Ultrasoundcardiographic Studies of the Mitral Valve Region in Young Infants with Mitral Atresia, Mitral Stenosis, Hypoplasia of the Left Ventricle, and Cor Triatriatum

By Nils-Rune Lundström, M.D.

SUMMARY

Reflected ultrasound has been used for investigation of the mitral valve region in early infancy. The material consists of two patients with mitral atresia, three patients with mitral stenosis and aortic valvular atresia, five patients with mitral and aortic valvular stenosis, one patient with cor triatriatum, and one patient with severe degree of aortic valvular stenosis with a normal mitral valve. The results are compared with ultrasoundcardiographic findings in ten infants without heart disease. A specific abnormal echo was obtained in cases with mitral atresia and cor triatriatum. The patients with a combination of mitral and aortic valvular lesions could be separated into two groups by means of ultrasoundcardiography. This separation reflected the functional state of the left ventricle based on the degree of mitral stenosis and hypoplasia of the left ventricle.

This noninvasive technic has been found useful as a complementary method for the investigation of the mitral valve region. It can even be performed in seriously ill infants.

Additional Indexing Words:
Hypoplastic left heart syndrome
Supravalvular ring in the left atrium

Among congenital heart malformations causing symptoms during the first month of life, those with a hypoplastic left ventricle constitute a large group. The main part of this group consists of various combinations of malformations of the aortic and mitral valves, and several classifications have been proposed for them. These malformations generally lead to congestive heart failure and death in early infancy. The clinical findings and the hemodynamic consequences of these malformations have been well described. Suggestions for a surgical palliative approach have been presented, and a report of successful surgical palliation has also been published. The diagnosis is provided by the clinical findings, but to enable detailed characterization of these malformations heart catheterization and angiocardiography have to be performed. The mitral valve region is involved in the malformation in most cases, and information about the functional state of the mitral valve should be of additional value.

Since the introduction of ultrasoundcardiography using reflected ultrasound, studies of the movement of the anterior mitral leaflet have been one of the most useful applications of this method as indicated by the many reports on acquired mitral heart disease in adults. A preliminary report on the use of reflected ultrasound for studying congenital
heart malformations in children has been published. In a study of patients with a single ventricle using this technic, a case of hypoplastic left ventricle was mentioned, but an investigation of the mitral valve region was not reported.

The aim of the present investigation was to evaluate the use of ultrasoundcardiography in the diagnosis of congenital heart malformations primarily or secondarily affecting the mitral valve and causing symptoms during the first month of life. For comparison ten normal infants of the same age were studied with the same technic.

**Methods**

**Material**

The material consisted of all patients observed during the period 1968 to 1970 with congenital malformations affecting the mitral valve region and causing symptoms during the first month of life and where ultrasoundcardiography was performed. For comparison one patient with severe congenital valvular aortic stenosis, normal mitral valve, and enlarged left ventricle is included.

Some clinical data on the patients, including specified diagnosis and the age of the patients when they first demonstrated symptoms and their age at death, are presented in table 1. They all had symptoms of severe congestive heart failure resistant to anticongestive treatment. All except three (cases 3, 7, and 12) were investigated by heart catheterization and angiocardiology. Two cases underwent surgery: in case 8 valvotomy of the aortic valve and in case 6 attempted surgery of a preductal coarctation. Both patients died after the operation. The other ten patients also died, and autopsy was performed on all of them.

**Autopsy Findings**

Some relevant anatomic findings obtained at autopsy are given in table 1. The body type was situs solitus in all with a normally placed heart and with normal relations between the great arteries. In all cases the right ventricle was enlarged. The tricuspid and the pulmonary valves were normal. In cases 1, 4, and 8, an atrial septal defect was found, and in the rest the foramen ovale was patent. The interventricular septum was normal except in case 1 (single ventricle) and case 2 (ventricular septal defect). Cases with severe mitral stenosis showed a mitral valve

### Table 1

**Survey of Material: Age at First Symptom and at Death and Some Findings at Autopsy**

<table>
<thead>
<tr>
<th>Case</th>
<th>Age at first symptom</th>
<th>Age at death</th>
<th>Degree of mitral stenosis*</th>
<th>Degree of hypoplasia of left ventricle</th>
<th>Additional anomalies</th>
</tr>
</thead>
<tbody>
<tr>
<td>1</td>
<td>10 days</td>
<td>1 1/2 month</td>
<td>—</td>
<td>—</td>
<td>Single ventricle</td>
</tr>
<tr>
<td>2</td>
<td>7 days</td>
<td>1 month</td>
<td>—</td>
<td>Severe</td>
<td>VSD</td>
</tr>
<tr>
<td></td>
<td></td>
<td></td>
<td></td>
<td></td>
<td></td>
</tr>
<tr>
<td>3</td>
<td>3 days</td>
<td>9 days</td>
<td>++</td>
<td>Severe</td>
<td>—</td>
</tr>
<tr>
<td>4</td>
<td>2 days</td>
<td>4 days</td>
<td>+++</td>
<td>Severe</td>
<td>—</td>
</tr>
<tr>
<td>5</td>
<td>1 day</td>
<td>7 days</td>
<td>+++</td>
<td>Severe</td>
<td>—</td>
</tr>
<tr>
<td>6</td>
<td>7 days</td>
<td>16 days</td>
<td>+</td>
<td>Severe</td>
<td>—</td>
</tr>
<tr>
<td>7</td>
<td>1 day</td>
<td>3 days</td>
<td>+++</td>
<td>Moderate</td>
<td>—</td>
</tr>
<tr>
<td>8</td>
<td>2 days</td>
<td>8 days</td>
<td>+</td>
<td>Moderate</td>
<td>—</td>
</tr>
<tr>
<td>9</td>
<td>1 day</td>
<td>3 days</td>
<td>+</td>
<td>0</td>
<td>—</td>
</tr>
<tr>
<td>10</td>
<td>10 days</td>
<td>23 days</td>
<td>+</td>
<td>0</td>
<td>Supravalvular ring in left atrium and coarctation of aorta</td>
</tr>
<tr>
<td>11</td>
<td>1 month</td>
<td>3 months</td>
<td>0</td>
<td>0</td>
<td>—</td>
</tr>
<tr>
<td>12</td>
<td>2 weeks</td>
<td>1 1/2 month</td>
<td>0</td>
<td>0</td>
<td>—</td>
</tr>
</tbody>
</table>

*Degree of mitral stenosis: 0 = no mitral stenosis; + = slight mitral stenosis consisting of hypoplastic mitral ring and short chordae tendineae; ++ = moderate mitral stenosis consisting of hypoplastic mitral ring and mitral valvular stenosis; +++ = severe mitral stenosis with pinpoint opening.

_Circulation, Volume XLV, February 1972_
consisting of a membrane with a very small central opening. In all the other cases with mitral stenosis except case 10, the mitral valve was hypoplastic, and the leaflets were thickened and deformed, often with short chordae tendineae. In case 10 a parachute deformity of the mitral valve existed together with a nonobstructive supravalvular ring in the left atrium, a valvular aortic stenosis, and a preductal coarctation of the aorta. These malformations were readily demonstrated on angiocardiography. They resembled those described by Shone et al.,19 with the exception that in the case reported here (case 10) the aortic stenosis was valvular. In case 11 the left atrium was divided by a membrane with a central opening 1 mm in diameter between the upper part, which received all the pulmonary veins, and the lower part, which communicated through the mitral ostium with the left ventricle. The left atrial appendix originated from the lower part of the left atrium, and the foramen ovale was positioned between the lower part of the left atrium and the right atrium. The membrane was well demonstrated on angiocardiography (see fig. 7).

The aortic valvular stenosis was very tight in cases 6, 7, 8, and 12 and of moderate degree in cases 9 and 10. In all but three cases, endocardial fibroelastosis was found in the left ventricle and was most pronounced in cases 5, 6, and 7 and especially in case 9. Endocardial fibroelastosis was not observed in the single ventricle of case 1 nor in the left ventricle in cases 2 and 11.

Ultrasound Examinations

These were performed with an ultrasonoscope.* This instrument produces 1,000 pulses/sec and utilizes a 2.25-megacycle transducer of 0.75-inch diameter. The technic of this examination in infants and children has been described in a previous paper from this laboratory.17


---

Figure 1

Line drawings of two recordings of echoes from the anterior mitral leaflet obtained from an infant 2 weeks old without heart disease. The two recordings were obtained at different heart rates: (A) 120/min and (B) 145/min. a = total amplitude of movement (12 mm); b = amplitude of opening movement in ventricular diastole (9.5 mm); c = amplitude of movement during ventricular systole (2.5 mm); d = distance moved in 0.5 sec during the early part of diastole, which gives a speed of movement of 100 mm/sec.
patients were examined in supine position during normal respiration lying within an incubator. A water-soluble gel was used to obtain airless contact between the transducer and the skin.

In normal infants an echo from the anterior mitral leaflet is easily obtained with the transducer in the fourth intercostal space 2 to 3 cm to the left of the midline and with the transducer in anteroposterior direction. With a heart rate less than about 120/min, a rapid opening movement in the beginning of diastole, an immediate closing movement toward a semiopen position, and a movement during atrial systole are readily identified (fig. 1A). This registration permits measurement of the speed of movement in posterior direction during the early part of diastole, and it has been found to correlate well with the severity of mitral stenosis in adults.\textsuperscript{13, 14} In infants, with their faster heart rates, the separation of the posterior movement in early diastole from the movement during atrial systole is often not possible. Here, instead, the normal pattern of movement is characterized by a fast opening movement during the early part of diastole and an immediate posterior movement to a semiopen position followed by a gradual closing movement to a completely closed position (fig. 1B).

With the transducer in the fourth intercostal space at the left sternal border and slightly laterally angulated, a double echo from the interventricular septum can be obtained. With the transducer in the same position but angulated about 30° to 45° medially and often slightly cranially, an echo from a tricuspid leaflet can be obtained. In normal infants this echo is only fragmentary in most cases, but the fast opening movement in the early part of diastole is always possible to identify. This echo is positioned anterior to the echo from the interventricular septum. In cases with a dilatation of the right ventricle, an echo from the anterior tricuspid leaflet is obtained more easily and is recorded with the transducer in anteroposterior direction at the left sternal border or slightly more lateral. The echo is usually complete and has the same pattern of movement as an echo from a normal anterior mitral leaflet.

With the transducer in the second or third intercostal space at the left sternal border and slightly medially angulated, occasionally cranially, two parallel echoes can be obtained (fig. 2). These echoes have been shown to originate from the anterior and posterior border of the aortic root.\textsuperscript{20, 21} Behind the posterior of these echoes and in front of the echo from the posterior heart wall, an echo-free space is found which has been identified as representing the left atrium.\textsuperscript{21, 22}

### Results

#### Normal Subjects

Data from ultrasoundcardiographic investigations of ten normal infants are given in table 2. The echoes obtained from the different anatomic regions studied show a rather small individual variation.

![Ultrasoundcardiogram](image)

**Figure 2**

Ultrasoundcardiogram obtained from a patient with aortic and mitral stenosis and a severely deformity of the mitral valve, supravalvular ring in the left atrium, aortic valcular stenosis, and coarctation of the aorta (case 10). Both recordings were obtained with the transducer in the third left intercostal space at the sternal border and with the transducer directed in medial-cranial direction. After obtaining the left recording, the transducer was angulated in a slightly more caudal direction, and the right recording was then made. As in all subsequent figures, echoes from anterior intracardiac structures are in the upper part and echoes from posterior intracardiac structures in the lower part of the recording.

\textit{Circulation, Volume XLV, February 1972}
Table 2

Ultrasoundcardiographic Findings in Material and Ten Normal Infants of Same Age

<table>
<thead>
<tr>
<th>Case</th>
<th>Echo from the anterior mitral leaflet or mitral region</th>
<th>Echo from tricuspid leaflet (total amplitude of movement)</th>
<th>Echo from interventricular septum</th>
<th>Additional findings</th>
</tr>
</thead>
<tbody>
<tr>
<td></td>
<td>Total amplitude of movement 0</td>
<td>Amplitude of opening movement in diastole</td>
<td></td>
<td></td>
</tr>
<tr>
<td>1</td>
<td>3.5 mm</td>
<td>0</td>
<td>13 mm</td>
<td>0</td>
</tr>
<tr>
<td>2</td>
<td>3 mm</td>
<td>0</td>
<td>15 mm</td>
<td>+</td>
</tr>
<tr>
<td>3</td>
<td>3 mm</td>
<td>1 mm</td>
<td>13 mm</td>
<td>+</td>
</tr>
<tr>
<td>4</td>
<td>4 mm</td>
<td>1 mm</td>
<td>10 mm</td>
<td>+</td>
</tr>
<tr>
<td>5</td>
<td>5 mm</td>
<td>2 mm</td>
<td>14 mm</td>
<td>+</td>
</tr>
<tr>
<td>6</td>
<td>4 mm</td>
<td>2.5 mm</td>
<td>12 mm</td>
<td>+</td>
</tr>
<tr>
<td>7</td>
<td>3 mm</td>
<td>1 mm</td>
<td>11 mm</td>
<td>+</td>
</tr>
<tr>
<td>8</td>
<td>7 mm</td>
<td>5 mm</td>
<td>12 mm</td>
<td>+</td>
</tr>
<tr>
<td>9</td>
<td>8.5 mm</td>
<td>5.5 mm</td>
<td>10 mm</td>
<td>+</td>
</tr>
<tr>
<td>10</td>
<td>7 mm</td>
<td>5 mm</td>
<td>10 mm</td>
<td>+</td>
</tr>
<tr>
<td>11</td>
<td>7 mm</td>
<td>5 mm</td>
<td>11 mm</td>
<td>+</td>
</tr>
<tr>
<td>12</td>
<td>10 mm</td>
<td>8 mm</td>
<td>10 mm</td>
<td>+</td>
</tr>
<tr>
<td>10 normal</td>
<td>10–14 mm</td>
<td>7–9 mm</td>
<td>12†</td>
<td>+ in all cases</td>
</tr>
<tr>
<td>infants*</td>
<td>(mean 11.7 mm)</td>
<td>(mean 8.3 mm)</td>
<td></td>
<td></td>
</tr>
</tbody>
</table>

*Aged 1 week to 1 month.
†Complete echo from tricuspid leaflet only obtained in one normal infant.

Tricuspid Region in Patients with Heart Malformations

An echo from the anterior tricuspid leaflet was found in all cases (fig. 3). These recordings were made with the transducer in the fourth left intercostal space, close to the left sternal border and in anteroposterior direction. In many of the patients, this echo could also be obtained with the transducer 1 to 2 cm further to the left in the same intercostal space. The total amplitude of movement of the echo from the anterior tricuspid leaflet is given in table 2. Posterior to the echo from the anterior tricuspid leaflet, echoes from the interventricular septum were found in all except case 1. The findings consisted of a broad single echo or two

Figure 3

Ultrasoundcardiogram obtained from a patient with aortic and mitral stenosis and a severely hypoplastic left ventricle (case 6). The left recording was made with the transducer in the fourth left intercostal space close to the left sternal border, and the right recording was obtained in the same intercostal space but about 1 cm to the left. Both recordings were done with the transducer in anteroposterior direction. Note the large amplitude of movement of the echo from the tricuspid leaflet compared with the echoes from the mitral leaflets; also echoes from both the anterior and posterior mitral leaflets are seen during ventricular diastole.
from the anterior mitral leaflet could obviously not be obtained. In the place normally occupied by the mitral valve region, an echo with a small amplitude of movement (3 to 3.5 mm) could be registered instead. The pattern of movement of this echo consisted of an anterior movement during ventricular systole and a posterior movement during ventricular diastole (figs. 4D and 5). No anterior opening movement during diastole was observed. In case 2 this echo was clearly separated from the echo obtained from the interventricular septum.

In all the cases where mitral leaflets existed, an echo from the anterior mitral leaflet could be obtained. The findings can be grouped into two categories:

In five patients (cases 3 to 7, table 2), the total amplitude of movement was very low (3 to 5 mm), the amplitude of opening movement in the early part of diastole being especially reduced (1 to 2.5 mm) compared with normals (7 to 9 mm). Moreover, the pattern of movement was clearly different from the normal situation and consisted of a slow anterior movement during ventricular systole followed by an abrupt but short anterior movement in the beginning of diastole (figs. 3 and 4C). During the rest of diastole, a slow posterior movement occurred, followed by a faster closing movement at the end of ventricular diastole. In one patient (case 6), echoes from both the anterior and the posterior mitral leaflets could be registered (fig. 3). The opening movement of the echo

parallel echoes 3 to 5 mm apart. In case 1 (with single ventricle), no echo from the interventricular septum could be obtained.

Mitral Region in Patients with Heart Malformations

In the two cases with mitral atresia, an echo could obviously not be obtained. In the place normally occupied by the mitral valve region, an echo with a small amplitude of movement (3 to 3.5 mm) could be registered instead. The pattern of movement of this echo consisted of an anterior movement during ventricular systole and a posterior movement during ventricular diastole (figs. 4D and 5). No anterior opening movement during diastole was observed. In case 2 this echo was clearly separated from the echo obtained from the interventricular septum.

In all the cases where mitral leaflets existed, an echo from the anterior mitral leaflet could be obtained. The findings can be grouped into two categories:

In five patients (cases 3 to 7, table 2), the total amplitude of movement was very low (3 to 5 mm), the amplitude of opening movement in the early part of diastole being especially reduced (1 to 2.5 mm) compared with normals (7 to 9 mm). Moreover, the pattern of movement was clearly different from the normal situation and consisted of a slow anterior movement during ventricular systole followed by an abrupt but short anterior movement in the beginning of diastole (figs. 3 and 4C). During the rest of diastole, a slow posterior movement occurred, followed by a faster closing movement at the end of ventricular diastole. In one patient (case 6), echoes from both the anterior and the posterior mitral leaflets could be registered (fig. 3). The opening movement of the echo

parallel echoes 3 to 5 mm apart. In case 1 (with single ventricle), no echo from the interventricular septum could be obtained.

Mitral Region in Patients with Heart Malformations

In the two cases with mitral atresia, an echo

parallel echoes 3 to 5 mm apart. In case 1 (with single ventricle), no echo from the interventricular septum could be obtained.
from the posterior mitral leaflet in the beginning of diastole was in the posterior direction.

In the remaining cases (cases 8 to 12, table 2), the echo from the anterior mitral leaflet showed a greater total amplitude of movement as well as a greater amplitude of opening movement in the beginning of diastole (5 to 8 mm). The pattern of movement consisted of a slow anterior movement during ventricular systole followed by a fast opening movement in the early part of diastole. In cases 8, 9, and 10, the movement during the following part of diastole was characterized by a rather slow posterior movement and ended with a fast closing movement at the end of diastole (fig. 4B). The movement of the echo from the anterior mitral leaflet in diastole in case 11 differed somewhat from that just described: the posterior movement was a little faster in the beginning of diastole (fig. 6) but not so fast as in normal cases (fig. 4A).

In case 12 the echo from the anterior mitral leaflet had a normal amplitude of opening movement in the early part of diastole. The movement during the rest of diastole, however, was characterized by a slow posterior movement followed by a fast closing movement at the end of ventricular diastole.

Left Atrium in Patients with Heart Malformations

In case 10 an additional abnormal finding was observed. With the transducer directed toward the aortic root, two parallel echoes from the anterior and posterior walls of the aortic root were observed and, behind the aortic root, an echo-free space representing the left atrium (fig. 2, left part). When the transducer was angulated slightly in the caudal direction, however, an abnormal echo appeared within the left atrium (fig. 2, right part). At autopsy a supravalvular ring in the left atrium was found to be so positioned that it was likely to have caused the abnormal echo.

In case 11 there was an abnormal echo posterior to the echo from the anterior mitral leaflet. This finding was confirmed at repeated investigations. The distance between the abnormal echo and the echo from the anterior mitral leaflet was 7 mm during ventricular systole. The pattern of movement of this echo

Figure 7

Angiocardiogram (levogram) in anteroposterior projection after contrast injection into the right ventricle obtained from the patient with cor triatriatum (case 11). Note the large left atrium with the dividing membrane (arrow).
consisted of a slow posterior movement during the first part and a more rapid posterior movement at the end of ventricular systole. In the early part of diastole, a rapid anterior movement occurred, followed first by a posterior and then an anterior small movement coinciding in time with atrial systole (figs. 4E and 6). In this case the relation between the mitral valve and the membrane in the left atrium was demonstrated on angiocardiogram (fig. 7); it was also confirmed at autopsy.

Discussion

The identification of intracardiac ultrasound echoes is based primarily on three circumstances: (1) the direction and depth at which the echo is obtained, (2) the relation to other ultrasound echoes of known origin, and (3) a characteristic movement of the echo in relation to the electrocardiogram or phonocardiogram. In addition, the ultrasoundcardiographic findings can be related to anatomic findings and/or findings at angiocardiography. Recently, a method of identification of echoes by a contrast method has also been published.21

Mitral Atresia

In the two cases of mitral atresia studied here, echoes were obtained from the region where the atretic mitral valve was expected to be localized. This echo was clearly separated from that of the posterior heart wall (fig. 5) and, in case 2, from that of the interventricular septum. The movement of the mitral valve ring or annulus has previously been studied by angiocardiography23 or ultrasoundcardiography.24 The pattern of movement of the mitral ring is characterized by an anterior movement during ventricular systole and a posterior movement during ventricular diastole. This pattern coincides very closely with the pattern of movement of the echo from the mitral valve region in cases 1 and 2. Furthermore, the findings at autopsy revealed no other structure than the atretic mitral valve region that could cause this echo. It seems likely, therefore, that the (abnormal) echoes observed in the cases of mitral atresia do originate from the atretic mitral valve region.

Mitral Stenosis Combined with Aortic Valvular Stenosis or Atresia

In all these cases the echo from the mitral valve region showed a pattern of movement clearly distinguishable from that observed in the cases of mitral atresia. The main distinguishing feature was a rapid anterior movement in the beginning of diastole. This rapid movement is characteristic of an echo from an atrioventricular leaflet.25 The echo apparently did not originate from the tricuspid valve since a separate echo from this leaflet was obtained closer to the sternal border and in a more anterior position. Since the rapid movement was in anterior direction, it is reasonable to assume that this echo originated from the anterior mitral leaflet. In one case simultaneous rapid movements were registered in the beginning of diastole both in anterior and posterior directions. At the end of diastole, simultaneous movements in the opposite direction were seen (fig. 3). This pattern makes it very likely that the observed echoes originated from the anterior and posterior mitral leaflets.

The ultrasoundcardiographic findings from the mitral valve region separate the material into several groups. In one group the echo from the anterior mitral region showed a much reduced total amplitude of movement, especially a small amplitude of opening movement in the early part of diastole. This group was anatomically characterized by a very tight mitral stenosis combined with a moderate or severe degree of hypoplasia of the left ventricle, or, alternatively, by a slight or moderate mitral stenosis combined with a severe degree of hypoplasia of the left ventricle. In both circumstances it is very likely that the hemodymanic function of the left ventricle was very limited.

The next group was characterized by a less reduced total amplitude of movement of the echo from the anterior mitral leaflet but with a distinctly abnormal pattern of movement. Anatomically, this group consisted of cases with slight mitral stenosis combined with a moderate degree of hypoplasia of the left ventricle or, alternatively, by a moderate...
degree of mitral stenosis combined with a normal-sized left ventricle.

Deviations from the patterns described in the groups above were found in cases 11 and 12. In case 11 a normal pattern but a slightly reduced amplitude of movement of the echo from the anterior mitral leaflet was obtained. Autopsy in this case revealed a normal mitral valve, a normal-sized left ventricle, and a membrane dividing the left atrium. In case 12 a normal amplitude but an abnormal pattern of movement of the echo from the anterior mitral leaflet was found despite a normal mitral valve at autopsy. There was, however, a very tight aortic stenosis with a very thick left ventricular wall and a moderate degree of endocardial fibroelastosis.

The abnormal pattern of movement of the echo from the anterior mitral leaflet described above in the patients with mitral stenosis combined with aortic valvular stenosis or atresia closely resembles that previously described in cases of acquired mitral stenosis and in older children with congenital mitral stenosis. A similar pattern of movement of the echo from the anterior mitral leaflet has been reported in cases with severe degree of obstruction of left ventricular outflow, and it has been attributed to reduced compliance of the left ventricular wall. A much reduced compliance of the left ventricular wall can be assumed to have existed in the cases reported here. This can reasonably be explained by a thick left ventricular wall and in many cases by a small left ventricular volume and possibly also by endocardial fibroelastosis.

The pattern of movement of the echo from the anterior mitral leaflet in cases of mitral stenosis can be distinguished from that obtained in cases with severe obstruction to left ventricular outflow if the speed of posterior movement in diastole before atrial systole is measurable (fig. 1). Normally, this speed of movement is more than 90 mm/sec, and in cases of mitral stenosis of clinical significance, it is below 30 mm/sec, and in cases with severe obstruction to left ventricular outflow it is also subnormal but not lower than about 40 mm/sec. In the patients described here, such a measurement was of course not possible because of the tachycardia. Despite this limitation ultrasoundcardiography can provide information about the functional state of the mitral valve and the left ventricle. This should be considered against the limited value of angiocardiography in evaluation of the mitral valve function in this age group.

**Cor Triatriatum**

An echo with the pattern of movement observed in case 11 (figs. 4E and 6) has not been described before. This echo was clearly separated from the echo from the posterior heart wall. It was also separated from the echo from the anterior mitral leaflet. The distance of 7 mm during ventricular systole between the abnormal echo and the echo from the anterior mitral leaflet excludes the posterior mitral leaflet as the origin of the echo. The pattern of movement of this abnormal echo is characterized by a slight movement during ventricular systole and a more pronounced movement during ventricular diastole, especially during atrial systole. This finding together with the position of the echo makes it very likely that it originates from a structure in the left atrium. Furthermore, a comparison of these findings with the angiocardiogram (fig. 7) and the autopsy observations makes it most likely that the abnormal echo originates from the membrane dividing the left atrium. Unfortunately the transducer was not directed toward the left atrium, a positioning which may have been technically better. However, the examination of this patient was made early in the present study.

**Supravalvular Ring in the Left Atrium**

The abnormal echo observed in case 10 is more vague and does not have any characteristic pattern of movement. The echo is, however, situated within the left atrium. A comparison with the findings at autopsy makes it likely that the echo originates from the supravalvular ring in the left atrium. The findings at ultrasoundcardiography do not give any definite suggestion as to diagnosis.
but may draw the attention to the left atrium for a further detailed analysis by angiocardiography.

As illustrated especially by cases 10 and 11, the region of the left atrium is well suited for investigation with ultrasoundcardiography. Similar experience has also been gained in adults; for example, left atrial myxomas are well demonstrated by this method.27

**Other Ultrasoundcardiographic Observations**

In all cases reported here, an enlargement of the right ventricle was found at autopsy. It had been found earlier that in cases with dilatation of the right ventricle a complete echo from the anterior tricuspid leaflet can be obtained with the transducer in anteroposterior direction.17 The observations reported here support this view. The observation that an echo from the interventricular septum could be obtained in all cases except those with a single ventricle is in agreement with earlier findings.18

**Conclusions**

This investigation shows that in patients with congenital heart malformations, ultrasoundcardiography performed in early life can provide additional information about the mitral valve region and the left atrium and may also be of great diagnostic value in some cases. The method is harmless, noninvasive, and can be used as a bedside investigation even in seriously ill infants. The method should thus be regarded as a complement to ordinary methods of investigation, for which it sometimes can serve as a guide.

**Addendum**

Since submission of this paper two more patients of this group have been studied. Both have died, one at the age of 3 months and the other at the age of 1 week. Autopsy was performed on both. In the first of these patients a mitral atresia and a single ventricle were found. The findings at ultrasoundcardiography closely resembled those obtained in case 1 in the present material. The other patient had an aortic valvular atresia, a moderate degree of mitral stenosis, and a severely hypoplastic left ventricle. The ultrasoundcardiographic findings in this patient closely resembled those obtained in case 3 in the present material.

The results obtained in these two additional patients thus agree quite well with those presented more extensively previously in this paper.

**References**


2. **LEV M**: Pathologic anatomy and interrelationship of hypoplasia of the aortic tract complex. Lab Invest 1: 61, 1952


4. **ERIKSSON BO, GRANT CA**: What is the "hypoplastic left heart syndrome?" Proc Ass Europ Paediat Cardiol 6: 29, 1970


8. **KANJUH VK, ELLIOT RS, EDWARDS JE**: Coexistent mitral and aortic valvular atresia. Amer J Cardiol 15: 611, 1965

9. **KAPLAN S**: Diagnosis and management of hypoplastic left heart syndrome and preductal coarctation. Proc Ass Europ Paediat Cardiol 6: 22, 1970


15. **EFFERT S**: Pre- and postoperative evaluation of mitral stenosis by ultrasound. Amer J Cardiol 19: 59, 1967

16. **SEGAL BL, LIKOFF W, KINGSLEY B**: Echocardi-
ography: Clinical application in mitral regurgitation. Amer J Cardiol 19: 50, 1967

Circulation, Volume XLV, February 1972
Ultrasoundcardiographic Studies of the Mitral Valve Region in Young Infants with Mitral Atresia, Mitral Stenosis, Hypoplasia of the Left Ventricle, and Cor Triatriatum
NILS-RUNE LUNDSTRÖM

Circulation. 1972;45:324-334
doi: 10.1161/01.CIR.45.2.324
Circulation is published by the American Heart Association, 7272 Greenville Avenue, Dallas, TX 75231
Copyright © 1972 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/45/2/324

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/