Neuroendoscopy in the Youngest Age Group

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Key words
- Aqueduct
- Endoscopy
- Fourth ventricle
- Hydrocephalus
- Infants
- Intracranial pressure

Abbreviations and Acronyms
CPC: Choroid plexus coagulation
ETS: Endoscopic third ventriculostomy
IIHS: International Infant Hydrocephalus Study
MRI: Magnetic resonance imaging
VPS: Ventriculoperitoneal shunt

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■ OBJECTIVES: **The aim of this report is to review current data on the role of neuroendoscopy in infants. Specific emphasis will be given to the International Infant Hydrocephalus Study (IIHS). Previous studies, available information, and future directions are discussed.**

■ METHODS: **The IIHS is a major international endeavor comparing the results of endoscopic third ventriculostomy (ETV) to ventriculoperitoneal shunting in infants younger than 2 years of age. It is a prospective, randomized study, with a "parental preference" option, that recruits infants with aqueductal stenosis without a history of prematurity or other associated brain anomalies. The primary outcome measure is neurocognitive outcome at 5 years of age. In addition to IIHS data, we also looked at results of neuroendoscopy in infants with other indications, such as fourth ventricular outlet obstruction, Dandy Walker syndrome, etc.**

■ RESULTS: **The IIHS study includes more than 40 centers on all continents. To date, we have recruited more than 150 infants to the study. At this point we can only release limited data, namely that the complication rates are similar between the two arms. More patients are needed to finalize the study, with an endpoint of 250 children.**

■ CONCLUSIONS: **Neuroendoscopy in infants can be performed with reasonable morbidity. The preferred indications in infants are still not totally refined, especially vis-a-vis shunting procedures. More international, multicenter efforts are required to clarify these points.**

UNDERSTANDING TREATMENT OPTIONS FOR PEDIATRIC HYDROCEPHALUS

Because it is one of the most common neurosurgical pathologies, pediatric hydrocephalus is the leading cause of neurosurgery for children in the United States. The ventriculoperitoneal shunt (VPS) has been the classic treatment for pediatric hydrocephalus since the early 1960s, serving as the "bread and butter," or foundation, of pediatric neurosurgery. Shunts have modified the prognosis of hydrocephalus from a lethal disease to a curable disease, with a relatively good prognosis as determined by the etiology (8, 15, 94, 137).

Hydrocephalus is a heterogeneous disease. Shunts are able to resolve almost all cases of hydrocephalus, whatever the etiology, with almost no contraindications. Many different types of shunts have been developed and are in use, including pressure-regulated, volume-regulated, externally-regulated, shunt assists, and dual-switch valves (44, 47, 57-61, 101, 115, 116, 163, 165, 170, 171, 207, 244, 246, 257, 266, 270). When shunts first appeared in our field, the advantages were clear and far outweighed the disadvantages; they enabled a relatively normal life with a relatively simple procedure. It took some time to realize and acknowledge that the shunt failure rate is significant, that complications are common, and that children with shunts are dependent upon surgical maintenance throughout their lives (8, 109, 128-132, 205, 215, 229 265).

Shunt complication rates are unacceptably high. Children with shunts have an increased likelihood of seizures, they can develop slit ventricle syndrome, and some of them suffer from under- or overshunting (18, 20-22, 53, 54, 72, 78, 140, 147, 189, 191, 197, 203, 209, 216, 217, 218, 245). With all these complications in mind, the arrival of neuroendoscopy on the scene was greeted with great enthusiasm. Neuroendoscopy was seen as a means of solving the challenges of hydrocephalus without the issues of the hardware.

Endoscopic third ventriculostomy (ETV) was designed primarily for hydrocephalus cases in which there is a blockage at the level of the aqueduct of Sylvius. In these cases, the endoscope is guided to the floor of the third ventricle, and an opening is created between the third ventricle and the interpeduncular cistern. This is a straightforward diversion procedure; no hardware is usually left in place, and fluid can egress from the third ventricle to the base of skull and ultimately arrive at the normal absorption sites at the convexity of the brain.

ETV: TECHNICALLY CHALLENGING, VARIABLE TECHNIQUES

ETV has never been definitively identified with a single standardized technique. The same basic procedure is implemented with quite a bit of technical variability in different medical centers. Technical nuances include the use of rinsing fluid, use of navigation, scope types (rigid or flexible), techniques for creating and widening the hole in the base of the third ventricle, and even basic concepts of how to close the skin and open the bone (41, 151, 210, 234).

ETV (and all others neuroendoscopic operations) are advanced procedures that are heavily dependent on sophisticated technology. ETVs require a learning curve, substantial experience, and careful coaching of young neurosurgeons. Every case should be carefully discussed between the participating neurosurgeons and the indications and contraindications analyzed after a close inspection of the specific microanatomical details on magnetic resonance imaging (MRI). The professional discussions must be accompanied by a discussion with the family of the available alternatives, their advantages, and disadvantages.

Morbidity from ETV may be underreported. The nightmare of every neuroendoscopist is massive bleeding, mainly arterial, during the procedure. Perforation of the basilar artery has been reported from even the best of medical centers (4, 12, 19, 29, 46, 63, 110, 127, 169, 196, 206, 211, 222, 223, 250). Smaller bleeds, mainly of venous origin, usually stop by themselves with either simple rinsing or a short burst of mono- or bipolar coagulation. Tissue damage during insertion and manipulation of the endoscope, subdural hematomas, endocrinological abnormalities, infections, cranial neuropathies, and other complications are also reported (16, 19, 34, 50, 52, 66, 74, 99, 112, 168, 181, 223).

Although most failures from ETV occur in the early period after the procedure, late obstruction of the stoma may lead to increased intracranial pressure and even sudden death (39, 40, 55, 76, 98, 118, 159, 173, 178, 182). It is therefore strongly advised that patients who undergo a successful ETV should be clearly told that they are not cured from the hydrocephalus, that symptoms can reappear, and that they may have dangerous consequences (153). These patients should be followed on an ongoing basis, and the medical center should have an open-door policy that encourages the patients to call or come back if any related symptoms are appearing. It is still not known whether those patients with no flow-void at the third ventricle stoma on postoperative MRI may be at a greater risk to develop a clinical syndrome and should be followed even more closely.

EVIDENCE-BASED MEDICINE: SETTING THE STANDARD FOR ETV

Studies focusing on ETV in the pediatric age group began to appear in the 1980s and 1990s, and continue to appear in the literature to this day (19, 23, 31-33, 51, 56, 62, 68, 84, 86, 88, 96, 107, 122, 124, 149-152, 178, 180, 210, 230, 233, 242, 243, 263, 271). However, even with all the series that have been published to date, it is hard to extract meaningful research data or operative guidelines. There are too many inconsistencies in the basic "ground rules" used by these researchers (141). For example, success rates of ETV are usually defined as one or more of the following factors: the disappearance of hydrocephalus symptoms, no signs of intracranial hypertension evident, and/or a technically successful procedure. Perhaps partially as a result of this wide range of definitions for the term "success," large disparities are found when one examines the results of ETV in children. Success rate vary greatly, ranging from a low success rate of 35% in a series from Toronto (243) up to a high of 83% to 89% in other series (45, 107, 108, 124).

Analyzing the differences between successful series and series with less-promising results shows that most of the differences can be traced to a gap in the early failure rate. Early ETV failures could be attributable to use of the wrong technique, different selection criteria in recruiting the patients or in defining failure, and also the multifactorial etiology of the hydrocephalic process itself (37, 38). It is essential, therefore, to define a uniform set of selection and failure criteria to objectively and meaningfully compare results among different centers.

Since the 1990s, ETV has been recognized as a valid alternative to shunt implants, mainly for patients with obstruction at the level of the aqueduct, the tectal plate, and the pineal region. ETV quietly developed into a mainstream, common procedure in pediatric neurosurgery without any prospective randomized trials (and certainly no multicenter trials) proving its efficacy compared with shunt procedures. Unfortunately, it seems apparent today that a classic randomized trial is no longer possible because most of us treating these patients would not agree to expose a classic candidate for ETV to randomization between two alternatives.

As ETV technology continues to evolve and improve, and as we collectively accumulate more experience and confidence with ETV, indications for ETV have broadened, introducing more challenges in understanding the pathophysiology of hydrocephalus and in proving the efficacy of a new procedure (ETV) over the more standard alternative (shunts). This was one of the reasons that in 2001 we established the International Study Group for Neuroendoscopy. The goal of this organization (more recently transformed into the International Federation for Neuroendoscopy) is to promote neuroendoscopy research and education.

There are many pathologies for which treatment with ETV is debatable. These include hydrocephalus in infants, patients with meningomyelocele and Chiari malformation, Dandy-Walker malformation, fourth ventricular outlet obstruction, during tumor surgery, and patients who have had a hemorrhage or an infection in their past (7, 14, 25, 26, 30, 35, 49, 53, 73, 77, 79-81, 100-103, 111, 114, 119, 123, 162, 164, 166, 174-177, 183, 184, 186, 187, 193, 200, 202, 212, 214, 221, 231, 232, 240, 241, 248, 249, 255, 258, 259, 261, 268, 269).

Over the course of 10 years of collaboration within the International Federation for Neuroendoscopy, we have learned to appreciate the advantages of cooperative multicenter studies. Our first attempt was with a study on repeat ETV for those patients for whom the original ETV initially succeeded. We pooled our experiences with 20 patients recruited from four centers (233). Another collaboration involved a multicenter study on the efficacy of ETV in patients who had previously experienced an infection and/or hemorrhage. For this study we pooled our experiences with 101 patients from seven medical centers around the world (232). We are currently analyzing the results of the International Neuroendoscopy Biopsy Study,