Atrial septal aneurysm in children

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Atrial septal aneurysm (ASA) has rarely been described in children. In this study, we evaluated the incidence and natural course of this anomaly in children. ASA was found in 30 patients (1%); 16 patients had type 1R and 14 patients had type 2L ASA. Twenty patients with ASA were followed: in four the ASA disappeared and in three patients with type 2L an atrial septal defect (ASD) developed during follow-up. The most common associated lesion was patent foramen ovale (PFO).

We conclude that ASAs in children are not rare lesions, and that these aneurysms, particularly type 2L aneurysms, should be followed for occurrence of ASD.

Key words: atrial septal aneuryms, echocardiography, children.

An atrial septal aneurysm (ASA) is a localized protrusion of the interatrial septum into the right or left atrium, or both, caused by bulging of the primary component of the septum through the oval fossa[1] Interatrial septal aneurysm has been detected with increasing frequency since the advent of two-dimensional echocardiography. The incidence of interatrial septal aneurysm has been reported to be 1.7% to 4.9% in children and infants[2,3]. Several cardiac defects have been noted to be associated with such aneurysms, including atrial septal defect and prolapse of the leaflets of the mitral and tricuspid valves[1,4,5]. Arrhythmias have also been reported both in postnatal patients and fetuses[1,6,7]. Its pathogenesis is controversial, but it is believed to depend on two predisposing factors: abnormal hemodynamic conditions and inherent structural weaknesses of the neonatal atrial septum[1,2]. The occurrence of an ASA after spontaneous closure of an atrial septal defect has also been reported.

The purpose of the study was to determine the prevalence of ASA and associated cardiac lesions in pediatric patients and the natural course of the defect, and to analyze the motion pattern of ASA.

Material and Methods

Echocardiographic examinations were performed in 3000 consecutive patients suspected of having cardiac abnormalities at the Pediatric Cardiology Unit between January 1993 and October 1998. Patients were included in the study if the interatrial septum could be satisfactorily visualized from both the subcostal and apical four chamber views. Ages of subjects ranged from 2 days to 11 years. Echocardiographic studies were performed by the same pediatric cardiologist using either a TOSHIBA SSA 160A with 2.5, 3.25, and 3 MHz or an ATL-HDL 3000 CW with 3-5 and 4-7 MHz transducers.

Each patient was evaluated for cardiovascular disease clinically and by chest radiography, electrocardiography (ECG), two-dimentional echocardiography (2DE), and color-flow Doppler echocardiography. Two types of atrial septal aneurysms were defined: type 1R if the bulging were in the right atrium only, and type 2L if the bulging were in the left atrium only (Figs 1 and 2)[8]. Each patient with an ASA was evaluated for coexistent atrioventricular valve prolapse. Evaluation of the interatrial shunts and diagnosis of accompanying cardiac anomalies were performed by Doppler echocardiography and contrast echocardiography (with D-galactose 300 mg/ml).

Patients with ASA were reevaluated serially by clinical examination, 2DE, and Doppler echocardiography. At each follow-up visit, the size and motion pattern of the ASA were compared to its appearance at initial diagnosis.

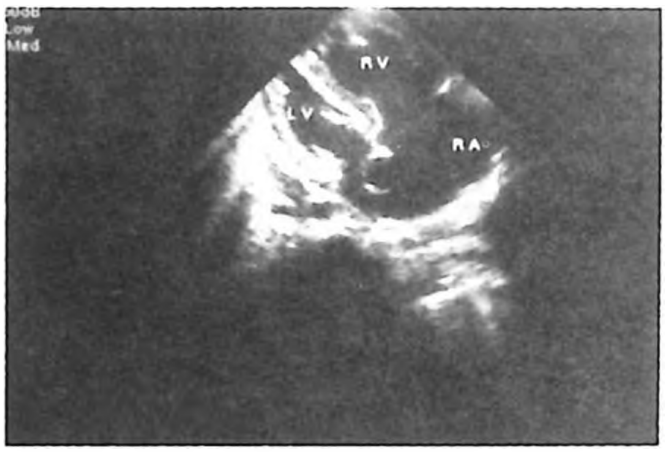

Fig. 1. Case 1. Two-dimensional echocardiogram in apical four-chamber view. Atrial septal aneurysm protruding into left atrium is noted.

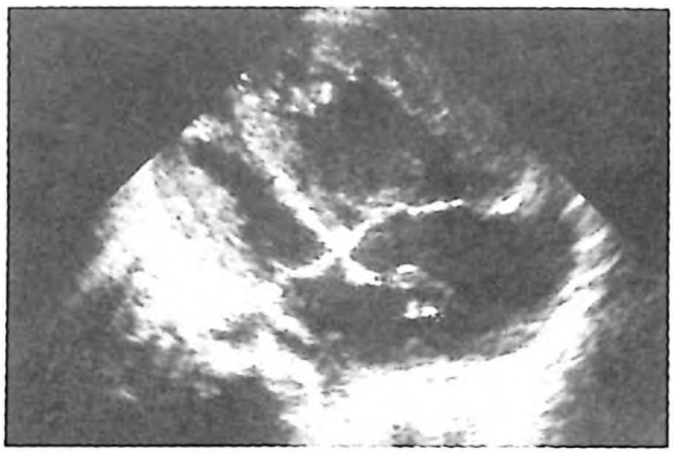

Fig. 2. Case 22. Two-dimensional echocardiogram in apical four-chamber view. Atrial septal aneurysm protruding into the right atrium is noted.

Results

Atrial septal aneurysm was found in 30 of 3000 patients (1%). There were 13 females and 17 males. The age at diagnosis of these ranged from 2 days after birth to 11 years. The follow-up period of these patients ranged from 4 to 31 months. Follow-up echocardiography was performed every four or five months once an aneurysm had been detected.

Sixteen patients had type 1R and 14 patients had type 2L ASA. Initially five patients showed an isolated aneurysm of the atrial septum. Three of these patients with type 2L ASA developed an atrial septal defect (ASD) during the follow-up period, one at the 6th month, one at the first year and one at the 18th month. All three patients were operated, for the defect. When associated with ASD, the direction of the ASA motion and that of the shunt found by Doppler echocardiography were similar. All other patients showed associated cardiac abnormalities (93%). The most common lesions were patent foramen ovale (PFO, 36.6%) and ventricular septal defect (VSD, 23.3%). The clinical and echocardiographic findings of the patients with ASA are summarized in Table I. No patient had associated atrioventricular valve prolapse. In 10 of 30 patients, follow-up tests were not performed. Four of these patients were lost to follow-up, one died due to respiratory failure and five patients were recently diagnosed. Disappearance of ASA was found in four of our patients (13.3%).

Table I. Characteristics of Patients With Atrial Septal Aneurysm

Patient	Age at Dx	Sex	Associated cardiovascular disease	Direction of ASA protrusion	Length of follow-up (mo)	Follow-up		Extracardiac findings
						ASA Dis	ASA Flow	
1[a]	10 mos	M	ASD	LA → RA	11 mos	0	No change	
2	7 mos	M	PFO	LA → RA	9 mos	0	No change	
3	52 days	M	PFO	RA → LA	4 mos	+	Dis	
4	9 mos	F	Cardiomyopathy	LA → RA	4 mos	+	Dis	
5	5 mos	F	PDA	RA → LA	7 mos	0	No change	
6	2 days[d]	M	PDA, VSD	RA → LA	31 mos	+	Dis	
7	7 mos	M	VSD	RA → LA	27 mos	0	No change	
8	3 mos	M	VSD	RA → LA	24 mos	0	No change	
9	9 days	M	PFO, VSD	RA → LA	16 mos	0	No change	OI
10	40 days	M	PDA	RA → LA	NF			MRD
11	16 mos	F	PFO	RA → LA	24 mos	0	No change	
12	4 mos	F	PFO	LA → RA	23 mos	0	No change	
13	3 years	F	PFO	RA → LA	NF			
14	3 mos	M	PFO	LA → RA	12 mos	0	No change	
15	3 mos	M	None	RA → LA	Ex			
16	43 days	M	PFO	LA → RA	6 mos	0	No change	
17	10 days	F	TF	RA → LA	20 mos	0	No change	Down syndrome

Table I. Continued

Patient	Age at Dx	Sex	Associated cardiovascular disease	Direction of ASA protrusion	Length of follow-up (mo)	Follow-up		Extracardiac findings
						ASA Dis	ASA Flow	
18	2 mos	F	VSD	LA → RA	Recently			
19	3 days	F	PFO	RA → LA	14 mos	+	Dis	
20	11 years	F	TF	LA → RA	NF			
21	5 mos	M	VSD	LA → RA	Recently			Down syndrome
22	5 mos	M	PFO	LA → RA	16 mos	0	No change	Down syndrome
23	2 mos	F	TF	LA → RA	Recently			
24	18 days	M	VSD	LA → RA	Recently			
25	9 mos	F	None	LA → RA	NF			
26	6 mos	F	PFO, TF, PS	LA → RA	Recently			
27	18 mos	M	TF	RA → LA	24 mos	0	No change	CAH
28	34 days	F	None	LA → RA	Recently			
29[b]	16 mos	M	ASD	LA → RA	16 mos	0	No change	
30[c]	7 years	M	ASD	LA → RA	3 years	0	No change	

ASA : atrial septal aneurysm.
ASD : atrial septal defect.
Dis : disappearance.
Dx : diagnosis.
LA : left atrium.
NF : no follow-up.
PDA : patent ductus arteriosus.
PS : pulmonary stenosis.
RA : right atrium.
TF : tricuspid failure.
VSD : ventricular septal defect.
PFO : patent foramen ovale.
CAH : congenital adrenal hyperplasia.
OI : osteogenesis imperfecta.
MRD : multiple renal dysplasia.
Ex : Exitus.
a : ASD ocurred at the 6th mo.
b : at the first year.
c : at the 18th month.
d : This patient is term infant.

Associated noncardiac anomalies were found in seven patients: four Down's syndrome, one osteogenesis imperfecta, one multicystic renal dysplasia, and one congenital adrenal hyperplasia (3-β hydroxysteroid dehydrogenase deficiency).

Discussion

Atrial septal aneurysm prevalence might have been underestimated in the past because of absence of symptoms and lack of noninvasive techniques for its diagnosis. 2DE is currently the best method for in vivo diagnosis of ASA[1,6,9,10]. Several studies have described ASA and its incidence in adults and children[1-3,6]. The reported incidence of ASA in the literature varied depending on the age of the study population and on the dignostic method used. Hanley et al.[1] found a higher incidence of ASA in children less than nine years old, very low incidence in adolescents and young adults, and increased incidence in older patients. Wolf et al.[2] identified ASA in 1.7% of infants and children who had undergone an initial diagnostic 2DE evaluation. Shiraishi et al.[3] reported the prevalence as 0.9% in children and 4.9% in infants. Brand et al.[11] showed an ASA incidence of 1% in infants and children. Rice and colleagues[7] found a very high incidence (64%) of ASA in fetal echoes of patients who were referred for the evaluation of fetal arrhythmia, compared to a 26% incidence in the fetuses referred for ruling out congenital heart disease. The prevalence of ASA detected by echocardiographic examination in adults is 0.2-0.5%[1-9]. This discrepancy between infants and adults implies the atrial septum is easily visualized in infants compared to adults, and that some of the aneurysms in infancy may disappear spontaneously with growth. ASA incidence was 1% in our study and similar to the general population is still unknown, since both present and previous studies specifically examined individuals with suspected cardiovascular disease.

Most previously published reports of ASA in children were associated with cardiac anomalies, such as ASD[1,10], tricuspid atresia[13], hypoplastic right heart syndrome[14] and transposition of the great vessels[16]. In adults, the associated cardiac defects were mitral valve prolapse[9], mitral stenosis or regurgitation[1], aortic stenosis and ASD. Rarely ASA occurs as an isolated pathologic finding. Pathologic examinations of ASA suggest a frequent association of this entity with interatrial communication. Mügge et al.[16] found that interatrial shunts, in particular foramen ovale, were the most common

abnormalities associated with ASA in adults. Barbosa et al.[17] in a mixed series of adults and children reported a 50% ratio of ASD among ASA patients. Brand et al.[11] showed in children that the most common associated lesion was atrial septal defect (69%); other associated cardiac lesions were VSD (28.5%), pulmonary stenosis (PS) (14%), patent ductus arteriosus (PDA) (11.5%) and coarctation or interruption of the aorta with subaortic membrane (6%). In our study, 93% of patients with ASA had additional cardiac anomalies; 7% occurred as an isolated lesion. Most frequently these included PFO (36.6%), although more complex defects were also found. Other associated cardiac lesions were VSD (23.3%), PDA (10%), (F) (16.6%), ASD (10%), and cardiomyopathy (3.3%). As noted in preious reports, ASA was often associated with interatrial communication (46.6%, 11 PFO, 3 ASD). In adults, thrombus formation, systemic emboli, atrioventricular and pulmonary vein obstructions, and atrial tachyarrhythmias were described in patients with ASA. No such complications were noted in our patients.

In our study, there were four children with Down's syndrome among ASA patients. Barbosa et al.[17] reported one Down's syndrome in 14 ASA patients. Further investigations are necessary to reveal the true relationship and incidence of ASA in patients with chromosomal abnormalities.

Several studies have suggested that interatrial septal aneurysms may be an initiating mechanism of atrial arrhythmis in adults and children. Miga et al.[18] suggested that ASAs are not a predisposing factor for the development or recurrence of atrial arrhythmias in infants. They appear to be an incidental finding without significant long-term complications. In our study, none of the patients had a history of arrhythmias.

Various mechanisms of the formation of ASAs have been postulated[1,2,10]. Wolf et al.[2] suggested that ASA formation in infants is related to abnormal hemodynamic conditions in the atrium (structural heart disease or arrhythmias) and inherent structural weaknesses of the neonatal septal tissue. The occurrence of an ASA after spontaneous closure of an ASD has also been reported[11]. Moreover, association of ASA with mitral valve prolapse or Marfan's syndrome implies that congenital connective tissue laxity is also responsible for ASA formation[12]. No patient in this study had atrioventricular valve prolapse or evidence of a systemic connective tissue disorder that might account for ASA formation. These findings suggest that such an aneurysm develops from other causes.

The majority of ASAs in infants will involute or completely resolve with age and intracardiac growth once normal atrial hemodynamics have been established. Brand et al.[11] found that the rate of the disappearance of ASAs was 45% in their patient. The aneurysm disappeared in most patients who had spontaneous closure of an associated ASD. Awan et al.[10] also described a case of spontaneous closure of an ASD with disappearance of ASA. In our study, ASA disappeared in four patients (13.3%). We observed that an ASD occurred in three patients during follow-up initially having type 2L ASA. Contrary to the literature, we observed that ASA, particularly type 2L aeurysms, may cause ASD.

In conclusion, ASA is not rare among children and it must be followed since type 2L ASA may cause ASD. In addition, interatrial shunts are frequent in ASA patients, and should be evaluated by Doppler and contrast echocardiography.

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