Objective: To evaluate the incidence and types of congenital heart defects associated with meningomyelocele.

Design: All neonates who underwent meningomyelocele repair and had a perioperative echocardiogram from July 1990 to October 1998 were studied. Medical records were reviewed for age, weight, clinical cardiac examination results, meningomyelocele location, and associated noncardiac defects. Heart defects were identified from reviewing echocardiographic reports and videotapes.

Results: At meningomyelocele surgery, the 105 patients (53 female; 52 male) ranged in age from 1 to 20 days and in weight from 0.6 to 4.1 kg. Congenital heart disease was detected in 39 patients (37%). A secundum atrial septal defect was the most common defect (24%). A ventricular septal defect was found in 10 patients, 2 patients had anomalous pulmonary venous return, and 1 each had tetralogy of Fallot, bicuspid aortic valve, coarctation, and hypoplastic left heart syndrome. A patent ductus arteriosus and patent foramen ovale were not considered abnormal in these neonates. The cardiac examination was abnormal in only 5 of the 39 patients with heart defects (sensitivity = 13%). The presence of associated noncardiac defects (in addition to meningomyelocele) and location of the meningomyelocele (cervicothoracic vs lumbar) did not affect the incidence of heart disease. Of the patients with heart defects, girls were more frequently affected (25 of 39 vs 14 of 39, P < .05).

Conclusions: Congenital heart defects are common in neonates, especially girls, with meningomyelocele and are unrelated to meningomyelocele location or associated noncardiac defects. Because the clinical examination is insensitive for detecting heart defects in this group, screening echocardiograms are warranted. This information has important implications for ventriculoatrial shunting, urinary tract instrumentation (antibiotic prophylaxis), and neurosurgical procedures (venous air embolism).


Editor's Note: In this age of cost-conscious care, it's nice to have a study like this that points out the value of performing a relatively expensive diagnostic test in certain newborns.

Catherine D. DeAngelis, MD

MENINGOMYELOCELE is the most severe form of dysraphism involving the vertebral column. Although its exact etiology is unknown, a multifactorial origin seems likely.1 Previous studies report that a high number of live-born infants with a meningomyelocele have at least 1 major malformation involving the central nervous, skeletal, cutaneous, or genitourinary system.1,2 The few previous investigations of associated cardiovascular malformations have suggested an increased incidence of conotruncal defects (Kousseff syndrome) and ventricular septal defects.3-7

The purpose of this study was to determine the incidence and types of congenital heart defects occurring in patients with meningomyelocele.

RESULTS

Of the 130 patients diagnosed with meningomyelocele during the study period, 105 satisfied the inclusion criteria. The characteristics of the study group are summarized in the Table. At the time of surgery, they ranged in age from 1 to 20 days and in weight from 0.6 to 4.1 kg. There was no significant difference in the number of boys and girls. Most patients had a meningomyelocele in the lumbar area. Additional noncardiac defects were present in 85%, including 65 patients with hydrocephalus, 36 with urinary tract involvement (neurogenic bladder, hydronephro-
PATIENTS AND METHODS

Neonates diagnosed with meningomyelocoele between July 1990 and October 1998 were identified by searching the hospital computer database. All neonates undergoing meningomyelocoele repair (open meningomyelocoele) within the first month of life were included. Because it is part of the perioperative protocol for meningomyelocoele repair at our institution, all patients had a perioperative complete 2-dimensional and Doppler echocardiogram.

The patient records were reviewed for demographic data, meningomyelocoele location, and associated noncardiac defects. Heart defects were identified by reviewing echocardiographic reports and videotapes. Only the primary cardiac diagnosis was recorded for each patient. A true secundum atrial septal defect was diagnosed when all the following were present: (1) a defect in the atrial septum larger than 4 mm without a tissue flap to cover the opening, (2) t-artifact at each edge, and (3) a low-velocity shunt (<1.5 m/s) across the defect. A patent foramen ovale and patent ductus arteriosus were considered to be normal for this age group.

All data are expressed as mean ± SD. Nonparametric data were compared using the χ² or Fisher exact tests. Statistical significance was inferred at P<.05.

The study was approved by the institutional review board at Primary Children’s Medical Center, Salt Lake City, Utah.

Patient Characteristics (n = 105)*

<table>
<thead>
<tr>
<th>Characteristic</th>
<th>Value</th>
</tr>
</thead>
<tbody>
<tr>
<td>Mean ± SD age, d (range)</td>
<td>2.5 ± 2.7 (1-20)</td>
</tr>
<tr>
<td>Mean ± SD weight, kg (range)</td>
<td>3.0 ± 0.7 (0.6-4.1)</td>
</tr>
<tr>
<td>Sex</td>
<td></td>
</tr>
<tr>
<td>M</td>
<td>52</td>
</tr>
<tr>
<td>F</td>
<td>53</td>
</tr>
<tr>
<td>Meningomyelocoele location</td>
<td></td>
</tr>
<tr>
<td>Lumbar</td>
<td>93</td>
</tr>
<tr>
<td>Cervicothoracic</td>
<td>12</td>
</tr>
<tr>
<td>Additional noncardiac defects</td>
<td>89</td>
</tr>
</tbody>
</table>

*Data are expressed as number of patients unless otherwise indicated.

There were no significant differences in the incidence of heart defects between the groups with and without noncardiac defects (Figure 1). Similarly, the incidence of heart defects was not significantly different between the group of neonates with lumbar and the group with cervicothoracic meningomyelocoeles (Figure 2). Heart defects were significantly more common (25 of 39) in girls (Figure 3).

Meningomyelocoeles occur at the end of week 4 of development if the neural tube fails to close spontaneously. If overproduction of spinal fluid occurs at this stage of development, the neural tube distends and fluid can infiltrate the surrounding mesoderm, destroying neural crest...
The neural crest cells play an important role in forming mesodermal organs such as the heart, urinary tract, and skeleton; thus, their destruction may prevent normal development of both the spinal cord and the heart. In addition, both the heart and central nervous system rely on homocysteine for their normal development. The neural crest cells play an important role in forming mesodermal organs such as the heart, urinary tract, and skeleton; thus, their destruction may prevent normal development of both the spinal cord and the heart. In addition, both the heart and central nervous system rely on homocysteine for their normal development.

Previous studies have demonstrated a 5% to 10% incidence of congenital heart defects in patients with neural tube defects. These studies differ from ours, however, because they included all neural tube defects rather than just open meningomyeloceles. The incidence of congenital heart defects in our study was 37% of patients with an open meningomyelocele (considerably higher than the 1% risk given for the general population), with atrial septal defects being the most common abnormality. We also found that newborn girls with meningomyelocele are more frequently affected. This may be explained by the higher incidence of atrial septal defects in the general female population as well. Because neither meningomyelocele location nor the presence or absence of additional nondescardiac defects were similar between those with and without heart defects, they cannot be used to predict patients at higher risk. In addition, because most of the defects are clinically silent at this age, early diagnosis can be made only by echocardiographic screening.

The identification of even minor heart defects has several important implications for this group of patients. First, spinal fluid is often overproduced in patients with a meningomyelocele, resulting in hydrocephalus after repair. If the hydrocephalus requires treatment, it is important to be aware that surgical procedures may prevent normal development of the spinal cord and the heart. In addition, both the heart and central nervous system rely on homocysteine for their normal development. The neural crest cells play an important role in forming mesodermal organs such as the heart, urinary tract, and skeleton; thus, their destruction may prevent normal development of both the spinal cord and the heart. In addition, both the heart and central nervous system rely on homocysteine for their normal development.

This study is limited by being retrospective, but we were able to obtain complete data from the clinical records. Although it may be difficult to differentiate between a secundum atrial septal defect and a patent foramen ovale, every attempt was made to include only true secundum atrial septal defects by adhering to strict criteria and reviewing all videotapes independent of the report. If there was a question, the defect was considered a patent foramen ovale.

Thus, congenital heart defects are common in neonates with meningomyelocele, especially in female patients. Because neither meningomyelocele location nor the presence of associated nondescardiac defects can be used to predict patients at risk for heart defects, and the clinical examination is insensitive for detecting heart defects in this group, screening echocardiograms are warranted. This information has important implications for patients who may require a ventriculostomal shunt, those at risk for endocarditis from urinary tract instrumentation, and those at risk for venous air embolism with certain neurosurgical procedures.

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REFERENCES