The Persistent Ostium Primum Atrial Septal Defect

By S. Gilbert Blount, Jr., M.D., Oscar J. Balchum, M.D., Ph.D., and Goffredo Gensini, M.D.

Five patients with an ostium primum type of atrial septal defect were studied by clinical and laboratory methods and the diagnosis was verified either at the time of open-heart surgery and/or at autopsy. The atrioventricular valves formed the floor of the defect and were deformed in 4 of the 5 patients. Mitral insufficiency was present as a result of this valvular deformity in these four patients. This valvular deformity resulted in a high pitched blowing systolic murmur, audible over the apical and lower left sternal areas, and left axis deviation and indications of left ventricular hypertrophy, determined by electrocardiography. The deformity of the mitral valve is considered to be the basis for these variations from the typical findings found in the patient with an ostium secundum type of atrial septal defect and make the differential diagnosis of the persistent ostium primum possible under these circumstances.

RECENT developments in the surgical therapy of congenital heart disease include the closure of atrial septal defects by various techniques, the method employed at this institution consisting of suture closure under direct-vision, open-heart surgery. The more frequent centrally located ostium secundum type of defect usually offers little technical difficulty to complete closure. The situation unfortunately differs greatly in the presence of the ostium primum type of atrial septal defect. Thus it is imperative and of more than academic interest to anticipate prior to surgery the type of atrial septal defect that may be encountered.

There are several factors that readily account for the difference in ease of technical closure and that determine the success and ultimate outcome of the procedure. Atrial septal defects of the so-called primum type are usually associated with abnormalities of varying degree of the atrioventricular valves. This is secondary to the maldevelopment of the embryonic common atrioventricular canal. All gradations of involvement are possible from the common atrioventricular valve with patency of the upper portion of the ventricular septum to minor involvement of either the mitral or tricuspid valves with the ventricular septum remaining intact. Realization of the presence of these defects of the atrioventricular valves and of the ventricular septum makes the technical difficulties to closure readily appreciated. Review of the literature has not revealed any information relative to this differential diagnosis.

A group of 85 patients with atrial septal defects have been studied at this institution during the past four years. The ostium primum type of defect has been established in five of these patients, either at operation or at postmortem examination. While it is realized that no single finding is definitely diagnostic of a persistent ostium primum, certain distinctive features are apparent and a review of these findings has led to the belief that the ostium primum type defect can frequently be distinguished from the ostium secundum type defect. Postmortem examination or direct inspection of the atrial septum at the time of open heart surgery together with an evaluation of the adequacy of valvular function at operation has offered the opportunity of correlating the clinical findings with the anatomic features in the individual case.

It is the intent of this communication, there-
fore, to review the findings upon which this differential diagnosis is based.

Embryology

At an early stage in the development of the heart the common atrium is separated from the common ventricle by a constricted passage termed the atrioventricular canal. The septum primum first appears from the superior-posterior aspect of the common atrium, growing caudally to reach the atrioventricular cushions. The latter are protuberances forming on the anterior and posterior walls of the atrioventricular canal, which fuse to divide this canal into right and left atrioventricular canals. These cushions form the septal cusps of the mitral and tricuspid valves, and a portion of the upper part of the interventricular septum. The remainder of the interventricular septum develops from the lower part of the common ventricle and extends upward to meet and fuse with the atrioventricular cushions. The septum primum joins with the fused atrioventricular cushions, forming the right and left atria. Normally this fusion is complete and the upper portion of the septum primum undergoes degeneration, just prior to its union with the fused atrioventricular cushions; an opening called the ostium secundum results. The septum secundum starts to form from the wall of the right atrium between the septum primum and the entrance of the superior vena cava and grows downward until its lower edge overlaps the foramen secundum. The passage thus formed between the two atria is termed the foramen ovale, and is bounded by the lower part of the septum primum below and the inferior border of the septum secundum above.

Failure of the septum primum to join with the fused atrioventricular cushions leaves an opening, the persistent ostium primum. This defect is usually large and is often accompanied by abnormalities in the development of the endocardial cushions resulting in associated malformations of the mitral and tricuspid valves and at times a defect of the upper portion of the ventricular septum. The mitral valve is more frequently involved than the tricuspid.

Material

Five patients with an ostium primum type of atrial septal defect were selected from a group of 85 patients with atrial septal defects which have been studied by clinical and laboratory methods, including cardiac catheterization. The size and location of the defect and the status of the atrioventricular valves were determined at the time of direct-vision, open-heart surgery in two patients and at postmortem examination in three patients. The age of these patients ranged from 27 months to 10 years. A summary of the history and clinical findings is as follows:

Case 1. S. A., was a 4 year old boy. The mother’s pregnancy was without incident and the patient was the product of a normal delivery with a birth weight of 6 pounds and 7 ounces. Development was normal, although he was always small for his age. He was an active child but tired easily with prolonged exertion. There was no definite history of frequent upper or lower respiratory infections. A heart murmur was first detected at the time of a routine physical examination at the age of 4 years.

The physical examination revealed that the neck veins were not distended nor pulsating. There was a slight prominence of the precordium, and upon palpation a hyperactivity of the heart was detected along the lower left sternal border. No shocks nor thrills were palpable, but percussion revealed the heart to be definitely enlarged to the left. Auscultation over the second intercostal space at the left sternal border revealed that the second heart sound was accentuated and moderately unduplicated. A loud (grade 3 on a grade 6 basis) rough systolic murmur was heard over the entire precordium and posteriorly over the left lung base, of maximal intensity in the second interspace at the left sternal border. A low-pitched mid-diastolic murmur was audible in the area just to the left of the lower sternum. The lungs were clear and there was no cyanosis, clubbing nor peripheral edema and the femoral pulsations were within normal limits. Neither the liver nor spleen were palpable.

At operation a large right atrium and right ventricle were found. The atrial septum was practically nonexistent, with only a slight ridge being present superiorly and posteriorly.

At postmortem examination (fig. 54) there was a large defect in the lower portion of the atrial septum. The mitral and tricuspid valves were deformed. The aortic leaflet of the mitral valve and the medial leaflet of the tricuspid valve were divided into two parts by a continuous cleft. The medial leaflet of the tricuspid valve had a larger (two-thirds) posterior and a smaller (one-third) anterior segment. The lat-
ter was devoid of chordae tendineae and formed a
ruffled fold. The aortic leaflet of the mitral valve
was divided into a large (two-thirds) anterior and a
smaller (one-third) posterior segment. The chordae
tendineae of the latter segment were attached to the
adjacent endocardium and not to the papillary
muscles. The anterior portion of the aortic leaflet of
the mitral valve and the medial leaflet of the tri-
cuspid valve were continuous across the floor of
the atrial septal defect. All of the leaflets of the
mitral and tricuspid valves had flattened irregular
gray-white nodules near their free ends, which
measured up to 4 mm. in diameter. The pulmonary
veins entered the left atrium in a normal manner.

Case 2. R. L., was a 27 month old boy. The
mother's pregnancy and delivery were normal. The
infant's birth weight was five pounds and five
ounces. Development and weight gain were poor,
and he experienced frequent upper and lower
respiratory infections. Breathing was labored and
dyspnea was manifest upon mild exertion. A heart
murmur was first noted upon routine physical
examination at the age of 4 months. The patient
was in failure at this time and was digitalized. The
physical examination revealed a slight bulging of
the precordium with an excessively forceful beating
of the heart being felt along the lower left sternal
border. A shock was palpable in the second inter-
costal space at the left sternal border, and a thrill
was present in the fourth intercostal space extend-
ing from the sternal border toward the apex. The
heart was enlarged to percussion, its margin extending to
the left anterior axillary line. Auscultation revealed
that the second sound in the second left intercostal
space was greatly accentuated and definitely re-
duplicated. A grade 4 rough blowing systolic
murmur was audible over the entire precordium, and
was of maximum intensity in the fourth intercostal
space at the left sternal border, and was louder in the
fifth than in the second intercostal space. A
separate higher pitched systolic murmur was audible
over the apical area. A gallop rhythm was present,
due to the presence of a low pitched mid-diastolic
murmur which was audible along the lower left
sternal border and also over the apical area. Neither
cyanosis nor clubbing was noted. Peripheral edema
was not detected, but the liver edge was palpable
4 cm. below the right costal margin, although no
rales were audible. The femoral pulsations were
normal in amplitude.

The right atrium, right ventricle and pulmonary
artery were found to be markedly enlarged at the
time of operation. A systolic thrill was palpable
over the right atrium. A low atrial septal defect of
the ostium primum type was estimated to measure
3 by 2 cm.

Postmortem examination revealed that both
ventricles were markedly enlarged, the right more
than the left. The right atrium was greatly dilated
while the left atrium was small. A large defect was
present in the inferior central portion of the atrial
septum, with the atrioventricular valves forming
the floor of the defect. The medial tricuspid leaflet
was short and bound down to the ventricular septum.
The other leaflets were broadened, and the chordae
tendineae were thickened and short. The line of
attachment of the posterior leaflet of the mitral
valve was short and blended into the inferior border
of the atrial septal defect. A broadened commissure
was present between the anterior and posterior leaf-
lets. The anterior leaflet was normal. Several chordae
tendineae extended from the anterior papillary mus-
cle to the superior portion of the ventricular septum,
and were thicker and shorter than normal.

Case 3. E. B., was a 10 year old boy. The mother's
pregnancy was without incident, and he was the
product of a normal full-term delivery. The birth
weight was 7 pounds and 8 ounces. His physical
development was apparently normal, although he
was mentally retarded. He began having epilepti-
form attacks during an episode of whooping cough
at the age of one and one-half years, and a heart
murmur was first heard at this time. There was no
definite history of upper or lower respiratory infec-
tions. He was always active and vigorous, without
limitation of exercise tolerance.

The physical examination revealed that the chest
was symmetrical. A forceful beating of the heart
was noted along the left lower sternal, and a thrust
was palpable in the second left intercostal space.
There were no thrills nor shocks palpable.
The heart was enlarged upon percussion, its border
extending to 1 cm. beyond the left mid-clavicular
line. Auscultation revealed that the second sound in
the second left intercostal space was slightly in-
creased in intensity and widely reduplicated. A grade
3 rough systolic murmur was audible over the ante-
rior chest and was of maximum intensity in the
second left intercostal space at the sternal margin.
A prominent low-pitched mid-diastolic murmur was
audible in the region between the lower sternum
and the apex of the heart. There was no cyanosis,
clubbing of the digits nor peripheral edema, and
the liver was not palpable. The femoral pulsations
were of normal amplitude.

This patient succumbed during an epileptiform
seizure which occurred on the day prior to operation.

Necropsy revealed the right atrium and right
ventricle to be greatly enlarged (fig. 5, B). An atrial
septal defect was present resulting in a roughly el-
liptical opening which was 3 cm. long by 2 cm. wide
and located in the lower portion of the atrial septum
immediately above the atrioventricular valves. The
foramen ovale was anatomically patent. The atrio-
ventricular valves were normal.

Case 4. J. R. was a 4 year old boy. The mother's
pregnancy and delivery were normal. Birth weight
was 7 pounds and 5 ounces. Development was nor-
mal, although growth was slow. At the end of one
year he weighed only 17 pounds. There were frequent and prolonged upper respiratory infections. Dyspnea and fatigue were manifest on slight exertion. The presence of a heart murmur and cardiac enlargement were first detected at 1 year of age.

Inspection revealed definite prominence and hyperactivity of the precordium. No thrills were palpable, but a faint shock was present in the second left intercostal space. Percussion revealed the heart to be enlarged to the left anterior axillary line. The point of maximum impulse was diffuse and palpable in the fifth and sixth intercostal spaces. Auscultation revealed the second sound in the second left intercostal space to be greatly increased in intensity and slightly reduplicated. A grade 3 to 4 harsh, systolic murmur was audible along the left sternal border, and a systolic murmur of a slightly higher-pitched quality was heard in the region of the fourth intercostal space at the left sternal border. A short low-pitched mid-diastolic murmur was audible over the apical area. There was no cyanosis nor clubbing of the digits. Peripheral edema was not present, the liver was not palpable, and the lungs were clear. The femoral pulsations were of normal amplitude.

The right atrium and right ventricle were greatly enlarged at operation. The left atrium appeared to be normal in size. The pulmonary artery was twice the size of the aorta. A thrill was palpable over the posterior portion of the right atrium. No septum could be palpated by finger exploration of the interior of the right atrium. When the exploring finger was inserted through the atrial defect a regurgitant jet through the mitral valve was felt, indicating mitral insufficiency. No attempt at closure was made.

Case 5. M. O'C., was a 28 month old girl. The mother's pregnancy was normal and the patient was the product of a normal full-term delivery. Her birth weight was 5 pounds and 9 ounces. Development and weight gain were poor and she suffered many upper and lower respiratory infections, including three episodes of pneumonia. She was very active, but experienced fatigue and dyspnea with exertion. Enlargement of the heart, cardiac murmurs and heart failure were first detected at the time of hospitalization for pneumonia at the age of 14 months.

Physical examination revealed prominence and hyperactivity of the precordium. There was a sustained thrust between the left lower sternum and the midclavicular line. A faint shock was palpable in the second left intercostal space at the left sternal border, but no thrills were present. Percussion revealed the heart to be enlarged to the left anterior axillary line. Upon auscultation the second heart sound in the second intercostal space at the left sternal border was widely reduplicated and increased in intensity. A soft blowing grade 2 to 3 systolic murmur was audible at the base of the heart, which was of maximum intensity in the second left intercostal space and well transmitted toward the shoulders. There was in addition a high-pitched blowing systolic murmur, grade 3 to 4 in intensity, audible over the apical area and transmitted toward the axilla. A short, low-pitched murmur was heard in early diastole, being of maximum intensity over the lower left parasternal area. No cyanosis, clubbing of the digits, nor peripheral edema was present, and no rales were audible over the posterior chest. The femoral pulsations were of normal amplitude.

Palpation of the exposed heart at the time of operation revealed a thrill to be present over the posterior portion of the right atrium and the pulmonary artery. A definite regurgitant jet was felt by the exploring finger when it was passed through the defect to the area of the mitral valve, indicating definite mitral insufficiency. Inspection revealed the atrial septum to consist of only a narrow rim in the superior and posterior portion of the atrium.

**Discussion**

*History.* The history of these five patients revealed that their birth weights were normal, but that growth and development were retarded. Upper and lower respiratory infections were a prominent feature of their history and the exercise tolerance was significantly decreased with the development of congestive failure in three patients. Therefore the history and symptomatology present in the patient with an ostium primum type of atrial septal defect are in general similar to those of the patient with an ostium secundum defect, and are the symptoms of patients with large left to right shunting of blood.

Their symptoms, however, are manifested earlier in life and are in general more incapacitating.

The increased severity of these symptoms is considered to reflect the larger volume of left to right blood flow which frequently occurs in these patients. The insufficiency of the atrioventricular valves also contributes to the increased work of the heart in the patient with a persistent ostium primum defect.

*Physical Examination.* The physical findings likewise resembled those of patients with the ostium secundum type of atrial septal defect. However, there were additional features of the cardiac examination which led to the consideration of the diagnosis of an ostium primum defect.
The first group of findings is considered to reflect the greater derangement of the hemodynamic pattern present in this defect. The heart was usually of greater size as reflected by the bulging precordium and other physical signs of cardiomegaly. The systolic murmur was of greater intensity, rougher in quality, and more widely transmitted than the murmur usually present in the patient with a secundum type of defect. This is the result of the greater degree of functional stenosis secondary to the higher pulmonary flows frequently found in patients with this type of defect. The second heart sound, audible in the second left intercostal space, tended to be louder in these patients and a palpable shock was present in this area in three of the five patients. This sound was reduplicated in all patients but widely split in only two patients. This contrasts with the patient with a secundum type defect where the primary feature of the second heart sound is the reduplication and not the intensity. The increased intensity of the second heart sound is considered to reflect the greater pulmonary flow and pressure frequently present in patients with the ostium primum type of defect. The constant mid-diastolic murmur present over the tricuspid area in these patients results from the high volume of flow across the tricuspid valve and the resulting functional tricuspid stenosis. It is again emphasized that this murmur does not necessarily indicate organic stenosis of the mitral valve as the mitral orifice was not narrowed in any patient in this group.\(^3\)

The second important finding that should lead to the suspicion of the presence of an ostium primum type defect is the systolic murmur located at the apical area and/or along the lower left sternal border. This murmur has a high pitched blowing quality, and is frequently loud and widely transmitted. The presence of this murmur suggests an ostium primum type of defect and is considered to be due to the malformation and resulting insufficiency of the mitral and/or tricuspid valves.

**Fluoroscopy.** The fluoroscopic examination revealed the vascularity of the peripheral lung fields to be greatly increased. The main pulmonary artery and the right and left pulmonary arteries were enlarged and the amplitude of their pulsations was increased (figs. 1 and 2). The aorta was inconspicuous in these patients. The heart was considerably to markedly enlarged in all patients and in general the overall heart size was greater than the heart size of patients of this same age group having a secundum defect. The right atrium and the right ventricle were markedly enlarged. This enlargement of the right ventricle is mainly the result of dilatation and therefore the true size of the main pulmonary artery is frequently underestimated due to the dilatation of the outflow tract of the right

---

**Fig. 1.** Postero-anterior roentgenograms of (A) patient S. A., case 1, (B) patient R. L., case 2 and (C) patient J. R., case 4.
ventricle. The detection of differential chamber enlargement is often difficult if not impossible in the presence of marked cardiac enlargement; nevertheless, there was suggestive evidence of enlargement of the left ventricle in four of the five patients. This enlargement of the left ventricle is considered to reflect the insufficiency of the mitral valve which is frequently deformed in the persistent ostium primum defect. This is in contrast to the patient with a secundum type of atrial septal defect where the atrioventricular valves are not involved and the chambers of the left heart are always normal or small (fig. 3).

Electrocardiograms: The electrocardiogram is of diagnostic value in indicating the presence of an ostium primum type of atrial septal defect (fig. 4).* Left axis deviation was noted in the electrocardiograms of three of the five patients, in two of which there was horizontal electrical position and in the remaining one a vertical electrical position. The precordial leads indicated an incomplete right bundle branch block in four of the five patients, and in the remaining patient were clearly indicative of right ventricular hypertrophy. There was additional evidence suggesting right ventricular hypertrophy in the four patients having an incomplete right bundle branch block. The precordial leads revealed findings suggesting left ventricular hypertrophy in four of the five patients. The presence of left ventricular hypertrophy is difficult to determine from the electrocardiogram in the presence of hypertrophy and conduction disturbance of the right ventricle and these statements are influenced by comparison with the electrocardiograms of patients with known ostium secundum type defects.

The P waves were considered to be abnormal in the tracings of three of the five patients. There was considered to be evidence of right atrial enlargement in three patients (R. L., J. R., and M. O'C.) and suggestive evidence of left atrial enlargement in two of these patients. There was a normal sinus rhythm in all patients and the rates were within normal limits. There was evidence of first degree heart block and prolongation of the QRS interval in three patients according to the standards proposed by Ziegler.*

The analysis of the electrocardiograms in

* Table 1 in which the electrocardiographic findings are summarized is being omitted at the request of the Editor. This table will be furnished upon request.
Fig. 4. Electrocardiograms of patients S. A. case 1, E. V. B., case 3, and J. R. R. case 4.
these patients is of particular value when compared with the tracings of 23 patients with established ostium secundum type of atrial septal defects.7 There were no instances of left axis deviation nor horizontal electrical position of the heart in any patient in this latter group. There was no suggestion of left ventricular or left atrial enlargement in any of these 23 patients, and in none was there an R wave in 

\( V_a \) of 21 mm. or greater. Thus, in patients with an ostium primum type defect the electrocardiogram differed strikingly from that seen in the ostium secundum defect, the sole exception being case 3 (E. B.); and it is of significance to note that this is the only one of the five patients in whom there was no involvement of the mitral valve (fig. 5B).

These variations from the typical electrocardiogram which is present in the patient with a known secundum type of defect are again considered to reflect the presence of mitral insufficiency and the resulting enlargement of the left ventricle.

**Physiologic Data.** Evaluation of the physiologic data revealed no features of distinct differential diagnostic value when compared with the data from patients with known ostium secundum defects (table 2). There was a suggestion that the volume of the left to right shunt was greater in patients with an ostium primum defect and that the pulmonary artery pressure was higher; however in the individual case these findings were of no definite diagnostic value.

The position of the catheter as it crosses over into the left atrium may be a helpful sign. A low position of this crossing would tend to suggest an ostium primum defect, further support of this contention being the easy entrance of the catheter into the left ventricle. The withdrawal of the catheter from a high pressure area containing highly oxygenated blood directly into a low pressure area with blood of a significantly lower oxygenation might also suggest the possibility of an ostium primum defect. Two patients had slight peripheral arterial unsaturation, an infrequent finding in patients with a secundum defect. There was no evidence of an additional left to right shunt at the ventricular level in any of these patients.

The features just discussed may be considered to be suggestive, but never diagnostic in the individual case. It is considered that the slight differences noted may be accounted for either on the basis of absence of the lowest portion of the atrial septum, the size of the defect, and the presence of mitral insufficiency.

**Anatomic-Pathologic Data.** The atrial septal defects were large, and the atroventricular valves formed the floor of the defect in all patients. This was established at postmortem examination in three instances and at the

![Fig. 5. Views of Left Atrium; Mitral Valve and Left Ventricle in (A) patient A. S. case 1 and (B) patient E. B. case 3.](http://circ.ahajournals.org/ViewPDF/9591372/10.1161/01.CIR.33.4.506)
time of direct-vision, open-heart surgery in the remaining two patients. The area of the foramen ovale was noted in all instances and there was patency in one patient (case 3).

The atrioventricular valves were deformed in two of the three hearts available for post-mortem examination (fig. 5 A). The mitral valve was more significantly involved in each instance.

The remaining patient (E. B.) of this group was found to have an ostium primum type defect at postmortem examination. However, he was the only patient that demonstrated no anatomical abnormalities of the atrioventricular valves. It is considered to be of interest and significance that in this patient there were no findings either on physical examination, in the electrocardiogram or upon fluoroscopy to indicate the possible presence of an ostium primum type defect. This patient, then, further supports the contention that abnormalities of the atrioventricular valves and, in particular, defects of the mitral valve give rise to the findings indicating the presence of the ostium primum type defect.

The location of the defect in the atrial septum is not considered to be of significance in the development of the distinctive pattern of the patient with this type of defect.

Although the anatomic details of the mitral valve are not known in the two patients still living, both mitral valves were functionally insufficient at the time of surgery. Prior to cessation of circulation the area of the defect and atrioventricular valves were palpated by the surgeon, Dr. Henry Swan. A regurgitant jet of blood was detected coming from the left ventricle back through the mitral valve into the right atrium as well as into the left atrium.

**SUMMARY AND CONCLUSIONS**

The five patients comprising this group all had large defects of the atrial septum. The defect was situated in the most caudal portion of the septum so that the atrioventricular valves in each instance formed the floor of the defect. These valves were abnormal in 4 of the 5 patients, the mitral valve in 4 and the tricuspid valve in 2. The mitral valve was considered to be insufficient in all four of these patients, and in two of the four it was definitely established at the time of surgery that this valve was incompetent. There was no evidence that the ventricular septum was defective in any of the five patients in this group.

These patients have thus been classified as having the persistent ostium primum type of atrial septal defect. It is realized that some will consider these patients to represent examples of a partial atrioventricular communis type of defect. We do not deny that this is a logical

### Table 2.—Cardiac Catheterization Data

<table>
<thead>
<tr>
<th></th>
<th></th>
<th></th>
<th></th>
<th></th>
<th></th>
<th></th>
<th></th>
<th></th>
<th></th>
<th></th>
<th></th>
<th></th>
<th></th>
<th></th>
<th></th>
<th></th>
<th></th>
<th></th>
</tr>
</thead>
<tbody>
<tr>
<td>Case 1 S. A. 4 yrs.</td>
<td>10.66</td>
<td>10.29</td>
<td>10.41</td>
<td>13.41</td>
<td>12.35</td>
<td>13.57</td>
<td>13.85 *</td>
<td>3.44</td>
<td>8.87</td>
<td>3.60</td>
<td>L to R</td>
<td>12.70</td>
<td>5.60</td>
<td>3.87</td>
<td>7.10</td>
<td></td>
<td></td>
<td></td>
</tr>
<tr>
<td>Case 2 R. L. 27 mos.</td>
<td>10.46</td>
<td>12.42</td>
<td>11.77</td>
<td>13.02</td>
<td>12.75</td>
<td>14.09</td>
<td>14.32</td>
<td>2.55</td>
<td>8.82</td>
<td>4.65</td>
<td>L to R</td>
<td>15.60</td>
<td>8.30</td>
<td>3.17</td>
<td>7.30</td>
<td></td>
<td></td>
<td></td>
</tr>
<tr>
<td>Case 3 E. B. 10 yrs.</td>
<td>12.90</td>
<td>12.48</td>
<td>12.69</td>
<td>14.54</td>
<td>15.39</td>
<td>15.92</td>
<td>17.02</td>
<td>17.35</td>
<td>16.71</td>
<td>4.02</td>
<td>12.50</td>
<td>3.91</td>
<td>L to R</td>
<td>11.90</td>
<td>3.76</td>
<td>8.50</td>
<td>8.14</td>
<td></td>
</tr>
<tr>
<td>Case 4 J. B. 4 yrs.</td>
<td>9.7</td>
<td>11.1</td>
<td>10.8</td>
<td>11.4</td>
<td>11.65</td>
<td>11.8</td>
<td>94.1%</td>
<td>95.9%</td>
<td>92.4%</td>
<td>2.1</td>
<td>96.2%</td>
<td></td>
<td></td>
<td></td>
<td></td>
<td></td>
<td></td>
<td></td>
</tr>
<tr>
<td>Case 5 M. O'C. 28 mos.</td>
<td>9.22</td>
<td>10.28</td>
<td>9.48</td>
<td>12.34</td>
<td>12.62</td>
<td>12.54</td>
<td>LA entered but no sample</td>
<td>13.17</td>
<td>88.8%</td>
<td>3.31</td>
<td></td>
<td></td>
<td></td>
<td></td>
<td></td>
<td></td>
<td></td>
<td></td>
</tr>
</tbody>
</table>

* Left ventricle also entered.
and acceptable designation. However, inasmuch as there was no clinical nor physiologic evidence in these five patients, nor anatomic evidence in the three patients postmortem that the ventricular septum was involved, we have chosen to designate this defect as the persistent ostium primum type of atrial septal defect.

Clinically, these patients tended to have larger hearts and to become symptomatic earlier than the patient with the ostium secundum type of defect. Physical examination revealed findings suggesting the presence of mitral insufficiency. The electrocardiogram frequently disclosed the presence of left axis deviation and evidence of left ventricular enlargement and, thus, differed from the rather characteristic tracing of the secundum type of defect. Fluoroscopy revealed no diagnostic features, although the heart tended to be larger and often there were findings suggestive of enlargement of the left ventricle. Cardiac catheterization yielded no diagnostic findings but the low position of the catheter as it crossed into the left atrium and at times the ease with which the catheter entered the left ventricle were suggestive findings. Therefore, these patients did not present a pathognomonic clinical picture, but there were certain features that differed from the findings in patients with a secundum type defect sufficiently to suggest the presence of the ostium primum type of atrial septal defect.

The location of the defect of the atrial septum is not considered to have a significant role in the production of the distinctive clinical picture. The anomalies of the mitral valve with the resulting mitral insufficiency are considered the basis of this differential diagnosis. These features are the presence of a murmur indicating mitral insufficiency, not present in the patient with a secundum type defect. The evidence of left ventricular enlargement frequently demonstrated by the electrocardiogram and fluoroscopy, and present at postmortem examination in two of the three hearts is the result of this mitral insufficiency.

Further evidence to support this conclusion is offered in the evaluation of case 3 (E. B.), who presented an entirely different clinical picture from the other four patients. The symptoms of this patient were minimal, the heart was only slightly enlarged (fig. 3) and there were no additional murmurs present other than those typical of a patient with a secundum type of atrial septal defect. The electrocardiogram was entirely different from those of the remaining four patients and it revealed only the changes indicative of a partial right bundle branch block. Thus, the clinical picture presented by this patient differed in no way from that of the patient with a secundum type of atrial septal defect. A clinical diagnosis of an ostium primum defect was not entertained in this patient and the diagnosis was made only after postmortem examination. The important fact is that this was the only patient that did not reveal congenital defects of the mitral or tricuspid valves.

This important exception further substantiates the belief that it is the involvement of the mitral valve with resulting mitral insufficiency that tends to differentiate the ostium primum type defect from the ostium secundum type of atrial septal defect.

**Summario in Interlingua**

Cinque patientes con un defecto atrio-septal del typo del ostio prime esseva studiate per methodos clinic e laboratorial. Le diagnose esseva verificatu al tempore de chirurgia cardiac aperte e/o al tempore de l'autopsia. Le valvulas atrioventricular representava le fundo del defecto e esseva deformate in 4 del 5 patientes. Insufficiencia mitral esseva presente como resultato de iste deformitate valvular in omne iste 4 casos. Le deformitate valvular resultava in un sufflante, altisonante murmure systolic, que esseva audibile supra le area apical e le area sinaltero sinistro-inferior, e in un deviation sinistrorse del axe e indications de hypertrophia sinistro-ventricular in le electrocardiogramma. Nos opinia que le deformitate del valvula mitral es le base de iste deviationes ab le constatationes typic pro patientes con defectos atrio-septal del typo del ostio secundum e rende possibile le diagnose differential de persistente ostio prime sub le conditiones delineate in le presente reporto.
REFERENCES


The Persistent Ostium Primum Atrial Septal Defect
S. GILBERT BLOUNT, JR., OSCAR J. BALCHUM and GOFFREDO GENSINI

Circulation. 1956;13:499-509
doi: 10.1161/01.CIR.13.4.499
Circulation is published by the American Heart Association, 7272 Greenville Avenue, Dallas, TX 75231
Copyright © 1956 American Heart Association, Inc. All rights reserved.
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:
http://circ.ahajournals.org/content/13/4/499

Permissions: Requests for permissions to reproduce figures, tables, or portions of articles originally published in Circulation can be obtained via RightsLink, a service of the Copyright Clearance Center, not the Editorial Office. Once the online version of the published article for which permission is being requested is located, click Request Permissions in the middle column of the Web page under Services. Further information about this process is available in the Permissions and Rights Question and Answer document.

Reprints: Information about reprints can be found online at:
http://www.lww.com/reprints

Subscriptions: Information about subscribing to Circulation is online at:
http://circ.ahajournals.org//subscriptions/