The incidence of congenital hydrocephalus varies in different populations. This is especially true for hydrocephalus associated with meningomyelocele, the incidence of which varies from about 40 out of 1000 births in some parts of Wales and Northern Ireland, to about 0.2 out of 1000 births in the Japanese population. The incidence of all other forms of hydrocephalus is nearly 1 for every 1000 births.

By definition, the term hydrocephalus is applied to any condition in which enlargement of the ventricular system occurs as a result of an imbalance between production and absorption of cerebrospinal fluid (CSF). There are two basic types of hydrocephalus: (1) obstructive hydrocephalus in which there is interference with the circulation of the CSF within the ventricular system itself; and (2) communicating hydrocephalus in which the CSF pathways inside the ventricular system are open and ventricular fluid is able to move freely into the spinal subarachnoid space, whereupon there is an interference with absorption of CSF.

It is interesting to note that there are a number of other conditions during infancy that can cause an abnormal enlargement of the cranial vault. Some of the conditions include:

1. Megalencephaly.
2. Chronic subdural effusion.
3. Cerebellar, pineal region and third ventricular tumors.
4. Secondary to midline brain tumor, or cerebrocerebral atrophy in degenerative and metabolic brain diseases.

Preoperatively a variety of diagnostic procedures may be done on the infant, many of which require general anesthesia. It should be noted that these diagnostic procedures carry an appreciable morbidity of their own. These procedures include ventriculography, angiography, pneumoencephalography, myelography, and computerized axial tomography.

Though the treatment for hydrocephalus has improved in recent years, there are still many problems to be resolved. The result is treatment that is only partially successful. The major goal in the treatment of the hydrocephalic infant is to reestablish the equilibrium between CSF production and resorption. Some of the methods of therapy include the following:

1. Administration of acetazolamide in a dose of 50-75 mg/kg/day. (This diminishes CSF production by one third and is useful in mild cases.)
2. Utilization of choroid-plexectomy, a surgical method for decreasing CSF production. (This technique carries with it a high mortality rate.)
3. Insertion of a Torkildsen shunt to bypass...
the obstruction. (This technique is not very successful in infants because the areas used in the bypass are too small.)

4. Creation of a ventriculoatrial shunt which returns the fluid directly into the blood stream. (This technique is often complicated by infection.)

5. Utilization of ventriculoperitoneal shunts. (These offer less serious complications but are easily occluded.)

6. Creation of a lumboureteral shunt. (This technique is rarely used because it requires the removal of a kidney.)

Pre-anesthetic considerations

Prior to intubation, it is important for the anesthetist to set up the following guidelines:

1. The infant is generally placed in the supine position with his/her head turned toward the side.

2. The infant must be kept completely immobile.

3. The anesthetist should position himself and his equipment in such a way that the surgeon has freedom to work about the infant’s head.

4. The anesthetist should be prepared to provide anesthesia for an indefinite amount of time, depending upon how the operation proceeds.

5. A request should be made for a reduction of the patient’s cerebral blood flow and maintenance of a “relaxed” brain, as described by Smith in *Anesthesia for Infants and Children*.

6. Provisions should be made for the availability of rapid, extensive blood replacement.

7. Allowances in the anesthetic regimen should be made for the use of vasoconstricting drugs to reduce bleeding from the scalp.

Some children, especially when the intracranial pressure is raised, have an inspiratory stridor, which is due to an indrawing of the aryepiglottic folds during inspiration. On occasion, the inspiratory obstruction is so severe that tracheal intubation is necessary. It seems that in hydrocephalic children, control of the airway is unusually precarious and may be lost when consciousness is only slightly impaired.

Causes of intracranial pressure

Extensive studies of neurophysiology and neuropharmacology have been conducted in recent years and have been primarily concerned with adults. But, some basic concepts have been established that may be applied to pediatric age groups. The effect of anesthetic agents on the normal and abnormal brain has been investigated with highly practical results. Information gained concerning regulation of intracranial pressure (ICP) has been of particular importance to anesthesiology.

Precise measurement of cerebral blood flow (CBF) and the cerebral metabolic rate for oxygen (CMRO₂) has not been made throughout all stages of development. It is believed that both CBF and CMRO₂ are relatively low at birth and then climb to maximal levels in infancy or early childhood. Using the nitrous oxide uptake technique, Kennedy and associates reported a mean CBF of 106.4 ml/g/min and a mean CMRO₂ of 5.17 ml/g/min in children 3-10 years of age. The CMRO₂ is reduced in hydrocephalus and other lesions. Also it should be noted that CBF falls if the cerebrospinal fluid (CSF) exceeds 380-450 mm H₂O.

Oxygenation and cardiac function are of primary concern during all operative procedures. In neurosurgery, ICP and brain volume are additional factors of primary importance, varying dangerously with the type of lesion and operative trauma, as well as anesthetic management.

ICP in neonates is in the range of 80 mm CSF, subsequently rising parallel to systemic blood pressure throughout childhood. One may see an elevation of the CSF to as high as 600 mm as a result of physical, physiologic, pathologic or pharmacologic stimuli. Some examples of these causes are listed in Table I.

### Table I

<table>
<thead>
<tr>
<th>Stimuli which cause an elevation of CSF</th>
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</thead>
<tbody>
<tr>
<td><strong>Physical</strong></td>
</tr>
<tr>
<td>Struggling during induction</td>
</tr>
<tr>
<td>Choking</td>
</tr>
<tr>
<td>Vomiting</td>
</tr>
<tr>
<td>Bucking on the endotracheal tube</td>
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<tr>
<td><strong>Physiologic</strong></td>
</tr>
<tr>
<td>Hypercarbia</td>
</tr>
<tr>
<td>Hypoxia</td>
</tr>
<tr>
<td><strong>Pathologic</strong></td>
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<tr>
<td>Intracranial tumor</td>
</tr>
<tr>
<td>Hemorrhage</td>
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<tr>
<td>Cerebral edema</td>
</tr>
<tr>
<td>Vascular anomalies</td>
</tr>
<tr>
<td>The two types of hydrocephalus</td>
</tr>
<tr>
<td><strong>Pharmacologic</strong></td>
</tr>
<tr>
<td>Ketamine (unless its response is blunted by a preadministration of a counteracting drug such as thiopental)</td>
</tr>
<tr>
<td>Most inhalation agents (can be counterbalanced by hyperventilation prior to the use of the agent and preliminary induction with thiopental or Innovar®)</td>
</tr>
</tbody>
</table>

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Throughout the operation it is highly desirable to maintain a moderate degree of hyperventilation to prevent the dangerous progression of cerebral acidosis, edema and ischemia. The desired range of PaCO$_2$ is considered to be 25-35 mmHg.\textsuperscript{6} The anesthetist must be constantly alert for the signs of increased ICP. Although the signs are not as easily recognized in the anesthetized patient, there are some significant changes that will give warning. These include:

1. Increased vascular engorgement on the surface of the brain.
2. Visible pulsation and swelling on the surface of the brain.
4. Occasional tachycardia.
5. Dysrhythmias.
6. Slow and/or irregular respirations.

**Regulation of intracranial pressure**

Once ICP has been recognized as a problem, the next step is to institute a management regimen. The following steps should be taken to reduce ICP:

1. Check to see that the infant is not hypercarbic.
2. Measure the arterial or venous pH and PCO$_2$ for definite evaluation of respiratory adequacy. PaCO$_2$ measurements between 25-35 mmHg, PVCO$_2$ measurements between 40-45 mmHg, and a pH reading of between 7.45-7.55 are adequate evidence of the absence of hypercarbia.
3. Elevate the patient's head 10°.
4. Increase anesthetic level if the patient is "light."
5. Administer steroids and diuretics.

It should be noted at this point that there are two steps that can be taken preoperatively if the surgeon so requests. They are:

1. The insertion of a lumbar puncture needle. (The amount of drainage is controlled by a clamp on the tubing.)
2. The maintenance of spontaneous respirations by:
   - Giving little or no preoperative sedation.
   - Inserting an endotracheal tube with the use of a topical anesthetic.
   - Requesting the surgeon to use a local anesthetic prior to incising the skin.
   - Maintaining the child on nitrous oxide and halothane mixtures.
   - Assisting respirations as necessary.
   - Monitoring the adequacy of ventilation with blood gases.

**Lesions of the central nervous system**

There are a variety of lesions of the central nervous system that involve the abnormal development of the spine, vertebrae or skull through a protrusion or exposure of the nervous elements; these are often associated with serious functional defects.\textsuperscript{6} The lesions which fall into this category are spina bifida, myelomeningocele, dysraphism, meningocele, and encephalocoele.

Once again, in these cases a key factor which must be considered pre-, intra-, and post-operatively is ICP. With these anomalies, the infants face a lifetime of pressure reducing maneuvers. The anesthetist must be prepared to treat this problem of pressure reduction whenever the patient is considered for surgery. Initially, if surgery is indicated, it is generally done within the first 24 hours after birth. The length of the procedure varies from between one to four hours, depending on the size of the defect.

Preoperatively, as in the hydrocephalic patient, the anesthetist should check for ICP and begin treatment immediately. Intraoperatively, general endotracheal anesthesia is indicated. Management of this patient will include intubation under intramuscular succinylcholine, then maintenance with nitrous oxide and halothane. In these cases, vigilant monitoring should include evaluation of blood loss, cardiorespiratory disturbances, ICP, blood pressure, and temperature. By careful monitoring, the anesthetist will be ready to manage any of the above complications. Postoperatively, these patients must be monitored as cautiously as intraoperatively.

**Prognosis**

According to Vaughn, the prognosis of infantile hydrocephalus has been significantly but not dramatically improved by the introduction of shunt operations. Untreated, 50-60% of infants with hydrocephalus succumb to the disorder itself or to intercurrent illnesses. If the process becomes arrested, about 40% of the infants are of normal intelligence. With good and continued neurosurgical and medical management, about 70% can be expected to live beyond infancy; of these, about 40% will retain intellectual capability, and about 60% will have significant intellectual and motor handicaps. The prognosis of infants with both hydrocephalus and spinal canal anomalies is considerably worse.\textsuperscript{6}

**REFERENCES**


ADDITIONAL READING

AUTHOR
Douglas Pokorny, CRNA, is a graduate of Rush-Presbyterian-St. Luke's Hospital School for Nurse Anesthetists. He is currently enrolled at Sangamon State University, completing a BS in anesthesia. He is a staff anesthetist at Edward Hospital, Naperville, Illinois.