Birth defects account for more than 21% of all infant deaths, and continue to be the leading cause of infant mortality. Neural tube defects, among the most common birth defects in the United States, are congenital malformations of the brain and spinal cord. These are classified as open or closed. They are considered open if neural tissue is exposed or covered only by a membrane, and are considered closed if the defect is covered with normal skin.

Cranial dysraphism (i.e., anencephaly, exencephaly, encephalocele) comprises one half of all neural tube defects. The rest occur along the spine. Myelomeningocele, also known as spina bifida cystica, is the most common congenital anomaly of the CNS, with a prevalence rate of 4.4 to 4.6 cases per 10,000 live births.

**Cause**

The embryologic basis of the clinical variation in neural tube defects is poorly understood. The clinical manifestations of the defect vary, and depend on the location of the failure of the neural tube to close.

Because the development and closure of the neural tube are completed within 28 days of conception, myelomeningocele occurs at the end of the fourth week of development if the neural tube fails to close spontaneously. A closed tube may reopen, however.

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myelomeningocele may occur anywhere along the neuraxis, the lumbo-
sacral region accounts for most cases. Myelomeningocele in the mid-
lumbar region can disrupt the conus medullaris, producing lower motor
neuron abnormalities, adversely affecting bladder and bowel control and
strength and posture in the lower extremities.\textsuperscript{37, 63} Cervical and upper
thoracic myelomeningocele account for less than 5% of all myelomening-
goceles.\textsuperscript{46}

**Associated Anomalies**

Although 20\% of affected patients have additional congenital ana-
omalies, chromosomal abnormalities, single gene mutations, and teratogenic
causes are identified in less than 10\% of cases.\textsuperscript{27}

Children with spina bifida almost invariably have an associated
Chiari II hind brain malformation, consisting of a small posterior fossa
and downward displacement of the cerebellar vermis below the foramen
magnum into the cervical spinal canal with elongation of the brainstem
and obliteration of the fourth ventricle.\textsuperscript{34}

Hydrocephalus frequently (85\%) is present in patients with myelo-
meningocele. Hind brain herniation with obstruction of the outflow of
the cerebrospinal fluid from the fourth ventricle is believed to be the
cause of the hydrocephalus.\textsuperscript{17} Overproduction of spinal fluid leads to
distension of the neural tube, fluid infiltration of the surrounding meso-
derm, and destruction of the neural crest cells.\textsuperscript{20} The neural crest cells
play an important role in forming mesodermal organs, such as the heart,
urinary tract, and skeleton; and their destruction may prevent normal
development of the spinal cord and the heart.\textsuperscript{14} Congenital heart defects,
including atrial septal defect, ventricular septal defect, anomalous pul-
monary venous return, tetralogy of Fallot, bicuspid aortic valve, coarcta-
tion, and hypoplastic left heart syndrome, occur in 37\% of patients with
open myelomeningocele; atrial septal defects are the most common.
Girls with myelomeningoceles are affected more frequently,\textsuperscript{51} which may
reflect the higher incidence of atrial septal defects in the general female
population.\textsuperscript{52}

Other associated defects include spinal arachnoid cysts,\textsuperscript{29, 50} defects
of the abdominal wall,\textsuperscript{49} split cord malformations (which have a reported
incidence of 6\% in patients with myelomeningocele\textsuperscript{28}), and possible
association with malignant hyperthermia.\textsuperscript{2}

Latex allergy and latex sensitization are well documented in patients
with myelomeningocele.\textsuperscript{10, 18, 30, 56, 64, 65} Between 10\% and 73\% of patients
with myelomeningocele have a history of clinical allergy to latex of
varying intensity.\textsuperscript{31, 59} The high incidence in these patients is believed to
be related to the degree of exposure to latex, and possibly is caused by
factors unique to patients with myelomeningoceles.\textsuperscript{4, 19, 21, 41, 64} The dis-
ease-associated propensity to produce latex-specific IgE antibodies with
consequent sensitization, is related primarily to spina bifida disease itself
and secondarily to the number of exposures to latex-containing products.
Sensitization to latex antigens may occur after the first contact, arguing for measures to avoid latex from birth.\textsuperscript{58} Although it is not known whether a delay in first contact with the latex allergens could postpone or reduce latex sensitization in children with myelomeningocele, the suggestion that genetic factors are responsible speaks for the initiation of measures to prevent exposure to latex at birth.

**Clinical Presentation**

Lesions of the lumbosacral area are associated with a better functional prognosis than those involving the thoracic area. Other predictors of function, in addition to location, include the size of the lesion, the amount of exposed neural tissue, the presence of kyphoscoliosis, lower extremity activity in utero, the timing of neurosurgical treatment, and the occurrence of postnatal complications.\textsuperscript{54}

Spina bifida and anencephaly are the two most common forms of neural tube defects.\textsuperscript{13} Anencephaly is uniformly lethal. In contrast, 80\% to 90\% of infants with spina bifida survive to adulthood.\textsuperscript{9} The survivors are likely to face life-long and devastating physical disabilities\textsuperscript{15} stemming from the neurologic defect itself or from its surgical repair. These disabilities include tethered spinal cord, paralysis, syringomyelia, syringobulbia, Arnold-Chiari II malformation, herniation of the cerebellum and hind brain, and associated hydrocephalus.\textsuperscript{5} There may be sequelae of neurologic deformities, leading to deformities of the spine and limbs; bladder, bowel, and sexual dysfunction, and learning disabilities.\textsuperscript{4,12}

**Diagnosis**

The early diagnosis of myelomeningocele is facilitated, in most cases, by the measurement of maternal serum $\alpha$-fetoprotein levels and the use of amniocentesis and high-resolution ultrasonography in the mid-second trimester of pregnancy.\textsuperscript{6}

Patients are evaluated preoperatively to ascertain the abnormalities resulting from the congenital defect. Children with myelomeningocele require spinal ultrasonography or MR imaging to rule out associated lesions, such as diastematomyelia, tethered cord, and hydromyelia. Imaging of the head to detect the presence of hydrocephalus and Chiari malformations is performed at birth.\textsuperscript{47}

Subsequent CT scanning or MR imaging is indicated to evaluate the size of the ventricles, especially preoperatively. The patients also are assessed for the presence of other associated anomalies.

A cardiology consultation followed by an ECG in patients with suspected cardiac lesions is recommended so that a definitive diagnosis can be reached for long-term management.

Abnormality of the urinary tract, mainly in the form of hydroureteronephrosis and vesicoureteral reflux, is seen in 10\% to 30\% of children
with myelomeningocele. If untreated, 40% to 90% of patients subsequently suffer significant upper urinary tract deterioration. Urologic evaluation is performed in all children with myelomeningocele to assess the risk for upper urinary tract disease, and includes renal ultrasonography and video urodynamics.47

Management

Early Maternal Management

The reduction in the incidence of neural tube defects has been ascribed to increased consumption of folic acid by expectant mothers and to the early detection of myelomeningocele and the termination of pregnancies.3

Prenatal: Intrauterine Surgery

Until recently, the neurologic deficits associated with myelomeningocele were attributed to the primary defect in closure of the neural tube. Recent work, however, supports a two-hit hypothesis, which, in addition to the primary embryologic error, postulates a secondary injury to exposed neural tissue throughout gestation.24 The secondary injury is believed to be caused by chronic mechanical injury and amniotic fluid-induced chemical trauma that damage the exposed unprotected fetal neural tissue progressively during gestation. Timely in utero repair of spina bifida in fetal sheep stops the ongoing spinal cord destruction and rescues neurologic function by the time of birth.35

Fetal surgery in humans has been successful at 23 weeks' gestation for a spina bifida lesion extending from the T-11 to S-1 level, with associated Arnold-Chiari type II malformation and borderline hydrocephalus. The baby was delivered before term by cesarean section. A postnatal MR image confirmed the integrity of the repair and the absence of the Arnold-Chiari type II malformation and hydrocephalus. Fetal surgical repair of severe spina bifida early in gestation can preserve neurologic function by arresting neural destruction. Prenatal repair also may allow preservation of function because of the plasticity of the developing nervous system. The resolution of the Arnold-Chiari malformation after the in utero repair suggests that this brain malformation is a secondary consequence of spina bifida. Such in utero repair may obviate the complications of Arnold-Chiari malformation, including the need for spinal fluid shunt procedures after birth.1

The results of these in utero repair procedures,1, 5, 35, 57 although encouraging, still are controversial because of the increased obstetric complications, such as oligohydramnios, premature rupture of membranes, preterm labor,6 and earlier gestational age at delivery.57 The increases in obstetric complications emphasize the importance of striking a balance between the potential benefits to the fetus and the risks for
the mother. The potential for decreasing the burden of disability on the patient, family, and society from congenital myelomeningocele may justify the risks for antepartum intervention.

POSTNATAL MANAGEMENT OF SURGERY

Preoperative Evaluation

**Neurologic Evaluation.** Causes for concern in the perioperative period relate not only to the size and anatomic position of the neurologic lesion (e.g., encephalocele, a large thoracic myelomeningocele) but also to its possible effect on breathing and control of the airway. Other frequently associated anomalies include Arnold-Chiari malformation and progressive hydrocephalus. Clinical features of brainstem dysfunction may include stridor, dysphagia, cranial nerve dysfunction, and central hypoventilation.

**Cardiac Consultation.** A preoperative ECG is recommended, especially in female patients, because neither the myelomeningocele location nor the presence of associated noncardiac defects can be used to predict patients at risk for heart defects. Clinical examination may be unreliable. This information has important implications for patients who may require a ventriculoatrial shunt, those at risk for endocarditis from urinary tract instrumentation, and those at risk for venous air embolism with certain neurosurgical procedures.

**Respiratory Evaluation.** Abnormalities of control of ventilation manifest themselves in the form of hypoventilation, sleep apnea, and prolonged breath holding as a result of structural derangement in the pontomedullary respiratory control center or in its afferent and efferent pathways. Respiratory complications also have resulted from bilateral vocal cord paralysis. Aspiration is always a concern because of possible lack of pharyngeal coordination, poor sucking reflex, and absent gag reflex.

A short trachea from reduced numbers of tracheal cartilaginous rings occurs in 36% of patients with myelomeningocele, who may have 15 or fewer cartilage rings than do normal patients (17.01 ± 1.28). The result is a high tracheal bifurcation and the possibility of accidental bronchial intubation. Preintubation chest radiographs should be obtained at a kilovoltage sufficient to produce evaluable air bronchograms reliably, in the hope of reducing the risk of accidental bronchial intubation and its possible complications, including bronchial stenosis. There is also a 14% to 20% incidence of radiographic absence of the 12th rib in patients with myelomeningocele. These patients have retarded growth and ossification of the ribs. They characteristically demonstrate radiographic absence in early life and hypoplasia of the 12th rib later in life.
Preparation for Surgery

The following should be performed in preparation for surgery:

1. Evaluation of signs and symptoms attributable to the neurologic deficit and any possible signs of increased intracranial pressure caused by Arnold-Chiari II malformation, including the need for drainage of cerebrospinal fluid by ventriculostomy before the definitive procedure
2. A CT scan to demonstrate the size of the ventricles
3. An ECG with cardiac consultation to evaluate possible associated cardiac anomalies
4. Air bronchogram to demonstrate the length of the trachea
5. Evaluation of possible airway problems during induction of anesthesia because of anatomic site or size of lesion and the potential for respiratory involvement from abnormal control of ventilation, impaired chemical drive, or possible bilateral vocal cord paralysis
6. Consideration of tracheotomy for airway management in the postoperative period, especially for patients who do not respond to cerebrospinal fluid shunting procedures
7. Preoperative chest radiograph to exclude the possibility of aspiration pneumonia caused by gastroesophageal reflux, absent gag and cough reflexes, or lack of oropharyngeal coordination with consequent continued spillage of oropharyngeal secretions into the trachea
8. The defect is well padded and protected in the preoperative period to avoid complications from compression, cerebrospinal fluid leak, or bleeding into the defect during movement of the patient
9. Patients who have thoracolumbar myelodysplasia with abdominal muscle weakness, inefficient cough, and decreased ability to clear secretions also may require postoperative ventilation after lengthy surgery
10. Reliable intravascular access is secured for replacement of blood and evaporative losses during repair of large myelomeningoceles; premature patients with large defects have difficulty in maintaining body temperature and fluid balance
11. Preoperative hematocrit is measured, and blood is made available for possible transfusion during operative repair of large defects
12. Sensitization to latex antigens may occur after the first contact, arguing for measures to avoid latex from birth

Management of Anesthesia

Surgery for repair of the myelomeningocele usually is performed in the early neonatal period to prevent infection, cerebrospinal fluid leak-
age, and the development of hydrocephalus with consequent cerebral dysfunction.

**Monitoring.** All surgical procedures in the early neonatal period are performed in warmed operating rooms. The patient is placed on a sheepskin blanket under warming lights before the operation. The patient's limbs and uninvolved body area are covered to maintain body temperature. All fluids are warmed by passage through a fluid warmer from the beginning of the procedure.

The choice of monitoring depends on the age and condition of the patient, the presence of any associated anomalies, and the planned surgical procedure. Latex precautions are observed in the selection of all equipment and drugs in the perioperative period. Routine monitors include ECG, pulse oximeter, indirect blood pressure, esophageal stethoscope, temperature probe, and a peripheral nerve stimulator to monitor neuromuscular blockade during the induction and intubation. A direct arterial line for monitoring of blood pressure, at least two good peripheral intravenous lines, and a urinary catheter are recommended for extensive and invasive surgical procedures. Surgeons may require any residual neuromuscular blockade to be reversed when intraoperative nerve stimulators are used during the repair of the myelomeningocele.

**Premedication.** Sedation is not required, and is contraindicated because hypoventilation may increase intracranial pressure and may cause brainstem herniation in the presence of the Chiari malformation with grave consequences, including cardiac and respiratory arrest.

**Anesthesia.** Latex precautions are strictly observed in the perioperative period. Preoxygenation with a nonlatex mask of appropriate size is followed by the intravenous induction of anesthesia with thiopental, 2 to 3 mg/kg, a short-acting muscle relaxant (e.g., rocuronium, 0.6 mg/kg, or rapacuronium, 2 mg/kg), atropine, and endotracheal intubation. Intubation is performed in the sick neonate while he or she is awake. Positioning tiny patients with associated congenital anomalies and a large encephalocele or myelomeningocele is challenging. The patient is placed in the supine position with the defect well protected by a donut tailored to the size of the anomaly. The body is raised, and the pressure points are padded. A large anomaly may necessitate the use of the lateral position to avoid any compression during induction of anesthesia and securing of the airway. Anesthesia is maintained with a low concentration of inhalational anesthetic. Narcotics are administered in small doses for only lengthy and invasive procedures. The eyes are taped, all monitors and intravascular lines are well secured, and the patient is turned to the prone position. The patient is prepared for surgery while still under the warming lights, and is draped quickly. Blood loss, fluid balance, and temperature are monitored closely; and all fluids and blood products are administered through a fluid warmer. The neonate's trachea may be extubated after short and uncomplicated procedures when there is confirmation of neurologic integrity. The patient should be fully awake before extubation, and should demonstrate cough and gag reflexes.
POSTOPERATIVE CARE

Infants who are at risk for postoperative hypoventilation or apnea are monitored with pulse oximeter and apnea monitors in a facility in which expert help is immediately available to secure the airway in an emergency. When the patient's trachea remains intubated, a postoperative chest radiograph confirming the correct position of the endotracheal tube is performed to avoid accidental endobronchial intubation and consequent problems.

SUMMARY

Neural tube defects of the brain and spinal cord, among the most common birth defects in the United States, cause neurologic morbidity from the lesions themselves and from associated hydrocephalus and Arnold-Chiari malformation. Because the myelomeningoceles, and encephaloceles are repaired surgically within hours of birth, neonatal anesthetic management with attention to fluids, body temperature, intravenous access, and monitoring is important. Anesthesia for treatment of hydrocephalus and Arnold-Chiari malformation takes into consideration the presence of intracranial hypertension and the need for manipulation of vital structures in the posterior fossa.

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