

The Clinical Course of Patients With Atrial Septal Defects

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Abstract

Background: Atrial septal defect (ASD) is a common congenital heart disease.

Objectives: The aim of this study was to assess the clinical course of ASD, as well as its frequency of spontaneous closure and regression of diameter.

Methods: In a study conducted from 2000 to 2011, 192 consecutive patients with an isolated ASD were followed up regularly with periodic echocardiographic evaluations according to a standardized protocol. The study was conducted in two series of patients: infants and children and adults. Infants were classified into three groups based on ASD diameter: small (≤ 5 mm), medium (6 - 9 mm), and large (> 9 mm). In the same manner, the ASD size in children and adults was categorized into three groups: small (≤ 10), medium (10 - 20 mm), and large (> 20 mm).

Results: The patients consisted of 76 (39.6%) males and 116 (60.4%) females. The mean age of patients at diagnosis was 16.12 ± 15.66 years (range 1 week - 75 years). The mean follow-up duration was 70 ± 9 months (range: 25 - 119 months). The most prevalent age group was adult patients (> 20 years), and most ASDs were of medium size (1 - 2 cm). The most common complication was mild pulmonary hypertension (31 cases, 16.2%), and Eisenmenger phenomenon was the rarest complication (2 cases, 1%). Overall spontaneous closure of ASD occurred in 42 (21.9%) cases. Spontaneous closure in cases with small defects occurred in 18 of 22 (81%) infants and 24 of 81 children > 1 years of age. Regression of ASD size occurred in 2 (9.5%) infants and 3 (3.7%) children. No spontaneous closure was observed in cases with a defect size > 10 mm, and no spontaneous occlusion was detected in adolescents or adults. Seventy-two percent ($n = 139$) of patients needed surgical repair or transcatheter closure of the ASD.

Conclusions: It is concluded that atrial septal defects < 6 mm typically close spontaneously, and ASDs measuring 6 - 9 mm may regress in infants and children. An ASD > 1 cm has little chance of spontaneous closure and may need surgical or device closure. ASD diameter and age at diagnosis are the most important predictors of spontaneous closure and the need for possible surgical or device closure.

Keywords: Atrial Septal Defect, Natural Course, Clinical Course, Spontaneous Closure

1. Background

ASD is more common in females than in males (1, 2). Several studies have reported spontaneous closure of ASD. Besides the age of diagnosis, the size of the defect has been reported to be an important factor in the likelihood of spontaneous closure. The rate of spontaneous closure has been reported to range from 14% - 55% (3). One study reported that spontaneous closure of ASDs smaller than 6 mm is very likely, and operative closure for defects larger than 8 mm has a high probability (4). Spontaneous closure does not seem to be affected by clinical signs and symptoms, because it has been reported in infants with heart failure (3, 5).

2. Objectives

The aim of this study was to determine the clinical course of ASD, its frequency of spontaneous closure, and size changes.

3. Methods

This was a retrospective study of consecutive infants and children with isolated ASD at a tertiary hospital. The data included patient age and sex and type, size, and complications associated with ASD. These data were collected through outpatient and hospital records. In infancy, the defects were classified as small (> 3 mm to ≤ 5 mm), medium (> 5 mm to 9 mm), and large (≥ 10 mm). After infancy, the ASD size was interpreted as follows: less than

10 mm as small, 10 - 20 mm as medium, and larger than 20 mm as large. All echocardiography was performed by the same pediatric cardiologist. Subcostal four chamber and sagittal views were used to measure ASD size. For statistical analysis, the chi-square test and Fisher's exact test were used, and P values < 0.05 was taken as significant.

4. Results

Among 2,200 cases of congenital heart disease, 216 (9.8%) patients suffered from ASD. Twenty four patients with incomplete data were excluded from the study. The study cases consisted of 116 (60.4%) females and 76 (39.6%) males. The mean age at diagnosis was 16.12 ± 15.66 years (range: 1 week - 75 years). The mean follow-up duration was 70 ± 9 months (range: 25 - 119 months). Among 192 ASD patients, 22 cases were infants, 81 were children, 25 were adolescents, and 64 were adults. Approximately 93.2% of ASD cases were of the simple secundum type, 2.6% were primary, 1.6% were sinus venous, and 2.6% were a combination of different ASD types.

Table 1 indicates the frequency of ASD complications in different age groups. It is shown that the most common complication was pulmonary arterial hypertension, and Eisenmenger phenomenon was the least common complication. As is clear from Table 1, most complications were related to medium and large defects. Table 2 depicts the frequency of ASD size in different age groups as follows: small ASD primarily occurred in infants, medium ASD in children, and large ASD in adults.

Table 3 shows the distribution of the type of closure of atrial defect related to age group. It can be seen that spontaneous closure occurred in infancy and childhood. Table 4 shows the frequency of spontaneous closure of ASD in different age groups.

5. Discussion

The highest number of spontaneous ASD closures occurred in patients younger than 2 years. This finding did not support the role of age in closure because ASD size played the main role in spontaneous closure. Spontaneous closure of medium and large ASDs did not occur, and the defect was recognized in older ages. In the current study, ASD constitutes 9.8% of congenital anomalies, which is comparable to the results of other studies. The ratio of female to male patients in this study was 1:5, which is lower than that in some studies (2) but in closer agreement with other reports (1, 6).

Among 22 cases of small ASD, 81% closed during infancy. This result is higher than that of studies that reported spontaneous closure rates of 14% - 55% (3, 7), but

concurrent with studies reporting closure rates of 66% - 92% (8).

In this study, the overall prevalence rate of hypertension in the pulmonary artery was 16.2%, and its incidence increased with increasing age among the four age groups. Its incidences in infants, children, adolescents, and adults were zero, 6.2, 16, and 34.2%, respectively. In another study, the risk of moderate to severe pulmonary hypertension in adults was reported as 27% (9). Therefore, aging can be an important factor influencing the high incidence of pulmonary arterial hypertension. Furthermore, pulmonary hypertension in adult patients in the current study was higher than in other studies, which may be due to delays in diagnosis or referral.

In our study, spontaneous closure occurred only during infancy and childhood. However, in adolescence and adulthood, no spontaneous closure was observed. Therefore, ASD in adolescents and adults should be followed up and if necessary repaired surgically or with transcatheter intervention. In our study, reduction in the size of ASD occurred in 12.7% of infants and children with an ASD size of less than 9 mm. In this respect, McMahon et al. have reported a result of 14% (9).

The median age of the patients with spontaneous closure was 2.25 years, and the maximum age at spontaneous closure was 3.9 years. Our study confirmed previous reports (10, 11) indicating that spontaneous closure can occur beyond infancy. Therefore, the window of opportunity for selective surgery can be determined according to patient age. None of our patients with an ASD size of 9 mm or less needed surgery. In our study, 72.4% of patients needed surgery or transcatheter closure, a finding similar to the results (77%) of Hanslik et al. (10) Ozcelik et al. (11) reported no cases of spontaneous closure for 8 mm ASDs (12). However, in our study, 8 (4.1%) cases of spontaneous closure of 9 mm ASDs occurred in childhood. Demir et al. (7) reported a spontaneous closure rate of 7.5% in 8 mm ASDs. Overall, it can be concluded that spontaneous closure may occur rarely in ASDs measuring 8 - 9 mm.

Heart failure occurred in 3.1% of the cases; however, none of the heart failure cases were in infants, and most of them were in adults. In other studies, the majority of heart failure cases were also reported in patients older than 40 years (13, 14).

In our study, 93 (48.4%) cases were closed by transcatheter and 46 (24%) by surgery. In the transcatheter group, one case died due to anaphylactic shock caused by the contrast agent. In the surgery group, there were two cases of death due to massive pericardial effusion and chronic intractable right heart failure.

In the transcatheter group, 8.2% of patients experienced complications, and 10.4% of patients from the sur-

Table 1. Distribution of Complications in Different Age Groups^a

Type of Complication	Age Group				Total
	Infants	Children	Adolescents	Older Ages	
Pulmonary hypertension	-	5 (6.2)	4 (16)	22 (34.4)	31 (16.2)
Heart failure	-	-	1 (4)	5 (7.8)	6 (3.1)
Eisenmenger	-	-	-	2 (3.1)	2 (1)
Mitral valve regurgitation	-	1 (1.2)	2 (8)	4 (6.25)	7 (3.7)
Arrhythmia	-	-	-	2 (3.1)	2 (1)
Uncomplicated	22 (100)	75 (92.6)	18 (72)	29 (45)	144 (75)
Total	22 (100)	81 (100)	25 (100)	64 (100)	192 (100)

^aPearson's chi-square test was used, P value = 0.001.

Table 2. Defect Size in Different Age Groups^a

Defect Size	Age Group				Total
	Infants	Children	Adolescents	Adults	
Small	16 (72.7)	7 (8.6)	-	-	23 (12)
Medium	4 (18.2)	52 (64.2)	17 (68)	28 (43.7)	101 (52.6)
Great	2 (9.1)	22 (27.2)	8 (32)	36 (54.41)	36 (56.3)
Total	22 (100)	81 (100)	25 (100)	64 (100)	192 (100)

^aPearson's chi-square test was used, P value = 0.001.

Table 3. Type of Closure of the Atrial Defect in Different Age Groups^a

Type of Closure	Age				Total
	Infants	Childhood	Adolescents	Adults	
Spontaneous	18 (81)	24 (29.6)	-	-	42 (21.9)
Reduced size	2 (9.5)	3 (3.7)	-	-	5 (2.6)
Catheterization	-	37 (45.7)	18 (72)	38 (58.4)	93 (48.4)
Surgery	-	14 (17.3)	6 (24)	26 (41.6)	46 (24)
No specific follow-up work	2 (9.5)	3 (3.7)	1 (4)	-	6 (3.1)
Total	22 (100)	81 (100)	25 (100)	64 (100)	192 (100)

^aPearson's chi-square test was used, P value = 0.001.

gical group suffered complications. The rates of mortality and complications were not significantly different between the aforementioned groups.

In other studies, complications in the surgical group were reportedly higher than in the catheterization group (15-17). Nevertheless, many studies have reported a higher rate of success, fewer complications, and shorter hospital stays for ASD closure with an Amplatzer device (18-22). Therefore, the closure of ASD through a catheter can be considered as an alternative method of surgical repair.

In our study, 2 (1%) patients had supraventricular arrhythmias, one adult patient had fibrillation, and another adult patient had an atrial flutter, indicating co-morbid condition rates similar to those found in other studies (2). It can be concluded that age is an important factor in the development of arrhythmia.

5.1. Conclusions

It can be concluded that babies suffering from ASDs measuring less than 5 mm do not require treatment be-

Table 4. Assessing the Frequency of Spontaneous Closure of Atrial Defects in Different Age Groups

Age, y	No. (%)
< 1	18 (42.9)
1.1 - 2	14 (33.3)
2.1 - 3	7 (16.7)
3.1 - 4	3 (7.1)
Total	42 (100)

cause spontaneous closure will likely occur in the first year of life. Infants with an ASD size of 5 - 9 mm may need echocardiography at the ages of 12 and 24 months. The probability of closure in ASDs 9 mm or larger is low, and parents should be reminded of the probable need for closure through surgery or catheterization. Children suffering from ASDs larger than 1 cm have a lower chance of spontaneous closure.

Footnote

Authors' Contribution: Study concept and design: Mostafa Behjati-Ardakani, Mohammad Golshan, Sedigheh Akhavan-Karbasi, and Seyed-Masood Hosseini; analysis and interpretation of data: M-Behjati-Ardakani; drafting of the manuscript: Mostafa Behjati-Ardakani, Mohammad-Amin Behjati-Ardakani, and Mohammadtaghi Sarebanhassanabadi; critical revision of the manuscript for important intellectual content: Mostafa Behjati-Ardakani, Mohammad Golshan, Sedigheh Akhavan-Karbasi, and Seyed-Masood Hosseini; statistical analysis: Mostafa Behjati-Ardakani and Mohammadtaghi Sarebanhassanabadi.

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