Initial management of hydrocephalus associated with Chiari malformation Type I–syringomyelia complex via endoscopic third ventriculostomy: an outcome analysis

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Object. The aim of this study was to evaluate the efficacy of endoscopic third ventriculostomy (ETV) in patients with Chiari malformation Type I (CM-I) and hydrocephalus with or without syringomyelia.

Methods. The authors identified, in a prospective endoscopy database, 16 adults and children (age range 2–68 years) with CM-I and hydrocephalus that had been managed with ETV. They reviewed the clinical features and radiographic findings for all patients. Fifteen patients underwent ETV as a primary treatment, whereas 1 patient underwent the procedure at the time of shunt failure. All patients had symptomatic hydrocephalus with either aqueductal or fourth ventricular outflow obstruction. The mean duration of follow-up was 42 months.

Results. Fifteen patients (94%) remain shunt free following ETV for CM-I. Five (83%) of the 6 patients with a syrinx had improvement or resolution of the syrinx following ETV. Six patients (37.5%) underwent foramen magnum decompression for persistent CM-I– or syrinx-related symptoms. There was no cerebrospinal fluid leakage or intracranial pressure-related problem following foramen magnum decompression.

Conclusions. Endoscopic third ventriculostomy provides a durable method of treatment for hydrocephalus associated with CM-I. It is effective as a primary treatment, and the authors advocate its use as a replacement for routine ventriculoperitoneal shunt insertion in these patients. Management of the hydrocephalus alone is often sufficient and may obviate decompression, although a significant proportion of patients will still need both procedures.

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Key Words • Chiari malformation Type I • endoscopic third ventriculostomy • hydrocephalus

Chiarimalformation Type I, defined by herniation of the cerebellar tonsils below the foramen magnum, is associated with hydrocephalus in 7–10% of cases.12 The origin of the hydrocephalus is often fourth ventricular outflow obstruction; however, a proportion of cases will also involve aqueductal stenosis.

In the presence of hydrocephalus, CSF diversion surgery, most commonly VP shunting, has been considered the best initial treatment option. Following treatment of the hydrocephalus, a number of patients will not demonstrate progressive neurological deterioration and therefore will avoid subsequent foramen magnum decompression.13 The resolution of CM–syringomyelia complex symptoms following ETV was first reported in 1996 by Nishihara et al.14 However, it has been suggested that in the presence of CM, third ventriculostomy may be contraindicated due to compression of the prepontine cistern.2 To date, there have been no large studies of ETV outcome in CM-I with hydrocephalus. We evaluated a series of 16 patients with CM-I and hydrocephalus managed with initial ETV to determine the outcome in terms of both shunt independence and avoidance of subsequent hindbrain decompression.

Clinical Materials and Methods

From a prospectively maintained endoscopy database, we identified 16 patients with CM-I and symptomatic hydrocephalus, treated with ETV between 1998 and 2006. All charts were reviewed for presenting symptoms that were classified as related to ICP, CM, or syrinx. All preoperative and follow-up MR imaging studies were reviewed to assess the level of descent of the cerebellar tonsils, associated aqueductal stenosis, and the presence of a spinal syrinx.

All patients underwent third ventriculostomy using a flexible endoscope (Karl Storz), which was introduced via a peel-away sheath. The stoma in the floor of the third ventricle was created using a light touch balloon (NMT Neurosciences), and the prepontine space was inspected for the presence of a second membrane, which was fenestrated if present. The senior author (C.M.) performed all procedures. Any intra- and postoperative complications were recorded. No patients underwent the placement of alternative CSF diversion devices such as an external ventricular drain or Ommaya reservoir at the time of ETV.

The primary outcome measure was shunt independence at the last follow-up, and secondary outcome measures in-
cluded progression to hindbrain decompression, complications of hindbrain decompression caused by elevated ICP, and radiographically demonstrated resolution of the syrinx.

Results

Patient Population

Sixteen patients with CM-I and hydrocephalus underwent ETV. All patients had ventriculomegaly together with symptoms and signs of elevated ICP. (Note that we perform ICP monitoring in patients with CM-I and small ventricles only if symptoms suggestive of increased ICP are also present.) There were 9 male and 7 female patients, with a mean age of 31.9 years (range 2–68 years) at presentation. The mean duration of follow-up was 42 months (range 6–96 months). One patient had Noonan syndrome, and 1 patient had previously had a head injury. No other patient had a history of neurological disease.

Fifteen patients underwent primary ETV, and 1 patient underwent ETV at the time of shunt malfunction. No patient in the series had undergone any previous surgical intervention to address the CM-I.

Symptom Categories

For the purposes of analysis we classified presenting symptoms into 3 groups: Group 1, elevated ICP-related symptoms (for example, global headache, vomiting, visual obscuration, papilledema, cranial nerve VI palsy, development of strain-related headache following hindbrain decompression); Group 2, classic CM-related symptoms plus elevated ICP (for example, cough-induced occipital headache, ataxia, or facial or upper limb symptoms in the absence of a syrinx); and Group 3, syringomyelia plus elevated ICP (for example, dissociated sensory loss or upper limb weakness and wasting). All patients demonstrated symptoms of raised ICP.

Neuroimaging Studies

All patients underwent T1- and T2-weighted MR imaging studies of the brain and spine with phase-contrast CSF flow studies. A neuroradiologist (K.D.) reviewed all images. All patients demonstrated CM-I with a mean 15-mm herniation of the cerebellar tonsils below the basion-opisthion line. All patients had ventriculomegaly. Twelve patients were characterized as having a small posterior fossa based on the presence of a low-lying confluence of sinuses, a reduction in the height of the occipital bone, and accentuation of the slope of the tentorium cerebelli with crowding of the posterior fossa contents (volumetric measurements were not obtained). Three patients also had aqueductal stenosis and 2 had a retrocerebellar arachnoid cyst. Six patients had a spinal cord syrinx. In the 4 patients who did not have posterior fossa osseous abnormalities, 2 had aqueductal stenosis and 1 had a retrocerebellar arachnoid cyst to account for the tonsillar ectopia; 2 of these 4 patients required subsequent foramen magnum decompression.

The mean level of tonsillar herniation following ETV was 13.3 mm, and the size of the ventricle reduced in only 5 patients. In the patients with aqueductal stenosis only 1 had evidence of posterior fossa osseous abnormalities, and the single ETV failure occurred in 1 patient with aqueductal stenosis.

Primary Outcome: Shunt Independence

Fifteen patients (94%) remained shunt free following ETV. The single case of ETV failure occurred in a 54-year-old woman who had previously undergone VP shunt placement for hydrocephalus associated with CM-I. She presented with headaches and right facial numbness caused by shunt obstruction as well as ventriculomegaly and aqueductal stenosis with CM-I on MR imaging. After ETV and shunt removal she continued to experience cough-induced headaches and facial numbness, and she underwent foramen magnum decompression 2 months after ETV. Her symptoms remained resolved for 2 years but then later recurred; a repeated ETV revealed a patent stoma, and a VP shunt was inserted.

Secondary Outcome: Symptom Resolution According to Symptom Category

Symptom resolution after ETV, according to symptom category, is featured in Table 1. All patients had symptoms of elevated ICP, and in 3 patients ICP-related symptoms were the only ones. Elevated ICP-related symptoms resolved in all patients following ETV. Eight patients had CM-associated symptoms in addition to those due to raised ICP, mainly cough-induced headache. Five patients presented with syringomyelia in addition to symptoms of increased ICP. Three patients with syringomyelia experienced improvement or resolution of their symptoms following ETV. Of the 8 patients with CM-I symptoms without syringomyelia, 4 experienced complete symptom resolution after ETV. The remaining 4 patients required subsequent foramen magnum decompression.

Progression to Hindbrain Decompression

Among the 16 patients, 6 (37.5%) required foramen magnum decompression following initial management with ETV. The mean time to decompression surgery was 6 months after the initial ETV (range 0.5–12 months). All 6 patients demonstrated minimal improvement in symptoms (other than those of raised ICP) following initial ETV, including strain-related headache. All 6 patients had resolution of strain-related headache following hindbrain decompression, and no patient suffered hydrocephalus-related complications at the time of decompression. All patients had improvement of facial and upper limb symptoms; however, 4 patients continue to have intermittent upper limb sensory disturbance.

Procedural Complications

In 1 patient a small extradural hematoma was demonstrated on postoperative imaging after ETV. This lesion was asymptomatic and thus was managed conservatively. There were no other complications related to ETV. In patients who had progressed to hindbrain decompression there were no complications as a result of raised ICP, such as CSF leakage; however, 1 patient had bilateral small subdural hygromas, which were managed conservatively and resolved.

Incidence of Syringomyelia

Six patients had a spinal cord syrinx (all cervicothoracic) on presentation. The largest syrinx extended from C-1 to T-
Chiari malformation Type I with hydrocephalus

9. The syrinx reduced in length and caliber following ETV in 4 patients, and there was complete resolution of the syrinx in 1 patient. These 5 patients had symptoms attributable to the syrinx, such as upper limb weakness or sensory changes. One patient had an asymptomatic syrinx, which remained unchanged after ETV.

Two patients (33%) with syringomyelia had persistent marked symptoms following ETV, despite radiologically demonstrated improvement in the appearance of the cavity, and thus underwent hindbrain decompression. Thereafter, symptoms improved, leaving upper limb sensory changes in 1 patient and upper limb dysesthesia in the other.

**Discussion**

The descent of the cerebellar tonsils below the foramen magnum was originally described by Chiari in 1891; however, the origin and pathophysiology of this condition remains poorly understood. In his 1891 monograph, Chiari originally surmised that hydrocephalus provided the driving force to push the cerebellar tonsils into the foramen magnum. However, overt hydrocephalus is only reported in 7–10% of patients with symptomatic CM-I. Some have asserted that CM-I is primarily a developmental anomaly of the posterior fossa with subsequent arachnoid adhesions. Therefore, any associated hydrocephalus may be an obstructive phenomenon due to fourth ventricular outflow obstruction.

Endoscopic third ventriculostomy has gained widespread acceptance as a durable alternative to ventricular shunt placement for obstructive hydrocephalus. In certain etiological groups, such as isolated aqueductal stenosis, success rates, as determined by shunt freedom, are in the order of 80–90%. Endoscopic third ventriculostomy presents an attractive alternative to ventricular shunt placement given the much lower risk of infection and long-term durability, which appear superior considering potential rates of shunt malfunction. Note, however, that late blockage of ETV stomas have been reported. The principal of a third ventriculostomy can be applied to obstructive hydrocephalus originating distal to the aqueduct; however, the reduction in the size of the prepontine subarachnoid space has been regarded as a limiting factor in these cases. Numerous isolated case reports of successful ETV in patients with CM-I have been published (Table 2), demonstrating the ability to safely form a stoma in the floor of the third ventricle despite the small size of the prepontine cistern. In addition, published cases have demonstrated success in terms of shunt freedom and symptom resolution. However, Fukuhara et al. have described CM-I as a risk factor for ETV failure, although their study included only 5 patients with CM-I. Our series of patients with CM-I represents the largest to date and reveals the efficacy of ETV in such a cohort of patients.

**TABLE 1**

*Outcome in 16 patients with hydrocephalus associated with CM-I–syringomyelia complex by symptom category*

<table>
<thead>
<tr>
<th>Symptom Category</th>
<th>No. of Patients/Total (%)</th>
<th>Symptoms Resolved w/ ETV Alone</th>
<th>Syrinx Rate (%)</th>
</tr>
</thead>
<tbody>
<tr>
<td>Elevated ICP</td>
<td></td>
<td></td>
<td></td>
</tr>
<tr>
<td>CM-I</td>
<td>8</td>
<td>7/8 (88)</td>
<td>4/8 (50)</td>
</tr>
<tr>
<td>Syringomyelia</td>
<td>5</td>
<td>5/5 (100)</td>
<td>3/5 (60)</td>
</tr>
</tbody>
</table>

*FMD = foramen magnum decompression.

**TABLE 2**

*Literature summary of cases of ETV for CM-I*

<table>
<thead>
<tr>
<th>Authors &amp; Year</th>
<th>No. of Patients</th>
<th>Presence of Syrinx</th>
<th>Success Rate (%)</th>
</tr>
</thead>
<tbody>
<tr>
<td>Metellus et al., 2002</td>
<td>1</td>
<td>yes</td>
<td>100</td>
</tr>
</tbody>
</table>
reported on a series of 22 patients with syringomyelia who were initially treated with ventriculostrial shunt placement; 20 patients experienced improvement in their symptoms. Ogilvy and Borges have described MR imaging evidence of a reduction in the size of a cervical syrinx following insertion of a VP shunt and associated symptomatic improvement. Such improvement has also been documented after ETV in isolated case reports. In the present study ETV resulted in radiologically demonstrated improvement or resolution of the syrinx in 5 patients (83%), although 2 patients (33%) required subsequent hindbrain decompression for ongoing syringomyelia symptoms.

Among our series of patients there was a low complication rate with no permanent sequelae. There were no episodes of CSF leakage, even at the time of hindbrain decompression, when one might expect CSF leakage if a ventricular system is not adequately decompressed and elevated ICP persists. Therefore, we advocate replacing ventricular shunt insertion with ETV in the management of CM-I associated with hydrocephalus. In up to 60% of cases ETV, with its very low complication rate, can provide definitive management and thus avoid unnecessary hindbrain decompression.

Conclusions

Endoscopic third ventriculostomy provides a durable form of treatment for hydrocephalus associated with CM-I. It is effective as the primary means of management, and we advocate replacing routine VP shunt insertion in patients with this combination of pathophysiology. Management of the hydrocephalus alone is often sufficient to alleviate symptoms associated with the CM-I and any associated syringomyelia can obviate subsequent decompression. Note, however, that a significant proportion of patients will still need both procedures, and complications due to elevated ICP are not seen at the time of decompression.

References


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