

The Role of Transesophageal Echocardiography in Adolescents and Adults with Congenital Heart Disease

NAVEEN SHEIKH, SAJAL KRISHNA BANERJEE, FAZLUR RAHMAN, ZAHID HOSSEN, C MAHMED, HARISUL HAQUE, SMA HABIB, MD. NOORNABI, MAMUQUEET, KHONDAKER, NIZAMUL HOSSAIN, ATM IQBAL HASAN, JAHANARA ARZU, MD MUSTASHIRUL HAQUE

Department of Cardiology, Bangabandhu Sheikh Mujib Medical University, Dhaka, Bangladesh

Address of Correspondence: Dr. Naveen Sheikh, Associate Professor, Department of Cardiology, BSMMU Dhaka, Bangladesh, E-mail: sheikhnaveen@yahoo.com.sg

Abstract:

There is frequent dropout of atrial septal echoes in the region of the fossa ovalis in the standard precordial echocardiographic imaging planes, that can be minimized by use of the subcostal imaging approach. The diagnostic sensitivity of this approach was reviewed in 154 patients (mean age 31 years, range 18 years to 45 yrs) with documented atrial septal defect in whom a satisfactory image of the atrial septum could be obtained. Subcostal two-dimensional and color Doppler echocardiography successfully visualized 93 (89%) of the 105 ostium secundum atrial septal defects, all 32 (100%) ostium Primum defects and 7 (44%) of the 16 sinus venosus defects. A defect was not visualized (false negative response) in 12 patients (11 %) with an ostium secundum defect and in 9 patients (56%) with a sinus venosus defect. In three of the former and five of the latter, a two-dimensional echocardiographic contrast examination and transesophageal echocardiography established the presence of the inter-atrial shunt. Forty eight patients (32 %) with clinical findings of uncomplicated atrial septal defect confirmed by two-dimensional, color Doppler and Transesophageal echocardiography underwent surgical repair of the defect without preoperative cardiac catheterization. There were no perioperative complications.

Introduction:

Transesophageal Echocardiography (TEE) is rapidly gaining acceptance as a semi-invasive diagnostic approach in adults in whom an adequate precordial ultrasound examination is impossible due to lung disease, chest deformity or mechanical ventilation (1). Transesophageal imaging is increasingly used as the approach of choice in the adult cardiac outpatient clinic for the investigation of atrial mass lesion, in the evaluation of mitral prosthetic valve function, in the assessment of endocarditis and its complication and in the evaluation of thoracic aortic pathology. However, little has yet been reported on its potential role in the evaluation of the complexities of congenital heart disease^{2,3}.

At present both transducer size and the semi-invasive nature of the procedure appear to preclude its use in sedated or non-anesthetized conscious children. Even when general anesthesia is used to allow probe insertion and placement, probe size will normally not allow children of less than 5 years to be investigated with safety. Fortunately, using a combination of precordial and subcostal high resolution cross-sectional imaging (i.e. 5.0 or 7.5 MHz) allied to spectral Doppler plus colour

flow mapping studies diagnostic information can be derived in virtually all unoperative infants and young children (i.e. those less than 5 years), and thus recourse to the alternative transesophageal approach is rarely required in this age group^{4,5}. However, as more children are operated on at a young age for complex heart disease a non-invasive technique is required for the sequential evaluation of the results of cardiac surgery and the identification of residual or newly acquired lesions. The precordial ultrasound window is frequently restricted in such operated patients with mid-line thoracotomies as a direct consequence of acquired fibrous adhesions following cardiac surgery. In addition, ultrasound imaging in all children becomes more difficult with age due to increasing chest and cardiac size and the natural reduction in the ultrasound window^{1,3}. Ultrasound studies of complex congenital heart disease in the adolescent and adult population present many difficulties. In addition, complex heart disease is frequently associated with either cardiac malpositions, where the heart is completely or partially obscured by the sternum and rib cage, significantly is spinal or thoracic cage abnormalities which make ultrasound examination difficult^{5,6}.

Table-I
Associated Cardiac Abnormalities in 154 Patients

With Atrial Septal Defect	
Mitral valve prolapse	20
Atrial septal aneurysm	2
Pulmonary stenosis (mild)	4
Mitral stenosis	2
Coarctation of aorta	1
Ischemic heart disease	2
Persistent left superior vena cava	1
Tricuspid insufficiency	1
Patent ductus arteriosus	1

Table-II

Type of defect	No. of patients	Patients with contrast study :no.	Patients with contrast study %	Patients with positive results no.	Patients with positive results%
Secundum	105	46	44	44	96
Primum	32	14	44	14	100
Sinus venosus	16	10	62	10	100
Coronary sinus	1	1	100	1	100

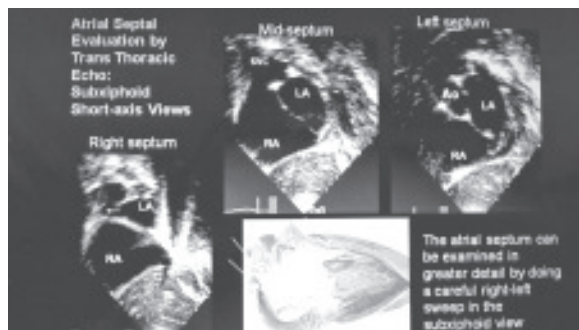


Fig.-1: Atrial septal evaluation by 2D echo from subcostal views (MIOT e-Textbook of Echocardiography in congenital heart disease chapter:III)

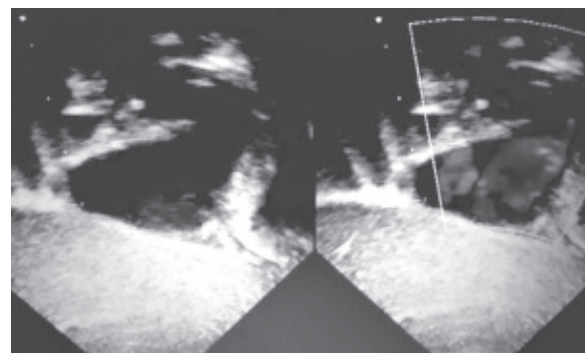


Fig.-3 : Ostium primum ASD



Fig.-2: Two-dimensional and color Doppler echocardiographic images (still frame photographs) of ostium secundum atrial septal defect, as viewed from the subcostal approach.

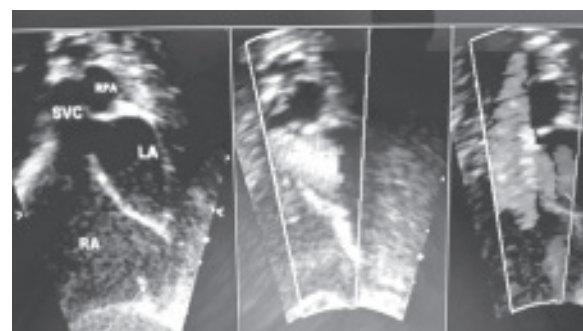


Fig.-4: Atrial Septal Defects: Sinus Venosus ASD :Subxiphoid short axis views.

Discussion:

Two-dimensional and color Doppler echocardiographic examination of the atrial septum utilizing the subcostal approach is the preferred method for the confident, noninvasive diagnosis and categorization of atrial septal defects. (7,8) Two dimensional echocardiographic contrast and color Doppler examinations complement the technique and enhance diagnostic accuracy. (9,12) The use of two-dimensional echocardiography has allowed direct visualization of the atrial septum (Figure:1). However, in the standard parasternal and apical imaging planes, because the direction of the echocardiographic beam is nearly parallel to the atrial septum, echo dropout, primarily in the region of the midatrial septum (fossa ovalis), is common, making these imaging planes unreliable for the echocardiographic diagnosis of atrial septal defect (1-3). In comparison, the subcostal approach permits the echographic beam to be directed nearly perpendicular to the atrial septum, allowing complete visualization of the entire atrial septum. Because a large series describing the usefulness and sensitivity of the two-dimensional and color Doppler echocardiographic subcostal approach (Figure:1) for the diagnosis of various types of atrial septal defects has not been reported, (10,11) we describe our experience with a broad spectrum of patients using this technique. Our results show that the subcostal technique is very reliable for the diagnosis and characterization of the common types of atrial septal defect. In certain circumstances, contrast echocardiography are useful adjunctive diagnostic procedures. On the basis of our experience, we also describe the proper technique for the subcostal echocardiographic examination of the atrial septum, Its limitations and the indications for and usefulness of contrast echocardiographic and Doppler examinations.

Methods:

Patients were included in the study on the basis of satisfactory two-dimensional echocardiographic examinations from parasternal and apical positions, but in particular from the subcostal approach for visualization of the atrial septum. Two-dimensional and color Doppler echocardiograms of 171 patients with atrial septal defect were reviewed. All patients were seen between April 2011 and April 2015 in BSMMU echo room. A total of 154 patients met these criteria. Seventeen patients (10%) were excluded from the study because in nine patients the image quality was poor and in eight patients no subcostal examination was done. Of the 154 patients, 58 were male and 96 were female, their ages ranged from 18 to 45 years

(mean 31 years). Patients who had an atrial septal defect as part of other major complex congenital heart abnormalities were excluded. Confirmation of ASD was made by both surgery and cardiac catheterization in 115 cases, surgery alone in 24 cases and cardiac catheterization alone in 15 cases. There was no perioperative mortality in any of 139 patients who underwent surgical repair.

Echocardiography.

The two-dimensional and color Doppler echocardiographic equipment used in the study included 3.5 MHz. The atrial septum was analyzed and the defects were categorized according to specific anatomic location (Fig. 1). Sinus venosus atrial septal defects are frequently missed and difficult to visualize with conventional two-dimensional and color Doppler echocardiographic views. The studies were recorded on videotape for storage and subsequent playback analysis.

Contrast echocardiography: Two-dimensional echocardiographic contrast studies were performed in 20 patients. The objective was to document a shunt at the atrial level. An echographic contrast study was considered positive if there was complete dense bolus opacification of the right atrium. After opacification of the right atrium, the immediate appearance of "microbubbles" in the left atrium (10) or the visualization of a negative "washout" jet in the right atrium (left to right shunt) (11,12) defined the presence of an interatrial shunt.

Results:

Ostium secundum defect (Fig. 2A and B). One hundred five patients (68%) had an ostium secundum defect. The mean age of this group was 37 years (range 18 years to 45 yrs); 34 were male and 71 female. Eighty-six (82%) underwent subsequent diagnostic cardiac catheterization, and 96 (91%) had surgical repair. Nineteen patients (18%) underwent operative closure of the defect without prior cardiac catheterization. An atrial septal defect (a persistent echovisible defect in the region of the fossa ovalis) was visualized by two-dimensional echocardiography in 93 of the 105 cases. In most patients, the defect was larger than 1.5 cm. In every instance when the atrial septal defect was visualized, it was properly categorized as a secundum type.

In 12 (11%) of the 105 patients, the defect was not visualized (false negative result). Eleven of the 12 patients had subsequent anatomic confirmation of the defect at surgery, in the remaining patient the defect was substantiated at cardiac catheterization and transesophageal echocardiography. The age of the patient did not influence the success rate of visualization of the

atrial septal defect. Echocardiographic contrast studies were performed in five of the patients, and in three an interatrial shunt was diagnosed (positive dye study). In two patients, contrast studies were nondiagnostic because of incomplete opacification. Two patients had Doppler examinations, both of which were positive. Therefore, in five (71 %) of seven patients in whom a defect was not directly visualized, the addition of a contrast study or pulsed Doppler echocardiography allowed a correct diagnosis of a shunt at the atrial level.

Ostium primum defect (Fig. 3): Thirty-two patients had an ostium primum defect. The mean age of this group was 20 years (range 20 yrs to 40 yrs years); 12 were male and 20 female. Thirty-one patients (97%) had subsequent cardiac catheterization, and 26 (81 %) underwent surgical repair. In each case, the defect was correctly diagnosed by two-dimensional and color Doppler echocardiography (diagnostic sensitivity 100%). The typical echocardiographic features of an ostium primum defect (Fig. 3C) were considered more diagnostic from apical and modified parasternal four chamber views (inflow views). Characteristic inferior displacement of the atrioventricular valves and exclusion of a ventricular septal defect were appreciated better from the apical views. On the basis of its characteristic location and appearance. The ostium primum defect was correctly categorized in every instance. *In six patients an associated ostium secundum defect was present but not appreciated by the echocardiographer.* In five of these six patients, the associated secundum defect was either small (4 to 6 mm) or fenestrated.

Sinus venosus defect (Fig. 4). Using modified subcostal and right parasternal longitudinal views, sixteen patients were found to have a sinus venosus atrial septal defect which was diagnosed by modified subcostal view. The combination of subcostal and right parasternal longitudinal imaging views will improve the detection of sinus venosus atrial septal defects

Sixteen patients had an atrial septal defect of the sinus venosus type. The mean age of this group was 41 years (range 19-45); 12 were male and 4 female. Of the 16 patients, 12 (75%) underwent cardiac catheterization and 4 did not. All 16 patients underwent surgical correction. The defect in the superior part of the atrial septum was visualized by two-dimensional echocardiography in 7 of the 16 (diagnostic sensitivity 44%), and in all 7 instances it was properly categorized as of the sinus venosus type. Right ventricular volume overload was present in 15 of the 16 patients, and right ventricular enlargement without

paradoxical septal motion was observed in the remaining patient.

Fifteen (94%) of the 16 patients with sinus venosus defect had associated partial anomalous pulmonary venous connection draining into the superior vena cava just above or at the junction of the right atrium and the superior vena cava (13 patients), directly into the right atrium (1 patient) or into the upper superior vena cava (1 patient). An anomalous pulmonary vein was visualized by two-dimensional echocardiography in one patient.

Coronary sinus defect. One patient in this series, a 50 year old woman, had a coronary sinus defect (confirmed at surgery). Preoperatively, this patient was suspected of having a nonvisualized ostium secundum atrial septal defect. Although the defect was not visualized by echocardiography, the results of two-dimensional echocardiographic contrast study were positive.

2-D ECHO CONTRAST STUDY:

Seventy-one patients had echocardiographic contrast studies and 69 (97%) of these had abnormal findings, most showing bidirectional interatrial shunts (Table 3). Sixteen patients had an atrial septal defect of the sinus venosus type. The mean age of this group was 41 years (range 5 to 66); 12 were male and 4 female. Of the 16 patients, 12 (75%) underwent cardiac catheterization and 4 did not. All 16 patients underwent surgical correction. The defect in the superior part of the atrial septum was visualized by two-dimensional echocardiography in 7 of the 16 (diagnostic sensitivity 44%), and in all 7 instances it was properly categorized as of the sinus venosus type. Right ventricular volume overload was present in 15 of the 16 patients, and right ventricular enlargement without paradoxical septal motion was observed in the remaining patient.

Of the nine patients in whom a sinus venosus defect was not visualized which were confirmed by transesophageal echocardiography.

The increased diagnostic sensitivity was attributed to a more detailed subcostal examination of the atrial septum, including anteroposterior scanning in the region of the superior atrial septum (Figure 1).

Fifteen (94%) of the 16 patients with sinus venosus defect had associated partial anomalous pulmonary venous connection draining into the superior vena cava just above or at the junction of the right atrium and the superior vena cava (13 patients), directly into the right atrium (1 patient) or into the upper superior vena cava (1 patient). All were diagnosed by Transesophageal echocardiography. An

anomalous pulmonary vein was visualized by two-dimensional echocardiography in one patient.

Coronary sinus defect:

One patient in this series, a 62 year old woman, had a coronary sinus defect (confirmed at surgery). Preoperatively, this patient was suspected of having a nonvisualized ostium secundum atrial septal defect. Although the defect was not visualized by echocardiography, the results of two-dimensional echocardiographic contrast study were positive. Two-dimensional echocardiographic contrast studies.

Seventy-one patients had echocardiographic contrast studies and 69 (97%) of these had abnormal findings, most showing

bidirectional interatrial shunts (Table 3). Apical and parasternal four chamber and short-axis views were most frequently utilized for visualization of the negative contrast and short-axis views were most frequently utilized for visualization of the negative contrast.

Associated disorders: Thirty-four patients had associated cardiac disorders that were diagnosed by two-dimensional echocardiography. In the group with a secundum defect, echocardiographic mitral valve prolapse was found in 20 patients (19%) (Table 4). In the group with a primum defect, a cleft mitral valve was visualized in all.

Patients undergoing operation without prior cardiac catheterization: Of the 154 patients, 24 (16%), with a mean age of 30 years (range 18 to 45), underwent successful surgical repair of atrial septal defect without preoperative cardiac catheterization. In this group of patients with typical clinical findings of atrial septal defect, the two-dimensional echocardiogram was used as the major confirmatory test (with or without contrast or Doppler studies). The atrial septal defect was visualized in 23 patients (96%), and the remaining patient had typical echocardiographic findings of right ventricular volume overload and mild to moderate pulmonary hypertension. Nineteen patients (79%) in this group had an ostium secundum defect, four had a sinus venosus defect and one had an ostium primum defect. Twelve (63%) of the 19 patients with an ostium secundum defect, 3 (75%) of the 4 patients with a sinus venosus defect and the 1 patient with an ostium primum defect underwent surgical repair on the basis of typical clinical findings (and diagnostic two dimensional echocardiographic findings)

without other confirmatory tests. None of the patients had a history of angina or myocardial infarction or had findings suggestive of associated valvular heart disease or severe pulmonary hypertension. At operation, the associated anomalous pulmonary venous connection in patients with a sinus venosus atrial septal defect was successfully repaired in all.

Reference:

1. Wren C, O'Sullivan JJ. Survival with congenital heart disease and need for follow up in adult life. *Heart* 2001;85:438-43.
2. Houston A, Hillis S, Lilley S, et al. Echocardiography in adult congenital heart disease. *Heart* 1998;80(suppl):S12-26.
3. Stumper O. Imaging the heart in adult congenital heart disease. *Heart* 1998;80:535-6.
4. Hirsch R, Kilner PJ, Connelly MS, et al. Diagnosis in adolescents and adults with congenital heart disease. Prospective assessment of individual and combined roles of magnetic resonance imaging and transesophageal echocardiography. *Circulation* 1994;90:2937-51.
5. Tynan MJ, Becker AE, Macartney FJ, et al. Nomenclature and classification of congenital heart disease. *Br Heart J* 1979;41:544-53.
6. Kroruk G, Hutterer B, Mossbacher H. Diagnosis of atrial left-to-right shunt, by cross-sectional contrast echocardiography. *Z Kardiol* 1981;70:138-45.
7. Weyman AE, Wann LS, Caldwell RL, Hurwitz RA, Dillon JC, Feigenbaum H. Negative contrast echocardiography. a new method for detecting left-to-right shunts. *Circulation* 1979;59:498-505.
8. Dillon JC, Weyman AE, Feigenbaum H, Eggleton RC, Johnston K. Cross-sectional echocardiographic examination of the interatrial septum. *Circulation* 1977;55:115-20.
9. Lieppe W, Scallion R, Behar VS, Kisslo JA. Two-dimensional echocardiographic findings in atrial septal defect. *Circulation* 1977;56:447-56.
10. Schapira IN, Martin RP, Fowles RE, Popp RL. Single and two dimensional echocardiographic features of the interatrial septum in normal subjects and patients with an atrial septal defect. *Am J Cardiol* 1979;43:816-9.
11. Tajik AJ, Seward JB, Hagler DJ, Marr DD, Lie JT. Two-dimensional real-time ultrasonic imaging of the heart and great vessels: technique, image orientation, structure identification, and validation. *Mayo Clin Proc* 1978;53:271-303.
12. Lange LW, Sahn DJ, Allen HD, Goldberg SJ. Subxiphoid cross-sectional echocardiography in infant, and children with congenital heart disease. *Circulation* 1979;59:513-24.