Echocardiography in the Diagnosis of Hypoplasia of the Left or Right Ventricles in the Neonate

By Richard A. Meyer, M.D., and Samuel Kaplan, M.D.

SUMMARY

Echocardiographic studies were performed on six patients with autopsy-proved aortic atresia and hypoplastic left ventricle and two patients with surgically proved tricuspid atresia and hypoplastic right ventricle. The findings were compared to those from the echograms of 50 normal newborns who served as a control group. The ultrasound measurements obtained in the normal newborns were as follows: mean right ventricular end-diastolic dimension, 1.3 cm (range, 1.0 to 1.7), mean left ventricular end-diastolic dimension, 1.6 cm (range, 1.2 to 2.0), mean left atrial dimension, 0.9 cm (range, 0.6 to 1.3), and mean left ventricular outflow tract dimension, 1.0 cm (range, 0.7 to 1.2).

The mean velocities of closure of the mitral and tricuspid valves during passive filling of the ventricles were 53 mm/sec and 43 mm/sec. The ranges were 36 to 80 mm/sec for the mitral valve and 34 to 56 mm/sec for the tricuspid valve.

In the patients with aortic atresia, the significant findings were a larger than normal right ventricular chamber (mean, 2.5 cm), a left ventricular chamber measuring less than 0.9 cm, and an absent or grossly distorted mitral valve echo. By contrast the patients with tricuspid atresia had very small right ventricular dimensions less than 0.6 cm, no demonstrable tricuspid valve echo, and a larger than normal left ventricle (mean, 2.3 cm). The above findings were diagnostic and were not confused with the normal newborn or other forms of congenital heart disease.

Additional Indexing Words:
Ultrasound cardiology Hypoplastic left ventricle syndrome Aortic atresia
Tricuspid atresia

ECHOCARDIOGRAPHY has been used frequently in adults to record mitral and tricuspid valve motion,1-3 to define left ventricular outflow tract (LVOT) and aortic valve motion,4 to detect pericardial effusions,5 and to measure right and left ventricular dimension6 as well as stroke volume.7-8 Its use in pediatric patients has been limited.9-11 The accurate diagnosis of congenital heart disease in neonates presently depends upon cardiac catheterization and selective angiocardiography which are associated with a significant risk.12 Since echocardiography is noninvasive and carries no risk, its application in the diagnosis of neonatal congenital heart disease seems ideal.

The purpose of this study is to report the echocardiographic findings in normal newborns and to describe the findings in neonates with proven or clinically suspected congenital heart disease. Findings in patients with aortic atresia and hypoplasia of the left ventricle and tricuspid atresia are emphasized.

Method

Group Studied

Echocardiograms were obtained on three groups of patients. Group 1 was composed of 50 newborns (31 males and 19 females) judged to be normal by physical examination after entry to the University of Cincinnati newborn nurseries.
Their ages were 5 to 125 hours, and they served as the control group. Group 2 consisted of eight patients. Six (ages, 1 to 6 days) had aortic atresia and hypoplasia of the left ventricle confirmed at autopsy. This diagnosis was made when there was complete obstruction of the aortic valve with hypoplasia of the ascending aorta and left ventricle. Three underwent cardiac catheterization and aortography; three did not. The seventh and eighth patients of group 2 had tricuspid atresia with hypoplasia of the right ventricle proved by cardiac catheterization. Both patients had systemic-to-pulmonary arterial shunts. One died and the diagnosis of tricuspid atresia was confirmed at autopsy. The other is presently doing well.

Group 3 consisted of four patients (ages, 1 to 7 days) with symptoms and signs of aortic atresia on admission to the hospital but whose echocardiograms were not abnormal. The symptoms and signs consisted of poor to absent peripheral arterial pulses, gray color, hepatomegaly, cardiomegaly, and hypoglycemia. Three of the four had normal hearts on follow-up examination. The remaining patient had transposition of the great vessels proved by cardiac catheterization and underwent a Raskind balloon septotomy.

**Technic**

Ultrasound examinations were performed on patients who were supine. None of the patients was sedated. Occasionally a sugar nipple was required to keep the baby contented and quiet, but it did not interfere with the examination. Since the study was conducted in two separate hospitals, different ultrasonoscopes were employed.* The transducer, made by Nortec Corporation, was used with both units. It had an active diameter of ½ inch and an outer diameter of ¾ inch. The repetition rate when used with the Hoffrel unit was 1,000/sec and with the Unirad unit was also 1,000/sec. A water-soluble gel was used to obtain an airless contact between the transducer and the skin.

The normal echogram of the anterior mitral leaflet (as described by Edler1 in adults) was obtained in neonates by placing the transducer over the third or fourth intercostal space either at the left sternal border or over the sternum and directing the beam posteriorly. Since the sternum is not calcified in the neonate, it offers little interference in obtaining substernal echoes. In neonates with enlarged hearts, the transducer often had to be positioned in the xiphoid notch or in the fourth or fifth intercostal space to the left of the sternum in order to obtain the echo from the anterior leaflet of the mitral valve.

From the mitral position, slight lateral-inferior rotation of the transducer provided strong endocardial echoes from the free wall of the left ventricle and the epicardium of the right ventricle (fig. 1). The right ventricular epicardial echoes were recorded as an undulating echo immediately subjacent to the straight lines of the chest wall. If the epicardial echoes were not a distinct undulating line, then the fuzzy or stippled echoes posterior to the heavy straight lines were considered to represent the surface of the right ventricle. Identifiable echoes from the ventricular septum also appearing in this plane permitted measurement of right and left ventricular size. The right ventricular dimension was measured as the distance between the right ventricular epicardial echoes and the echoes from the right side of the septum. Left ventricular dimensions were measured as the distance between the left septal echo and the left ventricular endocardial echo. All measurements were made in end-diastole. From this position, medial rotation of the transducer in either a superior or inferior direction revealed an echo from an anterior tricuspid valve leaflet.

In the normal adult,2 the depth of the mitral valve echo is 6 to 8 cm. In the neonate, however, all the cardiac echoes were seen within 5 to 6 cm of the skin surface. Therefore, the scale was expanded and the depth compensation control or ramp control for distant echoes adjusted for maximum gain at 3 to 4 cm in order to obtain the strongest echoes from the mitral valve and left ventricular free wall. For the ventricular septum and tricuspid valve, the gain and ramp control for near echoes was increased accordingly. Clarity of the echoes was obtained by adjusting the reject or clipper control. It often was necessary to reduce the near gain or ramp in order to avoid obscuring important echoes in the near field such as the tricuspid valve or the anterior echo of the left ventricular outflow tract (LVOT).

To obtain an aortic root echo4 and hence the left ventricular outflow tract (LVOT), the strong mitral valve echo was located. The transducer followed the mitral valve echo medially and superiorly until the mitral ring echo was identified13 which represented the posterior limits of the aortic root or left ventricular outflow tract (LVOT) (fig. 2). The posterior aortic root echo and mitral ring echo are at the same depth in normal individuals. This echographic finding establishes mitral-aortic continuity.11 A similar echo lay anterior to the mitral ring echo and undulated synchronously with it. This echo

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*Hoffrel Ultrasoundoscope model 101 with slow sweep accessory was used for neonates with heart disease. Unirad 100 Series Diagnostic Echoscope was used for normal neonates.
Figure 1

Echogram of normal newborn demonstrating the systolic and diastolic dimensions of the right and left ventricles. The sweep period is 2 sec compared to 4 sec in the other figures. CW = chest wall; RVED = right ventricular end-diastolic dimension; LVED = left ventricular end-diastolic dimension; RS = right septal echo; LS = left septal echo; $D_s$ = diastolic dimension; $D_d$ = systolic dimension. Parts of the left septal echo and endocardial echo of posterior left ventricular wall were retouched for clarification.

represented the fibrous attachment of the tricuspid valve and membranous septum and defined the anterior limits of the left ventricular outflow tract (LVOT)\(^4\) (fig. 3b). Left ventricular outflow tract (LVOT) dimensions were measured from peak to peak in end-systole. Between the left ventricular outflow tract (LVOT) and the anterior myocardial echoes was the right ventricular outflow tract (RVOT).\(^4\) Another echo lay posterior to the mitral ring which displayed little motion and has been described previously\(^15\) as the posterior wall of the left atrium. When this echo did move, it was M shaped and moved anteriorly in diastole. Left atrial dimensions were obtained by measuring the distance between the mitral ring echo and the posterior left atrial wall at the end of ventricular systole (figs. 3b, 4).

Results

Figure 5 is an idealized diagram, which illustrates the characteristic waveforms of the reflected ultrasound from various cardiac structures and relates them to temporal events in the cardiac cycle. The waveforms of the mitral and tricuspid valves\(^2\) are identical. Point E (fig. 5) represents the most anterior position attained by the echo source, and C represents the most posterior position. Thus, the total amplitude of the anteroposterior movement of the anterior mitral or tricuspid leaflet between end-diastole and early diastole is from point C to point E. The depth of the
Mitral or tricuspid valve was measured as the distance between point C or each valve echo and the anterior chest wall. The speed of diastolic downstroke or velocity of closure of the valve is measured from E to F. Table 1 gives the echocardiographic findings in the 50 normal neonates.

Typical echograms of newborns are seen in figures 1, 3, and 4. The heavy white lines at the top of the echogram originate from the chest wall and average 1 cm in thickness. The chest wall echoes were electronically suppressed in some cases (fig. 7). In every normal newborn a mitral valve and a tricuspid valve echo were recorded. The mitral valve echo was seen at an average depth of 3.8 cm from the anterior chest wall and was in continuity with a semilunar valve presumed to be aortic. The mean amplitude of leaflet motion was 1.0 cm, and the E-F slope or velocity of closure measured 36 to 80 mm/sec with a mean of 53 mm/sec. In normal adults this value is 90 to 190 mm/sec. The tricuspid valve echo, seen anterior to the mitral echo, was a mean distance of 2.8 cm from the chest wall and had an excursion of 1.1 cm. The E to F slope measurable in 12 patients was 34 to 56 mm/sec (mean, 43 mm/sec). This value compares to 60 to 125 mm/sec found in the normal adult. Immediately posterior to the tricuspid valve echo was the ventricular septal echo which appeared more posterior in this view because of the angle of the transducer necessary to obtain the tricuspid valve echo. The straight-line echoes through the tricuspid valve echo were transducer artifacts.

The mean dimension of the left ventricular outflow tract (fig. 3b) was 1.0 cm, and the mean left atrial dimension was 0.9 cm. Frequently an echo from the posterior cusp of the aortic valve was seen within the left ventricular outflow tract (LVOT) during diastole. The mean value of the right ventricular end-diastolic dimension was 1.2 cm. The left ventricular end-diastolic dimension was 1.6 cm while the right ventricular end-diastolic mean was 1.3 cm.

Echograms of the patients with aortic atresia (fig. 6) and tricuspid atresia (fig. 7) were distinctly different from those of the normal newborns. The diagnostic features of aortic atresia were: a large (2.3 to 3.2 cm) right ventricular cavity, a minute (0.4 to 0.9 cm) left ventricular cavity, and absence of, or a poorly visualized, mitral valve echo. These findings contrasted sharply with those in patients with tricuspid atresia (table 2). In tricuspid atresia, the echograms demonstrated a minute (0.4 to 0.6 cm) right ventricular cavity, an absent tricuspid valve echo, and a large (2.0 to 2.6 cm) left ventricular cavity. The depth of the mitral valve from the anterior chest wall was normal. This was attributed to the fact that the small cavity of the right ventricle was surrounded by a thick
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Figure 3

Echocardiogram of normal newborn. (a) Sweep period 2 sec. (b) Sweep period 4 sec. CW = chest wall; RV cavity = right ventricular cavity; LV cavity = left ventricular cavity; RVOT = right ventricular outflow tract; LVOT = left ventricular outflow tract; post. cusp = posteror aortic cusp; LA = left atrium. Parts of the endocardial echo of the posterior left ventricular wall were retouched for clarification.

wall and that the left ventricular cavity was enlarged. Thus, the normal depth of the mitral valve is not unexpected. In patients with aortic atresia, the left ventricular outflow tract (LVOT) was unidentifiable; however, in patients with tricuspid atresia, aortic-mitral continuity existed and an outflow tract echo was recorded. The unaffected A-V valve in each disease appeared normal on the echograms.

The patients in group 3 clinically appeared to have aortic atresia upon admission to the hospital. Yet the echocardiographic findings of these patients did not resemble those of the patients with proved aortic atresia. Their echograms demonstrated normal ventricular chamber size and A-V valve motion as well as mitral-semilunar valve continuity and normal left ventricular outflow tract dimensions (table 3). The cause of the shocklike symptoms in three of the four patients with cardiomegaly is unknown. Treatment for sepsis resulted in marked clinical improvement and on follow-up examinations the hearts were normal. It is possible that infectious myocarditis was a part of the generalized sepsis. The posterior ventricle in the patient with transposition of the great arteries was large when compared to normal, and the left ventricular end-diastolic dimension (LVED) was in the same range as that of the patients with tricuspid atresia. This patient had an intact ventricular septum and no angiographic evidence of obstruction to the outflow of the posterior ventricle or a patent ductus arteriosus. The cause of the large LVED dimension is not clear.

Discussion

Lundstrom and Edler found that the velocity of the anterior mitral leaflet closure during early diastole in 45 normal children (age, 1 week to 15 years) was 90 to 140 mm/sec. In our normal patients the speed of
Mitral Valve

Tricuspid Valve

Figure 4

Echogram of normal newborn showing the depth and waveform of the mitral and tricuspid valves. M.V. = mitral valve; T.V. = tricuspid valve. Mitral and tricuspid valve echoes were retouched for clarification.

closure of the anterior mitral leaflet was much slower, 36 to 80 mm/sec. Even though the

Table 1

Data on Ultrasound Cardiograms (Echocardiograms) of 50 Normal Neonates

<table>
<thead>
<tr>
<th>Group 1</th>
<th>Range</th>
<th>Mean</th>
</tr>
</thead>
<tbody>
<tr>
<td>M = 31; F = 19</td>
<td></td>
<td></td>
</tr>
<tr>
<td>Age (hr)</td>
<td>5 - 125</td>
<td>44</td>
</tr>
<tr>
<td>Wt (kg)</td>
<td>2.3 - 4.9</td>
<td>3.2</td>
</tr>
<tr>
<td>Tricuspid valve</td>
<td></td>
<td></td>
</tr>
<tr>
<td>Depth (cm)</td>
<td>2.4 - 3.2</td>
<td>2.8</td>
</tr>
<tr>
<td>Velocity (mm/sec)</td>
<td>34 - 56</td>
<td>43</td>
</tr>
<tr>
<td>Excursion (cm)</td>
<td>0.8 - 1.4</td>
<td>1.1</td>
</tr>
<tr>
<td>Mitral valve</td>
<td></td>
<td></td>
</tr>
<tr>
<td>Depth (cm)</td>
<td>3.1 - 4.7</td>
<td>3.8</td>
</tr>
<tr>
<td>Velocity (mm/sec)</td>
<td>36 - 80</td>
<td>53</td>
</tr>
<tr>
<td>Excursion (cm)</td>
<td>0.6 - 1.2</td>
<td>1.0</td>
</tr>
<tr>
<td>LA diameter</td>
<td>0.6 - 1.3</td>
<td>0.9</td>
</tr>
<tr>
<td>LV outflow tract</td>
<td>0.7 - 1.2</td>
<td>1.0</td>
</tr>
<tr>
<td>LV end-diastolic dimension</td>
<td>1.2 - 2.0</td>
<td>1.6</td>
</tr>
<tr>
<td>RV end-diastolic dimension</td>
<td>1.0 - 1.7</td>
<td>1.3</td>
</tr>
</tbody>
</table>

Abbreviations: M = male; F = female; LA = left atrium; LV = left ventricle; RV = right ventricle.

heart rates of these neonates averaged 120 beats/min, the majority of patients exhibited an A wave following the E to F slope (fig. 2a). A ready explanation for the apparent discrepancy between the neonate and adult values is not forthcoming. Likewise, closure of the anterior tricuspid leaflet is slower in the neonate (34 to 56 mm/sec) than in the adult.

Echoes of the anterior tricuspid leaflet were easily and consistently obtained in neonates in contrast to adults or older children. This was attributed to lack of calcification of the sternum and to the proximity of the valve to the transducer. Thus, in patients suspected of having tricuspid atresia, inability to record a tricuspid valve assumes diagnostic significance, particularly if a careful search for the valve in the usual positions has been made. Chesler and associates\(^\text{10}\) suggested that the ultrasonic findings in tricuspid atresia with

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MEYER, KAPLAN

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In patients with aortic atresia and a hypoplastic left ventricle, a normal anterior mitral valve leaflet is usually not recorded since mitral stenosis or atresia is coexistent (fig. 6). Occasionally, a recognizable mitral echo may be recorded, but the amplitude of the echo is small and grossly deformed, and diastolic closure when measurable is slow (table 2).

The echocardiographic findings in the patients who had aortic atresia with hypoplastic left ventricle and tricuspid atresia with hypoplastic right ventricle (group 2) were compared to 25 other neonates (not described in this study) with a variety of congenital cardiac defects. Those defects included ventricular septal defects, endocardial cushion defects, tetralogy of Fallot, single ventricle, total anomalous pulmonary venous drainage, and coarctation syndrome. The echograms of the patients with aortic and tricuspid atresia were diagnostic and not confused with any of the above congenital cardiac diseases. Patients who clinically appear to have aortic atresia but whose echocardiograms do not confirm the clinical impression must have further investigation for correct diagnosis. This was demonstrated in the four patients in group 3 when renewed effort was made to establish the correct diagnosis after the demonstration of normal echograms.

Table 2

Echograms of Patients with Heart Disease

<table>
<thead>
<tr>
<th>Patient</th>
<th>Sex</th>
<th>Age at death (days)</th>
<th>LVED (cm)</th>
<th>RVED (cm)</th>
<th>Mitral valve echo</th>
<th>Tricuspid valve echo</th>
</tr>
</thead>
<tbody>
<tr>
<td></td>
<td></td>
<td></td>
<td>D (cm)</td>
<td>A (cm)</td>
<td>V (mm/sec)</td>
<td>D (cm)</td>
</tr>
<tr>
<td>A. Aortic atresia and hypoplastic left ventricle</td>
<td></td>
<td></td>
<td></td>
<td></td>
<td></td>
<td></td>
</tr>
<tr>
<td>1</td>
<td>M</td>
<td>2</td>
<td>0.8</td>
<td>2.3</td>
<td>4.0</td>
<td>0.4</td>
</tr>
<tr>
<td>2</td>
<td>M</td>
<td>2</td>
<td>0.9</td>
<td>2.2</td>
<td>4.0</td>
<td>0.5</td>
</tr>
<tr>
<td>3</td>
<td>M</td>
<td>6</td>
<td>0.4</td>
<td>2.6</td>
<td>NM</td>
<td>NM</td>
</tr>
<tr>
<td>4</td>
<td>M</td>
<td>3</td>
<td>0.4</td>
<td>2.8</td>
<td>NM</td>
<td>NM</td>
</tr>
<tr>
<td>5</td>
<td>M</td>
<td>24</td>
<td>0.8</td>
<td>2.6</td>
<td>4.1</td>
<td>0.3</td>
</tr>
<tr>
<td>6</td>
<td>M</td>
<td>6</td>
<td>0.6</td>
<td>3.2</td>
<td>3.8</td>
<td>0.4</td>
</tr>
<tr>
<td>B. Tricuspid atresia with hypoplastic right ventricle</td>
<td></td>
<td></td>
<td></td>
<td></td>
<td></td>
<td></td>
</tr>
<tr>
<td>7</td>
<td>M</td>
<td>3</td>
<td>2.0</td>
<td>0.4</td>
<td>3.0</td>
<td>1.0</td>
</tr>
<tr>
<td>8</td>
<td>F</td>
<td>Alive</td>
<td>2.6</td>
<td>0.6</td>
<td>3.9</td>
<td>1.5</td>
</tr>
</tbody>
</table>

Abbreviations: LVED = left ventricular end-diastolic dimension; RVED = right ventricular end-diastolic dimension; D = depth; A = amplitude; V = velocity; NM = nonmeasurable.

Circulation, Volume XLVI, July 1972
Echogram from a patient with aortic atresia and hypoplastic left ventricle (upper right) demonstrates a minute left ventricular cavity and grossly distorted mitral valve echo compared to a normal neonate (lower right). A photograph and diagram of a heart from such a patient are shown. RV = right ventricle; TV = tricuspid valve; LV = left ventricle; A.M.E. = anterior mitral valve echo; RA = right atrium; PA = pulmonary artery; LA = left atrium. In the pathologic specimen, the upper arrow points to the atretic mitral valve (MV) and the lower arrow points to the endocardium of the left ventricle (LV).

Unfortunately, transposition of the great vessels in the newborn cannot be differentiated echocardiographically from a normal heart since there is continuity between the mitral valve and the semilunar valve. In a neonate with signs of cyanotic cardiac disease and a normal echocardiogram, cardiac catheterization and angiography are general-

Table 3

<table>
<thead>
<tr>
<th>Patient</th>
<th>Sex</th>
<th>Age (days)</th>
<th>Diagnosis</th>
<th>LVED (cm)</th>
<th>RVED (cm)</th>
<th>Mitral valve echo</th>
<th>Tricuspid valve echo</th>
</tr>
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<tbody>
<tr>
<td></td>
<td></td>
<td></td>
<td></td>
<td>D (cm)</td>
<td>A (cm)</td>
<td>V (mm/sec)</td>
<td>D (cm)</td>
</tr>
<tr>
<td>a</td>
<td>M</td>
<td>1</td>
<td>NL</td>
<td>1.2</td>
<td>1.4</td>
<td>4.1 0.8 66</td>
<td>2.8</td>
</tr>
<tr>
<td>b</td>
<td>M</td>
<td>1</td>
<td>NL</td>
<td>1.8</td>
<td>1.4</td>
<td>3.5 0.9 46</td>
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<tr>
<td>c</td>
<td>M</td>
<td>14</td>
<td>NL</td>
<td>1.1</td>
<td>1.2</td>
<td>2.8 0.6 44</td>
<td>1.8</td>
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<tr>
<td>d</td>
<td>M</td>
<td>7</td>
<td>TGV</td>
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<td>1.4</td>
<td>4.0 0.8 40</td>
<td>3.0</td>
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</tbody>
</table>

* A 3-pound premature infant.

Abbreviations: LVED = left ventricular end-diastolic dimension; RVED = right ventricular end-diastolic dimension; D = depth; A = amplitude; V = velocity; NL = normal heart; TGV = transposition of the great vessels.
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TRICUSPID ATRESIA

Figure 7

Echogram in patient with tricuspid atresia showing minute right ventricular cavity compared to large left ventricular cavity. The chest wall echoes were electronically suppressed. The tricuspid valve echo is absent. The lateral view of a left ventriculogram and an idealized drawing are also shown. RV cavity = right ventricular cavity; IV septum = interventricular septum; LV cavity = left ventricular cavity; VSD = ventricular septal defect; Ao = aorta; PA = pulmonary artery; RV = right ventricle; LV = left ventricle.

Echogram in patient with tricuspid atresia showing minute right ventricular cavity compared to large left ventricular cavity. The chest wall echoes were electronically suppressed. The tricuspid valve echo is absent. The lateral view of a left ventriculogram and an idealized drawing are also shown. RV cavity = right ventricular cavity; IV septum = interventricular septum; LV cavity = left ventricular cavity; VSD = ventricular septal defect; Ao = aorta; PA = pulmonary artery; RV = right ventricle; LV = left ventricle.

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Circulation. 1972;46:55-64
doi: 10.1161/01.CIR.46.1.55
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

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