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Spontaneous Closure of Atrial Septal Defects

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Abstract. Atrial septal defects (ASDs) are found more frequently in the pediatric population than in adults, and improved diagnostic techniques with echocardiography (2DE) and Doppler facilitate diagnosis so that repair is possible at an optimal time. The purpose of our investigation was to study the size of ASDs at diagnosis, how size changes during follow-up, and to explore the relationship between size at diagnosis and need for surgery. We reviewed the medical records of all patients in Iceland with the diagnosis of ASD born between 1984 and 1993. ASD was confirmed by 2DE in all patients and defects smaller than 4 mm were excluded. ASD size was measured by 2DE from subxyphoid long and short axis views. There were 91 patients—29 males and 62 females. Four patients died from causes other than the heart defect and had not been operated upon. Seven patients with ASD primum and sinus venosus defects were excluded from analysis. There were 29 patients with a 4 mm defect, 17 patients with 5 or 6 mm defects, 8 patients with 7 or 8 mm defects, and 26 patients had defects >8 mm. In the 4 mm group, in 26 patients (89%) the ASD closed spontaneously or decreased in size, and 1 patient had been operated upon. In the 5 or 6 mm group, 15 of 19 ASDs (79%) had closed spontaneously, and 2 patients (9.5%) had been operated upon. In the 7 or 8 mm group, 1 of 6 ASDs (16.6%) had closed spontaneously and 3 had been closed surgically. In the >8 mm group, 1 of 24 ASDs had closed spontaneously and 20 (91%) had been closed surgically. We conclude that defects smaller than 6 mm in diameter are very likely to close spontaneously although follow-up is necessary. Defects larger than 8 mm have a high probability requiring operative closure.

Key words: Atrial septal defects — Spontaneous closure

Atrial septal defect (ASD) is the second most common cardiac defect in children and adults [1, 12, 13, 15]. ASDs often escape diagnosis in childhood since the patients often have minimal symptoms and the auscultatory findings are often mistaken for an innocent heart murmur. Therefore, the patients are often adolescents or adults by the time the diagnosis is made [1, 12]. There are some investigations that deal with ASDs in younger children who have symptoms from the defect, thus indicating that ASDs can in fact lead to heart failure in infancy [3, 8, 9, 11, 14, 15, 18].

Anatomically the ASDs are classified as primum ASD, secundum ASD, sinus venosus ASD, and coronary sinus ASD [4, 5, 10]. The secundum ASDs are the most common and are located in the region of the fossa ovalis in the atrial septum [1, 10].

Most patients with ASDs of the primum and sinus venosus types need surgery, although very small defects might be left unoperated. Several studies have shown that secundum ASDs can undergo spontaneous closure. Large secundum ASDs need surgical closure, although the size of ASD is not well documented nor is the need for surgical closure. For large secundum ASD elective surgery is generally recommended at 4–6 years of age [7, 12, 13, 17]. Spontaneous closure does not seem to be affected by symptoms since spontaneous closure has been reported in patients with heart failure in infancy [2, 12, 13, 17]. Early publications dealing with spontaneous closure of ASD report the incidence of spontaneous closure to be 14–55% [7, 12, 13, 17]. Recently, the spontaneous closure rate has been given as between 66% and 92% [16].

Factors that influence spontaneous closure are (1) the size of ASD at diagnosis and (2) the age at diagnosis. Mody [13] found that ASDs closure is frequently in children diagnosed before 1 year of age, and Cockerham et al. [4] reported an increased closure rate in patients diagnosed before 2 years of age. Radzik et al. [16] concluded that the size of ASD at diagnosis is the best predictor of spontaneous closure. Radzik et al.'s paper dealt mostly with small defects and very few large ones.

The purpose of our study was to investigate the size distribution of ASDs in our population, the rate of spontaneous closure of ASDs, and the closure rate for different sizes of defects and to determine the probability of

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operation in relation to the size of the defect at diagnosis. This cohort of patients consisted of all children born in Iceland between 1984 and 1993 diagnosed with ASD. Thus, this is a cross section of an entire national population.

Materials and Methods

Patients

The cohort consists of all children born in Iceland during a 10-year period (1984 through 1993) diagnosed with ASD. We reviewed all medical records of patients diagnosed with secundum and primum ASD and sinus venosus ASD. We also reviewed two-dimensional echocardiograms (2DEs) of these patients. The 2DE was performed at diagnosis and was repeated from 3 months to 2 years following the diagnosis. Five patients with very large defects comprised one 2DE study and were operated on the basis of the results of the initial study. These patients were all 4 years old or older at the time of diagnosis.

The inclusion criteria for the study were as follows:

1. Minimal size of ASD was 4 mm. According to Radzic et al. [16], defects of 3 mm or less close spontaneously. The images were obtained in the subxyphoid long and short axes.
2. The ASD was not part of another associated defect that needed treatment (e.g., tetralogy of Fallot, transposition of the great arteries, tricuspid atresia, large ventricular septal defect [VSD], and endocardial cushion defect). ASDs that were associated with minor cardiac defects not meriting treatment (e.g., mild valvar pulmonary stenosis and small VSD) were included.

Thus, we were able to obtain information and 2DE studies on all patients diagnosed in Iceland during the 10-year period who had one or more 2DE examinations.

The following data were recorded: age at diagnosis and results of 2DE, i.e., size of ASD. For patients for which we had serial 2DE, we recoded the change in the size of ASD over time.

2DE and Doppler studies were available for all patients for the time period from 1984 to 1987, when we used a General Electric machine that used a freeze frame and recorded the smallest diameter. In 1987 a HP Sonos 500 unit was used and this was upgraded to a Sonos 1000 in 1990. We were able to improve the images considerably when color Doppler was introduced in 1987. However, no measurements were made directly on the color jet. The atrial septum was examined in subxyphoid long and short axes and measurements of the ASD were performed.

The size of the right ventricle (RV) was examined in all patients for evidence of volume overload, i.e., enlarged right atrium or RV. This was a subjective assessment by the echocardiographer, but no measurements were made. In the patients examined by the HP Sonos 100 unit, Q_p:Q_s ratio was calculated using the software package developed in our unit. Since we did not have these data for all patients the data was not included in this paper.

Analysis of Data

Patients with primum ASD and sinus venosus defects were excluded from analysis as were patients who died. The remaining 80 patients with secundum ASD were analyzed with regard to spontaneous closure.

Table 1. Size of defect and median age at diagnosis

Size of defect (mm)	Median age at diagnosis (months)
4	1.0
5–6	3.0
6–7	5.6
>8	14

Statistics

Statistical analysis was carried out as appropriate using the chi-square analysis, p values of <0.05 were considered statistically significant.

Results

Patients

An ASD was found in 91 patients—29 males and 62 females (a male:female ratio of 1:2.2). From January 1, 1984, to December 31, 1993, there were 43.809 live births in Iceland. Thus, the incidence of ASD was 2.1 per 1000 births. Four patients died, all from causes other than the heart disease; none of them had been operated on for the defect. These 4 patients were excluded. The age at diagnosis ranged from 1 week to 10 years, with a mean age of 12.1 ± 13 months.

Anatomy

There were 84 patients with a secundum ASD (92.5%), 4 patients with a sinus venous ASD (4%), and 3 patients with primum ASD (3.5%).

Age at Diagnosis

The median age at diagnosis differed depending on the size of the defect. In the group of patients with 4-mm defects the median age at diagnosis was 1 month, whereas in the group of patients with defects larger than 8 mm in diameter the median age at diagnosis was 14 months ($p = 0.001$). Table 1 shows the correlation between size of the defect and age at diagnosis.

Echocardiography

There were 80 patients with secundum ASD in whom we analyzed data. Throughout the follow-up period all the echocardiographic studies were performed by the same

Table 2. Summary of ASD size changes

Size of ASD (mm)	Number of patients	Number of patients with spontaneously closed ASD (%)	Number of patients decreased in size (%)	Number of patients unchanged in size	Number of patients in whom ASD increased in size and/or was closed surgically (%)
4	29	18 (62)[a]	8 (27)	2 (6%)	2 (6)
5–6	17	11 (64)	5 (29)	0	1 (7)
7–8	8	1 (12.5)	4 (50)	0	3 (37.5)
>8	26	0	2 (8)	0	24 (92)

[a] The percentages refer to the ratio within each size category.

person and on the same echo machine and were reviewed at the same time.

Size of ASD. There were 29 patients with a 4-mm defect, 17 patients with 5- or 6-mm defect, 8 patients with 7- or 8-mm defect, and 26 patients had a defect larger than 8 mm in diameter. Follow-up information was available for all patients.

1. 4-mm ASDs: When we examined the 4-mm defects 18 of 29 (62%) had closed spontaneously (mean follow-up 36 ± 12 months) and 8 had decreased in size (28%) to what could be regarded as a patent foramen ovale. Two defects remained at 4 mm and one had actually increased in size from 4 to 8 mm on follow-up.
2. 5- or 6-mm ASDs: There were 17 patients with a mean follow-up of 38 ± 16 months. In 11 patients the defects closed during the follow-up period, in 5 patients the defects decreased in size, and in 1 patient it increased in size: this patient was operated upon.
3. 7- or 8-mm ASDs: There were eight patients in this group with a mean follow-up of 48 ± 15 months. In one patient the defect closed spontaneously, and in four patients the defects decreased in size. Two patients were operated upon and in one patient the defect increased in size and will need to be operated upon.
4. >8-mm ASDs: There were 26 patients in this group with a mean follow-up of 35 ± 12 months. The ASD did not close spontaneously in any of these patients, although it did decrease in size in 2 patients (from 9 to 4 mm). The remaining 24 patients have undergone surgical closure of the defect. The changes in size of the ASDs are summarized in Table 2.

The probability of surgical closure of ASD is very low for the 4- to 6-mm defects but >90% in the >8-mm size group. The probability of spontaneous closure is presented in Fig. 1: there is a >90% probability of spontaneous closure in the 4-mm group and there were no spontaneous closures in the >8-mm group.

Discussion

In our population, ASD is the second most common cardiac defect after VSD. The incidence of ASD is somewhat higher than previously reported: 2.1 per 1000 live births. This most likely reflects improved diagnosis and application of 2DE to the diagnosis and evaluation of heart murmurs. The male:female ratio is similar in our study to that previously reported, with ASD being more common in females [1, 8].

The anatomy of the ASD was also similar in our population, with secundum ASD by far the most common defect and sinus venosus ASD and primum ASD less common, found in fewer than 10% of our patients.

In accordance with the study by Radzic et al. [16], we excluded 3-mm and smaller defects from our analysis, and we regarded these as patent foramen ovale since they almost invariably close before 18 months of age.

In this study larger defects compared to smaller defects were found in considerably older patients. The reason for a referral to a pediatric cardiologist for the majority of patients is the presence of a heart murmur. Since the heart murmur of ASD is often quite inconspicuous and patients often have a history of repeated respiratory infections, it is possible that the murmur was not detected when the patients were examined previously for such an infection, thus delaying diagnosis. The larger defects were found later, possibly indicating that at an older age most of the smaller defects have closed spontaneously and thus more patients have large defects.

When we examined the size of the defect in our patient population, it was interesting to note the number of large defects. In Radzic et al.'s [16] study, only 4% of the patients had ASDs larger than 8 mm, whereas 38% (33 of 87) of our patients had a defect of this size. Radzic et al. only deal with very young patients (diagnosed in

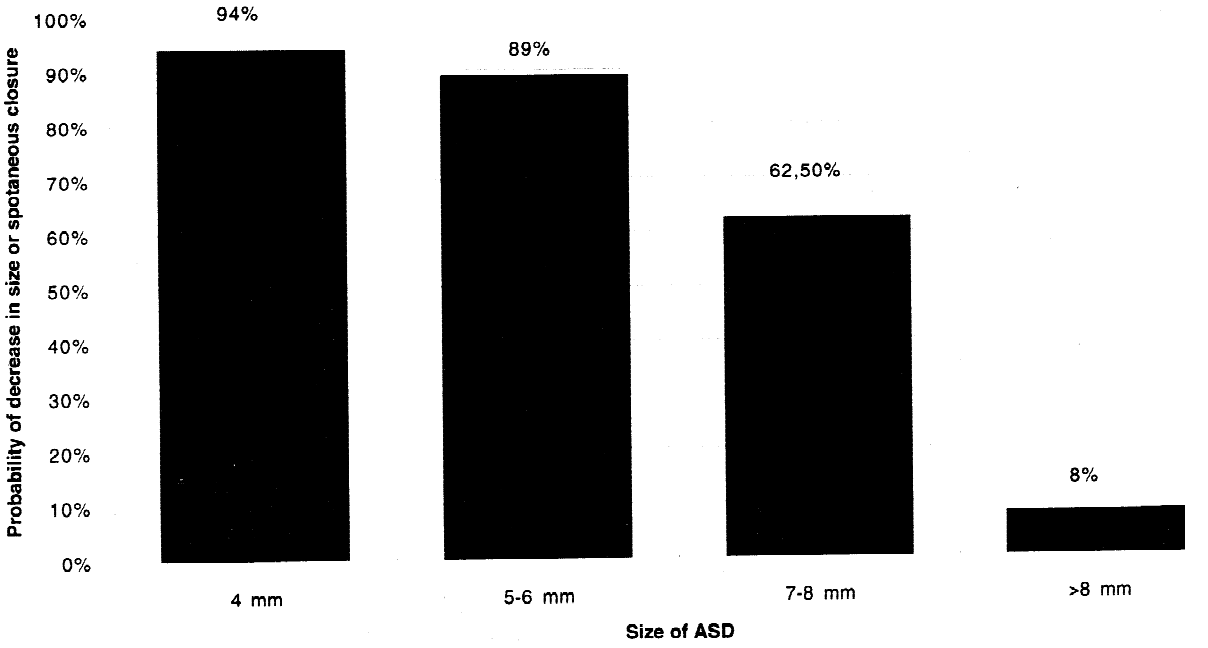

Fig. 1. The probability of spontaneous closure of ASD based on the size of the defect at diagnosis.

the first year of life) and because it is more common to find large ASDs in slightly older patients (median age 14 months in our study), a higher proportion of the patients in their study have small defects. On the other hand, our study dealt with all patients over a given period, thus reflecting reliable proportions.

The main trend is for small defects to close spontaneously and for the defects >8 mm to need to be closed surgically. However, occasionally there are defects that actually increase in size as the child grows. Thus, when a small ASD is found, it is prudent to repeat a 2DE at a later date, preferably before the age of 4 years.

The accepted indications for surgical closure of ASD are the presence of a defect with signs of volume overload such as a dilated right atrium and right ventricle [8]. In our group all patients with 6 mm or larger defects there was evidence of volume overload.

Several theories have been put forth regarding the mechanism of spontaneous closure. Cockerham et al. [4] and Edwards [6] suggested that the growth of the heart stretches the edges of an elliptoid defect, thus bringing the edges closer together. Cayler [3] suggested that continuous growth of the septum secundum occurs after birth and eventually closes the defect. The reason for an increase in the size of the ASD with growth is unclear. Possibly it is caused by stretching in the opposite direction to the short axis of the elliptoid shape of the defect, or it may be due to the continuous shunt keeping the defect open.

From our study we can predict the probability of an operation based on size of the ASD at the time of diagnosis (Fig. 1). For defects 4 mm in diameter the probability is very low, whereas for a defect larger than 8 mm at diagnosis there is a 95% probability of operation. Thus, we can better inform the parents about what to expect from the child's defect once we know the diameter of the ASD.

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