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## ATRIAL SEPTAL DEFECT IN INFANTS

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SINCE the first mention of a case of atrial septal defect in a report by Morgagni in 1765, over 300 cases had been recorded in the literature by 1957.<sup>1</sup> It might be estimated that since then probably an equal number of cases have been reported.<sup>2,3,4,5</sup> Atrial septal defects are thought to be, by most authors, among the commonest of congenital heart defects. Wood cites an incidence of 18%, Keith 7%, and in our material, atrial septal defects of the secundum variety represent 10.04% of the 3,786 patients studied in the last 10 years. In the discussion to follow, when referring to atrial septal defects, we will mean the ostium secundum variety and will exclude, by the accepted criteria,<sup>7,8,9</sup> endocardial cushion type lesions.

It would appear that atrial septal defects are compatible with a long life span with reports in the literature of patients living up to 70 and 80 years.<sup>10,11</sup> As a matter of fact, Wood stated (personal communication) that in inmates of institutions for the aged, the percentage of atrial septal defects was not appreciably different from that noted in the general population. The evidence at present suggests that approximately 50% of cases of uncomplicated atrial septal defects survive more than 40 years.

Although the natural history of the defect is well documented in older patients<sup>10,11</sup> not much is known about the course in infancy and early childhood since, unlike most congenital anomalies of the heart, it is decidedly unusual for an atrial defect to manifest itself early. In Braudo's series of 31 patients, only 16% had a history of mur-

mur at birth and in 23% a murmur was not discovered until age 5 years or older.<sup>12</sup> As a matter of fact, there has been only one publication so far reporting on infants with atrial septal defects of the secundum variety.<sup>5</sup>

The purpose of this presentation is to describe the clinical and physiologic picture of atrial septal defect in a sizeable group of infants seen at the Children's Hospital Medical Center, to report their course during some years' follow-up, and to search for any distinguishing features between these individuals and those in whom atrial defects are discovered later in life.

### MATERIAL AND METHODS

Autopsy material from January, 1955, to the present, September, 1962, was analyzed and of the better than 2,400 necropsies done at the Children's Hospital Medical Center during this period, there were no infants under 2 years of age who died of an uncomplicated atrial defect. Atrial defects were found as incidental lesions in infants succumbing from other diseases, but these were eliminated from the study because they never had any signs or symptoms referable to the atrial defect.

In analyzing the cardiac catheterization data during this same period, 1955 to 1962, 10 patients, 6 males and 4 females, appeared to fulfill the clinical and laboratory requirements for the diagnosis of secundum atrial defects. These were: (a) An increase in oxygen saturation between the superior vena cava and the right atrium of at least 15% in one set of samples or of 10% in two

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or more sets. (b) Absence of a right to left shunt (except Case 10, in whom the diagnosis was confirmed at surgery). (c) Electrocardiographic evidence of a clockwise frontal plane loop with a mean electrical axis to the right of  $0^\circ$ , thus excluding the endocardial cushion defect group. Three of these 10 patients were catheterized twice, and the period of follow-up ranged from 1 month to 7 years, averaging 26 months. Two of these patients have undergone surgical repair and all are living at the time of this writing.

## RESULTS

### Clinical Observations

Study of the family histories revealed congenital heart disease, a patent ductus arteriosus, in an aunt of one patient (Case 3).

Gestational and birth histories were unremarkable in all, and birth weights ranged from 5 lb (2.3 kg) to 8 lb 9 oz (3.71 kg).

Associated defects were found in four patients; pectus carinatum in one, hydrocephalus in another, micrognathia in a third, and pyloric stenosis in the fourth case.

Congestive heart failure was suspected, on the basis of tachypnea, tachycardia, and cardiomegaly, in 3 of the 10 cases, at 7 months, 12 months, and 13 months, respectively. In several instances, there was a question of co-existing pneumonia at the time congestive failure was thought to be present. That heart failure was probably present in these three babies was borne out, however, by the improvement in appetite and decrease in tachypnea and tachycardia which followed digitalization. Two of these patients continued to do well when digitalis was discontinued at 2 years and at 3½ years respectively. The third patient is now 4 years old and remains on digitalis (Case 7). No additional patients developed congestive failure during their follow-up.

A history of an increased number of severe respiratory infections was elicited in 3 of these 10 patients at the time the definitive diagnosis was made by cardiac catheterization, and one other patient catheterized early in life subsequently developed frequent respiratory infections.

Five children were in the 3rd percentile, or below, in weight at the time of the first cardiac catheterization. Two were in the 20th percentile, one in the 25th percentile, one in the 50th percentile, and the last in the 90th percentile. Three patients at or below the 3rd percentile in weight remained below the 10th percentile during follow-up periods averaging 31 months. Three patients who were above the 10th percentile in weight remained above the 10th percentile during follow-up, and in the remaining 4 patients not enough time has elapsed since their cardiac catheterization studies to assess their growth. The retardation in linear growth was less striking in that height was under the 50th percentile in all, but at or above the 10th percentile in eight.

A thrill, attributable to the gradient across the pulmonary valve, was palpable in three of the cases under discussion. The first heart sound was considered normal in all, while the intensity of the second sound at the second left interspace was described as increased in eight cases. Wide splitting of the second sound was heard in 7 of the cases, and in these the effects of respiration on the degree of splitting was commented upon in only 5 cases at the first examination; in 4 of them no respiratory variations were noted, and in the fifth the degree of splitting was thought to vary with the phases of respiration.

A systolic murmur, no louder than grade III in intensity, usually best noted at the second and third left interspace, was heard in all patients. The murmur was discovered in 8 patients before the age of 6 months and in one of each at 9 months and 21 months respectively.

An apical or lower left sternal border diastolic murmur, an inflow rumble,<sup>12,13</sup> was heard in 8 of the cases.

Figure 1 summarizes these findings.

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### Electrocardiographic Findings

Electrocardiograms were obtained in all cases, and the characteristic tracing of right axis deviation in the standard limb leads

with an rsR' pattern in the right precordial leads was found in most.<sup>4,15</sup>

There was an rsR' pattern in V<sub>1</sub> in 7 cases, an rsr' pattern in one, a qR pattern in another, and a pure R in the last case.

The mean electrical axis ranged from +95° to +110° (Fig. 2) and there was right ventricular hypertrophy in 6 cases evidenced by voltages in the right precordial leads exceeding the normal ranges determined by Alimurung and Massell.<sup>16</sup>

The direction of the frontal plane loop, as determined by a vectorial analysis of the scalar electrocardiogram, was inscribed in a clockwise fashion in the 9 patients in whom this analysis was possible. In the 10th, the mean electrical axis was perpendicular to the frontal plane.

Subsequently, the electrocardiograms remained essentially unchanged and only one patient whose electrocardiogram was at first normal developed right ventricular hypertrophy (Case 8).

#### X-ray Findings

X-rays were obtained on all patients at the time of cardiac catheterization. There was mild cardiac enlargement in 3 patients and moderate enlargement in 7 (Fig. 3). The pulmonary artery appeared normal in 2, mildly prominent in 4, and moderately prominent in the last 4 patients. The size of the pulmonary artery was difficult to assess because of the presence of thymic tissue in some and because the main pulmonary artery appeared to lie more medially than usual in some of the younger cases.

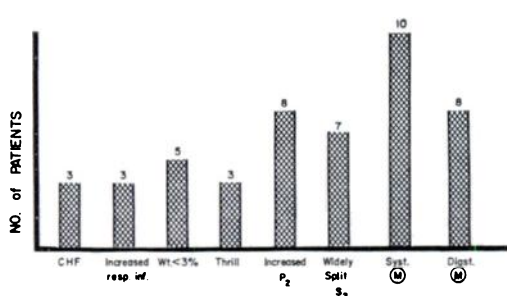


FIG. 1. Summary of the signs, symptoms, and physical findings in 10 infants with secundum atrial defects. CHF, congestive heart failure; M, murmur.

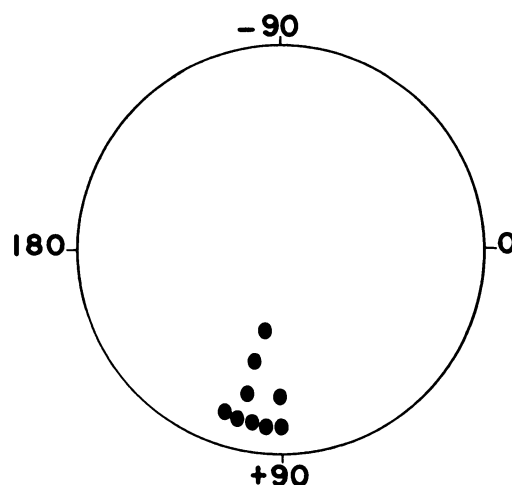


FIG. 2. The mean electrical axes determined by vectorial analyses of the scalar electrocardiograms. In one of the 10 patients, the mean QRS axis was indeterminate.

Pulmonary vasculature was felt to be slightly engorged in five patients and moderately engorged in five.

With follow-up, there was little change in the x-ray appearance except for one patient (Case 8) whose moderate enlargement became more marked.

#### Cardiac Catheterization Findings

Cardiac catheterization under mild sedation (Demerol, Phenergan, and Chlorpromazine mixture) demonstrated, by oxygen saturation data, an appreciable left to right shunt in all patients (Table I). A rise in oxygen saturation at the right atrial level might theoretically be due solely to anomalous pulmonary venous drainage, but the existence of an atrial defect was proven on cineangiograms by left atrial injections in 7 cases. In the remaining 3, the presence of an atrial septal defect was proven at surgery in one (Case 6), and by oxygen saturation data and virtual identity of left atrial and right atrial pressures in the other two (Cases 7 and 9).

In 4 cases (1, 2, 6, and 9), there was a small rise in oxygen saturation from the right atrium to the pulmonary artery raising the suspicion of an additional shunt at the ventricular level. This increase in saturation was not felt to be significant and was attributed to poor mixing in the right

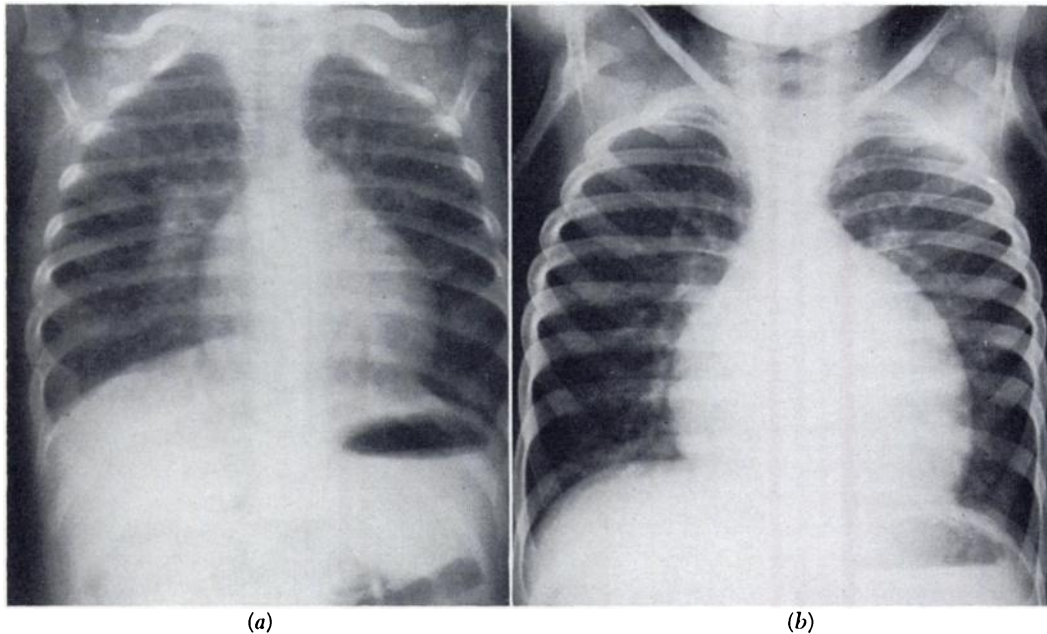


FIG. 3. Postero-anterior radiograms of patients 1 and 6, respectively, showing mild cardiac enlargement in (a), moderate enlargement in (b), and pulmonary vascular engorgement in both.

atrium. Ventricular defects were ruled out in three of these (1, 2, and 6) by left ventricular injections of radio-opaque dye which revealed no right ventricular filling. In the remaining patient (Case 9), the min-

ute rise in oxygen saturation plus the identity of left atrial and right atrial pressures tended to rule out the presence of a ventricular septal defect.

There was a small right to left shunt at

TABLE I  
CATHETERIZATION DATA

Patient	Age at Cath.	Saturations %					Pressures mm HG								Cineangiograms	
		SVC	RA	RV	PA	SA	SA	LA	RA	RV	PA	QP/QS	Pulm. Resist.	Syst. Resist.	Pulm. valve Gradient	LA injection demonstrating atrial L→R shunt
1 S.W.	5 mo.	60.7	83	86.9	86.4	96.5	90/	6 →4	32/5	28/10	3:1	1	—	4	Yes	Yes
2 S.S.	8 mo.	61	82.3	83.1	85.8	91.8	100/37	12 →7	59/8	30/17	3:1	<1	13	29	Yes	Yes
3 D.B.	8 mo.	62.9	78.7	77.6	78.3	92.9	90/45	6 →3	24/3	19/10	2.1:1	<1	16.4	5	Yes	Not done
4 J.T.	9 mo.	64.3	88.3	83.6	82	91.5	110/54	8 →6	41/8	28/4	2.5:1	<1	12	13	Yes	Yes
Cath #1	9 mo.	65.4	82.5	82.5	83.7	94.6	95/58	11.5→4.5	42/7	29/4	2.7:1	2	10.5	13		
5 B.L.															Yes	Not done
Cath #2	2 1/2 yr.	71.6	81.7	81.2	82.5	95.4	104/50	9 3	20/3	12/8	1.8:1	<1	15.2	8		
Cath #1	13 mo.	70.1	87.2	83.5	78	97	85/62	— 5	45/6	51/15	1.5:1	2.8	20	14		
6 C.V.								Pc mean							Not done	Yes
Cath #2	6 1/2 yr.	70.3	84.6	87.8	89.1	96.2	114/68	8 2	44/8	37/10	3.6:1	1.3	19.5	7		
7 F.F.	18 mo.	70.3	87.2	87.8	87.8	96.5	110/60	7 5	50/7	24/10	3:1	<1	16	26	Not done	Not done
8 P.M.	18 mo.	73	88	86.1	88.3	96.7	94/40	3 2.5	44/6	26/10	3:1	<1	10.6	18	Yes	Yes
9 D.W.	21 mo.	50.3	78.2	79	79.3	93.8	108/72	0 0	28/2	18/8	3:1	<1	33	10	Not done	Not done
Cath #1	2 yr.	75.7	88.3	87.5	89.1	95.4	110/60	7 6	43/6	28/10	2.9:1	1.5	23.8	15		
10 R.F.															Yes	Yes
Cath #2	5 1/2 yr.	65.1	89.9	88.6	89.1	93.8	106/66	9 →5	34/4	23/15	4:1	1.2	23	11		

→ withdrawal pressures

the atrial level in one patient only in whom the systemic arterial saturation was 93.8%, while the pulmonary vein sample was 98.4% saturated (Case 10). The diagnosis of a secundum defect in this patient was confirmed at operation when a large defect, 3.5 by 1.5 cm, was found posteriorly in the septum.

In Case 6, first studied in 1955, the relatively small pulmonary flow was calculated on the basis of one pulmonary artery sample only. That the size of the shunt may then have been underestimated is suggested by the fact that a repeat study, 5 years later and including several pulmonary artery samples, revealed a 3.6:1 pulmonary to systemic flow ratio. This was in keeping with the x-ray changes as well as the operative finding of a 3 cm-long defect in the inferior aspect of the atrial septum near the coronary sinus.

The pulmonary to systemic flow ratios were all over 2:1, except for Case 6 mentioned above. There was no appreciable pressure gradient between the atria in 8 patients. However, in 2 (Cases 2 and 5), there was an unexplained difference of 5 mm and 7 mm of mercury between the mean pressures of the two atria. We have as yet no information on the size of the defects in these two infants in whom the shunts were almost twice the size of the systemic flows.

Gradients across the pulmonary valve existed in all, with a range of from 4 to 29 mm of mercury. In light of the large pulmonary flow in each, the gradients were considered to be functional. Pulmonary artery pressure was not elevated, and pulmonary resistance in all was in the normal range. Values under one unit were found in six infants.

Repeat studies were undertaken in 3 patients from 2 to 5½ years later. Increased pulmonary flows were again found, and no major change in pressures was noted.

#### COMMENT

Since the diagnosis of secundum atrial defects has been infrequently made in in-

fancy, it seemed worthwhile to compare the clinical and hemodynamic features of these 10 infants with those well described in older children with atrial septal defect. It should be recognized that these findings may not be present in other infants with atrial defects who were not brought to the attention of the cardiologist. As in the children, our patients were found to have a hyperdynamic right ventricular impulse, a systolic murmur of moderate intensity at the upper left sternal border, and a widely split second sound. The splitting was uninfluenced by respiration in all but one of these, and a diastolic rumble was heard in most of the cases. Electrocardiograms and x-rays revealed the classical findings of the secundum defect.

Cardiac catheterization data revealed mild right ventricular hypertension and low pulmonary artery pressure despite the moderately large left to right shunts. A functional gradient existed across the pulmonary valve. The high pressure gradient between the atria in two cases is without explanation. As there is little information on the hemodynamics of secundum defects in infancy, the significance of this unusual gradient has yet to be determined.

It is generally believed that persistence of the fetal pattern of right ventricular and pulmonary artery hypertension prevents shunting of blood from left to right through an atrial defect early in life. Since the findings in these 10 cases were similar to those in the older patient, it can only be postulated that in these infants right ventricular compliances somehow approached levels usually found later in life.

#### SUMMARY

1. Ten patients between the ages of 5 months and 2 years with the secundum variety of atrial septal defect were diagnosed by cardiac catheterization.

2. The hemodynamic and physiologic data classically associated with secundum atrial defects were found except for the unexplained existence in two cases of a pressure gradient between the atria.

3. Most of these patients were relatively asymptomatic, and the few who developed congestive heart failure were controlled medically until the optimum age for elective repair was reached.

4. All 10 patients are alive at the time of this writing.

#### REFERENCES

1. Wagner, J., and Graham, G.: Atrial septal defect in children. *Brit. Heart J.*, 19:318, 1957.
2. Davidsen, H. G.: *Atrial Septal Defect*. Copenhagen: Ejnar Munksgaard, 1960.
3. Bedford, D. E.: The anatomical types of atrial septal defect. *Amer. J. Cardiol.*, 6:568, 1960.
4. Wood, P.: *Disease of the Heart and Circulation*. London: Eyre and Spottiswoode, 1956.
5. Hastreiter, A., Wennemark, J., Miller, R. A., *et al.*: Secundum atrial septal defects with congestive heart failure during infancy and early childhood. *Amer. Heart J.*, 64:467, 1962.
6. Keith, J. D., Rowe, R. D., and Vlad, P.: *Heart Disease in Infancy and Childhood*. New York: Macmillan Co., 1958.
7. Scott, R. C.: The electrocardiogram in atrial septal defects and atrioventricular cushion defects. *Amer. Heart J.*, 62:712, 1961.
8. Burchell, H. B., DuShane, J., and Brandenburg, R.: The electrocardiogram of patients with atrio-ventricular cushion defects. *Amer. J. Cardiol.*, 6:575, 1960.
9. Liebman, J., and Nadas, A. S.: The vectorcardiogram in the differential diagnosis of atrial septal defect in children: *Circulation*, 22:956, 1960.
10. Bedford, D. E., Papp, C., and Parkinson, J.: Atrial septal defect. *Brit. Heart J.*, 3:37, 1941.
11. Colmers, R. A.: Atrial septal defects in elderly patients. *Amer. J. Cardiol.*, 1:768, 1958.
12. Braudo, J. L., Nadas, A. S., Rudolph, A. M., *et al.*: Atrial septal defects in children. A clinical study with special emphasis on indications for operative repair. *PEDIATRICS*, 14:618, 1954.
13. Leatham, A., and Gray, L.: Auscultatory and phonocardiographic signs of atrial septal defects. *Brit. Heart J.*, 18:193, 1956.
14. Nadas, A. S., and Alimurung, M. M.: Apical diastolic murmurs in congenital heart disease. *Amer. Heart J.*, 43:691, 1952.
15. Barber, F. M., Magidson, O., and Wood, P.: Atrial septal defect with special reference to the electrocardiogram, the pulmonary artery pressure, and the second heart sound. *Brit. Heart J.*, 12:277, 1950.
16. Alimurung, M. M., Joseph, L. G., Nadas, A. S., *et al.*: The unipolar precordial, and extremity electrocardiogram in normal infants and children. *Circulation*, 4:420, 1951.

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