abdominal Vessels	Visceral Heterotaxia	Clinical Course
Left isomerism (polysplenia syndrome):				
1 (3y, M)	Levocardia post repair; partial AV canal	Azygous cont of IVC to R SVC; aorta on L	Liver on L; stomach on R; gallbladder midline	A&W
2 (6w, M)	Levocardia: complete AV canal	Hemiazygous cont of IVC to L SVC; aorta on L	Liver on L; stomach on R	A&W
3 (1w, M)	Dextrocardia: univentricular heart, common atrium, and common AV valve; interruption of aortic arch, type B	Hemiazygous cont of IVC to L SVC	Liver on R; stomach on L	Deceased
4 (6d, F)	Levocardia: univentricular heart, common atrium, and common AV valve; interruption of aortic arch, type B	Azygous cont of IVC to R SVC	Liver on L; stomach on R	Deceased
5 (26y, F)	Levocardia	Azygous cont of IVC to R SVC; aorta on L	Liver on L; stomach on R; gallbladder on R	A&W
6 (6y, M)	Levocardia, VSD	Azygous cont of IVC to R SVC; aorta on L	Liver on L; stomach on R; gallbladder on L	A&W
7 (2y, F)	Levocardia, ?VSD	Azygous cont of IVC to R SVC; aorta on L	Liver on R; stomach on L; gallbladder on R	A&W
Right isomerism (asplenia syndrome):				
8 (4y, F)	Levocardia: DORV with PS; TAPVC to R atrium; common AV valve	IVC on L; R arch crosses to L	R arch, type I; liver on L; stomach on R	A&W; L B-T shunt
9 (3d, M)	Dextrocardia: DORV with large VSD and PS, common atrium, and common AV valve	IVC or R; R aortic arch	R arch; liver midline; stomach midline	Deceased; postmortem at 9 m
10 (4y, M)	Dextrocardia: univentricular heart with PS, common atrium, and common AV valve	IVC crosses to R; R aortic arch	R arch, type I; liver on L; stomach on L	Deceased; postmortem
11 (14m, M)	Levocardia: univentricular heart, common atrium, and common AV valve	IVC on L; aorta on L	L arch; liver on L; stomach on L	A&W
12 (10m, F)	Dextrocardia: TGA (AV conc, VA disc); pul atresia; multiple VSDs; common atrium	IVC on L; aorta on L	L arch; liver on L; stomach on R	Deceased
13 (4y, M)	Levocardia: univentricular heart; pul atresia	IVC on L; aorta on L	L arch; liver on L; stomach on R	A&W; R B-T shunt
14 (3y, M)	Levocardia: univentricular heart; pul atresia; PAPVR; two AV valves	IVC on R; aorta on R	R arch, type I; liver on R; stomach on L	A&W; L B-T shunt
15 (1d, F)	Levocardia: univentricular heart; pul atresia; common AV valve	IVC crosses to L; aorta on L	L arch; liver midline; stomach on R	Deceased
Situs inversus:				
16 (6y, M)	Univentricular heart; two AV valves	IVC on L; aorta on R	Liver on L; stomach on R; spleen on R	A&W; R B-T shunt
17 (5y, F)	AV disc; VA disc with pul atresia; VSD (ventricular inversion)	IVC on L; aorta on R	Liver on L; stomach on R	A&W; R Waterson shunt
18 (4y, F)	AV disc; VA disc with pul atresia; VSD (ventricular inversion)	IVC on L; aorta on R	Liver on L; stomach on R; spleen on R	A&W; L B-T shunt
19 (6y, M)	Univentricular heart: pul atresia; two AV valves; accessory chamber	IVC on L; aorta on R	Liver on L; stomach on R	A&W; R B-T shunt
20 (5y, M)	Levocardia: AV disc; VA disc with pul atresia; VSD	IVC on L; aorta on R	Liver on L; stomach on R; spleen on R	A&W; R Waterson shunt, L B-T shunt

Note.—y = year; w = week; d = day; AV = atrioventricular; VSD = ventricular septal defect; cont = continuation; IVC = inferior vena cava; SVC = superior vena cava; R = right; L = left; A&W = alive and well; DORV = double outlet right ventricle; TAPVC = total anomalous pulmonary venous connection; PS = pulmonic stenosis; TGA (AV conc) = atrioventricular concordance; VA disc = ventriculoarterial discordance (complete transposition of the great arteries); pul = pulmonary; B-T shunt = Blalock-Taussig shunt; R arch, type I = right aortic arch type I with mirror image branching; PAPVR = partial anomalous pulmonary venous return; AV disc = atrioventricular discordance; VA disc = ventriculoarterial discordance (corrected transposition of the great arteries).

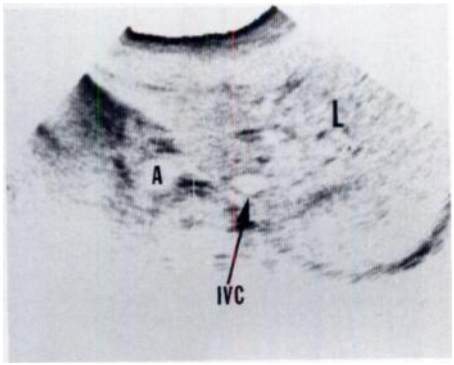


Fig. 1.—Total situs inversus, case 19, 6-year-old boy, transverse sonogram. Left-sided liver (L) and left-sided inferior vena cava (IVC, *arrow*). Aorta (A) to right of spine.

Results

Situs Inversus

The five patients with total situs inversus (table 1) demonstrated mirror imaging on chest and abdominal radiographs. The sonographic examination in each case showed mirror image positioning of the abdominal viscera and vessels. In each case, the liver was located on the patient's left with the inferior vena cava to the left of the spine and the aorta descending to the right of the spine (fig. 1). In each case, the stomach was located on the patient's right, and, in three cases, sonography demonstrated the presence of a spleen in the patient's right upper quadrant (cases 17, 19, and 20). These five cases had severe cardiac anomalies,

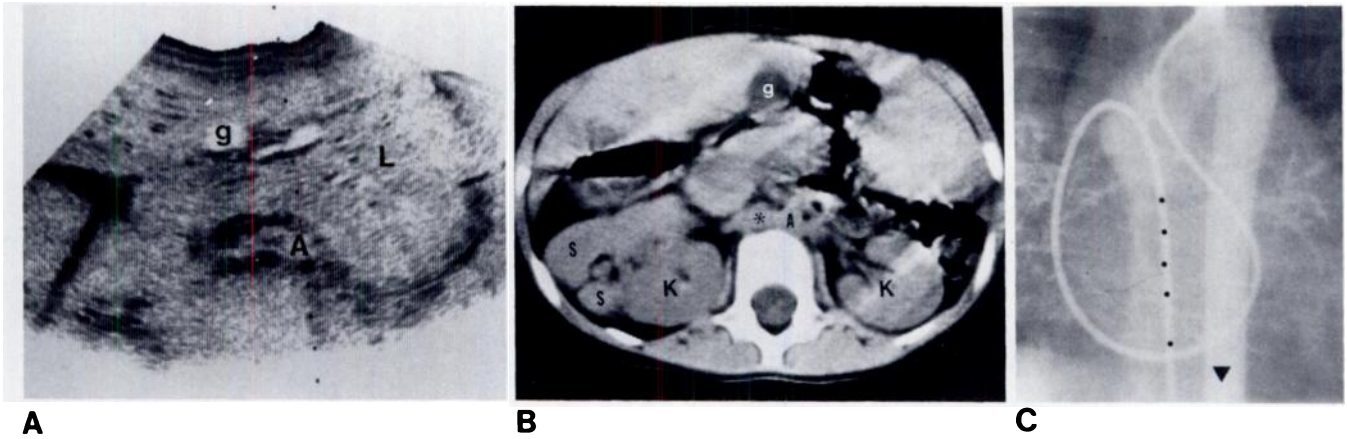


Fig. 2.—Left isomerism (polysplenia syndrome), case 1, 3-year-old boy. **A**, Transverse sonogram. Liver (L) to left with aorta (A) to left of spine. Inferior vena cava not seen; independent drainage of hepatic veins into atrium. Midline gallbladder (g). **B**, CT scan without contrast enhancement. Transverse liver and midline gallbladder (g). Structures thought to represent splenules (S). Aorta (A) with retrocrural azygous vein (*asterisk*). Kidneys (K). **C**, Correlation with vascular anatomy initial cardiac catheterization. Femoral vein catheter (*dots*) passes through azygous vein with contrast material injected into normal descending left-sided aorta (*arrowhead*).

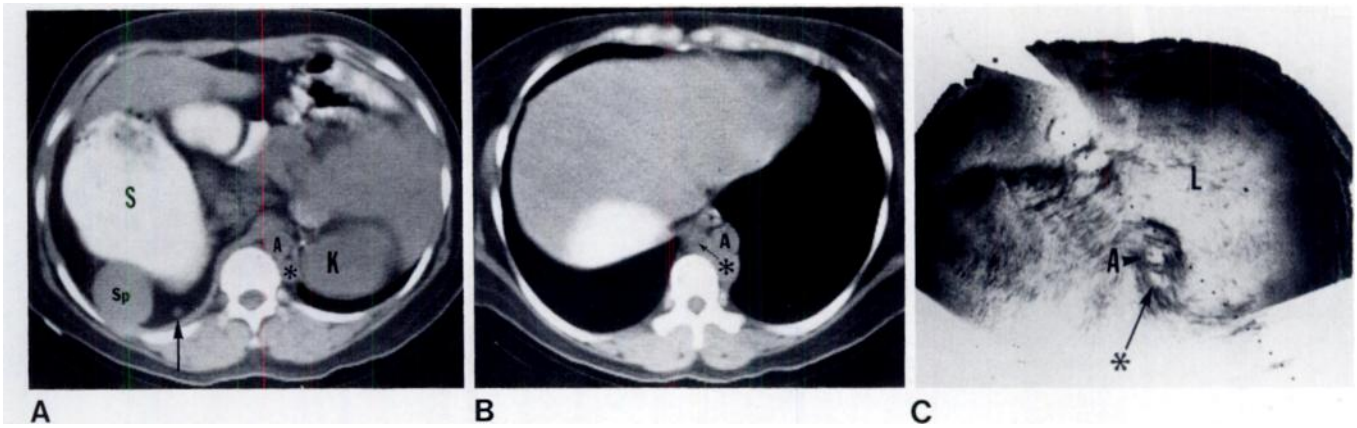


Fig. 3.—Left isomerism (polysplenia syndrome), Case 5, 26-year-old woman with right upper quadrant abdominal pain. **A**, CT scan with oral contrast. Right-sided stomach (S) with three structures thought to represent multiple splenules (Sp, *arrow*) in right upper quadrant. Liver predominantly in left upper quadrant. Aorta (A) on left but no inferior vena cava in usual position adjacent to caudate lobe of liver and anterior to crus of diaphragm. Instead, there is retrocrural left-sided vessel (*asterisk*); also, left kidney (K). **B**, Higher in abdomen. Course of hemiazygous vein (*asterisk*) as it crosses behind aorta (A) from left to right (*arrow*) to form azygous vein, which ascends to enter right superior vena cava. **C**, Vascular anatomy on comparable sonogram. Aorta (A, *arrowhead*) and hemiazygous vein (asterisk, *arrow*).

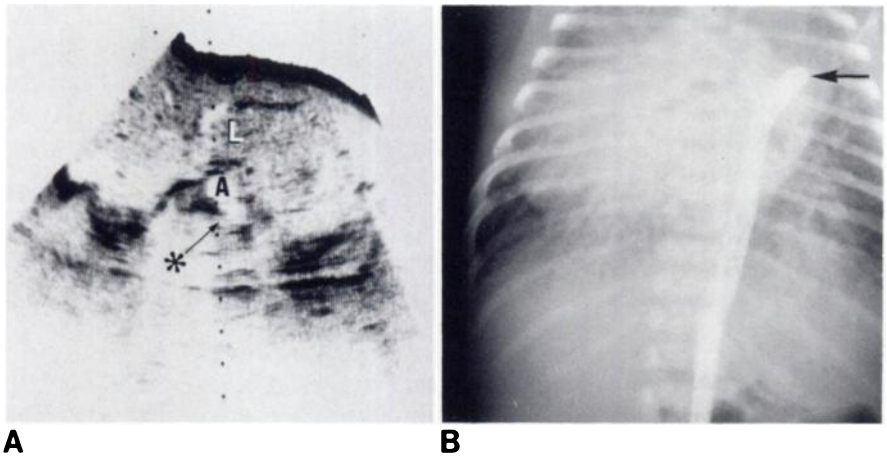


Fig. 4.—Left isomerism (polysplenic syndrome), case 3, 1 week-old-boy with congestive heart failure and situs ambiguus. **A**, Transverse sonogram before cardiac catheterization. Large transverse liver (L). Aorta (A) to left of spine and venous structure (*asterisk*, *arrow*) presumed to be left inferior vena cava or hemiazygous vein to left below aorta. Normal inferior vena cava not visualized entering atrium. **B**, Foot vein injection before cardiac catheterization to confirm sonographic findings. Left inferior vena cava with hemiazygous continuation to left superior vena cava (*arrow*). Polysplenia syndrome with hemiazygous continuation of the inferior vena cava was diagnosed from chest film and abdominal sonogram.

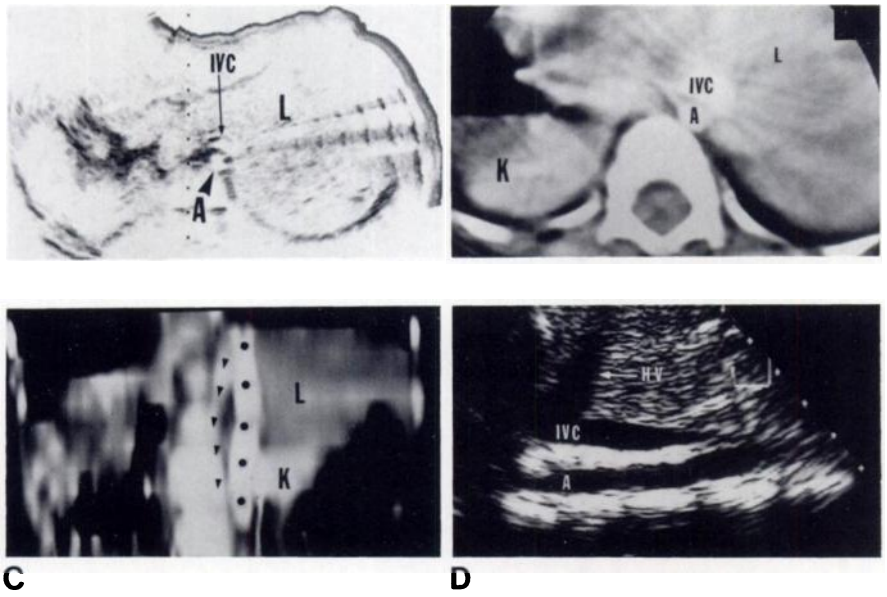


Fig. 5.—Right isomerism (asplenia syndrome), case 8, 4-year-old girl. **A**, Transverse sonogram 0.5 cm below xiphoid. Crossing of inferior vena cava (IVC, *arrow*) and aorta (A, *arrowhead*) to left of spine. In all cases of asplenia syndrome, aorta and inferior vena cava cross midline or are located on same side of spine. Vascular antomy easily demonstrated. Liver (L) on left. **B**, CT scan at comparable level. Inferior vena cava (IVC) and aorta (A) with contrast enhancement. Kidney (K) on right with left-sided liver (L). No splenic tissue seen. **C**, Coronal reconstruction in plane through IVC and aorta. Actual course of inferior vena cava (*dots*) and aorta (*arrowheads*). Aorta and inferior vena cava, contrary to normal relationship, are on same side of upper abdomen and often superimpose or cross in frontal projection. Liver (L) and left kidney (K). **D**, Longitudinal two-dimensional sector scan at 1 cm to left of midline. Hepatic vein (HV) and inferior vena cava (IVC) enter atrial structure with aorta (A) directly below caval segment on same side of spine. Real-time imaging showed vascular pulsations.

which are generally seen in only 3%–5% of patients with total situs inversus.

Left Isomerism (Polysplenia Syndrome)

Abdominal sonography demonstrated an abnormal position of the liver or abdominal heterotaxia in five (71%) of seven patients (table 1). In each case, absence of the inferior vena cava at the level of the liver was noted (fig. 2A). The longitudinal sonographic examination also demonstrated lack of an inferior vena cava entering the right atrium with independent drainage of the common vein or hepatic veins into the atrium [9, 10]. Five of the seven patients with left isomerism or polysplenia syndrome underwent cardiac catheterization and the vascular anatomy was correlated with the sonography and CT findings (fig. 2).

CT was performed on two patients (cases 1 and 5), and the positions of the abdominal vessels and multiple splenules were demonstrated [11, 12] (figs. 2B, 3A, and 3B). Sonography and CT (figs. 3A and 3C) were comparable. The vascular anatomy can be traced into the upper abdomen and chest showing crossover of an abnormal vessel or azygous vein into the chest (figs. 3A and 3B). There was good correlation between the vascular anatomy demonstrated on sonography and the vascular findings at angiocardiography (fig. 4).

Right Isomerism (Asplenia Syndrome)

In all cases of asplenia syndrome, the aorta and inferior vena cava were in juxtaposition on the same side of the spine [2, 5, 7] (fig. 5A) (table 1). This can be seen with sonography or CT of the abdomen. In addition, with CT, the presence or absence of splenic tissue can be determined (figs. 5A and 5B). Coronal reconstruction of the CT images at the level of the major abdominal vessels will depict their

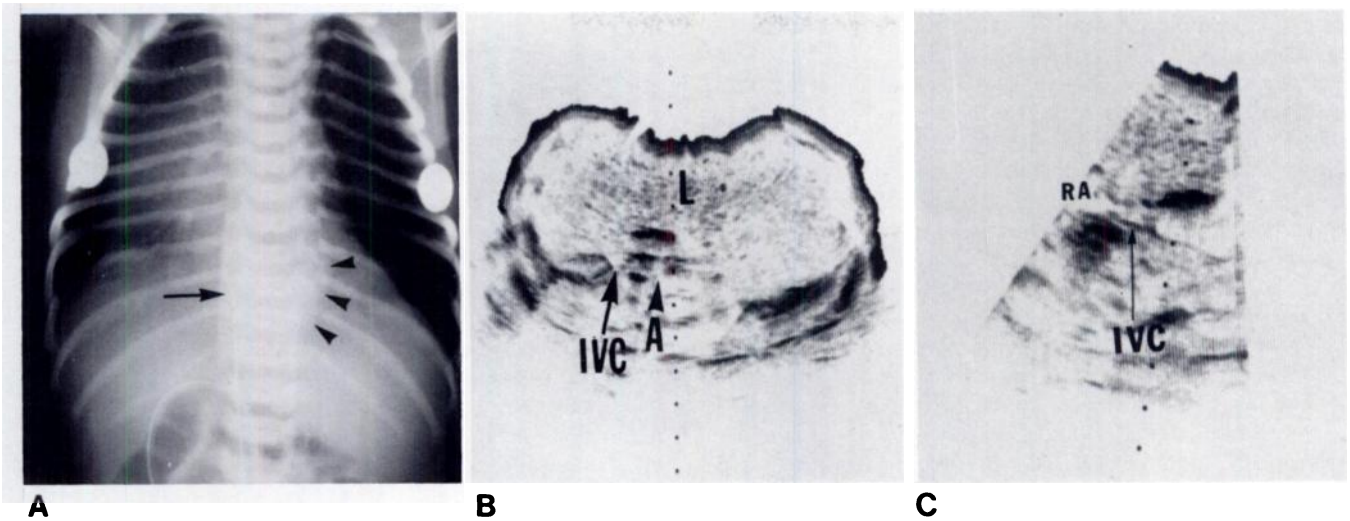
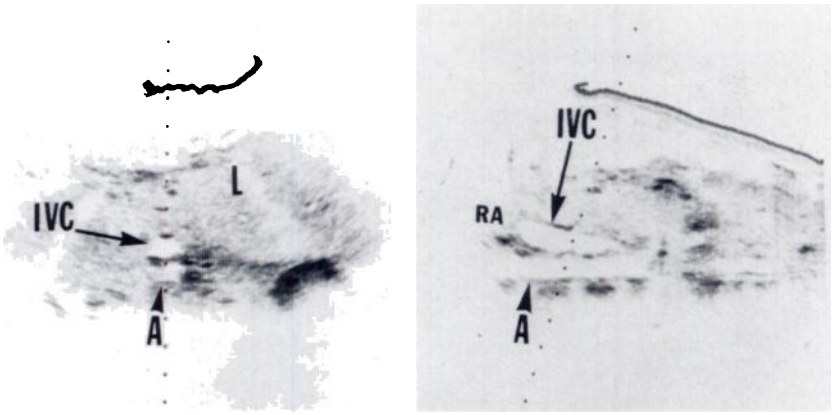


Fig. 6.—Right isomerism (asplenia syndrome), case 9, 3-day-old cyanotic boy. **A**, Situs ambiguus with decreased pulmonary vascularity, dextrocardia, and abdominal heterotaxia with transverse liver. Umbilical artery catheter (*arrow*) to right of spine in upper abdominal aorta. Density to left of spine (*arrowheads*) was thought to represent microgastria, which was demonstrated with an esophagogram. Inferior vena cava shadow not seen in lateral projection. **B**, Transverse sonogram. Midabdominal liver (L). Dense echo complex represents midline stomach. Vascular anatomy in newborn can be difficult to resolve with confidence. However, aorta (A, *arrowhead*) and inferior vena cava (IVC, *arrow*) appear to lie to right of spine, consistent with asplenia syndrome. **C**, Longitudinal scan 1 cm to right of midline. Inferior vena cava (IVC, *arrow*) enters atrial structure (RA). This allowed cardiologist to place a catheter from femoral vein into atrium rather than considering brachial vein approach. On the basis of sonograms and radiographs diagnosis of right isomerism was made before cardiac catheterization.

Fig. 7.—Right isomerism (asplenia syndrome), case 10, 4-year-old boy. **A**, Transverse sonogram. "Piggyback" vascular anatomy with inferior vena cava (IVC, *arrow*) anterior to aorta (A, *arrowhead*). Both structures to right of spine. L = liver. **B**, Longitudinal scan. Inferior vena cava (IVC, *arrow*) enters right atrium (RA) directly on top of abdominal aorta (A, *arrowhead*) 1 cm to right of midline. This anomaly should not be confused with normally visualized tubular anechoic crus of diaphragm seen ventral to abdominal aorta on longitudinal scans in normal patients. Without angulation of transducer, inferior vena cava can be seen to cross aorta or travel on same side of spine with aorta in most cases of right isomerism. Real-time imaging shows vascular pulsations.



course in the abdomen (fig. 5C). These findings can be correlated with the longitudinal or sagittal sonographic examination, which showed the inferior vena cava and aorta to the same side of the spine in a "piggyback" fashion with the inferior vena cava entering the right atrium (fig. 5D). This inferior vena cava–right atrial communication indicates the location of the "right-sided atrium" in patients with right isomerism or asplenia syndrome.

Sonography was performed in all patients and, even in infancy, the position of the aorta and inferior vena cava could be detected on the same side of the spine (Fig. 6). In addition, inferior vena cava–right atrial communication could be identified on the longitudinal sonogram. Abdominal sonography correlated well with the angiocardiographic findings in all cases (fig. 7).

Discussion

Greater numbers of situs abnormalities will be encountered in the future due to the increasing use of sonography and CT for abdominal evaluation. The key to the diagnosis of these visceroatrial anomalies lies in the recognition of derangement of the great vessels of the abdomen. This presents a particular challenge to the sonographer, since organ identification rests on a normal (situs solitus) vascular pattern. With this difficulty in mind, real-time sonographic evaluation of the aorta and inferior vena cava become critical in order to localize the aorta and inferior vena cava. With CT, contrast infusion may be necessary in some instances to document the true vascular nature of some situs abnor-

malities to avoid confusion with adenopathy or other paraspinal pathology.

Patients with *situs inversus* can be evaluated with abdominal sonography or CT. These patients usually do not have congenital heart disease, however, our cases showed severe cardiac malformations and the confirmation of situs inversus via abdominal sonography was extremely helpful. In all cases, there was mirror image positioning of the viscera and vascular structures. Therefore, sonographic examination of the abdomen will differentiate total situs inversus from situs ambiguus.

Left isomerism or the *polysplenia syndrome* may be seen in adulthood and some cases demonstrate minor or no signs of congenital heart disease. Therefore, if sonography or CT is the first imaging method, these abnormal vascular heterotaxic patterns should be recognized as such [11–14].

With left isomerism, interruption of the inferior vena cava between the renal veins and hepatic veins with azygous/hemiazygous continuation is usually seen. This sign may be suggested from evaluation of the plain chest radiograph with absence of an inferior vena cava shadow on the lateral projection and a widening of the paravertebral pleural reflection in the right or left paratrachel area, which may indicate a dilated azygous or hemiazygous vein [15–18]. This venous anomaly may appear as a mediastinal mass in older patients [16, 17, 19]. When this anomaly is suspected on the plain chest film, it is important to confirm the absence or presence of inferior vena cava–atrial communication with a sonography (or CT) of the abdomen before cardiac catheterization. Abdominal sonography will show the exact location of the abdominal aorta as well as absence of the inferior vena cava at the level of the liver with independent drainage of the confluence of hepatic veins into the right atrium [9, 10]. With real-time imaging capabilities, this anomaly can be demonstrated even in very small infants.

In addition, since patients with polysplenia syndrome frequently have abdominal heterotaxia, meaning the anomalous placement of organs and major blood vessels within the abdomen, both sonography and CT are useful in the diagnosis of visceral heterotaxia. By definition, the polysplenia syndrome has two or more spleens [2–4]. CT examination of the abdomen will demonstrate azygous or hemiazygous continuation of the inferior vena cava with a vein localized behind the crus of the diaphragm, as well as demonstrating multiple splenules that are always located along the greater curvature of the stomach [11–13]. Partial or complete failure of rotation of the intestinal tract is common and the mesentery may be located in the midline. The liver and occasionally unusual location of the gallbladder can be visualized [2] (figs. 2A and 2B) (table 1).

Patients with *right isomerism* or *asplenia syndrome* frequently have severe congenital heart lesions (table 1). The plain chest radiograph in this syndrome is frequently difficult to evaluate in the very cyanotic and sick newborn (fig. 6A).

Sonography, even in the very ill patients, will demonstrate the location of the inferior vena cava and the site of the inferior vena cava–atrial communication. It is important to demonstrate this anatomy prior to cardiac catheterization to insure that a catheter can be passed from the femoral route into the heart. Patients with interruption of the inferior vena cava (with azygous/hemiazygous continuation) should be evaluated from an antecubital approach in many cases.

In asplenia syndrome, the anomalous relationship of the inferior vena cava and abdominal aorta is also present, with both vessels usually coursing on the same side of the spine [2, 5, 7]. This can be demonstrated with sonography or CT. However, there are rare case reports of interruption of the inferior vena cava in the asplenia syndrome [6, 20, 21].

Patients with right isomerism (asplenia syndrome) have cardiac malformations commonly including atrioventricular canal, univentricular heart (single ventricle), pulmonary stenosis or atresia, transposition of the great arteries, and anomalous pulmonary venous connection [1–6]. In contrast, patients with left isomerism (polysplenia syndrome) often have ventricular septal defect, double outlet right ventricle, left-sided obstructive lesions, and anomalies of systemic veins more often than pulmonary venous return [1–6]. The severity of the cardiac anomalies is usually greater with the asplenia syndrome than with polysplenia syndrome. The presumptive but not definite diagnosis of asplenia syndrome can also be made by evaluation of the peripheral blood smear, which may show Howell-Jolly and Heinz bodies with absence of the spleen [4, 5]. It is important to recognize these visceroatrial situs abnormalities to suggest the prognosis and possible surgical palliation or correction. In addition, patients with asplenia syndrome are at a greater risk of sepsis with prophylactic antibiotics recommended [22, 23].

The plain radiographic signs of asplenia and polysplenia have been well defined [1–6, 20]. In polysplenia, the chest radiograph frequently shows bilateral hyparterial bronchi with corresponding bilateral pulmonary artery anatomy like that of a normal left lung. On the lateral chest radiograph, these arteries project posterior to the trachea [6], and the inferior vena cava shadow may be absent. In asplenia, the radiographic signs include bilateral eparterial bronchi with the corresponding pulmonary artery anatomy like that of a normal right lung. The pulmonary arteries are anterior to the tracheobronchial tree on the lateral projection [6]. Occasionally, one can see bilateral minor fissures secondary to bilateral trilobed lungs with right isomerism. The inferior vena cava can usually be seen on the erect lateral chest radiograph, but it may be difficult to detect in infancy or with cardiac enlargement and sonography will be helpful.

Radionuclide venography associated with liver-spleen radionuclide imaging has been diagnostic in evaluating the heterotaxic syndrome [24, 25]. However, in our experience, there have been several false-positives in asplenia syndrome and false-negatives in polysplenia syndrome [5, 20]. In addition, polysplenia syndrome can be diagnosed by angiography which will demonstrate multiple splenules and a common celiomesenteric artery [26], which can be detected with abdominal sonography in some patients.

We have found sonography and CT correlated with chest radiographs to be the most accurate means of completely diagnosing the type of visceroatrial situs prior to cardiac

catheterization with confirmation by angiocardiography or, ultimately, pathology. As sonography, and to a lesser exent CT, are used for the initial abdominal evaluation, incidental visceroatrial situs abnormalities will be encountered and should be recognized.

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