Diagnosis of Atrial Situs by Transesophageal Echocardiography

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In a prospective investigation, direct visualization of both atrial appendages was attempted during transesophageal echocardiographic studies in 132 patients with congenital heart disease. High quality cross-sectional images delineating the unique morphologic details of both atrial appendages were obtained in every patient. Abnormal cardiac position such as dextrocardia (four patients) or mesocardia (two patients) did not pose any problems for transesophageal assessment of both atrial appendages. Thus, direct diagnosis of atrial situs was possible in every patient. Atrial situs solitus was present in 127 patients studied. Three patients were found to have situs inversus, one had left atrial isomerism and one had right atrial isomerism. No patient with juxtaposed atrial appendages was encountered. All patients had prior subcostal ultrasound scans for assessment of the morphology and relation of the suprarenal abdominal great vessels and the related patterns of hepatic venous drainage.

Patients with abnormal atrial situs had correlative high kilovoltage filter beam radiography for assessment of bronchus morphology. The results of situs determination obtained by either method were in agreement.

In this series, transesophageal echocardiography allowed the direct and accurate visualization of both atrial appendages and the determination of atrial situs in all patients studied. Transesophageal echocardiography may prove to be the most reliable in vivo technique for determination of atrial situs.

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The determination of atrial situs is the initial fundamental diagnostic step for sequential chamber localization in the diagnosis of complex congenital heart disease (1–3). In clinical practice, atrial situs is usually determined noninvasively by either radiographic definition of the bronchus morphology (4–6) or assessment of the morphology of the abdominal great vessels and their relation to one another and to the spine (7). These two techniques used in combination yield reliable results in the majority of patients (8,9). A complete abdominal ultrasound investigation including the definition of the splenic status should be performed in all cases where abnormal situs is suspected. However, because there are reports (10–12) of discordance between the arrangement of the atria and the thoracic and abdominal organs, all indirect diagnostic approaches for the definition of atrial situs have potential limitations.

The ideal diagnostic investigation for the clinical definition of atrial situs would allow identification of the unique morphologic characteristics of each atrial chamber. Because both the systemic and pulmonary venous connections in complex congenital heart disease can be anomalous, their determination is of little value for the diagnosis of atrial situs. The anatomic characteristics of the christa terminalis or of the right atrial venous valves are too subtle to allow differentiation on either precordial echocardiographic or angiographic grounds. Therefore, only the visualization of both atrial appendages, with their constant unique morphologic characteristics (13), would allow a direct diagnosis of atrial situs. In newborns, precordial echocardiography sometimes permits demonstration of both atrial appendages with a series of parasternal short-axis views (7). However, with increasing size of the patient and the natural reduction in the precordial ultrasound window, direct visualization of both atrial appendages from the precordium becomes virtually impossible.

Transesophageal echocardiography has gained widespread clinical acceptance in adult practice during recent years (14–16). The technique allows high quality imaging in almost all patients. In particular, the cardiac chambers closest to the esophagus can be assessed with much more

detail than is usually possible from the precordium. Experience with this new diagnostic technique in the study of congenital heart disease is still limited (17,18). However, our recent experience in adolescents and adults with such disease suggested that both atrial appendages could be demonstrated in every patient. Therefore, transesophageal echocardiography may be expected to be a reliable (albeit semiinvasive) diagnostic tool for the direct morphologic diagnosis of atrial situs.

Methods

Study patients. Complete transesophageal echocardiographic studies were performed in 132 patients with congenital heart disease as part of a larger study evaluating congenital cardiac lesions from the esophageal approach. The age at study ranged from 3.7 to 68 years (mean 28.2). The atrial appendage morphology was assessed and atrial situs was defined in every patient as part of a complete study protocol. Transesophageal studies were performed either on an outpatient basis (123 adolescent and adult patients) or during routine cardiac catheterization (9 children). Approval by the hospital ethical committee of Erasmus University Rotterdam, was obtained before the start of the study protocol and informed consent was obtained from the patients or their parents before individual studies.

Transesophageal studies. Studies were successful in all 132 patients. For the 123 outpatient studies, the patients fasted for ≥4 h. No antibiotic prophylaxis was administered; mild sedation using a short-acting benzodiazepine was used in seven patients. After local anesthesia of the pharynx, the probe was introduced into the hypopharynx; then, with the patient being asked to swallow, was advanced to the lower part of the esophagus. A continuous electrocardiographic (ECG) tracing was used for monitoring during the studies; no other monitoring technique was employed in outpatients. Although isolated supraventricular premature beats were occasionally seen, no patient had sustained supraventricular tachycardia and cardioversion (either medical or electrical) was not required in any patient. The adolescent and adult patients were studied with the use of various 5 to 5.6 MHz transesophageal single plane probes (maximal tip dimension 13 × 15 mm) and either a Toshiba SSH 160 A or a Vingmed CFM 700 ultrasound system.

Studies in the nine children were performed during routine cardiac catheterization with the patient under general anesthesia as part of a prospective study on the role of transesophageal echocardiography in the pediatric patient with complex congenital heart disease. In these patients, a specially dedicated 5 MHz pediatric transesophageal probe was used with an Aloka SSD 870 ultrasound system. The small dimensions of the probe (maximal tip dimension 7 × 8 mm) allowed investigation of children of >10 kg without any complications (for example, signs of bleeding, esophageal trauma).

Imaging technique. The initial step of every investigation was assessment of atrial appendage morphology by cross-sectional imaging. The probe was advanced within the esophagus to the level of the atria and then an optimal scan position was chosen by varying the level of insertion and by rotation of the probe. Care was taken to obtain almost transverse sections of both atrial appendages by slight up-down and sideward movement of the tip of the transducer by use of the probe steering mechanism. Only these transverse sections allow simultaneous demonstration of both the junction with the atrial cavity and the internal morphology of the atrial appendage. The right atrial appendage is usually visualized when scanning in a basal short-axis view, which is obtained by following the superior vena cava into the right atrium. The cavity of the right atrial appendage is shown in transverse sections to the right of the aortic root and above the tricuspid valve. After anticlockwise rotation of the probe, the left atrial appendage is seen to the left of the aorta just above the mitral valve and anterior to the orifice of the left upper pulmonary vein.

Correlative investigations. All patients had prior complete transthoracic ultrasound investigations, including subcostal scans for assessment of the morphology and the relation of the suprarenal abdominal great vessels and the related patterns of hepatic venous drainage. Patients with detected abnormal atrial situs had additional correlative high kilovoltage filter beam radiography for assessment of bronchus morphology.

Results

Atrial appendage morphology. Transesophageal echocardiography allowed direct visualization of the morphology of both atrial appendages in all 132 patients studied. The morphologically right atrial appendage was demonstrated to have a short blunt appearance and a broad junction with the right atrial cavity. In contrast, the morphologically left atrial appendage was demonstrated to have a long, narrow and crenelated appearance and a narrow junction with the atrial cavity. The delineation of these unique features consistently allowed the differentiation of both atrial appendages. Thus, the determination of atrial situs was possible in every patient studied. Cardiac malrotation or malposition, present in six patients (four with dextrocardia, two with mesocardia), did not prevent complete assessment of appendage morphology in any patient, although variations in the examination technique had to be adopted to scan the relevant imaging planes.

Atrial situs solitus was present in 127 patients. The right-sided appendage was identified to be of right morphology and the left-sided appendage of left morphology in all patients. There was no case of discrepancy between the transesophageal diagnosis and the previous clinical, ultrasonographic and radiographic diagnosis or the anatomic findings at previous surgical repair.
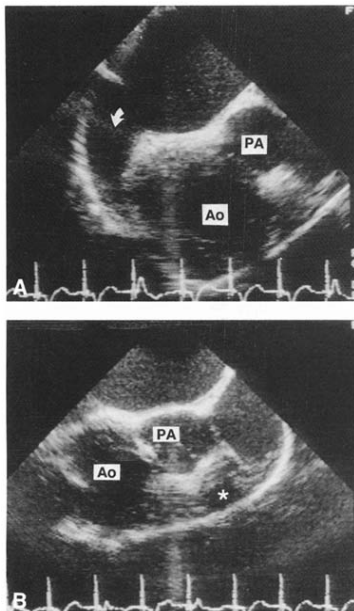
Figure 1. Situs inversus in a patient with congenitally corrected transposition of the great arteries and dextrocardia. **A,** The right-sided atrial appendage has a long and narrow cavity; the junction with the atrial cavity **(arrow)** is narrow. Thus, the right-sided appendage is of left atrial appendage morphology. **B,** The left-sided atrial appendage **(asterisk)** has a broad junction with the atrial cavity; the appendage cavity itself is wide and has a blunt ending. These morphologic characteristics define the left-sided appendage to be of right atrial appendage morphology. Ao = aorta; PA = pulmonary artery.

Figure 2. Left atrial isomerism in a patient with congenitally corrected transposition of the great arteries and dextrocardia seen from within the esophagus. Both the right-sided **(A)** and left-sided **(B)** appendage have a narrow junction **(straight arrow in A, curved arrow in B)** with the atrial cavity; the appendage cavity is long and narrow. These morphologic features identify both atrial appendages to be of left atrial appendage morphology; thus, the patient is diagnosed to have left atrial isomerism. Abbreviations as in Figure 1.

Atrial situs inversus was demonstrated in three patients, all of whom had congenitally corrected transposition (atrioventricular [AV] discordance and ventriculoarterial [VA] discordance). One of the patients had dextrocardia. The right-sided atrial appendage in these patients was identified to be of left morphology (Fig. 1A) and the patient's left-sided appendage of right morphology (Fig. 1B). All three patients had high kilovoltage filtered beam radiography for identification of bronchial situs and a repeat subcostal ultrasound scan for 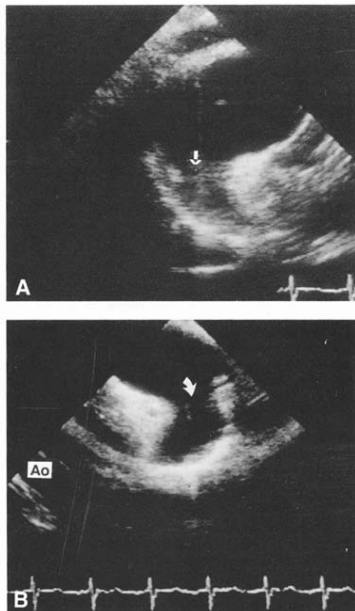demonstration of the abdominal vessel morphology and splenic status. Both techniques demonstrated mirror image arrangement of the thoracic and abdominal organs. No discrepancies between the results of atrial situs determination by these indirect investigations and the transesophageal studies were found. Precordial echocardiography did not allow demonstration of either atrial appendage in any of these patients.

Atrial isomerism. Bilateral morphologically left atrial appendages were demonstrated in one patient with dextrocardia and congenitally corrected transposition (Fig. 2). Both appendages were found to have a narrow junction with the corre-

sponding atrial cavity, clearly identifying them to be of left morphology. This result of the transesophageal study was in agreement with the diagnosis made on the basis of filtered beam radiography, showing bilaterally left bronchi (long curved bronchi with late bifurcation). A previous subcostal ultrasound investigation of the upper abdomen had shown drainage of the hepatic veins to a single subdiaphragmatic confluence into the right-sided atrium. There was a hemiazygos continuation of the inferior vena cava. Thus, this patient, on the basis of the subcostal ultrasound investigation alone, could well have had situs solitus with interruption of the inferior vena cava. Dextrocardia prevented a complete precordial echocardiographic investigation and, in particular, the demonstration of either atrial appendage. The transesophageal study clearly demonstrated left atrial isomerism with left ventricular topology (19) and discordant VA connections.

Bilateral morphologically right atrial appendages, thus right atrial isomerism, were clearly demonstrated in a 7 year old child with a normally positioned heart. The broad junction with the venous component of either atrium and a short blunt shape of both appendages led to their identification as being of right morphology (Fig. 3). The child was found to have a left persistent superior vena cava, which drained into the roof of the left-sided atrium, interposed between the left-sided atrial appendage and the left upper pulmonary vein (Fig. 3B). Visualization of either atrial appendage from the parasternal or subcostal windows was impossible during precordial studies. Previous filter beam radiography had demonstrated bilateral short eparterial bronchi with early bifurcation, suggesting right atrial isomerism. Subcostal echocardiography identified an inferior right vena cava that was anterior to the abdominal aorta, but hepatic venous drainage remained unclear. Absence of the coronary sinus and complete drainage of all hepatic veins into the right-sided inferior vena cava could be demonstrated only by transesophageal echocardiography. The transesophageal study further showed a complete AV septal defect, right ventricular topology and double-outlet right ventricle with the aorta in front and to the right of an unperforated pulmonary ostium.

Discussion

Abnormal atrial situs is by itself of little clinical importance. The complex associated malformations that are almost always present with either right or left atrial isomerism are the determining factors for (surgical) management and natural history (9,13,20). However, identification of atrial situs has to be the initial step for the complete diagnosis of all congenital heart malformations (3).

Conventional diagnosis. High kilovoltage filter beam radiography for identification of bronchial situs, introduced by Deanfield et al. (6), yields reliable information of atrial situs in the vast majority of patients. The low radiation dose and
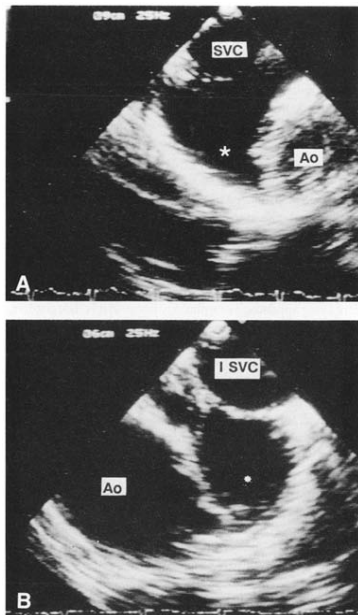

Figure 3. Right atrial isomerism in a child with common atrioventricular valve, right-sided ventricular topology and double-outlet right ventricle. **A,** The right-sided appendage has a broad junction with the atrium; the cavity (**asterisk**) is wide and has a blunt ending and is thus of right atrial appendage morphology. **B,** The left-sided appendage has a wide cavity (**asterisk**) and a blunt ending. The junction with the atrial cavity is narrowed because of interposition of a persistent left superior vena cava (l SVC) that drains into the left-sided morphologically right atrium. Ao = aorta.

the noninvasive character of this investigation allow its routine use in patients with congenital heart disease. Subcostal echocardiographic investigation of the suprarenal great vessels, as first described by Huhta et al. (7), is the most extensively used screening method for detecting abnormal atrial situs. Again, reliable information is obtained in the majority of patients. Both techniques, which are frequently used together, combine the advantages of noninvasiveness and high sensitivity and specificity, as cases of discordance between the arrangement of the atria and the thoracic and

abdominal organs are rare (10–12). Furthermore the techniques are not restricted to selected patient groups and yield information regardless of patient age. However, the unique morphologic details of the right or the left atrial appendage cannot be demonstrated.

Transesophageal diagnosis. The high quality images obtained by transesophageal cross-sectional imaging in every patient of this study allowed consistent differentiation of both atrial appendages by detailed demonstration of their unique morphologic characteristics. The morphologically right atrial appendage is visualized as a short blunt cavity that has a broad junction with the smooth-walled atrial chamber. In contrast, the left atrial appendage is demonstrated to be a long, narrow and largely crenelated cavity with a narrow junction with the venous component of the left atrium. The junction of the atrial appendage with the atrial cavity is the most reliable feature for identifying appendage morphology because it is independent of any degree of atrial distension, which may result from valvular dysfunction or intracardiac shunting. Abnormal cardiac positions (such as dextrocardia or mesocardia) required slightly modified transducer manipulation, but never represented a limitation for the transesophageal approach. In general, the probe manipulation technique in patients with dextrocardia is a mirror image of the series of probe maneuvers carried out in patients with a normally positioned heart. If present, a persistent left superior vena cava will be found interposed between the left atrial appendage and the left upper pulmonary vein. The misinterpretation of this extra "cavity" as an atrial appendage is excluded by following the course of the individual structures. Identification is furthermore aided by combined color flow mapping and pulsed wave Doppler studies.

Clinical implications. Although our findings suggest that transesophageal echocardiography should allow direct identification of both atrial appendages in every patient, its role solely in determining atrial situs will remain limited in clinical practice because it is an unpleasant semi-invasive technique. Only recently, small dedicated pediatric transesophageal probes have been developed that allow the safe investigation of small children under general anesthesia or heavy sedation. At present, transesophageal studies in patients with congenital heart disease appear to be indicated in those symptomatic patients with a poor precordial ultrasound window who have problems related to either systemic or pulmonary venous return, atrial lesions, abnormalities of the AV junction or pathologic changes of the left ventricular outflow tract. In all patients with congenital heart disease studied by transesophageal echocardiography, the determination of atrial situs should be carried out routinely because it is an easy and reliable method.

Transesophageal echocardiography appears to be the most reliable in vivo technique for the identification of atrial situs because it consistently allows the delineation of the unique morphologic details of both atrial appendages. It may be considered the ultimate diagnostic technique in cases where ambiguity of situs determination persists after combined radiographic and transthoracic ultrasound studies.

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