

## Hydrocephalus in Uganda: the predominance of infectious origin and primary management with endoscopic third ventriculostomy

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**Object.** The aim of this prospective study was to investigate the causes of hydrocephalus in Uganda, the efficacy of endoscopic third ventriculostomy (ETV) in this environment, and whether existing parameters could be used to guide patient selection.

**Methods.** Three hundred consecutive children, 81.3% of whom were younger than 1 year of age, underwent ventriculostomy preceding ETV as an initial treatment for hydrocephalus. In 179 patients (60%) the hydrocephalus was caused by a cerebrospinal fluid infection; in 76% of patients the infection had occurred in the 1st month of life. In 229 patients (76.3%) ETV was performed; 2% of patients were lost to follow up after less than 1 month and the surgical mortality rate was 1.8%. The first ETV was successful in 115 patients (52%); the mean follow-up period was 15.2 months. The mean time to repeated operation following a failed ETV was 1.5 months. Sixty-five patients underwent a second endoscopy; 37 underwent a second ETV, of which 14 procedures (38%) were successful (mean follow-up period 12.25 months). The overall success rate for ETV was 59%. Among patients older than 1 year of age, the procedure was successful in 22 (81%) of 27 with postinfectious hydrocephalus (PIHC) and 18 (90%) of 20 with nonpostinfectious hydrocephalus (NPIHC). The success rate of ETV among those patients younger than 1 year of age was 59% (60 of 101) for patients suffering from PIHC and 40% (21 of 52) for those suffering from NPIHC. Age correlated with success for NPIHC ( $p = 0.0002$ ) and PIHC ( $p = 0.0421$ ). The success rate of the surgery for patients with myelomeningocele and hydrocephalus who were younger than 1 year of age was 40% (eight of 20). The success rate of the surgery for PIHC in infants younger than 1 year of age was 70% (44 of 63) among patients with aqueductal obstruction but 45% (14 of 31) among patients with aqueductal patency ( $p = 0.0254$ ). Fourth ventricular size as demonstrated on cranial ultrasonography or computerized tomography scanning predicted whether the aqueduct was patent ( $p = 0.0001$ ).

**Conclusions.** Infection is the most common cause of hydrocephalus in Uganda. In all children older than 1 year of age and in those younger than 1 year of age with PIHC and aqueductal obstruction, which was reliably predicted by cranial ultrasonography, ETV was effective.

**KEY WORDS** • endoscopic third ventriculostomy • hydrocephalus • neonatal meningitis • ventriculitis • myelomeningocele • developing country • pediatric neurosurgery

**T**HE incidence of hydrocephalus in East Africa is very high. The use of shunts in a developing country—even if the difficulties of cost and availability are surmounted—presents unique problems. The complications of shunt malfunction and infection are manageable when competent neurosurgical care is available on an urgent basis; in a situation like that in Uganda, however, ready access to such care is impossible for most patients because of financial and logistical barriers. Long-term shunt dependency is more dangerous under these circumstances than it is in the developed world.

*Abbreviations used in this paper:* AIDS = acquired immunodeficiency syndrome; BA = basilar artery; CSF = cerebrospinal fluid; CPX = choroid plexus cauterization; ETV = endoscopic third ventriculostomy; HIV = human immunodeficiency virus; NPIHC = nonpostinfectious hydrocephalus; PIHC = postinfectious hydrocephalus; VP = ventriculoperitoneal.

In a developing country, ETV presents an attractive option for potentially treating hydrocephalus in a permanent way without the use of a shunt and its attendant expense, risks of infection and malfunction, and the need for life-long maintenance. The usefulness of ETV has been clearly demonstrated in cases of aqueductal stenosis in older children and adults;<sup>13,15,17</sup> however, questions have lingered concerning its use in infants,<sup>6,13,15,29</sup> in cases of hydrocephalus secondary to infection,<sup>6,7,16,22</sup> and in those associated with a myelomeningocele.<sup>6,11,29</sup> The majority of our patients present for treatment when they are younger than 1 year old, and the most common cause of hydrocephalus appears to be infections such as ventriculitis and meningitis. From the outset, therefore, the usefulness of ETV in our setting was uncertain. Nonetheless, the difficulty and danger of maintaining shunts in the environment of a developing country provided compelling reasons to study the efficacy of ETV as the initial treatment for hydrocephalus of all origins in children of all ages.

The CURE Children's Hospital of Uganda, a referral mission hospital for pediatric neurosurgery, opened in January 2001. Within the first 26 months after opening 468 patients underwent surgery for the initial treatment of hydrocephalus, and 864 operations for hydrocephalus were performed. After ETV became available in our hospital, a decision was made to study this procedure prospectively as the primary treatment for all children presenting with hydrocephalus, regardless of patient age or origin of the condition. Children presenting with hydrocephalus underwent ventriculostomy as a prerequisite for undergoing ETV, the performance of which was the goal. This article focuses on the results of that approach in the first 300 children in the series, most of whom were infants.

### Clinical Material and Methods

#### *Data Collection and Clinical Evaluation*

Patient information was collected prospectively and entered into a database. Clinical information included history, neurological examination, head circumference, and characteristics of the fontanel (when present). Laboratory data included HIV status and analysis of CSF culture results. Neuroimaging information included cranial ultrasonography performed by the author in the majority of patients. The capabilities necessary to produce printed copies of the scan and onscreen measurements such as cortical mantle thickness and frontal horn index only became available late in the series. When ultrasonography was not available, a computerized tomography scan was obtained in Kampala (a 6-hour roundtrip) at no cost to the patient. Data recorded at the time of the endoscopy included the ETV site, whether flow was noted through the stoma, the reason for any abandoned ETV attempt, the appearance of the fourth ventricle on preoperative imaging and that of the aqueduct at the time of endoscopy, and a description of surgical findings.

#### *Patient Population and Selection for ETV*

Between June 2001 and March 2003, 300 children with hydrocephalus underwent ventriculostomy preceding the attempted ETV. The mean age of the patients was 15.1 months (range 0.07–255 months). Moreover, 188 patients (62.7%) were 6 months of age or younger and 244 (81.3%) were 12 months of age or younger. Only 56 patients (18.7%) were older than 1 year of age. There were 167 boys (55.7%) and 133 girls (44.3%) (male/female ratio 1.26).

In the first 5 months of the study, 28 of 108 children were eliminated as candidates preceding ventriculostomy, 12 because of disease origin (myelomeningocele or Dandy-Walker malformation), 10 because of severely distorted ventricular anatomy found on cranial ultrasonography, four because of severe enlargement of the fourth ventricle, and two because of very young age. These patients underwent shunt treatment without receiving an ETV. After our experience with the first 80 patients who did undergo ventriculostomy, however, the decision was made to perform ventriculostomy initially with the intention of performing an ETV in all children presenting with hydrocephalus regardless of age, cause of hydrocephalus, or distortion of ventricular anatomy on preoperative imaging.

#### *Surgical Procedure*

The equipment used for the procedure included a flexible endoscope, telecam, xenon nova light source, a Bugby electrocautery wire (all Karl Storz, Tuttlingen, Germany), and a Sony Trinitron monitor (Tokyo, Japan).

No disposable items were used (such as sterile sleeves to drape the camera or peel-away sheaths to introduce the scope) because of their unavailability and prohibitive cost. The scopes were sterilized in Cidex solution for a minimum of 16 hours after each use. The camera and cables were sterilized in Cidex solution for a minimum of 30 minutes between each use and then again overnight. The scope, camera, and cables were rinsed thoroughly in sterile water prior to use and placed in the operative field, resting on the drapes over the patient's chest.

Patients were prepared as if they were to undergo placement of a right frontal VP shunt. Through a small curved coronal incision, the dura mater was opened sharply in the lateral fontanel (or through a burr hole), the cortex was cauterized, the blunt-ended trocar of a No. 14 French peel-away catheter introducer (which we sterilize and reuse) was passed 1 cm through the pia mater and removed, and the scope was passed into the lateral ventricle. Typically, little CSF is lost during the procedure, with the exception of that taken for analysis. Continuous irrigation was not necessary.

The foramen of Monro was identified and the third ventricle was entered. The aqueduct was inspected to determine patency (Fig. 1). The third ventricular floor was inspected to identify the dorsum sellae, the mammillary bodies, and the BA complex. A determination was made regarding whether the floor could be safely fenestrated without injury to the other structures, and when possible, it was fenestrated with a Bugby wire just behind the dorsum sellae, using brief pulses of electrocautery on the surface followed by blunt penetration through the floor. The midline was used unless the position of the BA indicated that a position slightly lateral to it should be selected. The wire was used to dilate the opening gradually by gently stretching the tissues. The scope was passed through the opening in the floor and, when necessary, additional membranes were penetrated until the scope could be passed into free cisternal space, ideally with visibility of the BA, cranial nerves, and brainstem. The scope was withdrawn from the ETV stoma, and the presence or absence of CSF flow was noted.

For cases later in the series, if the third ventricular floor could not be safely fenestrated, the lamina terminalis was used as an alternative site for ventriculostomy. In a similar fashion, the Bugby wire was used to penetrate into the subarachnoid space anterior to the optic chiasm, and the presence or absence of CSF flow was noted. With experience, both sites began to be used when safe to do so. Often, if the floor appeared draped over the BA and inaccessible for ETV, it would be elevated after an ETV of the lamina terminalis was performed, which would cause the interpeduncular cistern to fill with CSF, thus making it possible to perform an ETV of the third ventricular floor.

#### *Postoperative Follow-Up Period*

It was my intention that patients undergo follow-up examinations at 1 week and 3, 6, and 12 months postopera-

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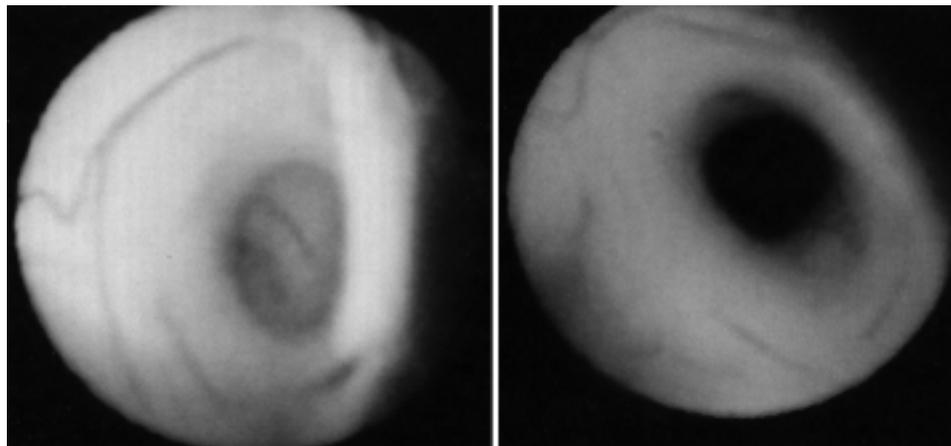


FIG. 1. Endoscopic view of a closed aqueduct (*left*) and an open aqueduct (*right*).

tively. This schedule was not always feasible, however, because of the difficulties of transportation for the majority of our patients. In addition, follow-up examinations would sometimes occur off schedule when illnesses such as malaria motivated a patient's return to the hospital. Patients lost to follow up were aggressively sought whenever possible with home visits, often deep in the villages, by our social work staff. In follow-up examinations, head circumference, fontanel, symptoms, neurological findings, and developmental progress were assessed. Cranial ultrasonography was also performed to evaluate the ventricles.

### Criteria for Success

Success was defined ultimately in this study as the avoidance of VP shunt placement. Criteria for success included a shift in head circumference growth to a normal or less-than-normal rate as plotted on a standard growth chart; decompression of the anterior fontanel; relief from symptoms of elevated intracranial pressure such as irritability, vomiting, and headache; decreased spasticity and/or improved

gait; and resolution of ocular symptoms such as sunsetting or sixth nerve palsy (Fig. 2).

### Statistical Analysis

Analysis of the data between groups was performed using the two-tailed Fisher exact test. A probability value less than 0.05 was considered statistically significant.

## Results

### Patient Population and Selection for ETV

In 229 (76%) of 300 patients ETV was successfully completed; these patients ranged in age from 0.07 to 255 months (mean age 15.9 months). Moreover, 60.6% of patients were 6 months of age or younger and 78.6% were 1 year old or younger (Table 1).

### Clinical Evaluation of the Severity of Hydrocephalus at Presentation

The majority of patients suffered from severe ventriculomegaly at the time of presentation, a condition which often results from delayed treatment. Of those children younger than 36 months old, 63% demonstrated a head circumference 2 cm or greater than the 95th percentile for their age, and in 29% the measurement was more than 6 cm above the 95th percentile. Cortical mantle thickness was measured preoperatively in 64 infants in the latter part of the series. The mean preoperative cortical mantle thick-



FIG. 2. Photograph showing a typical patient in this series with severe macrocephaly and sunsetting of the eyes.

TABLE 1  
Summary of patients by age in whom ETV was completed

Age (mos)	No. of Patients
0-3	78
>3-6	61
>6-12	42
>12-24	17
>24	31
total	229

ness was 1.64 cm (range 0.56–3.26 cm); 73% of patients demonstrated a thickness of less than 2 cm.

#### *Causes of Hydrocephalus*

The cause of hydrocephalus in each patient was defined as PIHC, NPIHC, or hydrocephalus associated with myelomeningocele. In the present study, patients in whom hydrocephalus was associated with a lesion obstructing the aqueduct, such as tumor (four patients) or cyst, giant aneurysm, or mesencephalic cavernous malformation (one patient each), were included in the NPIHC group, along with those in whom the cause of hydrocephalus was unknown or congenital, such as in cases of congenital aqueductal stenosis, Dandy–Walker malformation, or hydrocephalus associated with an encephalocele. Children suffering from hydrocephalus associated with a myelomeningocele were considered separately.

#### *Postinfectious Hydrocephalus*

The designation of PIHC was determined according to several factors: age at onset of hydrocephalus; history of febrile illness with or without seizures and its proximity in time to onset of clinically evident hydrocephalus; ultrasonography or computerized tomography findings of septations or loculations, anatomical distortion, or intraventricular deposits (Fig. 3); and findings at the time of ventriculocopy that were consistent with prior ventriculitis. A designation of PIHC was assigned on the basis of the following: 1) no history consistent with hydrocephalus at birth; and 2) either a history of febrile illness and/or seizures preceding the onset of clinically apparent hydrocephalus, or alternative findings such as imaging and endoscopic results indicative of prior ventriculitis.

Among the 300 patients in the present study, 179 (60%) suffered from PIHC as defined earlier. Of these 179 patients, 139 (78%) demonstrated at the time of endoscopy evidence of inflammatory changes within the ventricles, including intraventricular septations, scarring of the third ventricular floor with anteroposterior tenting of the floor, distortion of intraventricular anatomy, studding of the ependymal surfaces and third ventricular floor with inflammatory exudates or hemosiderin, and obstruction of the aqueduct or foramen of Monro by scar or inflammatory exudates (Fig. 4).

Without a doubt, PIHC has been the single most common cause of hydrocephalus in children presenting to us for treatment since our unit was opened. Between January 2001 and March 2003, 265 (57%) of 468 patients presenting with hydrocephalus, including the 179 in the present study, were considered to have PIHC according to the criteria outlined here. Of these 265 patients, 157 were boys and 108 were girls (male/female ratio 1.45), 161 (61%) presented with a history of febrile illness with convulsions, and 90 (34%) a history of febrile illness alone. In seven patients illness marked only by the onset of convulsions was evident and no fever was reported. In the cases of six patients, the mother was unaware of any fever or convulsions that preceded the onset of the hydrocephalus; however, all six demonstrated other evidence of previous ventriculitis. In three patients severe multiloculated hydrocephalus was revealed on preoperative imaging. Three other patients underwent ventriculocopy that revealed se-

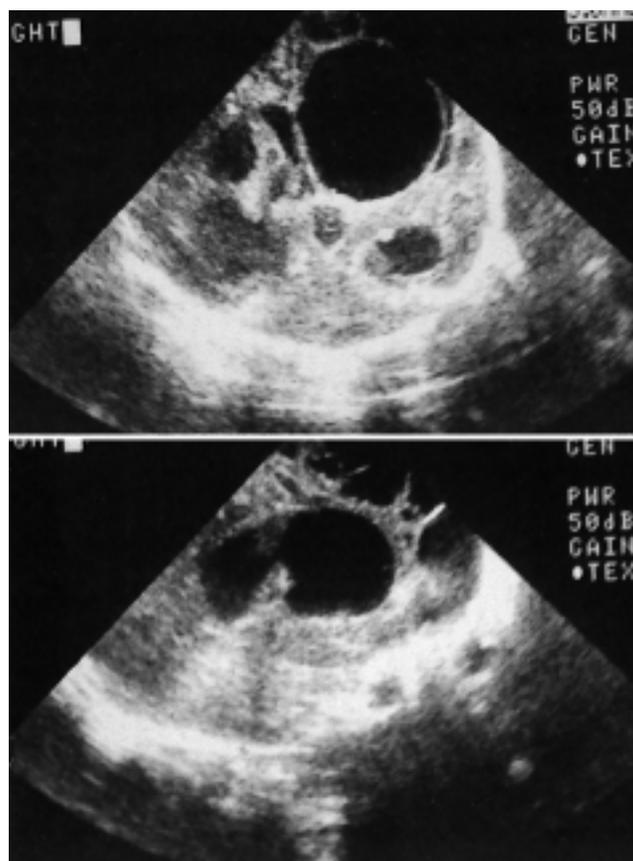


FIG. 3. Coronal (*upper*) and sagittal (*lower*) cranial ultrasonography studies obtained in an infant with PIHC, demonstrating inflammatory deposits in the right ventricle, periventricular parenchymal cystic changes, and septations. Note nonvisualization of the fourth ventricle.

vere postinflammatory changes in each. In 245 cases the caretaker gave a reliable account of the age at onset of the illness. The mean age at onset was 1.27 months (age range 0–48 months). In 186 patients (76%), however, the illness occurred within the 1st month of life (Table 2); PIHC appeared to be secondary to neonatal infection in 76% of patients. The mean time from onset of the illness to first observable symptoms indicating the onset of hydrocephalus was 0.72 months (range 0–4.75 months); onset in only four patients (1.6%) exceeded 3 months. The short time between the illness and the reported onset of hydrocephalus in a previously well child further supports infection as the cause of the condition.

#### *Analysis of CSF*

A CSF sample was routinely obtained in patients undergoing endoscopy for ETV. Those patients with PIHC were more likely to have abnormal CSF results; however, given the duration of time between initial illness and treatment of the resulting hydrocephalus, it is not surprising that many patients with PIHC exhibited relatively normal profiles. Patients with PIHC tended to have higher CSF cell counts and protein levels than did those with NPIHC. The difference between the two groups regarding the number of cases in which measures greater than 100 cells/ $\mu$ l were recorded

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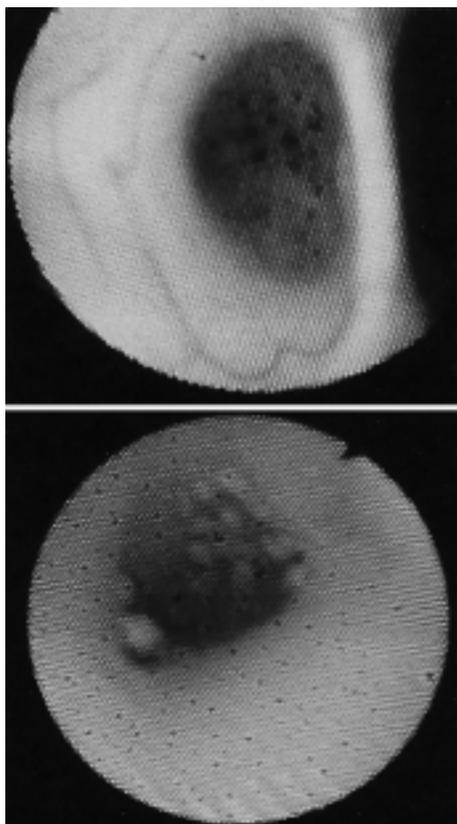


FIG. 4. Endoscopic view of an obstructed aqueduct with punctate hemosiderin deposits (posterior commissure on right) (*upper*) and an obstructed aqueduct studded with inflammatory deposits (*lower*).

was statistically significant (13 of 151 patients with PIHC and one of 74 patients with NPIHC,  $p = 0.0391$ ) as was the difference for the number of patients demonstrating protein content measurements greater than 100 mg/dl (40 of 151 patients with PIHC and seven of 72 patients with NPIHC,  $p = 0.0046$ ). Laboratory analysis of the CSF samples proved nondiagnostic in all but one patient, whose sample was found to contain Gram-negative rods. All of the samples were cultured, but no bacterial growth was detected.

### The Role of HIV Status and Causes of Hydrocephalus

Because of the relatively high prevalence of HIV and AIDS in Uganda, relationships between HIV and the three disease categories of hydrocephalus were explored. All surgical patients are routinely screened to determine HIV status by using the Determine HIV-1/2 test (Abbott Labora-

tories, Abbott Park, IL). In our study, positive results were routinely confirmed with enzyme-linked immunosorbent assay, and those patients with equivocal results were excluded from this analysis. A total of 454 patients who underwent surgery for hydrocephalus in our unit demonstrated unequivocal HIV or AIDS screening results; in 3.1% of these patients screening results were positive.

Given the likelihood of a higher incidence of positive results in younger patients due to the transmission of maternal antibodies, the results were assessed with regard to age. Among patients presenting between 0 and 3 months of age, the HIV screening results were positive in one (1%) of 99 patients with PIHC, in none (0%) of 42 with NPIHC, and in four (9.8%) of 41 treated for hydrocephalus secondary to myelomeningocele. The difference between patients with myelomeningocele and NPIHC approached statistical significance ( $p = 0.0551$ ), and the difference between those with myelomeningocele and PIHC was significant ( $p = 0.0258$ ). The difference in numbers of patients between those with seropositive myelomeningocele and all other patients in this age group (PIHC and NPIHC combined) was also significant ( $p = 0.0095$ ). There was no difference between NPIHC and PIHC in this age group ( $p = 1.000$ ). There was no significant difference among groups of patients older than 3 months. These results fail to indicate any association between HIV and PIHC; it might be inferred, however, that open neural tube defects may increase the likelihood of mother-to-child transmission of HIV.

### Abandonment of the Initial ETV Attempt

Of the 300 patients undergoing ventriculoscopy preceding the intended treatment of hydrocephalus by ETV, 219 (73%) underwent a successful ETV at the initial procedure. In 55 patients (18%) the attempted ETV was abandoned and a VP shunt was placed. In 21 patients (7%) a reservoir was placed when the initial ETV attempt was abandoned due to poor visibility resulting from turbid or xanthochromic CSF (19 patients) or inflammatory scar (two patients). Bilateral CPX was performed in three patients when ETV was not possible technically, and in two patients the ETV was abandoned but no alternative procedure was performed at the time.

The reasons for abandoning the initial ETV attempt in 81 patients are summarized in Table 3. Among the 30 patients with severe distortion of the ventricular anatomy resulting from inflammation, 11 also demonstrated multiloculated ventricles that were endoscopically communicated during the initial operation by fenestrating the septations prior to placing the ventricular catheter under endoscopic vision.

Among the 11 patients in whom ETV was abandoned due to congenital anatomical abnormalities, the following were encountered: four infants with myelomeningoceles, in two of whom a thick third ventricular floor was evident; one patient in whom the foramen of Monro was absent; one in whom obstruction to the access to the floor occurred due to a large interthalamic adhesion with no subarachnoid space behind the lamina terminalis; one with an encephalocele in whom neither the floor nor the lamina terminalis was identifiable; one with holoprosencephaly and fused thalami in whom no access to the third ventricle was evident; one with Dandy-Walker malformation in whom a cystic elevation of the floor filled the third ventricle; two

TABLE 2

Summary of patient ages at the onset of illness resulting in PIHC

Age at Onset of Illness (mos)	No. of Patients (%)
≤1	186 (76)
>1-3	36 (15)
>3-6	18 (7)
>6	5 (2)

TABLE 3  
Summary of conditions contributing to abandonment of first ETV attempt

Condition	No. of Patients
distorted anatomy (PIHC)	30
turbid CSF	26
congenital anomaly	11
thick ventricular floor	6
proximity to BA	5
scarred cistern	2
BA aneurysm	1

with hydranencephaly, one with fused thalami in whom no access to the third ventricle was available and the other in whom identifiable landmarks within the third ventricle were present; one with bilaterally absent foramina of Monro; and one child in whom the foramen of Monro was unilaterally absent, the floor of the third ventricle was too small, and the lamina terminalis and chiasm could not be identified.

Among the 21 patients who underwent reservoir placement for turbid or amber CSF at the initial endoscopy, the outcomes following the second endoscopy after the CSF had sufficiently cleared were as follows: eight patients underwent ETV and one ETV plus bilateral CPX; five underwent placement of a VP shunt when ETV was abandoned; one underwent VP shunt placement without subsequent endoscopy and one underwent subdural–peritoneal shunting; and in five patients the hydrocephalus arrested without the need for further treatment following serial reservoir tapping. The patients who underwent placement of a VP shunt after the second endoscopy suffered severe anatomical distortion as a result of the original ventriculitis; in two patients multiloculated ventricles required fenestration of septations and endoscopic placement of the catheter. One patient in whom ETV had been abandoned in favor of VP shunt placement following the initial endoscopy underwent ETV at the time that a subsequent VP shunt was removed because of infection. Ten patients in whom the procedure had been abandoned at the time of the initial endoscopy underwent ETV eventually; a total of 229 (76%) of the 300 patients ultimately underwent ETV. Shunting or shunt placement was performed in 61 patients, 13 of whom underwent endoscopic creation of a communication between loculated ventricles; CPX was performed in three and no additional procedures were performed in seven patients.

*Correlation of Fourth Ventricular Size on Preoperative Imaging With Endoscopic Assessment of Aqueduct Patency*

Because we assumed that the patency of the aqueduct would influence the likelihood of surgical success, our ability to predict the status of the aqueduct preoperatively was evaluated. In most cases the aqueduct was observed at the time of endoscopy and these observations were correlated with the preoperative impression of the size of the fourth ventricle on the imaging study, which was most often cranial ultrasonography. Patients with hydrocephalus secondary to myelomeningocele are excluded from this analysis because of the Chiari II malformation. In 200 patients from either the PIHC or NPIHC group, both the aqueduct at en-

doscopy and the fourth ventricle on the preoperative imaging study could be assessed with confidence. The fourth ventricle was assessed using cranial ultrasonography with a combination of coronal, axial, sagittal and, when necessary, mastoid views.

A clearly obstructed or closed aqueduct was revealed at the time of ventriculoscopy in 93 (91%) of 102 patients with a small fourth ventricle (Fig. 3). Among the nine patients with obstruction of the aqueduct and an enlarged fourth ventricle, six suffered from PIHC. This finding may indicate obstruction at two or more locations—at the aqueduct and the fourth ventricular outlets or beyond. Of the 83 patients with an enlarged fourth ventricle on the preoperative imaging study (Fig. 5), 69 (83%) demonstrated an open aqueduct. Among 15 patients in whom the fourth ventricle was believed to be of normal size, the aqueduct was closed in six and open in nine. The correlation between the appearance of an enlarged or small fourth ventricle on preoperative imaging and the status of the aqueduct on direct inspection was statistically significant ( $p = 0.0001$ ) (Table 4).

*Overall Results of ETV*

Of the 229 patients who underwent a completed ETV, five (2%) were lost to follow up less than 1 month after the operation, and four (1.8%) of the 224 in whom sufficient follow up occurred died within 1 month of surgery.

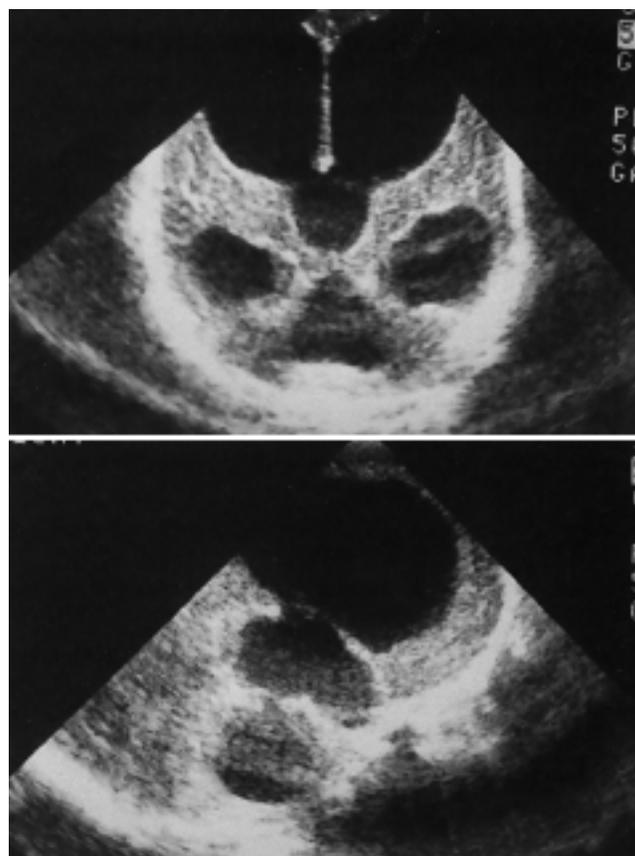


FIG. 5. Coronal (upper) and sagittal (lower) cranial ultrasonography studies demonstrating an enlarged fourth ventricle. Note the prominent foramen of Monro and sylvian aqueduct.

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TABLE 4  
Summary of fourth ventricular size and aqueductal patency

Status of Aqueduct	Size of the 4th Ventricle			Total
	Small	Large	Normal	
closed	93	9	6	108
open	3	69	9	81
narrow	6	5	0	11
total	102	83	15	200

Of the remaining 220 patients, treatment was successful in 115 (52%) after the first ETV; the mean follow-up period was 15.2 months (Table 5). Of these 115 patients, 91 (79%) were followed for more than 1 year with a mean follow-up period of 17.8 months (range 12–31 months), 12 (10.4%) were followed for 6 to 11 months, and 12 (10%) for less than 6 months. Of the 24 patients who were followed for less than 1 year, 12 died of unrelated causes between 2 and 11 months postoperatively, three were unable to be traced due to insecurity in Northern Uganda, and the remaining patients could not be found. Of the 105 patients in whom initial ETV failed, 65 underwent repeated endoscopy, 39 underwent shunt therapy, and one did not return for further treatment. The mean time from the initial attempted ETV to a repeated operation was 1.5 months (range 0.25–6 months). Fifty-four patients (53%) underwent a second surgery within 1 month or less, and 97 of the 105 patients in whom initial ETV failed (92%) presented with failure at 2 months or less postoperatively.

Of the 65 patients undergoing repeated endoscopy for failure of the initial ETV, 40 underwent reopening of a closed ETV. Of these 40 patients, none were lost to follow up and 3 patients died less than 1 month postoperatively. Of the remaining 37 patients who underwent a second ETV, the procedure was successful in 14 (38%); the mean follow-up period was 12.25 months (range 3–24 months). Of the remaining 25 patients who underwent a second endoscopy, the ETV was found to open or was unable to be reopened; 19 of these patients underwent VP shunt placement, four underwent choroid plexectomy, and two underwent placement of a reservoir.

In total, 129 (59%) of 220 patients who were followed up sufficiently ultimately underwent a successful ETV and did not require a shunt placement or an alternate procedure. This finding includes all patients regardless of age or cause of hydrocephalus. Analysis according to age, cause of hydrocephalus, and whether the aqueduct was open or closed at the time of endoscopy defined subsets of patients with distinct outcomes.

TABLE 5  
Summary of results of first ETV

Result	No. of Patients
success	115
failure	105
lost to follow up	5
death	4
total	229

Patients in each of the three categories of disease (PIHC, NPIHC, and myelomeningocele) were further subcategorized into Type A (< 1 year of age with an open aqueduct), Type B ( $\geq$  1 year of age with an open aqueduct), Type C (< 1 year of age with a closed aqueduct), or Type D ( $\geq$  1 year of age with a closed aqueduct) (Table 6). Patients in whom the aqueduct was either narrow but patent or not able to be evaluated were not assigned to a group; however, these patients are included in the analysis for total numbers of patients younger or older than 1 year of age.

### Results for Patients With PIHC

A total of 131 patients underwent ETV for PIHC, but two were lost to follow up and one died in the postoperative period, leaving 128, of whom 113 could be subdivided by both age and aqueduct status (Table 7). Among Type A patients, ETV was successful in 14 (45%) of 31. This finding includes two patients in whom a second ETV was successful and three in whom a second ETV failed. For Type B ETV was successful in seven of nine patients. Type C patients suffering from PIHC composed the single largest group in our patient population, and 44 (70%) of 63 Type C patients underwent a successful ETV, including five patients who underwent a successful second ETV and six in whom repeated operation failed. Eight of 10 Type D patients underwent successful ETV including two patients who underwent successful repeated operation and one in whom the second procedure failed. The difference overall in outcome between patients with an open aqueduct (Types A and B) and those with a closed aqueduct (Types C and D) was not statistically significant ( $p = 0.0639$ ).

Analyzing by age, 60 (59%) of 101 patients younger than 1 year of age and 22 (81%) of 27 patients older than 1 year of age underwent successful ETV (Table 8). Overall, ETV was successful in 82 (64%) of 128 patients with PIHC, including 10 patients who underwent a successful repeated operation. The mean follow-up period for these patients was 14.6 months (range 1–31 months). The difference in outcome between those patients younger than 1 year of age and those older than 1 year was significant ( $p = 0.0421$ ).

The patient populations for Types B and D were small for a meaningful comparison; nevertheless a statistically significant difference in outcome was found between Types A and C ( $p = 0.0254$ ): among patients with PIHC who were younger than 1 year of age, those with aqueductal obstruction experienced a significantly better outcome.

Patients with significant postinflammatory changes posed some of the most difficult technical challenges for the performance of ETV. Distortion of the anatomy often obscured landmarks, and access to the third ventricular

TABLE 6  
Classification of patient type by age and aqueductal patency

Type	Age (yrs)	Aqueduct
A	<1	open
B	$\geq$ 1	open
C	<1	closed
D	$\geq$ 1	closed

floor was sometimes made difficult by the presence of scar, hemosiderin, or inflammatory deposits.

*Results for Patients With NPIHC*

A total of 76 patients underwent ETV for NPIHC. One died in the postoperative period and three were lost to follow up, leaving 72 patients, of whom 65 could be subdivided by both age and aqueduct status (Table 7). Among Type A patients, ETV was successful in eight (31%) of 26; this finding includes three patients in whom a second ETV was successful and three in whom it was not. In four of six Type B patients, ETV was successful, and in both cases in which failure occurred a repeated operation also failed. In 11 (48%) of 23 Type C patients ETV was successful. Three of the cases in which failure occurred involved repeated operations; no successful ETV involved a repeated operation. All 10 Type D patients experienced successful outcomes from the initial ETV. Overall, the difference in outcome between patients with NPIHC with an open aqueduct (Types A and B) and those with a closed aqueduct (Types C and D) was statistically significant ( $p = 0.0481$ ).

When performing an analysis with respect to age, ETV was successful in 21 (40%) of 52 patients younger than 1 year of age and in 18 (90%) of 20 of patients older than 1 year (Table 8). Overall, 39 (54%) of 72 patients with NPIHC were successfully treated with ETV, including three patients who underwent a successful repeated ETV. The mean follow-up period for these patients was 15.6 months (range 1.5–29 months). The difference in outcomes between the two age groups was significant ( $p = 0.0001$ ).

The total number of Type B patients was small, but the differences in outcome between Types A and D ( $p = 0.0002$ ) and Types C and D ( $p = 0.005$ ) were significant. Interestingly, unlike in patients with PIHC, the difference between an open and closed aqueduct did not make a significant difference in the outcome among patients with NPIHC who were younger than 1 year of age, that is to say in Types A and C ( $p = 0.2536$ ).

*Results for Patients With Myelomeningoceles*

Twenty-seven patients with myelomeningocele underwent ventriculoscopy; a total of 22 underwent a completed ETV for initial management of their hydrocephalus. Of these 22, two died within 1 month of the operation and no

TABLE 8  
*Summary of results of ETV by cause of hydrocephalus and patient age*

Cause of Hydrocephalus	Patient Age (yrs)	No. Successful/Total (%)
PIHC	<1	60/101 (59)
	≥1	22/27 (81)
NPIHC	<1	21/52 (40)
	≥1	18/20 (90)
myelomeningocele	<1	8/20 (40)

patient was lost to follow up. No patient was older than 1 year of age. The mean age at the time of the initial ETV was 2.8 months (range 1–10 months). In four of 11 patients with open aqueducts (Type A) and in two of three patients with closed aqueducts (Type C), ETV was successful (Table 7). In one (20%) of five patients with a narrow or slit aqueduct ETV was successful, and the procedure was successful in an additional patient in whom the aqueduct could not be assessed. In all, eight (40%) of 20 patients with myelomeningocele were successfully treated with ETV; this finding includes one successful repeated operation (Table 8). The mean follow-up period was 12.9 months (range 3–18 months). Differences in outcomes between the small numbers of patients in Types A and C were not statistically significant.

*Anatomical Findings in Patients With Myelomeningoceles*

Among the 27 patients with myelomeningoceles who underwent ventriculoscopy, a number of anatomical abnormalities were noted (Table 9). The choroid plexus was atrophic or underdeveloped in three patients and absent in one. In eight patients the choroid plexus in the lateral ventricles was connected across the midline by a thin, vascular, sheetlike structure resembling an exposed tela choroidea membrane. In 10 patients the forniceal bodies were fused into a single thick midline structure (Fig. 6). The foramina of Monro were completely absent in one patient with no visible choroid plexus (as noted earlier) and were observed to be quite stenotic in another four patients. The aqueduct was clearly visible and appeared completely closed at its orifice in five patients, extremely narrow or slitlike in nine, and open at its orifice in 11. In one patient what appeared to be a small accessory aqueductal orifice slightly anterior to the main orifice was observed. The interthalamic adhesion was very thick in 17 patients, partially obscuring the floor or hampering entrance into the third ventricle in some cases. In five patients what appeared to be variable degrees of interhypothalamic adhesion, traversing the third ventricle and interconnecting its lateral walls, hampered access to the floor (Fig. 7). The floor of the third ventricle was thickened in 16 patients, and in six the brainstem and BA appeared to be crowded up against the clivus beneath the floor, resulting in an extremely narrow cisternal space.

*Failure With Patent ETV at Repeated Endoscopy*

In 17 patients ETV failed despite the creation of an opening to a good space (with no additional membrane or scar underlying the site) that was observed during repeated endoscopy. In 15 of these patients, the status of the aqueduct was known. Of these patients, six were in the PIHC Type

TABLE 7  
*Summary of results of ETV by cause of hydrocephalus and patient type*

Cause of Hydrocephalus	Patient Type	No. Successful/Total (%)
PIHC	A	14/31 (45)
	B	7/9 (78)
	C	44/63 (70)
	D	8/10 (80)
NPIHC	A	8/26 (31)
	B	4/6 (67)
	C	11/23 (48)
	D	10/10 (100)
myelomeningocele	A	4/11 (36)
	C	2/3 (67)

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TABLE 9  
Summary of findings at endoscopy in 27 patients with a myelomeningocele

Finding	No. of Patients (%)
thick interthalamic adhesion	17 (63)
thick ventricular floor	16 (59)
aqueduct closed or narrow	14 (52)
fused, thickened fornices	10 (37)
interconnected choroid plexus	8 (30)
narrow cistern	6 (22)
interhypothalamic adhesions	5 (19)
stenotic foramen of Monro	5 (19)
hypoplastic choroid plexus	4 (15)

A group, three were in the NPIHC Type A, two were in the NPIHC Type B, and one was in the myelomeningocele Type A group; that is, open aqueducts were apparent in all. Three patients—two with PIHC and one with NPIHC—were Type C (closed aqueduct); in each of these patients the only ETV site open was the lamina terminalis. Each of the 13 patients, therefore, in whom ETV was found to have failed despite an open ETV site in the floor of the third ventricle, demonstrated an open aqueduct. Of the 17 total patients 14 were younger than 1 year old. Stated another way, in no patient with aqueductal obstruction was the ETV considered to have failed if repeated endoscopy demonstrated that a patent opening was created in the floor of the ventricle. The apparent cause of failed ETV in those patients was in all cases obstruction of the ETV in the floor. From this finding it can be inferred that repeated endoscopy should be performed at the time of ETV failure in those patients with closed aqueducts.

### Effect of ETV Site on Outcome

Because Type C patients with PIHC composed the single largest homogeneous category of patients (63 patients < 1 year with PIHC and a closed aqueduct), this subgroup was used to assess whether the site of the ETV (third ventricular floor, lamina terminalis, or both) influenced outcome.

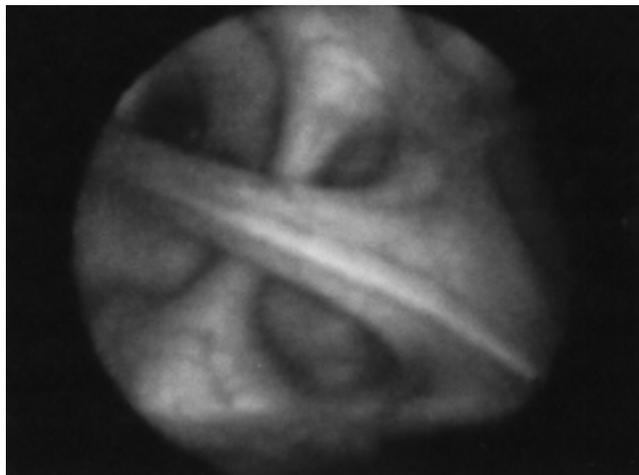


FIG. 6. Endoscopic view in which thick, fused forniceal bodies appear crossing perpendicular and superior to an enlarged interthalamic adhesion.

In an analysis of outcome data for the initial ETV, the procedure proved successful in 13 (72%) of 18 patients who underwent only a floor ETV, in 23 (70%) of 33 patients who underwent ETV in both the floor and lamina terminalis, and in three (25%) of 12 patients who underwent only a lamina terminalis ETV. The difference in outcomes between those patients who underwent a floor ETV and those who underwent ETV in both the floor and lamina terminalis was not statistically significant ( $p = 1.000$ ). The difference between those patients who underwent an ETV in the floor and those who underwent an ETV in the lamina terminalis was significant ( $p = 0.0236$ ), and the difference in outcome between lamina terminalis ETV only and floor ETV with or without lamina terminalis ETV was also significant ( $p = 0.0085$ ).

The outcomes of ETV only in the lamina terminalis among Type C patients with PIHC, therefore, did not appear to be as successful as the outcomes of ETV in the floor. Moreover, a second ETV in the lamina terminalis in combination with an ETV in the floor did not appear to offer an advantage.

### Correlation of Observable CSF Flow With Success

In 154 patients the presence or absence of CSF flow was recorded. Of 79 patients who experienced a good outcome, CSF flow through the ETV(s) was observed in 77 and minimal or no flow in two. Of 75 patients in whom the first ETV failed, good CSF flow through the ETV(s) was observed in 61 and minimal or absent flow in 14. In 14 (88%) of 16 patients ETV failed when CSF flow was not apparent. The difference in outcomes between those patients in whom flow was observed and those in whom flow was minimal or absent was statistically significant ( $p = 0.0011$ ).

### Mortality and Morbidity Rates

A total of 466 ventriculoscopic procedures were performed in all 300 patients. There were 10 postoperative deaths, defined as death due to any cause within 1 month of the operation. For all ventriculoscopy procedures, therefore, whether an ETV was completed or not, the overall surgical mortality rate was 2.1%. One death resulted from an intraoperative cardiopulmonary arrest following an uneventful and uncomplicated ETV. Four deaths were secondary to postoperative infection—two secondary to shunt infection following shunt placement at the time of an abandoned ETV attempt and two secondary to Gram-negative ventriculitis following an ETV. An additional two deaths following ETV might possibly have been related to postoperative infection: one patient experienced a 2-day period of fever and seizures and died at home because of lack of transportation to the hospital and the other died at home of an unknown febrile illness. One patient died at home of an unknown cause following placement of a reservoir when ETV was abandoned. One patient who underwent ETV at the time of VP shunt removal for infection died of pneumonia and sepsis. In one patient in whom ventriculoscopy revealed a large basilar tip aneurysm obstructing the floor, a VP shunt was placed with the ventricular catheter positioned endoscopically; the patient died later of what appeared to be an aneurysm rupture.

In 229 of 300 patients ETV was completed, and five pa-

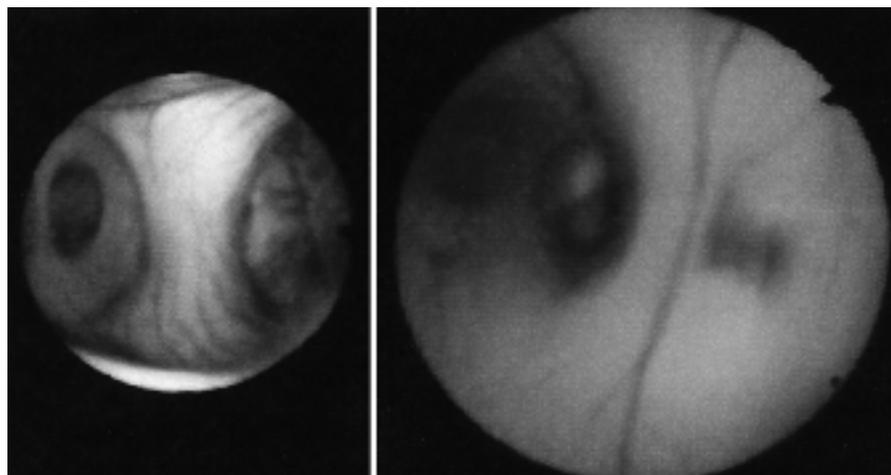


FIG. 7. *Left:* Endoscopic view of a large intermediate mass with the superior edge of the aqueduct visible at its posterior margin. The thick third ventricular floor and pituitary are seen to the left of (anterior to) the interthalamic adhesion, and an interhypothalamic adhesion is evident just anterior and superior to the pituitary. *Right:* Endoscopic view of the third ventricular floor after an ETV just anterior to an interhypothalamic adhesion that bisects the floor. Note another segment of thin floor and the mammillary bodies immediately posterior. The dorsum sellae and posterior edge of the pituitary can be seen through the ETV opening.

tients were lost to follow up. Of the 224 patients with a follow-up period longer than 1 month who underwent an initial ETV, four died; the surgical mortality rate for ETV was 1.8%. Of the 40 patients who underwent repeated ETV at the time of endoscopy for ETV failure, three died within 1 month of surgery. The total surgical mortality rate for ETV, including initial and repeated surgeries, was 1.9% (seven of 264 patients).

One patient with Gram-negative ventriculitis recovered, and early in the series, two patients required a second operation because of CSF leakage. There were no known incidents of hypothalamic dysfunction or cranial nerve palsy. There was no case in which the BA was injured. In one patient, significant intraventricular hemorrhage occurred after fenestration of the lamina terminalis and an external ventricular drain was temporarily required postoperatively. That patient ultimately underwent shunt placement.

Eighteen patients are known to have died after the 1-month postoperative period following ETV (first time or repeated procedure), at a mean of 7.4 months postsurgery (range 2–21 months). The majority (16) of these patients died in the 1st year postsurgery. In 11 cases the child died at home of an unknown cause. In three cases the reported cause of death was malaria, in one case it was an unknown febrile illness, and in one case each the cause was respiratory illness, brainstem tumor, and gastroenteritis.

### Discussion

#### *Causes of Hydrocephalus in Uganda*

Hydrocephalus is very common in East Africa, although its exact incidence is not known. It has recently been stated that the most common cause of the condition in the Central African countries of Zambia, Zimbabwe, and Malawi is congenital, associated with neural tube defects and aqueductal stenosis; the ratio of congenital to “postmeningitic” hydrocephalus in Zimbabwe was reported to be 2:1.<sup>1</sup> Au-

thors of a study from Saudi Arabia of infants with hydrocephalus attributed the cause to meningitis in only 14.8% of cases.<sup>10</sup> In contrast, our experience indicates that hydrocephalus secondary to CNS infection is actually the single most common cause of hydrocephalus in Uganda, accounting for 60% of cases. In the present study, the distribution of patients in each category of disease reflected that seen in our overall experience with the 468 patients with hydrocephalus who presented for treatment between January 2001 and March 2003. In this larger group, the cause of hydrocephalus was determined to be postinfectious in 265 cases (57%), nonpostinfectious in 136 cases (29%), associated with myelomeningocele in 61 cases (13%), associated with encephalocele in five cases (1%), and the probable result of neonatal intraventricular hemorrhage in one case.

Although malaria is very common in Uganda, the correlation of meningeal or ventricular inflammation and hydrocephalus to malaria is unknown. In patients with malaria, the CSF usually appears normal but may demonstrate moderately increased protein or moderate lymphocytosis.<sup>18</sup> Malaria is often the presumed diagnosis in febrile infants and in children from places such as Uganda where malaria is endemic and is often treated empirically, thereby possibly delaying the diagnosis and management of other febrile illnesses such as meningitis.<sup>23</sup>

A more probable cause of PIHC is meningitis. Neonatal meningitis has been previously associated with ventriculitis, aqueductal obstruction, ventricular loculations, and cerebral infarction.<sup>5,19,21</sup> Such conditions were commonly seen in our patients with PIHC. In 76% of children presenting with PIHC in this study, the infection that preceded the onset of the hydrocephalus occurred within the 1st month of life (the neonatal period). Sepsis and meningitis with onset in the first few days after birth are likely due to organisms acquired during delivery.<sup>2</sup> In Western literature, *Escherichia coli* and Group B streptococci are indicated as the most common causative organisms in neonatal meningitis;

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other Gram-negative enteric bacilli, *Listeria monocytogenes*, and *Staphylococcus aureus* are also of noted importance.<sup>2,5,24</sup> A study from Malawi of 61 neonates with meningitis showed the most common causative organisms to include *Streptococcus agalactiae* (23%), *Salmonella typhimurium* (15%), *Strep. pneumoniae* (11.5%), and other Gram-negative rods (11.5%).<sup>23</sup> The cause of ventriculitis in our patients was not known, and in Uganda the most common organisms causing neonatal meningitis are not reported. In our study, CSF samples were sterile at the time of surgery, although endoscopic biopsy of the intraventricular inflammatory exudates in five patients revealed polymorphonuclear leukocytes. Because the reported mortality rate of neonatal meningitis approaches 50%,<sup>5,23</sup> this disease is presumably a significant cause of death, as well as disability from primary brain injury, in Ugandan infants.

That the majority of cases of hydrocephalus in Ugandan children are postinfectious, and that the condition can be successfully managed with ETV are perhaps the most important findings of this study. Although these results cannot be extrapolated to other developing countries, it is at least reasonable to assume that PIHC secondary to neonatal ventriculitis or meningitis is an important cause of hydrocephalus throughout East Africa and perhaps in other developing nations as well. This finding represents a significant public health and preventive medicine problem because PIHC may be related to perinatal conditions that predispose children to neonatal sepsis. In our patient population, the majority of children are born at home in a rural setting and with no medical assistance. According to United Nations statistics for 2003, in only 39% of births in Uganda is a skilled attendant present at delivery. Obstetrical complications such as premature rupture of membranes, premature or protracted labor, and chorioamnionitis have been associated with neonatal sepsis.<sup>24</sup> Certain local practices, such as placing cow dung on the umbilical stump, may also contribute to infection. We found no correlation between HIV or AIDS and PIHC, however. Even when successfully and promptly treated, PIHC can be associated with primary parenchymal damage related to the initial infection that predisposes the child to developmental delay, spasticity, and visual impairment.

### *The Usefulness of ETV in a Developing Country*

The management of hydrocephalus throughout Sub-Saharan Africa is hindered by economic constraints and the difficulties patients and families face regarding transportation and access to proper care. Shunts, as well as the expertise to place them, are not available to most children. When shunts are available, most people cannot afford the device. Even when the placement of a shunt is possible, its use is more dangerous in this context than in developed countries because complications and shunt malfunctions are less likely to be properly addressed in a timely fashion, if at all. This information is particularly sobering in view of the prospective multicenter North American study that found a 43.6% incidence of shunt failure (which included an 8.1% infection rate) within the first 2 years of placement performed by presumably the best of hands.<sup>9</sup> Avoiding shunt dependency in an environment such as ours is an important goal. Moreover, in view of the limited economic and healthcare resources in developing countries, it has been ar-

gued that ETV may be more cost effective than shunt placement in terms of the long-term use of available resources.<sup>3</sup> A separate small study failed to support this hypothesis, and its authors argued that long-term follow up and larger numbers of patients are needed.<sup>12</sup>

Of 300 patients presenting for initial treatment of hydrocephalus by endoscopy preceding an attempted ETV, 151 (50.3%) ultimately underwent shunt placement because either they were unable to undergo ETV or the procedure failed. In this series, our initial experience, approximately half of all patients presenting for treatment, regardless of age, causes of hydrocephalus, or imaging findings, were treated without shunt placement.

In general, third ventriculostomy for noncommunicating hydrocephalus is the accepted, preferred treatment.<sup>13,15,17</sup> Its usefulness, however, has been questioned in cases involving infants,<sup>6,13,15,29</sup> and in hydrocephalus secondary to infection<sup>6,7,16,22</sup> or associated with myelomeningocele.<sup>6,11,29</sup> Because approximately 80% of patients presenting to us for treatment of hydrocephalus were younger than 1 year old, and because the cause in approximately 60% of patients appeared to be infection, these subgroups of patients were evaluated with a particular interest in determining the efficacy of ETV. Our observation, however, that in more than half of the patients with PIHC a small fourth ventricle on cranial ultrasonography indicated that many of these cases might be noncommunicating in nature, and thus amenable to treatment by using ETV.

In most African countries,<sup>1</sup> including Uganda, ETV has been previously unavailable. To my knowledge there has not been a prospective study of ETV as the primary treatment of all children with hydrocephalus in a developing country. In addition, this is the largest series of infants or of patients with PIHC treated with ETV of which I am aware.

The results of this study indicate that ETV can be performed with acceptable results in the environment of a developing country. The findings further demonstrate that ETV is useful in those patients who most commonly present to our practice, that is, young infants with hydrocephalus resulting from infection. If made available, ETV could provide a safer alternative to life-long shunt dependency as the primary mode of treatment for hydrocephalus in developing countries.

Patients tend to present to our unit with advanced hydrocephalus and severe ventriculomegaly. This tendency can be attributed to long intervals between the onset of the child's symptoms and his or her arrival at our facility. It is notable that of 468 children presenting for management of hydrocephalus (including the 300 in the present study) the mean time from clinical manifestations of hydrocephalus to presentation to our unit for treatment was 7.46 months. This finding highlights one of the challenges in the management of hydrocephalus in a setting such as ours. The reasons for such a dramatic delay include misconceptions about the problem (including animistic/spiritualistic interpretations and the early resort to "traditional" practitioners), a fatalistic worldview, discouragement from seeking help by members of the local community, hopelessness about accessing help because of lack of funds, and lack of transportation. In addition, parents often are not properly referred or are repeatedly put off when they seek medical attention for their child. Whatever the reasons, the tardy

presentation for treatment has a negative impact on ultimate outcome.

#### *Preoperative Selection Parameters*

The aim of this study was to investigate the usefulness of three readily available preoperative parameters (age, cause of hydrocephalus, and the status of the aqueduct) to classify patients in a way that would be predictive of the likelihood that ETV would be successful, thus providing simple criteria for patient selection. Because in the cases of PIHC and NPIHC fourth ventricular size on cranial ultrasonography correlated very well with whether the aqueduct was found to be patent at endoscopy, these parameters provide a simple and logical classification scheme: PIHC Types A, B, C, and D and NPIHC Types A, B, C, and D.

Not surprisingly, patients in whom ETV proved most successful were NPIHC Type D (patients > 1 year of age with aqueductal obstruction), but this was a small group (10 patients). The next three most successful patients were PIHC Types B and D (all patients with PIHC  $\geq$  1 year old) with 78 and 80% success rates, respectively, and PIHC Type C (infants < 1 year of age with a closed aqueduct) with a 70% success rate. For patients with PIHC, those younger than 1 year of age with a closed aqueduct had significantly better outcomes than those with open aqueducts ( $p = 0.0254$ ), but this was not the case in the NPIHC group ( $p = 0.2536$ ). Four of six NPIHC Type B and two of three myelomeningocele Type C patients underwent successful ETVs, but these subgroups are too small for meaningful commentary. Those subgroups with less than 50% success rates were NPIHC Type C (48%), PIHC Type A (45%), myelomeningocele Type A (36%), and NPIHC Type A (31%).

The number of patients older than 1 year of age in this study is too small to allow for conclusions regarding success of ETV by type and causes of hydrocephalus, although patients older than 1 year in every subcategory (except those with a myelomeningocele) demonstrated success rates between 67 and 100%. Some conclusions regarding ETV in infants younger than 1 year of age, however, can be drawn. The only patients younger than 1 year old in whom results similar to those in older patients can be expected are those with PIHC who have a closed aqueduct (Type C). In 70% of these patients, hydrocephalus was successfully treated with ETV. The procedure was effective in less than 50% of all other infants younger than 1 year of age.

Based on this experience and excluding patients with a myelomeningocele, who will be considered separately, we currently regard ETV as the procedure of first choice for all children older than 1 year regardless of the cause of hydrocephalus and for those children younger than 1 year of age with PIHC and aqueductal obstruction (PIHC Type C). The status of the aqueduct can be reliably predicted in these infants using preoperative cranial ultrasonography.

It is debatable whether proceeding with an ETV when the likelihood of success is 50% or less is justifiable. It might be recommended that ETV be delayed in infants younger than 1 year of age except for those in the PIHC Type C category. An ETV could be attempted later for subsequent shunt failure or infection after the child reaches 1 year of age. One might, however, argue to "give it a try"

when circumstances make shunt placement sufficiently undesirable or dangerous, as it is in cases in which a patient will be returning ultimately to a remote environment in which funds for transportation are scarce.

#### *Factors Prohibiting Completion of ETV at the First Endoscopy*

As operative experience increased in this series, it became much less common for the surgeon to abandon the ETV attempt in favor of shunt placement. As long as the CSF remains clear enough for adequate visibility and scarring from PIHC has not completely effaced the third ventricular landmarks or blocked access to it entirely, ETV can usually be completed despite the proximity of the BA or thickening of the third ventricular floor, factors that would have led to abandonment of ETV earlier in the series. Nearly all cases in which ETV is abandoned are now those in which a shunt is placed because intraventricular distortion from scarring is too severe or those in which a reservoir is necessitated due to poor visibility resulting from insufficient clarity of the CSF.

#### *The ETV in Cases of PIHC*

Although the numbers of patients are few, the previously reported experience with treating postmeningitic hydrocephalus by ETV has been disappointing and considered a contraindication to the procedure.<sup>6,7,16,22</sup> It has been considered axiomatic by some researchers that any history of meningitis or ventriculitis is a negative predictor of ETV success.<sup>25</sup> A retrospective multicenter review that included 42 patients undergoing ETV with a history of CSF infection was more encouraging, however,<sup>26</sup> as was a recent retrospective review that included five patients (age range 4.5–21 years) with PIHC.<sup>28</sup>

In the present study ETV was successful in 70 to 80% of patients in whom infection appeared to have caused the hydrocephalus; the exception to this finding occurred in Type A patients (< 1 year of age with an open aqueduct) in whom the success rate was 45%. The infectious origin of hydrocephalus in our patients often leads to ventriculitis with obstruction of the aqueduct; indeed, the majority of the patients with PIHC (64%) demonstrated aqueductal obstruction on direct inspection. It is also quite possible that for some cases in which the aqueduct is open, there may be obstruction to fourth ventricular outflow or extraventricular intracisternal obstruction, as some investigators have recently argued.<sup>20</sup> This may account for the success of ETV seen in many of the patients with open aqueducts. The pathophysiology of PIHC in our patients appears to differ from the common expectation of a communicating PIHC secondary to impaired CSF absorption, which would be unlikely to benefit from ETV.

In the case of multiloculated ventricles, fenestration for communication is necessary whether an ETV can be performed or a VP shunt is placed. In patients with this condition, therefore, endoscopy is almost always beneficial.

#### *The ETV in Cases of NPIHC*

Among infants younger than 1 year of age with NPIHC and a closed aqueduct ETV was less successful than among similar patients with PIHC. Given that most infants classi-

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fied as having NPIHC were assumed to be suffering from hydrocephalus of a congenital nature and that patients with PIHC did not, by definition, harbor hydrocephalus at birth, the difference in outcomes may lie in the fact that infants with PIHC have had an opportunity to develop subarachnoid spaces and CSF absorption capacity, whereas those with congenital hydrocephalus have not. It is no surprise that ETV was very successful (100%) in patients with NPIHC older than 1 year of age with a closed aqueduct (Type D), but for all other patients with NPIHC (Types A, B, and C) the procedure was not as successful as it was in patients with PIHC of the same type; this finding was not an anticipated outcome of the study.

### *The ETV in Hydrocephalus Associated With a Myelomeningocele*

The origin of hydrocephalus associated with a myelomeningocele is thought to be multifactorial and may be secondary to obstruction of CSF flow at the aqueduct, the fourth ventricular outlets, the craniocervical junction, or the arachnoid granulations;<sup>8</sup> ETV would appear, therefore, to be a logical treatment option for some of these patients. In a retrospective review of 69 patients harboring a myelomeningocele conducted over an 18-year period the overall success rate of ETV was 72%.<sup>29</sup> The majority (55 of 69) of the patients in that series, however, were older (mean age 11 years) and had previously undergone shunt placement. The success rate of those patients younger than 2 years of age was 53% and of those younger than 6 months (eight patients), only 12.5%. The usefulness of ETV as the initial treatment of hydrocephalus in infants with a myelomeningocele who are younger than 6 months old therefore appeared questionable.

Our experience with infants suffering from a myelomeningocele has been more encouraging. In 28 infants with a myelomeningocele, an ETV was completed in 22 and sufficient follow up occurred in 20. All 22 patients were younger than 1 year of age, with a mean age of 2.8 months (range 1–10 months). Fifteen patients were younger than 3 months old; ETV was successful in 50% of these cases. These youngest infants experienced considerably better outcomes than those whose cases were reported elsewhere.<sup>29</sup> The overall success rate for patients with myelomeningocele was 40%, which is identical to the success rate for patients with NPIHC younger than 1 year of age (21 of 52).

In eight patients with a myelomeningocele, performing an ETV in the third ventricular floor was deemed technically inappropriate and, therefore, ETV was performed only in the lamina terminalis. This procedure was successful in only one (14%) of seven patients in whom follow up was sufficient. The inclusion of lamina terminalis ETV increased the number of patients in whom ETV was performed; however, of those who underwent an ETV in the floor—regardless of whether an ETV of the lamina terminalis was also performed—seven (54%) of 13 underwent successful ETVs. This finding is consistent with the inferior results of lamina terminalis ETV among patients with PIHC compared with the results for ETV in the floor. When an ETV performed in the floor is technically feasible, this finding indicates that the procedure offers a reasonable chance (> 50%) of shunt independence from infancy for-

ward in patients with a myelomeningocele. Endoscopy for attempted floor ETV in patients with hydrocephalus associated with a myelomeningocele may be a reasonable approach, therefore, and it is currently our practice.

### *The ETV in Infants Younger Than 1 Year of Age*

Of the 229 patients who underwent a completed ETV, 78.6% were 1 year old or younger. For patients in this age group ETV has not been uniformly advocated,<sup>6,13,15,29</sup> although good results have been reported in small groups of infants with congenital aqueductal stenosis.<sup>7,16</sup> Authors of a recent report suggested that the cause of hydrocephalus was a more important determinant of ETV success than patient age.<sup>4</sup> The present study confirms that finding; although results are generally better in older patients, the young age of a patient does not alone prohibit good outcomes for ETV. The effect of age on outcomes for ETV varies according to the cause of the hydrocephalus and the status of the aqueduct—parameters that can be reasonably assessed prior to surgery.

### *The Usefulness of Repeated Endoscopy in Patients Presenting With ETV Failure*

Repeated endoscopy for ETV failure has been advocated previously.<sup>27</sup> In the present study it was found that, in contrast to those with open aqueducts, no patient with aqueductal obstruction was noted to have treatment failure in the face of a patent ETV in the floor on repeated endoscopy. This finding indicates the need for repeated endoscopy to assess the ETV for reopening in this group of patients. Among all patients who underwent reopening of a closed ETV following repeated endoscopy, 38% experienced successful outcomes. Repeating the ventriculoscopy does not significantly lengthen the duration of the procedure in the event that VP shunt placement is required, and it offers the additional advantage of facilitating endoscopic ventricular catheter placement if the ETV is found to be open or a repeated ETV is not possible. Repeated ventriculoscopy is recommended, therefore, in those patients presenting with ETV failure, particularly in patients with aqueductal obstruction.

### *Lamina Terminalis ETV*

At the start of this series, it was hoped that the lamina terminalis would provide an alternative site for ETV when fenestrating the floor was not feasible. The success rate of the lamina terminalis ETV among Type C patients with PIHC (the single largest group) was not as high as the rate for ETV in the floor, however, and the addition of a lamina terminalis ETV to a floor ETV did not appear to confer an advantage. Moreover, lamina terminalis ETV was not successful when performed in patients in the myelomeningocele group—the very patients in whom floor ETVs are often difficult.

### *Postoperative Death*

The surgical mortality rate was 1.9%; most deaths were due to infection. This finding might reflect a combination of factors common among our patients: poor nutritional status, poor hygiene, very young age, debilitated physical status (due to advanced hydrocephalus and, often, recent



FIG. 8. Malnourished infant with advanced hydrocephalus positioned for ETV.

recovery from another significant illness), and poor skin integrity (due to advanced hydrocephalus resulting in thin scalp and poor nutritional status) (Fig. 8). In addition, patients whose conditions deteriorate at home often delay in presenting for treatment, sometimes dying at home or presenting in an extremely weakened condition. These factors contribute significantly to the difficulties of the practice of pediatric neurosurgery in this environment.

The one patient who suffered an intraoperative cardiac arrest immediately following an uneventful ETV was resuscitated, but the child experienced another arrest and died later the same day. This incident may be similar to that in a recent case report citing this as a rare complication of ETV.<sup>14</sup>

The 16 patients who died during the 1st year (between 2 and 11 months) postsurgery contributed another 7% to the mortality rate beyond the initial postoperative time frame. This result reflects the risks to children at large in Uganda, where the infant mortality rate is currently 79 per 1000 live births and that for patients younger than 5 years old is 124 per 1000 live births.<sup>30</sup>

### Conclusions

On the basis of this study, the following conclusions can be made. 1) Ventriculitis or meningitis (primarily neonatal) appears to be the most common cause of hydrocephalus in Uganda and represents a significant preventive medicine/public health problem. 2) In a developing country such as Uganda, where environmental factors make shunt dependency more dangerous, ETV can be performed with acceptable results. 3) For all children with hydrocephalus who are older than 1 year of age, and for those patients younger than 1 year who are suffering from PIHC with

aqueductal obstruction, ETV is recommended as the first treatment of choice. 4) The status of the aqueduct can be reliably assessed preoperatively by observing the size of the fourth ventricle on cranial ultrasonography; therefore, the preoperative classifications presented in this report can be used easily in most centers. 5) The success rate for ETV in the floor is higher than in the lamina terminalis, and a lamina terminalis ETV performed in addition to an ETV in the floor does not increase success. 6) The absence of observable CSF flow through the ETV site at the time of the procedure correlates with failure.

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