Symptomatic Atrial Septal Defect in Infancy

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SUMMARY
Atrial septal defect of the ostium secundum variety (ASD) is uncommonly diagnosed in infancy and rarely results in congestive heart failure (CHF). Eleven patients with uncomplicated ASD who developed CHF between 6 days and 13 months of age are presented.

The diagnosis of ASD was documented by cardiac catheterization in all 11 patients. Left-to-right shunt ranged from 52 to 80%. Right ventricular pressure was near systemic levels in two patients and at least somewhat elevated in the other nine. Total pulmonary resistance (TPr) was within normal limits in all 11. No left ventricular obstructive lesions were found. Although one patient had unusual thickening of the left ventricular myocardium, and the left ventricular cavity was enlarged in two other cases, no specific abnormality in left or right ventricular function could be defined.

Spontaneous closure of the ASD did not occur in any of our patients. Since spontaneous closure even of a large ASD can occur, however, it seems advisable to defer elective surgical closure until after 2 years of age if successful medical management can be achieved. In those patients in whom early surgery is not necessary, the clinical course usually becomes that of a typical ASD by age 2 years.

Additional Indexing Words:
Congestive heart failure Congenital heart disease

Atrial septal defect (ASD) is one of the common cardiac malformations, probably the most frequent. However, ASD is rarely diagnosed and even less commonly causes symptoms in infancy. Throughout this manuscript, the term ASD refers to the typical ostium secundum defect; all patients with ostium primum ASD or endocardial cushion defect have been excluded.

In the past 8 years, we have evaluated 11 patients who presented with congestive heart failure (CHF) in infancy and who had an isolated ostium secundum ASD. These 11 infants comprise 3% of all cases of ostium secundum ASD seen at the University of Minnesota Hospitals since 1952 and 10% of those in whom the diagnosis of ASD was made in infancy.

Material and Methods

Clinical Features
The 11 infants presented to this hospital with CHF at ages ranging from 6 days to 13 months. CHF was defined clinically by the presence of tachypnea, dyspnea, hepatomegaly, and cardiomegaly in all 11. Ten of the 11 patients were given digoxin, and in each the symptoms noted above improved. The eleventh patient had definite mild CHF but was not placed on digoxin prior to cardiac catheterization. After catheterization, gradual improvement in clinical symptoms occurred without digoxin.

The physical findings in these infants were often atypical for atrial septal defect. In four, the murmur was holosystolic rather than ejection in quality. Only two patients had fixed splitting of S2 at the initial examination. P2 was accentuated in all 11 patients. Phonocardiograms were not available in any of the 11 patients.

On the other hand, a typical diastolic tricuspid flow murmurs were heard in 10 of the 11 patients. This murmur was misinterpreted in these patients because of a low index of suspicion for the diagnosis of ASD, and instead contributed to a presumptive diagnosis of patent ductus arteriosus (PDA) in three and total anomalous pulmonary venous connection (TAPVC) in one. In the patients suspected of having a VSD, this murmur was thought to be a mitral flow murmur.

Once a cardiac diagnosis is confirmed by cardiac catheterization, the auscultatory findings have a tendency to become "typical." In our patients, the auscultatory findings remained atypical until about 2
years of age. For example, three of the four patients with holosystolic murmur had an ejection quality murmur first noted at 17, 18, and 30 months of age, respectively. The patients with variable splitting of S2 did not develop fixed splitting of S2 until at least 2 years of age.

The decrease in intensity of P2 in these patients and the persistence of the tricuspid flow murmur thus resulted in the typical auscultatory picture of a large atrial septal defect in each patient at about 2 years of age.

Each patient had a 12-lead ECG at the first visit. Incomplete RBBB was present in five patients; four had other evidence of RVH. The remaining two had no evidence of ventricular hypertrophy. Transition to incomplete RBBB occurred in two patients after 2 years of age. The others were younger than 2 years at the last follow-up (three patients) or died (one). Right atrial enlargement, unusual in ASD, was present in five patients.

The thoracic roentgenograms showed cardiomegaly and increased pulmonary vascularity in all 11 infants (figs. 1, 2). A definite coincidental pneumonia was present in only one infant.

In none of the first nine patients was ASD considered as the primary precatheterization diagnosis. In fact, ASD was considered in the differential diagnosis in only one. The two patients seen subsequent to our increased interest in this problem both had a primary precatheterization diagnosis of ASD.

The reasons for our early failure to consider ASD as a diagnosis were: (1) failure to appreciate the occurrence of CHF in isolated ASD in infancy; (2) presence of holosystolic murmur; (3) absence of fixed splitting of S2; (4) accentuated P2; and (5) misinterpretation of the tricuspid diastolic murmur.

Cardiac Catheterization Data

The 11 patients were catheterized at ages ranging from 1 week to 17 months. In each instance, diagnostic catheterization was performed because of persistent symptoms of CHF despite appropriate medical management. The catheterization findings are summarized in table 1.

An atrial left-to-right shunt was confirmed by oximetry in all patients. Magnitude of the atrial left-to-right shunt ranged from 52 to 80%, the average shunt being 64%. The difficulties of accurate estimation of atrial left-to-right shunts are well known. Nevertheless, these figures are comparable to the size of shunts estimated in older patients with ASD. In the two patients in whom a later catheterization was performed, magnitude of the left-to-right shunt was unchanged 16.5 and 35 months later.

Angiographic confirmation of the atrial left-to-right shunt was accomplished in eight patients. Left-to-right shunts at the ventricular or great vessel level were excluded by angiography in eight of the 11 patients and by surgery or autopsy in the other three patients. In all 11 patients left-sided obstructive lesions were ruled out by angiography.

Right and left atrial pressures were measured immediately sequentially in each patient. Although a significant pressure difference between the atria was present in case 1, the right and left atrial pressures were essentially identical in the other 10 patients. The ventricular pressure tracings were not obtained simultaneously one to the other or simultaneously with atrial tracings, which may account for some of the discrepancies noted in table 1. Whenever a withdrawal tracing was obtained from LV → LA or from RV → RA, the end-diastolic ventricular pressure was comparable to the atrial pressure. In none of the 11 patients,
Figure 2

Serial PA thoracic roentgenograms in case 3 at the time of serial heart catheterizations. (Left) Age 6 weeks. CHF was present, and catheterization demonstrated a 75% left-to-right shunt. Moderate cardiomegaly and increased pulmonary vascularity are evident. (Right) Age 2 years. Despite clinical improvement, and no clinical evidence of CHF, the chest X-ray is unchanged. A 72% left-to-right shunt was present at recatheterization.

therefore, could a pressure difference be demonstrated across the mitral or aortic valves or in the aorta.

Pulmonary artery pressure was near systemic in two patients and somewhat elevated in most of the other patients. On the other hand, total pulmonary resistance (TPR) was within normal limits for age in all patients (fig. 3).1

Subsequent Clinical Course

The subsequent clinical course of these patients following diagnostic catheterization is summarized in table 1. Early surgery was performed in two patients with intractable CHF. Both patients died postoperatively, one 4 days following operation secondary to complications of air embolism and the other 19 days following operation secondary to heart block and pneumonia. It is important to note, however, that these two patients were operated on 7 and 9 years ago, respectively. Two patients have undergone elective surgical closure at the customary age for a typical ASD (age 4–6 years).

One patient, managed medically, died at home of undetermined cause 6 weeks after digoxin (Lanoxin) was discontinued. The other six patients are asymptomatic and awaiting elective surgical closure. All six patients, however, have persistent clinical findings of a large left-to-right shunt, and thoracic roentgenograms continue to show cardiomegaly and increased pulmonary vasculature (figs. 1, 2).

Discussion

Ostium secundum ASD is rarely included in a discussion of congenital cardiac defects in infancy. It has not been reported to be a cause of death in neonatal cardiac mortalities.2

Circulation, Volume XLVII, May 1973
## Hemodynamic Findings and Subsequent Clinical Course in 11 Patients

<table>
<thead>
<tr>
<th>Case</th>
<th>Age (months)</th>
<th>CHF</th>
<th>Cath</th>
<th>PA*</th>
<th>RV</th>
<th>LV</th>
<th>LA†</th>
<th>RA†</th>
<th>L-R shunt (%)</th>
<th>Site of selective angiography</th>
<th>Age digoxin stopped (yr)</th>
<th>Subsequent clinical course**</th>
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Abbreviations: cath = catherization; PA = pulmonary artery; LV = left ventricle; RV = right ventricle; LA = left atrium.

*Pressures given are systolic, diastolic, and mean.
†Pressures given are a wave, v wave, and mean.
‡Associated right-to-left shunt.
§Sinus venous defect found at surgery.
**All patients listed as asymptomatic have typical clinical findings of an ostium secundum ASD with a large left-to-right shunt.
Only a small number of infants with CHF secondary to an ASD are reported in the literature. The clinical findings were not typical of ASD during infancy but tended to become typical of classical ASD as the patients approached 2 years of age. This progression was also noted in our patients.

There is no explanation for the clinical finding of an initial holosystolic murmur in four of the 11 patients. The cardiac catheterization data do not support the presence of an additional hemodynamic defect, such as tricuspid insufficiency or ventricular septal defect, to explain the holosystolic murmur. Phonocardiography was not available in this group of patients.

The number of reported cases of ASD with CHF in infancy has been too small to define prognosis. The tendency has been, however, for these patients to respond to medical management and not require early surgical closure.

The series of 11 patients presented here provides further documentation that symptomatic ASD can occur as early as the first few days of life. The major importance of this group of patients, however, should be as a focus for understanding how they differ from the older child with a typical ostium secundum defect in whom the murmur is often not even heard until school age and who remains asymptomatic throughout childhood.

The possible explanations of CHF in infants with ASD include: (1) larger than normal left-to-right shunt; (2) presence of another left-to-right shunt; (3) earlier than usual decrease in pulmonary vascular resistance; (4) presence of a left-sided obstructive lesion; (5) abnormal ventricular compliance; or (6) abnormal atrial compliance.

Magnitude of Left-to-Right Shunt

Left-to-right shunt in this group of patients ranged between 52 and 80%. The average was 64%, which is a typical shunt for the older child with ASD. In the two patients who had a subsequent preoperative cardiac catheterization (1.5 and 3 years later, respectively), the magnitude of the left-to-right shunt had not changed.

Presence of a Second Shunt

The presence of an associated left-to-right shunt was thoroughly evaluated in all 11 patients. No additional intra- or extracardiac left-to-right shunt could be found by either anatomic or angiographic evaluation.

Early Decrease in Total Pulmonary Resistance (TPR)

An early decrease in TPR would be a logical explanation for the early appearance of a large left-to-right shunt. The values for TPR in our patients are shown in graphic form in figure 3. Although four patients fall below the mean value for TPR as plotted against body surface area, the standard deviation is such that all patients have a TPR which is within the normal range.

Left-Sided Obstructive Lesion

The presence of a significant left-sided obstructive lesion should be a major consideration in any infant with a large left-to-right shunt at the atrial level. Pagtakhan et al., for example, reported seven infants who presented between 3 days and 9 weeks of age with severe CHF and a large left-to-right atrial shunt. Each of these patients had a left-sided obstructive lesion, such as endocardial fibroelastosis, aortic atresia, or coarctation of the aorta. In the one survivor of this group, the ASD closed spontaneously 3 years after surgical correction of coarctation of the aorta in early infancy.

All patients in our series were thoroughly evaluated for a left-sided obstructive lesion. A pressure gradient across the mitral valve was not present in any instance. However, since the left atrium would be decompressed by the septal defect, the absence of such a gradient may not be conclusive. Nonetheless, no mitral obstruction was present by anatomic or angiographic evaluation in any of the 11 cases. The presence of a left ventricular or aortic obstructive lesion could also be excluded by hemodynamic data and angiography in all 11 patients.

Abnormal Ventricular Compliance

An abnormality in ventricular compliance remains as a potential cause. A greater than usual disparity between right and left ventricular compliance in infancy would account for early development of the large left-to-right atrial shunt. Such a disparity in ventricular compliance cannot be evaluated by end-diastolic pressure (EDP), since the filling pressure is the same for both ventricles when a large atrial communication is present.

Presumptive angiographic evidence exists in three patients to suggest abnormalities in ventricular function. The left ventriculogram in case 5 (fig. 4) illustrates a possible abnormality in ventricular function. As illustrated, the left ventricular myocar-
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Figure 4

Left ventriculogram obtained in case 5 at age 5 months (70% left-to-right shunt). Left ventricular end-diastolic pressure is 15 mm Hg. (Left) Ventricular systole. Note the wide distance between the left coronary artery and the ventricular cavity. The ventricular cavity is elongated. (Right) Ventricular diastole. The thick myocardium is again noted. Of special interest is the unusual inferior indentation into the left ventricular cavity.

dium is thickened, and the left ventricular cavity is elongated, with a marked inferior indentation being present.

Infants and children with chronic right ventricular overload secondary to a large atrial shunt have been shown to have abnormally low left ventricular end-diastolic volume and systemic output. In two of our patients, however, the left ventricular cavity

Figure 5

Left ventriculograms obtained in case 7 at age 8 months (52% left-to-right shunt). Left ventricular end-diastolic pressure is 5 mm Hg. (Left) Ventricular systole. The ventricle has contracted well. (Right) Ventricular diastole. The left ventricular end-diastolic volume is greater than expected in an uncomplicated ASD.
appears to be larger than normal (fig. 5) and certainly larger than usual in patients with typical ASD.

Whether the abnormal left ventriculograms in three of the seven patients who had left ventriculograms can be specifically related to abnormal left ventricular compliance cannot be determined. Although a disparity between right and left ventricular compliance would seem to be the best explanation for early development of a large left-to-right atrial shunt, this hypothesis cannot be further evaluated until it is possible to perform left ventricular volume studies in young infants such as those in our series.

Abnormal Atrial Compliance

Two atria communicating via a large ASD have equal pressures, but may not necessarily function as a single filling chamber. If the compliance of the atria are dissimilar, the one with the lesser compliance will receive less volume (pressures being equal). Although we have no direct evidence that this is the case, it has long been known that left atrial compliance may differ greatly in patients with mitral stenosis—those with high compliance develop large left atrial volumes and are therefore "protected" from pulmonary edema. The low-compliance left atria, on the other hand, accept increased volume only at increased pressure and therefore contribute to the development of pulmonary edema.

In a patient with a large, pressure-equilibrated ASD, the lower the left atrial compliance, the greater the left-to-right shunt. Likewise, if right atrial compliance is greater than left atrial compliance, a larger left-to-right shunt should occur. In either instance, consistent with Laplace's law, the right atrium might then be expected to enlarge and the left atrium to become smaller, thus perpetuating the development of an early, large atrial left-to-right shunt.

There is no clinical or catheterization evidence in any of our 11 patients to indicate either spontaneous closure or reduction in size of the ASD. However, it is now well established that spontaneous closure of even large ASD can occur. Cayler, for example, reported spontaneous closure of large ASD in 31% (four cases) of all infants with ostium secundum ASD catheterized over a 4-year period.

If possible, corrective surgery should be deferred until 2 years of age to allow for possible spontaneous closure. In nine of our 11 patients, medical management was sufficient even when CHF occurred as early as 6 days of age. Improvements in intracardiac surgery and postoperative care of the infant are such, however, that operative correction should be recommended for the occasional infant who may not respond to appropriate medical management.

Conclusions

Symptomatic ASD does occur in infancy but is uncommon. ASD should be included in the differential diagnosis of any large left-to-right shunt, especially if the systolic murmur is not harsh or loud, and the diastolic murmur is heard best at the lower left sternal border.

The explanations for the early occurrence of a large atrial left-to-right shunt cannot yet be determined. Although abnormalities in ventricular or atrial compliance may occur, current methodology is not adequate to quantitate such abnormalities in the sick infant.

Spontaneous closure did not occur in any patient in our series. In those patients in whom early surgery is not necessary, the clinical course tends to become that of a classical ASD by age 2 years.

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Circulation, Volume XLVII, May 1973
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Circulation. 1973;47:1042-1048
doi: 10.1161/01.CIR.47.5.1042
Circulation is published by the American Heart Association, 7272 Greenville Avenue, Dallas, TX 75231
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Print ISSN: 0009-7322. Online ISSN: 1524-4539

The online version of this article, along with updated information and services, is located on the World Wide Web at:
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