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Anesthetic considerations for congenital hydrocephalus

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Key points

Congenital hydrocephalus can present multiple challenges, including undiagnosed genetic abnormalities, prematurity-associated comorbities, and distortion of normal airway anatomy by macrocephaly. Neonatal neurophysiological differences can also affect intracranial pressure management. With proper preparation, one can optimize these issues and minimize the risks of anesthetic complications.

Abstract

Predictable differences in the pediatric versus adult airway generally make management more difficult, but neonatal intubation can also be complicated by the presence of congenital abnormalities. In this case, we review the anesthetic challenges presented by a 1 day old, full-term gestation female with a head circumference of 52.5 cm (> 99.9th percentile) undergoing insertion of ventriculo-peritoneal shunt for ventriculomegaly and massive congenital hydrocephalus. Perioperative management included consideration of the presence of unknown congenital or genetic anomalies, adequate positioning and planning for the risk of difficult airway, and preparation for possible high intracranial pressure.

Keywords: Airway management, hydrocephalus, intubation, laryngoscopy, macrocephaly

Introduction

Hydrocephalus in a neonate may be congenital or acquired. Congenital hydrocephalus occurs in 0.5 to 0.8 per 1000 live births (1, 2). Causes include neurologic pathologies like neural tube defects, genetic disorders, or idiopathic (1, 2). Many infants with congenital hydrocephalus are premature, and therefore problems associated with prematurity are also important concerns. Acquired hydrocephalus at birth may be a consequence of infection, intraventricular hemorrhage, trauma, and tumors (1). Neonatal hydrocephalus, whether it is congenital or acquired, can result in poor neurologic outcomes due to multiple shunt failures, as well as irreversible cellular damage and ischemia associated with ventriculomegaly (1). A mortality rate of 25% within the first week of life and 38% at 1 year of age has been reported (2). Here, we present a case of a 1 day old female with severe hydrocephalus and discuss the anesthetic implications. The patient's mother reviewed the case report and photographs and has given consent for publication.

Case report

A 1 day old, 40 week 1 day gestation female born with a head circumference of 52.5 cm (> 99th percentile) and weight of 4810 grams (> 97th percentile) (Figure 1). Maternal history was significant for smoking, herpes simplex virus, and history of depression. Severe hydrocephalus was noted on prenatal ultrasound, and the patient was delivered via an elective, uncomplicated Ceasarean section under spinal anesthesia (APGARs were 8 and 9). After delivery, physical exam demonstra-

ted macrocephaly with tense fontanelles, as well as significant frontal and parietal bossing. CT of the brain showed massive enlargement of the lateral ventricles with a rim of brain tissue lining the anterior calvarium, an enlarged third ventricle, and an enlarged CSF space in the dorsal midline posterior fossa (Figure 2). The patient was scheduled to undergo insertion of ventriculoperitoneal (VP) shunt for ventriculomegaly and hydrocephalus. The exact cause of the hydrocephalus was unknown at the time of surgery. In preparation for induction, careful attention was paid to positioning the patient for intubation. A ramp of sheets was used to elevate the patient's body and the head was placed on a gel ring in order to obtain the sniffing position (Figure 3). An inhalation induction with sevoflurane in oxygen and nitrous oxide was performed and a 3.0 cuffed ETT was placed using a Miller 0 (Grade 1 view). The patient was maintained on sevoflurane and rocuronium during the procedure and the VP shunt was placed uneventfully. At the end of the case, the patient remained intubated at the surgeon's request and was transferred to the intensive care nursery in stable condition (Figure 4).

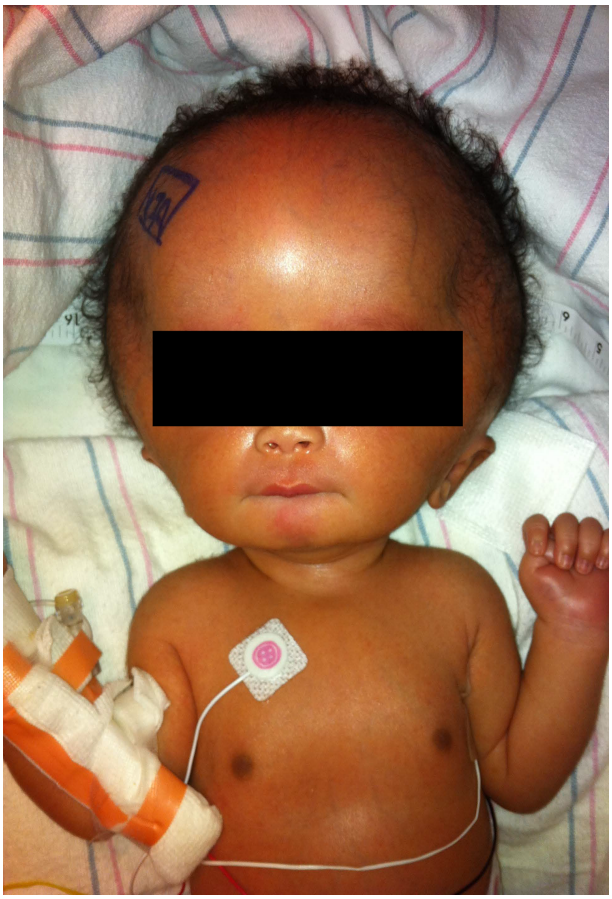

Figure 1. A 1 day old, 40 week 1 day gestation female born with a head circumference of 52.5 cm (>99th percentile) and weight of 4810 gm (>97th percentile), prior to intubation.

Discussion and conclusions

Congenital hydrocephalus may be obstructive, communicating, or both, and is commonly associated with myelomeningocele, Arnold-Chiari or Dandy-Walker malformation, arachnoid cysts, and vascular malformations (1). It is also found in various syndromes, such as X-linked hydrocephalus and trisomies 13, 18, 9, and 9p, and may be associated with other anomalies in approximately 50% of cases, which is a rate comparable to that of omphalocele (2, 3). When obtaining a preoperative history and developing an anesthetic plan for these patients, the possibility of associated comorbities and neurologic abnormalities should be considered, even if they are undiagnosed at the time of surgery.

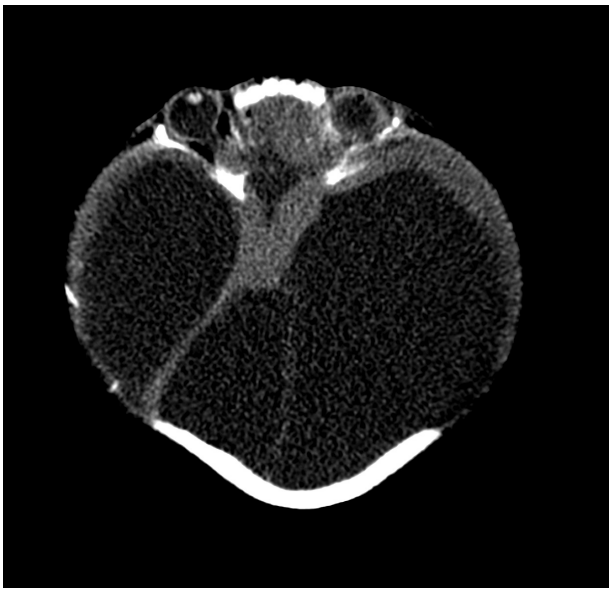

Figure 2. Preoperative CT of the head demonstrates impressively enlarged lateral ventricles, third ventricle, and CSF space in the posterior fossa.

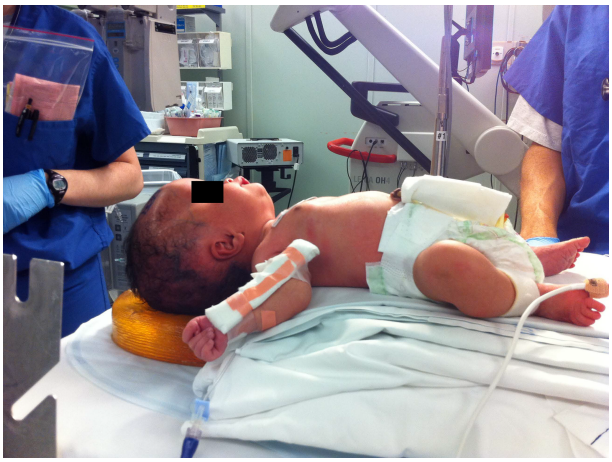

Figure 3. Positioning for intubation involved a ramp of sheets to elevate the patient's body and placing the head on a gel ring in order to obtain the sniffing position.

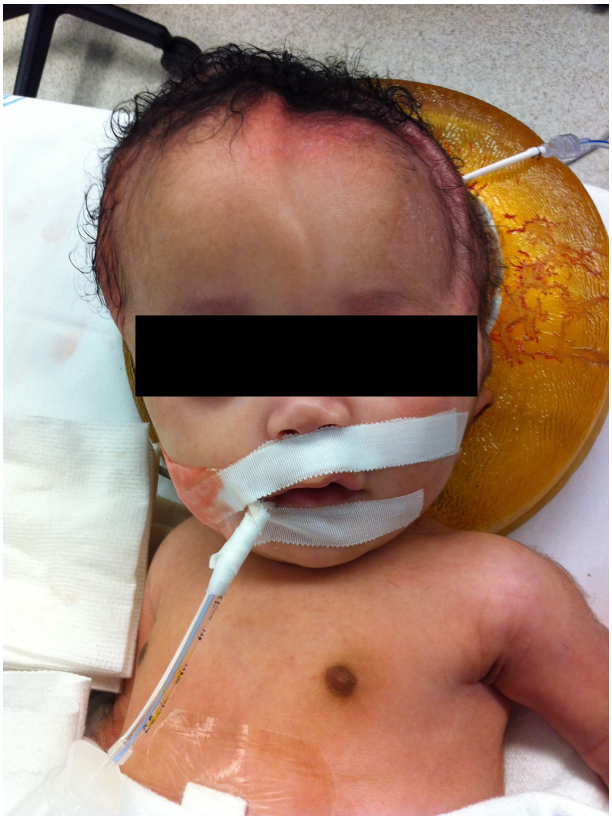

Figure 4. The patient remained intubated and was transferred to the intensive care unit postoperatively. There was a significant reduction in head circumference after VP shunt placement.

When obtaining a preoperative history and developing an anesthetic plan for these patients, the possibility of associated comorbities and neurologic abnormalities should be considered, even if they are undiagnosed at the time of surgery. A good physical exam prior to surgery should document any neurologic deficits, as well as any signs of increased intracranial pressure (frontal bossing, dilated scalp veins, cranial nerve palsies). Additionally, many neonates with congenital hydrocephalus are premature with low birth weight, and therefore anemia, coagulopathy, jaundice, respiratory disease, and persistent fetal circulation are also important concerns for anesthesia. Our patient's hydrocephalus was considered idiopathic and she continues to be treated for hydrocephalus, seizures, and developmental delay.

Intraoperatively, it is likely that positioning and airway management will be challenging. The pediatric airway is often considered more difficult than the adult airway due to five main anatomical differences: a larger tongue, a cephalic-located larynx, anteriorly slanted vocal cords, a short, omega-shaped epiglottis, and the narrowest portion located in the subglottic region at the level of the cricoid cartilage (4). In congenital hydrocephalus, macrocephaly may distort the normal anatomy of the skull, making airway management even more difficult. The large occiput places the neck in extreme flexion and the large forehead may obscure the line of sight in laryngoscopy, so elevating the body with pillows or towels is necessary in order to facilitate laryngoscopy (5). Our patient's head circumference was 52.5 cm, which corresponds to the 50th percentile for a 10 year old, yet her airway anatomy resembled that of a newborn. This provided a challenging combination for intubation. She had an enormous occiput, frontal bossing that did not obscure sight lines, disarticulated cranial bones with tense fontanelles, and a distorted jaw. The distorted jaw may have allowed additional space in which to displace the tongue. Our positioning of the patient allowed us to optimize the oral, pharyngeal, and laryngeal axes, yielding a grade 1 view.

Additionally, intracranial hypertension or decrease in intracranial compliance almost always accompanies hydrocephalus in infants and children (6). There are a few differences in neonatal neurophysiology that affect ICP management. First, the normal range of ICP in neo-

nates is 2-6 mmHg, which is lower than adults (6, 7). Additionally, cerebral perfusion pressure (CPP), cerebral blood flow (CBF), and MAP values are lower in infants than in children or adults (CPP: 40-60 mmHg for infants, 50-70 mmHg for adults; CBF: 40 ml/100g/min for infants, 100 ml/100g/min for children, 60 ml/100g/min for adults) (8). This results in a smaller difference in CPP between the lower limit of autoregulation and normal MAP values, leading to decreased autoregulatory reserve in infants (8, 9). Third, while the neonate has a flexible skull that can enlarge in response to expanding ventricles if hydrocephalus develops slowly, there is a gradual decrease in compliance as the tissues distend, and acute increases in cranial volume are not compensated, which can lead to herniation (6, 8). Even in chronic hydrocephalus, the compressed and stretched brain tissue adjacent to the ventricles caused by intracellular dehydration will contribute to a higher ICP (1, 8). In infants, clinical findings of increased ICP are nonspecific, such as vomiting, lethargy, irritability, bulging fontanelle, or poor feeding, so it may be difficult to determine preoperatively if a neonate has increased ICP. These patients are also at risk for vomiting and pulmonary aspiration (8). Preparations for elevated ICP should be undertaken, including avoidance of hypercarbia and hypoxia, treatment of hypotension, hyperventilation, elevation of the head of bed, avoidance of PEEP to prevent venous congestion, and administration of mannitol and osmotic agents. Hydrocephalus and distortion of the brainstem can result in bradycardia, hypertension, and changes in respiratory rate, and the anesthesiologist should be prepared to immediately respond to these changes.

Postoperatively, the neonate is at risk for chronic neurologic deficits. Between 28 and 78% of patients with congenital hydrocephalus suffer from developmental or neurologic delays (1). This is usually attributed to the high failure rate of CSF shunts, but it is also known that hydrocephalus causes irreversible cellular damage by acutely compressing and stretching the periventricular tissue, leading to ischemia and hypoxia (1). Chronically, it can result in gliosis, neuroinflammation, demyelination, axonal degeneration, metabolic impairment, altered blood-brain barrier transport, altered neuronal connectivity, and cell death (1). The degree of impairment is inversely correlated with the age of onset, and also depends on the magnitude and duration of ventriculomegaly (1). The neonate with congenital hydrocephalus can present multiple challenges for the anesthesiologist, but with proper preparation, one can optimize these issues and minimize the risks of anesthetic complications.

Disclosure

Institutional Review Board approval was obtained for this case report and the patient's mother consented to publication. There was no funding for this study. The authors report no conflicts of interest.

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