Are Screening Echocardiograms Warranted for Neonates With Meningomyelocele?

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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 ventriculooatrial 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. 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. The few previous investigations of associated cardiovascular malformations have suggested an increased incidence of conotruncal defects (Kousseff syndrome) and ventricular septal defects.

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

The patient records were reviewed for demographic data, meningomyelocele 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.

<table>
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<th>Patient Characteristics (n = 105)*</th>
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<td>Mean ± SD age, d (range)</td>
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*Data are expressed as number of patients unless otherwise indicated.

sis, vesicoureteral reflux), 26 with skeletal abnormalities (club feet, scoliosis, hip dysplasia), 3 with intestinal abnormalities (cloacal dystrophy, rectal agenesis, rectal prolapse), and 2 with an identifiable clinical syndrome (Di-George, Cornelia de Lange).

The echocardiogram was abnormal in 39 patients (37%). Abnormalities included a secundum atrial septal defect in 25 (24%), ventricular septal defect in 10 (10%), and anomalous pulmonary venous return in 2 (2%). Tetralogy of Fallot, bicuspid aortic valve, coarctation, and hypoplastic left heart syndrome each occurred in 1 patient. Although a patent ductus arteriosus was found in 50 patients and a patent foramen ovale in 32, they were considered to be normal at this age and those patients were not included in the group with cardiac defects.

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 meningomyeloceles (Figure 2). Heart defects were significantly more common (25 of 39) in girls (Figure 3).

Only 5 of the 39 patients with echocardiographic evidence of congenital cardiac defects had abnormal cardiac examination results (performed by the neonatologist, pediatrician, or neurosurgeon), for a sensitivity of 13%.

Meningomyeloceles 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...
quent manipulations of the genitourinary tract or from fre-
gomyelocele who have associated renal defects and may
endocarditis is very important for patients with menin-
gomyelocele and those at risk for venous air embolism with cer-
tinal neurosurgical procedures.

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REFERENCES

15. Harris MM, Yeman TA, Davidson A, et al. Venous air embolism during cranio-

Figure 3. Relationship of heart defects to the sex of the patient. Asterisk indicates P < .05.