SYMPTOMATIC ATRIAL SEPTAL DEFECT IN INFANCY

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ABSTRACT

Isolated ostium secundum atrial septal defect (OS-ASD) may rarely lead to severe symptoms in infancy. Over a period of four years, 12 infants admitted to our hospital with severe congestive heart failure had an isolated OS-ASD. Their echocardiographic findings, clinical course, and outcome were outlined. All 12 infants had atypical findings including a holosystolic murmur parasternally and absence of wide fixed splitting of the second heart sound, and none were clinically suspected to have an ASD. M-mode echocardiography in all infants showed a greater than normal diastolic right ventricular internal diameter in the absence of pulmonary hypertension, indicating an increased right ventricular distensibility. Ten out of 12 infants responded well to medical decogestive treatment, and none developed pulmonary hypertension. Two infants who failed to improve with drugs, successfully underwent surgical closure, while two other infants revealed clinical and echocardiographic evidence of spontaneous closure.

Key words : *Atrial septal defect, Infancy, Ostium secundum.*

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An isolated ostium secundum atrial septal defect (OS-ADS) is a common cardiac malformation, accounting for 8% of congenital heart defects(1). It usually goes unnoticed as symptoms are absent, and physical signs are subtle. An important deviation from this usual pattern, are the symptomatic infants with OS-ASD, who comprise 5% of all patients with OS-ASD(2). This report outlines the clinical and echocardiographic profile, and outcome in 12 such cases who developed symptoms in infancy.

Material and Methods

Over a period of four years (1985-1988), 12 infants with OS-ASD presented with congestive heart failure. Details of history and clinical examination were recorded, and an *X*-ray chest and electrocardiogram were done in all cases. The diagnosis of OS-ASD was established on M-mode and two-dimensional echocardiography. All 12 infants were initially treated conservatively with chronic digoxin and diuretic therapy. Follow-up evaluation included clinical response, growth monitoring and six-monthly echocardiography. Two infants who responded poorly to medical thereapy underwent cardiac catheterization. The echocardiography findings in four of these infants were compared with those in four asymptomatic infants, incidentally detected to have an OS-ASD.

Results

The basic patient characteristics, mode of presentation and the clinical, *X*-ray and ECG findings are summarized in *Table I.* Six out of 12 infants failed to thrive, and two of these were preterm infants. All 12 patients had atypical findings in the form of a holosystolic parasternal murmur, and three had, in addition, a tricuspid diastolic flow murmur. None had a wide fixed splitting of

the second heart sound. ECG showed right axis deviation in all, with the frontal axis ranging from 45° - 210°. All infants showed a high R : S ratio with increased intrinsicoid deflection, and inverted T waves in V_1 and V_2. *Table II* gives details of the echocardiographic features in all these patients. The sizes of the OS-ASD ranged from 3-8 mm. M-mode echocardiography of the pulmonary valve showed presence of 'a' wave and normal EF slope in all, indicating absence of pulmonary hypertension. In four infants, fluttering of the tricuspid valve was seen, and none had any associated cardiac anomalies. All infants had diastolic right ventricular internal dimensions [RVID(D)] higher than normal (*Table II*). RVID(D) in four randomly chosen cases of symptomatic ASD was evidently greater than that in four asymptomatic infants with OS-ASD. However, the number of cases was too small for statistical validation. Cardiac catheterization done in two infants showed increased pulmonary : systemic blood flow ratios with normal pulmonary vascular resistance.

Ten of the 12 infants responded well to medical decongestive therapy with fairly good physical growth, and reduction in the frequency of respiratory tract infections; however, none developed pulmonary hypertension. The two infants who failed to improve with drugs, were subjected to corrective surgery, at 7 and 9 months of age, respectively, and are since doing well. Two patients on follow-up have shown clinical and echocardiographic evidence of closure (*Fig.)* while in three others the character of the murmur has changed from holosystolic to midsystolic.

TABLE I–*Patient Characteristics in Infants with Symptomatic Atrial Septal Defect*

Case	Age at diagnosis (months)	Symptoms	Murmur	P_2	ECG	CT ratio	Clinical diagnosis
1	2	CHF	HSM/TDFM	Single	RAD + RVH	0.57	PDA
2	2	CHF/FRI/FG	HSM	Single	RAD + RVH	0.59	VSD
3	4	CHF/FRI/FG	HSM	Single	RAD + RVH	0.64	VSD
4	1	CHF	HSM	Normal	RAD + RVH	0.62	VSD
5	3	CHF/FRI/FG	HSM/TDFM	Single	RAE + RVH	0.64	TAPVR
6	3	CHF/FRI/FG	HSM	Single	RAE + RVH	0.58	PS
7	2	CHF/FRI/FG	HSM/TDFM	Single	RAD + RVH	0.60	VSD
8	3	CHF	HSM	Single	RAE + RVH	0.65	VSD
9	2	CHF/FRI/FG	HSM/TDFM	Normal	RAD + RVH	0.60	VSD
10	2	CHF	HSM	Single	RAD + RVH	0.64	VSD
11	2	CHF	HSM	Single	RAD + RVH	0.64	VSD
12	1	CHF	HSM	Single	RAD + RVH	0.55	VSD

CHF - Congestive heart failure
FRI - Frequent respiratory tract infections
FG - Failure to grow
HSM - Holosystolic murmur
TDFM - Tricuspid diastolic flow murmur
RAE - Right atrial enlargement
RAD - Right axis deviation
RVH - Right ventricular hypertrophy

TABLE II—*Echocardiographic Findings in Symptomatic Infants with ASD*

Case No.	RVID(D)* (mm)	IVS motion	Tricuspid valve
1.	17	Flat	Diastolic flutter
2.	18	Paradoxical	Normal
3.	Not measured	Paradoxical	Normal
4.	22	Paradoxical	Normal
5.	32	Paradoxical	Diastolic flutter
6.	27	Paradoxical	Normal
7.	24	Paradoxical	Diastolic flutter
8.	18	Flat	Normal
9.	20	Paradoxical	Diastolic flutter
10.	18	Flat	Normal
11.	18	Paradoxical	Normal
12.	20	Flat	Normal

RVID(D) : Right ventricular internal diameter (diastole)
IVS : Interventricular septum
* Normal for age : < 15 mm

TABLE III—*Comparison of Size of Septal Defect and RVID in Symptomatic and Asymptomatic Infants with ASD*

Symptomatic			Asymptomatic		
Case No.	Age (mo)/ Size of defect	RVID(D) (mm)	Case No.	Age (mo)/ Size of defect	RVID(D) (mm)
1.	2/Moderate	17	(*i*)	2/Large	11
2.	3/Large	32	(*ii*)	5/Large	14
3.	3/Large	27	(*iii*)	9/Large	11
4.	2/Moderate	20	(*iv*)	4/Moderate	12

RVID : Right ventricular internal diameter (diastole)
Moderate defect : 5-7 mm
Large defect : >7 mm

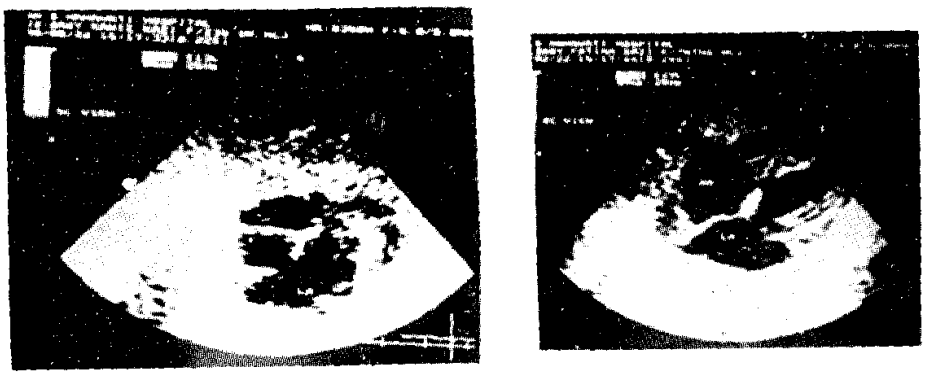

Fig. Showing natural closure of secundum defect on follow up.

Discussion

OS-ASD may rarely present in infancy with congestive heart failure. There has been no such report in the Indian literature. The symptomatic infant having a large left to right shunt through the ostium secundum defect poses an important diagnostic challenge. The cardiac auscultatory findings include, variable splitting of the second heart sound, increased intensity of pulmonic component, a parasternal holosystolic murmur and a tricuspid diastolic flow murmur. The holosystolic murmur reflects tricuspid regurgitation due to right ventricular failure and dilatation resulting in dilatation of the tricuspid annulus. As the right ventricle at this age occupies the apex, this murmur is heard at the left lower sternal border, mimicking a ventricular septal defect(5). Likewise the tricuspid diastolic flow murmur, can be misinterpreted as the diastolic component of a ductus murmur. Wide fixed splitting of the second heart sound is conspicuously absent(6).

The exact mechanisms by which this left to right, atrial level shunt makes some patients symptomatic at an earlier age has not been elucidated. Neither the magnitude of the shunt, nor delayed fall in pulmonary vascular resistance, satisfactorily explain the premature onset of right ventricular failure(7). However, a greater than usual right ventricular distensibility may be a cause for early left to right shunting. Normally, during infancy, the right ventricle due to its hypertrophy, is less distensible than the left ventricle. It has been suggested that a decreased amount of non-contractile cellular elements, or reduced myocardial proline concentration may be responsible for the increased distensibility in symptomatic patients(8). This has been reflected in our findings of increased RVID(D) in all symptomatic patients, in contrast to normal dimensions in asymptomatic infants, irrespective of the size of the septal defect.

Vigorous medical management, including digitalis, diuretics and antibiotics, is usually adequate to maintain them through the critical first year of life. It has been seen that, as they grow older, the pulmonary infections subside and a growth spurt occurs(4). Early surgery may be required in only a small subgroup of patients with resistant failure. Interestingly, some of the OS-ASD can undergo spontaneous closure even up to the age of 8 years(9,10), as was seen in two cases in the present series.

In conclusion, OS-ASD should be considered in the differential diagnosis of infants presenting in cardiac failure, with failure to thrive, or cyanosis related to pulmonary infection. It is preferable to manage such infants conservatively, as it is not uncommon for them to experience spontaneous closure.

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NOTES AND NEWS

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IAP Mysore is hosting the XII Karnataka State Conference of IAP on 5th, 6th and 7th November, 1993 at Mysore. The CME will be held on 5th November, 1993 and the Conference on 6th and 7th November, 1993.

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