Morphological variations of fossa ovalis atrial septal defects (secundum): feasibility for transcatheter closure with the clam-shell device

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Abstract

Objective—To assess the morphology of fossa ovalis atrial septal defects (FOASD; secundum atrial septal defect) to determine in what proportion percutaneous closure with the clam-shell device might be feasible.

Design—Review of the intraoperative description of the morphology and size of FOASD.

Patients—106 consecutive patients with FOASD.

Setting—A regional children's cardiac referral centre.

Main outcome measures—Determination of morphology and size of FOASD.

Results—Morphological variations in the FOASD were found: (1) central FOASD, 70 (66%); (2) superior FOASD, 4 (3.7%); (3) inferior FOASD, 8 (7.6%); (4) posterior FOASD, 2 (1.9%); (5) subtotal absence of atrial septum, 3 (2.8%); (6) completely fenestrated FOASD, 10 (9.4%); (7) partially fenestrated FOASD, 9 (8.5%). Depending on its site, the edge of the FOASD may be close to important atrial structures such as the orifices of the superior and inferior caval veins, coronary sinus, and right pulmonary veins. The shape of the FOASD was usually oval with the major diameter ranging from 10 to 50 mm with a mean (SD) of 27.8 (0.93) mm. The minor diameter ranged from 4 to 30 mm with a mean (SD) of 15.3 (5.9) mm. The ratio of the major to minor diameter ranged from 1.0 to 5.0 with a mean of 2.01. Based on intraoperative measurement and description, it is postulated that it should be possible to close about 50% of these defects with the clam-shell device.

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Atrial septal defect is the fifth most common congenital heart abnormality at birth and occurs with a frequency of 3.2/10 000 live births.¹ Because of the effects of chronic volume overload of the right ventricle, surgical closure had been recommended for all but the smallest of defects.² The safety and benefits of surgical treatment have been well documented.³

After the successful introduction of interventional cardiac catheter techniques to treat congenital heart disease, methods were evolved for closure of atrial septal defects percutan-

eously.⁴⁻⁹ Rachkind⁵ pioneered a technique with a hook lined umbrella device whereas King and Mills used an interlocking double umbrella device.⁶ More recently techniques have been developed to close atrial septal defects in the fossa ovalis percutaneously with a clam-shell device that is an adaptation of the persistent ductus arteriosus occlusion device.⁷ Preliminary reports suggest that not all patients with atrial septal defects are suitable candidates for this procedure and they have highlighted the importance of determining the morphology and size of the defect if complete closure is to be obtained.⁷⁻⁹ The morphology of this defect has been described in the past¹⁰⁻¹² but the variability of the morphology of the fossa ovalis atrial septal defect (FOASD) has not been highlighted. A previous study to redress this was based on necropsy and animal specimens.⁷ We have reviewed the morphology of FOASD in a population of surgical patients to determine the variability of anatomy of this simple defect and to evaluate in what proportion of these patients percutaneous closure with the clam-shell device might be feasible.

Patients and methods

The case notes of 106 consecutive patients (age, median 5.92, range 0.98-16 years) operated on for repair of FOASD at our institution were reviewed. The intraoperative description and size of all FOASD as noted by the surgeons were recorded. The description of the FOASD was made with reference to a previous report by Sweeney and Rosenquist.¹² As this study was based on intraoperative observation in which a description of left atrial structures was not usually recorded only those findings in relation to right atrial structures and to the right pulmonary veins are reported. The atrial structures that were consistently described by our surgeons were the superior and inferior caval veins, the orifice of the coronary sinus, and the orifice of the right pulmonary veins (fig 1).

Where available, the dimensions of the FOASD were recorded. If a defect was oval in shape, the major diameter is defined as the larger of the dimensions recorded whereas the lesser dimension is the minor diameter. With the dimensions obtained at surgery, the ratio of the major to minor diameter was calculated.

Feasibility of closure by the clam-shell device of each individual defect was assessed by determining its size and the relation of its edge to the contiguous atrial structures. Transcatheter closure with the clam-shell was deemed

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Figure 1 Original drawing of central fossa ovalis atrial septal defect as drawn by surgeons. (70 (66%) patients). Atrial structures consistently described by the surgeons were the superior caval vein (SVC), inferior caval vein (IVC), coronary sinus (CS) and right pulmonary veins (RPV).

possible if the major diameter of the defect was equal to or less than 25 mm and when the edge of the defect was not close to important atrial structures.^{7,8} The atrial structures that were considered to affect the feasibility of transcatheter closure were the superior and inferior caval veins, the right pulmonary veins, and the coronary sinus.

Results
MORPHOLOGY OF ATRIAL SEPTAL DEFECT
We found significant variations in the morphology of the FOASD and have classified them into seven types based on their anatomy (table 1).

(1) Central FOASD
Most of the FOASD were single defect confined to the central portion of the fossa ovalis (70 patients, 66%). The limbus and the margins were well formed with the edge of the defect situated well away from important adjacent atrial structures. Feasibility for transcatheter closure with the clam-shell device in these defects depends on the size of the defect (fig 1).

(2) Superior FOASD
In four patients (3.7%) the defects were situated in the superior aspect of the fossa ovalis but were still within the confines of its borders. These defects did not have the morphological features of a sinus venosus defect and the pulmonary venous drainage was normal in all. The superior limbus of these four defects was well developed and the edge of the defects was not close to the orifice of the superior caval vein. As these defects were not close to important contiguous atrial structures, transcatheter closure would be possible if its diameter were acceptable (fig 2a).

(3) Inferior FOASD
In eight patients the atrial septal defect extended inferiorly and was close to the orifice of the inferior caval vein in six patients (5.7%) and coronary sinus in two patients (1.9%). In these two patients, the edge of the defects was described by the surgeon as within 2.0 mm of the orifice of the coronary sinus. In these eight patients the application of the clam-shell could obstruct the orifice of these structures (fig 2b).

(4) Posterior FOASD
The posterior limbus of the atrial septum was severely attenuated in two patients (1.9%), so much so that the atrial septal defect extended posteriorly with its posterior edge being formed by the posterior atrial wall. This posterior edge was very close to the right pulmonary veins in both these cases. These defects were considered to be unsuitable for closure with the clam-shell device as the defects lacked a posterior rim for fixation of the device and also it could obstruct the right pulmonary veins (fig 2c).

(5) Subtotal absence of atrial septum
Three patients (2.8%) were considered to have subtotal absence of the atrial septum as the limbus and margins were so severely deficient that the atrial septum seemed to be absent. No other morphological features, however, suggested an atrioventricular septal defect. The major diameter of these three defects was greater than 25 mm therefore rendering them unsuitable for closure by the clam-shell device. Furthermore they lacked a rim for fixation of the device (fig 2d).

(6) Completely fenestrated FOASD
Ten patients (9.4%) had either multiple fenestrations in the floor of the fossa ovalis or multiple strands of atrial septal tissue criss-crossing the defect. We have considered that these defects may not be suitable for transcatheter closure as it is often not possible to determine size accurately before the procedure (fig 2e).

(7) Partially fenestrated FOASD
The remaining nine patients (8.5%) had a partially fenestrated FOASD. They had, in addition to a central defect in the fossa ovalis, multiple fenestrations in the septum primum that were still present over the posteroinferior part of the fossa ovalis. Unlike the completely fenestrated FOASD, however, transcatheter closure could still be considered if the major diameter is suitable (fig 2f).

Morphological variations and dimensions of fossa ovalis atrial septal defects (FOASD)

Types of FOASD	No of patients (%)	Dimension of major diameter		
		≤ 2.5 cm	> 2.5 cm	Unknown
Central	70 (66.0)	29	20	21
Superior	4 (3.7)	3	1	0
Inferior	8 (7.6)	2	5	1
Posterior	2 (1.9)	1	1	0
Subtotal absence of atrial septum	3 (2.8)	0	3	0
Completely fenestrated	10 (9.4)	3	6	1
Partially fenestrated	9 (8.6)	2	5	2
Total	106 (100.0)	40	41	25

SHAPE AND SIZE OF FOASD
Almost all the defects were oval in shape with the major diameter in the superior to inferior orientation. Dimensions of the defects were recorded by the surgeons in 81 patients (table 1); the major diameters ranged from 10 to 50 mm with a mean (SD) of 27.8 (9.3) mm whereas the minor diameters ranged from 4 to 30 mm with a mean (SD) of 15.3 (5.9) mm. The ratio of the major to the minor diameters

Figure 2 Variations of fossa ovalis atrial septal defect FOASD, superimposed on surgeon's original drawing. Fig (2a) superior FOASD 4 (3.7%) patients, fig (2b) inferior FOASD 8 (7.6%) patients, fig (2c) posterior FOASD 2 (1.9%) patients, fig (2d) subtotal absence of atrial septum 3 (2.8%), fig (2e) completely fenestrated FOASD 10 (9.4%) patients, fig (2f) partially fenestrated FOASD 9 (8.9%) patients.

ranged from 1.0 to 5.0 with a mean of 2.01. Thus the shape of the FOASD may vary from a slit-like defect with a high ratio to a more circular defect with a ratio approaching unity.

In the remaining 25 patients whose FOASD dimensions were not recorded by the surgeons, the defects were usually described by the surgeons as small single central defects.

SUITABILITY FOR TRANSCATHETER CLOSURE

Of the 81 patients with a FOASD in whom the dimensions of the defects were known, 40 defects had a major diameter of 25 mm or less and therefore should theoretically be suitable for transcatheter closure. Amongst this group, however, six defects were considered to have unfavourable anatomy; two defects were close to the orifice of the inferior caval vein, one close to the right pulmonary veins, and three others were completely fenestrated. We therefore concluded that in 34 out of the 81 patients (42%) with FOASD, it might have been possible to close their defects with the clam-shell device.

In the remaining 47 patients it was postulated that transcatheter closure with the clam-shell device could not be performed safely.

Discussion

Our study confirms the great variability in the anatomy of the FOASD in our patients. Even though the most common variation is a single central defect located within the confines of the fossa ovalis, some of the defects extended beyond its margins. This observation is in agreement with reports based on necropsy

specimens.^{7,13} Depending on the degree to which it extends beyond the confines of the fossa ovalis, the defect may be close to the coronary sinus, inferior caval vein, or the right pulmonary veins; placement of a clam-shell device may then obstruct these structures. Some defects were very large with considerable attenuation of the limbus. They are similar to the common atrium haemodynamically but anatomically they are different and do not possess the other features of an atrioventricular septal defect.¹⁴ It would not be possible to close these defects with the clam-shell device because of an insufficient rim for fixation of the device. The fenestrated atrial septal defect poses a problem for the cardiologist contemplating transcatheter closure. Its margins are not well defined by transthoracic echocardiography and may lead to inaccurate sizing. Perhaps when its anatomy and size can be more reliably evaluated with the newer imaging techniques, such as transoesophageal echocardiography, it may then be possible to close some of these defects with the clam-shell device.

One of the most important criteria for suitability of transcatheter closure with the clam-shell is the size of the defect.⁷ With the maximum diameter of available device at 33 mm and with a 40 mm device in development, defects with a maximum major diameter of 25 mm should theoretically be suitable for closure with this technique.⁷ There must, however, also be an acceptable rim of atrial septal tissue separating the edge of the defect from its contiguous structures for proper fixation of the device. Evaluation before the

procedure must therefore use a technique that will allow accurate assessment of the anatomy of the defect, its surrounding contiguous structures, and accurate measurement of its size.

Cross sectional echocardiography from the subxiphoid position has been shown to be very accurate in defining the morphology of atrial septal defects; so much so that cardiac catheterisation is no longer needed before surgery.¹⁵⁻¹⁷ The addition of Doppler colour flow mapping has enhanced the diagnostic accuracy of cross sectional echocardiography and may improve the accuracy of diagnosis of fenestrated FOASD.¹⁸ Unfortunately, due to the limitations of the subxiphoid and transthoracic window, not all the information can be obtained by routine cross sectional echocardiography in every patient. Transoesophageal echocardiography enables the operator to image the atrial septum in total with the added advantages that the ultrasound beam is perpendicular to the atrial septum and without interference from lung tissue.¹⁹ Therefore a superior image quality and perhaps a more accurate estimation of the size of the defect may be obtained. Hellenbrand *et al* have suggested that the concomitant use of transoesophageal echocardiography may also increase the efficacy and safety of the transcatheter technique for closure of an atrial septal defect because of more accurate placement of the device.²⁰ We have conducted a preliminary study of transoesophageal echocardiography with a biplane probe to define the morphology of the atrial septal defect and have reported its accuracy.²¹ Transoesophageal echocardiography with the biplane transducer in our opinion is the most accurate method for defining the margins and sizing of an atrial septal defect, and its use is mandatory for accurate placement of the device at the time of the procedure.

In our study we have principally based our assessment of suitability for using the clam-shell device on the size of the defect and an analysis of contiguous right atrial structures and the degree of proximity of the right pulmonary veins but the other left atrial structures should also be considered as conceivably they may also be obstructed by the device. It must be pointed out that surgical measurements were made with an empty and relaxed heart. In real life the hearts are filled and the defects may be more circular.²²

In conclusion, we found significant variability in the morphology of FOASD that may influence the feasibility of transcatheter closure with the clam-shell device. Based on the 81 defects in which the dimensions were recorded by the surgeons, it was postulated that only about 42% of FOASD may be suitable for transcatheter closure. If, however, we include the remaining 25 patients with unknown

diameter but were described as having small defects, the final proportion suitable for transcatheter closure would be slightly greater than 50%. Preclosure evaluation should include accurate assessment of the maximum size of the defect and also the proximity of important atrial structures. The technique of choice for this purpose is most likely to be combined transthoracic and transoesophageal cross sectional echocardiography with colour flow Doppler.

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