Hindbrain decompression for Chiari–syringomyelia complex: an outcome analysis comparing surgical techniques

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Abstract
The natural history of Chiari malformation and our ability to alter it is poorly understood, and reported results of hindbrain decompression show high recurrence rates. We report 11 years of experience of hindbrain decompression, to evaluate long-term outcome related to surgical technique. The results of patients who underwent hindbrain decompression between 1994 and 2005 were retrospectively analysed. We identified 96 patients from operative records and reviewed all clinical records for presenting symptomatology and examination findings. Decompression technique includes traditional decompression, duraplasty and bone—only decompression. Symptoms at the last available follow-up visit were defined by the assessing clinician as resolved, improved, unchanged or worse. There were 35 males and 61 females with a mean age of 33 years (range 6 – 62 years). The mean length of follow-up is 3.6 years with a range of 6 months to 9 years. Postoperative resolution or improvement in symptoms was seen in 75 patients (78%). Drop attacks and headaches were the most likely to respond to hindbrain decompression, showing improvement or resolution in 100 and 92% of cases. Dysaesthetic arm pain and weakness carried the worse prognosis with only 20% having symptom resolution. Sixteen patients had only bony decompression leaving the dura intact. In eight patients (66%) the headaches resolved following bony decompression alone. However the headaches were unchanged in 25% of cases. Dysaesthetic pain and weakness was unchanged in 60%. Restoration of CSF flow dynamics at the foramen magnum by surgical decompression does not consistently result in resolution of symptoms in all patients. Identification of predictors of successful outcome following decompression, coupled with early intervention and appropriate choice of procedure may result in improved outcomes. Although this is a retrospective study it suggests that bone only decompression should be reserved for patients with isolated headache.

Key words: Chiari malformation, foramen magnum decompression, outcome.
retrospectively analysed. We identified 96 patients from operative records and reviewed all clinical records, and pre- and postoperative MR imaging. There were 35 males and 61 females with a mean age of 33 years (range 6–62 years). There were 13 patients in our series under the age of 16 years.

Follow-up included clinical examination in the outpatient clinic and repeat MR imaging of the brain and spine. Symptoms at the last available follow-up visit were defined by the assessing clinician as resolved, improved, unchanged or worse. The mean length of follow-up is 3.6 years with a range of 6 months to 9 years.

**Radiological appearance**

Analysis of the preoperative MR images demonstrated 83 (86%) patients with Chiari 1 malformation (defined as tonsillar descent greater than 5 mm below the foramen magnum) and 13 (14%) with Chiari 2 malformation (defined as herniation of a portion of the medulla and associated dysraphism). Forty-six patients had an associated syringomyelic cavity on the preoperative MRI.

**Presenting symptomatology**

Patients presented with a varied complex of symptoms that were in order of frequency:

- headache 63 (66%);
- dysaesthetic pain or limb weakness 34 (35%);
- cranial nerve dysfunction 14 (15%);
- altered sensation 14 (15%);
- ataxia or unsteadiness 13 (14%);
- scoliosis 9 (9%);
- drop attacks 7 (7%; Table I).

Thirteen patients (13.5%) presented with classical cough induced headache as the sole symptom.

**Surgical strategy**

Patients who manifested hydrocephalus had this treated with either ventriculoperitoneal shunting or endoscopic third ventriculostomy prior to proceeding to hindbrain decompression. These patients only proceeded to decompression if they demonstrated progressive neurological deterioration. All patients underwent posterior fossa craniectomy measuring 2 × 3 cm, in continuity with the foramen magnum. Eighty-one patients had the posterior arch of C1 removed to achieve good exposure of the cerebellar tonsils, where the tonsils descended beyond C1.

In 16 patients bony decompression only was performed, leaving the dura intact, where this was felt to achieve adequate decompression. The dura was opened in the remaining 80 patients. The surgery was then tailored to the individual patient with the aim of establishing good CSF flow between the 4th ventricle and the cervical subarachnoid space. The cerebellar tonsils were stitched back or cauterized in 58 patients (72%), in the remaining 22 patients the tonsils were left intact. In those patients who underwent dural opening, 90 had the dural margin sutured up to muscle, creating an artificial pseudomeningocele. Six patients in this series had dural grafting to achieve dural expansion, based on surgeon preference. The graft material was a synthetic dural substitute (Preclude, W. L. Gore & Associates, Arizona) All patients who underwent dural opening in this series also underwent arachnoid opening.

**Complications**

The overall complication rate in our series was 27%. There was one postoperative mortality within 30 days due to sepsis. One patient (1%) had a postoperative stroke, which resolved except for a mild facial weakness. Nine patients (9.4%) developed postoperative pyogenic meningitis. Twelve patients (12.5%) had CSF leaks requiring wound resuture, antibiotics and in some cases placement of a lumbar drain. One patient subsequently required a ventriculoperitoneal shunt. Three patients (3.1%) developed superficial wound infection, which resolved with antibiotic treatment. No patients who underwent duraplasty experienced CSF leak.

**Overall outcome**

Overall, postoperative resolution or improvement in symptoms was seen in 75 patients (78%), no change was seen in 20 patients (20%) and two patients (2%) had worse symptoms postoperatively. In patients with syringomyelia (n = 46) 74% showed improvement or complete resolution of their presenting symptoms, in comparison 42 (84%) of those without syringomyelia had postoperative improvement or resolution of symptoms. Postoperative magnetic resonance imaging of the 46 patients with syringomyelia showed the cavity had collapsed in 19 following hindbrain decompression, 21 showed some reduction in calibre or length on follow-up imaging and six remained the same. No patients had progression of the syrinx postoperatively.

<table>
<thead>
<tr>
<th>Table I. Presenting symptoms</th>
<th>Number</th>
</tr>
</thead>
<tbody>
<tr>
<td>Headache</td>
<td>63</td>
</tr>
<tr>
<td>Dysaesthetic pain or weakness</td>
<td>34</td>
</tr>
<tr>
<td>Ataxia or unsteadiness</td>
<td>13</td>
</tr>
<tr>
<td>Scoliosis</td>
<td>9</td>
</tr>
<tr>
<td>Cranial nerve dysfunction</td>
<td>14</td>
</tr>
<tr>
<td>Drop attacks</td>
<td>7</td>
</tr>
<tr>
<td>Altered sensation</td>
<td>14</td>
</tr>
</tbody>
</table>
Outcome of presenting symptomatology

On analysis of the presenting symptoms, we found drop attacks and headaches were the most likely to respond to hindbrain decompression, showing improvement or resolution in 100 and 92% of cases, respectively (Table II). Dysaesthetic arm pain and weakness carried the worse prognosis with only 20% having symptom resolution and 23% experiencing improvement during follow-up. 3% of patients with dysaesthetic arm pain had worse symptoms postoperatively.

Outcome of scoliosis

In patients presenting with scoliosis, three were males and six females with a mean age of 16 years (range 7 – 33 years). In nine patients presenting with scoliosis postoperative MRI confirmed resolution or marked collapse of the syrinx in all patients. In one patient (11%) the scoliosis curve corrected following hindbrain decompression. In six patients (67%) no further progression of the curve was identified on follow-up and no subsequent intervention has been required. Two patients (22%) showed progression of the curvature necessitating a spinal instrumentation procedure.

Outcome of foramen magnum decompression in Chiari 2 malformation

Thirteen patients had Chiari 2 malformation with an age range of 10 – 54 years (mean 29 years). Four patients had syringomyelia and three presented with drop attacks, all of whom experienced no further attacks following decompression. The remaining 10 presented with headaches, upper limb weakness and dysaesthetic pain. Of these four remained the same and the other six patients (22%) showed progression of the curvature necessitating a spinal instrumentation procedure.

<table>
<thead>
<tr>
<th>Symptom</th>
<th>Resolved</th>
<th>Improved</th>
<th>Unchanged</th>
<th>Worse</th>
</tr>
</thead>
<tbody>
<tr>
<td>Headache</td>
<td>43 (68%)</td>
<td>15 (24%)</td>
<td>5 (8%)</td>
<td>0</td>
</tr>
<tr>
<td>(n = 63)</td>
<td></td>
<td></td>
<td></td>
<td></td>
</tr>
<tr>
<td>Altered sensation</td>
<td>6 (43%)</td>
<td>3 (21%)</td>
<td>5 (36%)</td>
<td>0</td>
</tr>
<tr>
<td>(n = 14)</td>
<td></td>
<td></td>
<td></td>
<td></td>
</tr>
<tr>
<td>Dyseaesthesia or weakness</td>
<td>7 (20%)</td>
<td>8 (23%)</td>
<td>18 (53%)</td>
<td>1 (3%)</td>
</tr>
<tr>
<td>(n = 34)</td>
<td></td>
<td></td>
<td></td>
<td></td>
</tr>
<tr>
<td>Cranial nerve dysfunction</td>
<td>8 (57%)</td>
<td>2 (14%)</td>
<td>4 (29%)</td>
<td>0</td>
</tr>
<tr>
<td>(n = 14)</td>
<td></td>
<td></td>
<td></td>
<td></td>
</tr>
<tr>
<td>Unsteadiness or ataxia</td>
<td>4 (31%)</td>
<td>8 (62%)</td>
<td>1 (7%)</td>
<td>0</td>
</tr>
<tr>
<td>(n = 13)</td>
<td></td>
<td></td>
<td></td>
<td></td>
</tr>
<tr>
<td>Drop attacks</td>
<td>7 (100%)</td>
<td>0</td>
<td>0</td>
<td>0</td>
</tr>
<tr>
<td>(n = 7)</td>
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<td></td>
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</table>

(69%) showed sustained improvement after hindbrain decompression, over a mean follow-up period of 4.6 years (range 1 – 9 years).

Outcome analysis based in surgical technique

Sixteen patients had only bony decompression leaving the dura intact. The majority of these patients had headaches (n = 12). In eight patients (66%) the headaches resolved following bony decompression alone. However, the headaches were unchanged in 25% of cases. Table III shows the outcome for all symptoms in this group. No patients in this group presented with ataxia or drop attacks, but sensory disturbances and cranial nerve dysfunction improved or resolved in all cases. Dysaesthetic pain and weakness was unchanged in 60%. One patient in this group had a spinal syrinx, which disappeared on follow-up imaging.

Fig. 1 shows the comparison of symptom resolution or improvement between the traditional dural opening procedure and bony decompression. Headaches were seen to improve more frequently in the dural opening group; however, the outcome for dysaesthesia is similar in both groups.

Six patients underwent dural grafting using synthetic graft. No patients suffered postoperative complications. Of these six patients, two had syringomyelia, in one the cavity collapsed following surgery and in the other the syrinx remains unchanged. In five patients symptoms were improved or resolved. One patient had no improvement in symptoms, but did not have progression of neurological deficit. None of these six patients have required revision surgery.

Recurrence

During the follow-up period 10 patients (10.4%) have experienced recurrence of symptoms similar to their preoperative symptomatology. Four patients had only bony decompression giving an overall recurrence rate of 25% in this group. The remaining six patients had dural opening giving a recurrence rate of 7.5%. The mean time to recurrence of symptoms is 26 months (range 6 months to 7 years). Postoperative MRI shows poor CSF flow at the foramen magnum.
in six patients and to date five have undergone revision surgery.

Discussion

There is evidence that underdevelopment of the mesodermal occipital somite is responsible for the small volume of the posterior fossa noted in patients with Chiari malformation, resulting in abnormal flow of CSF at the foramen magnum.6,7 It is, therefore, logical to extrapolate that enlargement of the dimensions of the posterior fossa and re-establishing flow of CSF at the craniocervical junction would result in improvement of symptoms. Badie et al.2 demonstrated that Chiari 1 patients with a decreased posterior fossa volume responded better to surgical decompression than those with normal posterior fossa volumes.2 However, in general, outcome is difficult to predict and is variable despite perceived adequate decompression. In our series, 78% of patients overall experienced a good outcome postoperatively (improvement or resolution of symptoms). This is comparable to previous reports of large series in the literature.8,9

The optimal surgical treatment for symptomatic Chiari malformation remains controversial. The goal of surgery is to restore normal CSF dynamics to the craniocervical junction. However, the extent of surgery required to achieve this has not been elucidated and many different operative techniques have been recommended. The establishment of good CSF flow from the 4th ventricle to the cervical subarachnoid space is the aim of hindbrain decompression. Williams described the traditional technique of creating an artificially enlarged cisterna magna to provide a reservoir for spinal pressure surges.4,10 His technique included suturing the dural edges to the divided suboccipital muscles with wide arachnoid dissection.10 Subsequently, modifications of this technique have been advocated, including duraplasty to avoid CSF leak and pseudomeningoele formation and dura-splitting (leaving the inner layer of dura and arachnoid intact) or bone only decompression to avoid CSF exposure, and subsequent arachnoid adhesion formation. The extent of decompression necessary to achieve normal CSF dynamics is variable between each patient and dependent not only on the level of tonsillar descent, the presence of a syrinx and the resulting symptomatology. Even when good CSF flow is felt to have been achieved at operation, with the extent of decompression tailored to the individual patient circumstances, a significant proportion will have no improvement in their symptoms. The use of intraoperative ultrasound to assess CSF flow following bony decompression to guide the need for progression to more invasive decompression has been reported but does not correlate with outcome.11

Bone-only decompression provides a minimally invasive method of restoring CSF circulation. In our series of 16 patients undergoing bone only decompression 74% of those with headache had resolution or improvement of symptoms and poor outcomes were seen in patients with dysesthetic pain or ataxia. This suggests patients presenting with more than simple classical Chiari type headache should undergo a formal dural opening procedure, which in our series is the majority of patients, as only 13.5% had headache as the only symptom. In addition, in our series the rate of recurrence of symptoms is significantly higher in the bony decompression group than in those undergoing dural opening.

Predicting those patients whose symptomatology will consistently improve with hindbrain decompression remains an elusive goal. McGirt et al. describe the use of cine phase contrast MRI imaging of CSF flow at the foramen magnum as a predictor of those who are likely to respond to hindbrain decompression, showing complete obstruction to CSF flow demonstrated preoperatively as an independent predictor of long term symptom resolution, regardless of the degree of tonsillar ectopia.12 In addition, they noted that patients with syringomyelia or generalized headaches were at increased risk of symptom recurrence.

There was an overall 30% complication rate in the dural opening group compared with 12.5% in the bony decompression group. This is compared with the literature which quotes a complication rate of up to 42% in patients where the dura is opened and a 10% complication rate where the dura is left intact.13–15 Indeed, postoperative pyogenic meningitis only occurred in the dural opening group without duraplasty.

We had one (1%) mortality in the immediate postoperative period. Several deaths have been reported after posterior fossa decompression for Chiari malformations, with a reported mortality rate of 3%.14,16 Many complications of hindbrain
decompression are related to CSF exposure to blood, muscle and cellular debris, resulting in adhesive arachnoiditis causing impedance to CSF flow. Whilst bone only decompression may avoid these complications, the outcome in relation to symptom improvement in our series is poor with 25% showing no change in their headaches and 60% showing no change in arm dysesthesia. Although the number of patients in the bone only group is small and, therefore, the results can be interpreted with some caution. Some authors advocate a dura-splitting approach to avoid CSF exposure but providing better decompression than removing only bone.17

In our series patients with dysesthetic pain were the least likely to improve, regardless of the surgical approach adopted. Williams noted that signs related to a syringomyelic cavity, such as muscle wasting and sensory loss seldom improve following hindbrain decompression. The longer the clinical history, the worse the prognosis. Some authors suggest that patients with symptoms lasting longer than 2 years have a worse prognosis.19 This is particularly true of syringomyelia, where irreversible neurological damage may persist after surgery even when resolution of the syrinx is seen. This may explain the poor outcome of patients with dysesthetic pain or weakness in this series. We found that posterior fossa syndromes such as headache, ataxia and drop attacks were more likely to improve following hindbrain decompression. This finding is comparable to other studies demonstrating 70 – 100% success rate in these cases.13,14,19,20

Radiological resolution or improvement in the syrinx was seen in 88% of cases. Previous studies have reported radiological and clinical improvement or stabilization of syringomyelia in 65 – 88% of cases following hindbrain decompression.15,16,21,22

With regards to scoliosis 78% of patients in our series showed resolution or stabilization of the curvature following hindbrain decompression. This is consistent with previously published literature with up to 71% of children showing improvement.13,23,24 In addition, patients with Chiari 2 malformation who are a notoriously difficult group to manage also show sustained improvement following hindbrain decompression, although a higher proportion will subsequently have symptom recurrence and require further procedures.

Conclusions

Chiari malformation is being increasingly diagnosed, but remains little understood in terms of its aetiology and symptomatology. Restoration of CSF flow dynamics at the foramen magnum by surgical decompression demonstrated both at operation and on postoperative imaging does not consistently result in resolution of symptoms in all patients. Identification of predictors of successful outcome following decompression, coupled with early intervention and appropriate choice of procedure may result in improved outcomes. Although this is a retrospective study it suggests that bone only decompression should be reserved for patients with isolated headache.

Conflict of interests

The authors have no conflict of interests to declare. CH, TP and GF devised the paper, CH, OR and HZ collected and analysed data. TP and GF act as guarantors.

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