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Treatment of the Chiari malformation with bone decompression without durotomy in children and young adults

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Abstract *Objective:* The purpose of this study was to provide more information on alternative operative approaches to the treatment of symptomatic Chiari type I and/or type II malformations in children and young adults. *Methods:* All patients were treated with craniocervical decompression without durotomy or duroplasty. The cervical decompression was carried on to the lowest level of tonsillar herniation. *Conclusions:* In 22 patients with symptomatic Chiari type I (4) and/or Chiari type II (18) malformation, there was total improvement in 12, partial improvement in 7, and no change in 3. There was no operative morbidity or mortality. Craniocervical decompression without durotomy may be a suitable treatment modality for symptomatic Chiari malformation in children and young adults.

Keywords Chiari malformation · Decompression · Durotomy · Children

Introduction

The treatment of the symptomatic Chiari malformation by surgery has been widely advocated, but the operative techniques that have been recommended are extremely diverse [3, 4, 5, 6, 7, 9, 10, 11, 12, 13, 14, 16, 18, 22, 23].

The operative interventions recommended are untethering of the distal cord [7], decompression of the cervical spine only with durotomy [14], suboccipital decompression, durotomy, and cervical laminectomy [3, 5, 7, 12], suboccipital decompression, durotomy, duroplasty, and cervical decompression [11, 13, 18], dural stripping [10], and exploration of the fourth ventricle [16]. In this paper we report a series of patients with Chiari type I and/or type II malformation who underwent a decompressive occipital craniectomy and cervical laminectomy without durotomy, as an efficient procedure to simplify the intervention and minimize operative morbidity and mortality.

Materials and methods

Patient population

Between 1989 and 1998, 22 consecutive patients with symptomatic Chiari malformation underwent operative treatment. There were 9 male and 13 female patients. The age range was 2–28 years

Table 1 Clinical findings

Signs/symptoms[a]	No. of patients[b]
Increased spasticity of lower extremities	8
Airway difficulties	6
Swallowing difficulties	5
Progressive nystagmus	5
Occipital-cervical pain ("headaches")	6
Weakness/dysesthesias of upper extremities	3
Progressive scoliosis	3
Progressive hearing difficulties	2
Neck arching	2
Tongue atrophy	1
Blurred vision	1

[a] In 22 patients with Chiari I and Chiari II malformation
[b] A given patient may have more than one of the above

(median 7 years). Fourteen patients had spina bifida and hydrocephalus and 8 did not. Chiari malformation type 1 was present in 4 patients, and type II in 18. Associated syringomyelia was present in 9 patients: 8 patients had Chiari type II and 1 had Chiari type I. All patients underwent neuroimaging by MRI prior to operative intervention. In all patients cranial neuroimaging was performed to document that the clinical deterioration was not caused by progressive hydrocephalus and/or by shunt malfunction. All patients but 1 underwent postoperative spinal MRI.

Clinical findings

Clinical deterioration was what mandated operative intervention. The clinical preoperative findings are listed in Table 1.

Chiari type I patients

There were 4 patients (2 females, 2 males) in this group. The age range at presentation was 4–16 years. One patient had syringomyelia. None of these patients had hydrocephalus. Two of the patients presented with progressive scoliosis and 1, in addition, had motor weakness and clumsiness of the upper extremities. Two presented with occipital cervical pain and arching of the neck.

Chiari type II patients

There were 18 patients (11 female, 7 male) in this group. The age range at presentation was 3–28 years. There were 14 patients with spina bifida and hydrocephalus. Eight patients presented with increased spasticity ("tightness") in the proximal lower extremities. Airway difficulties were present in 6 patients, swallowing difficulties in 5. Other findings were nystagmus (5), occipital-cervical pain ("headaches") (3), upper extremity weakness (2), hearing difficulties (2), progressive scoliosis (1), and tongue atrophy (1).

Operative treatment

The patients underwent operative intervention on the horseshoe headrest in the prone position with arterial line, venous line and Foley catheter. Wide suboccipital craniectomy and cervical laminectomy were performed to the lowest level of the hind brain – tonsillar herniation without durotomy. The cervical laminectomy was not carried out laterally. The level of the laminectomy was C1 in 3 cases, C1–2 in 16, and C1–3 in 3. All patients except for the 3 patients who had required tracheostomy preoperatively were extubated in the postanesthesia care unit immediately after the intervention. All patients were monitored in pediatric intensive care unit overnight. No blood transfusion was required for any of the patients in this series.

Results

Clinical course

The patients were followed up postoperatively for 26 months to 18 years (median: 9 years). Complete improvement of symptoms was noted in 12 patients, while 7 showed partial improvement and 3 remained unchanged.

Operative morbidity and mortality

No patients acquired postoperative CSF leakage, wound infection, pseudomeningocele, postoperative hydrocephalus, recurrent nausea, or the postoperative posterior fossa syndrome [11], cerebellar ptosis [12], or cervical instability.

There was no operative mortality. However, on follow-up 2 patients with Chiari type II malformation were found to have died, 1 of them 2 years and the other 3 years after surgery. These were 2 patients who had undergone tracheostomy preoperatively because of the severity of breathing and swallowing difficulties. Each had maintained the tracheostomy until death (accidental disconnection from the ventilator in 1 and suspected seizure and respiratory arrest in the other).

Operative interventions

The time needed for the operative interventions (incision to closure) was 105–155 min (median 130 min).

Length of hospital stay

The length of hospital stay for the 19 patients without tracheostomy was 3–8 days (median 4.0 days). The 3 patients with tracheostomy had a longer stay related to their airway management.

Long-term follow-up

Twenty patients did not have any recurrence of symptomatology in the follow-up period of this study (26 months to 18 years, median 9 years). The remaining 2 patients were the preoperative tracheostomy patients previously described.

Associated syringomyelia

There were 9 patients who had associated syringomyelia. One patient underwent simultaneous occipital and cervical decompression with a syrinx–peritoneal shunt. Those that had an extensive and/or progressive syringomyelia on follow-up MRI (performed 3–6 months following operation) underwent placement of a syrinx–peritoneal shunt [1, 2]. This was required in 3 patients.

In the single patient with Chiari type I with syringomyelia there was no progression after decompression; no shunt procedure has been necessary, and the clinical findings all improved (motor weakness and scoliosis). The 3 patients who underwent syrinx–peritoneal shunting had Chiari type II malformation. They had no complications from the procedure. None of these patients have required, to date, a shunt revision. Of the 4 remaining patients, 3 had no resolution or progression of the syringomyelia and have not undergone further operative interventions, and 1 has yet to have postoperative neuroimaging.

Clinical course and type of Chiari malformation

Chiari type I patients

The 4 patients in this group had an improvement of clinical symptoms (occipital-cervical pain and arching of the neck, progressive weakness and clumsiness of the upper extremities). Of the patients with scoliosis, 1 underwent a spinal fusion 3 months after occipital-cervical decompression, because of the extent of preoperative deformity. The other patient with scoliosis stabilized and there has not been any progression.

Chiari type II patients

There were 18 patients in this group. Some symptoms and clinical findings improved, while others remained unchanged; none were made worse. The 3 patients with occipital-cervical pain (headaches), the 2 with upper extremity weakness, and the 8 with increased spasticity (tightness) in the proximal lower extremities experienced improvement of all of these. Swallowing difficulties improved in 3 patients, remaining unchanged in 2. Breathing difficulties (upper airway) improved in 4 and remained unchanged in 2. In this group, 1 of the 3 patients with tracheostomy prior to decompression improved and was decannulated, but the other 2 were not.

Of the 5 patients with coarse nystagmus, there was some improvement in 3 with residual milder nystagmus. In 2 patients, there was no improvement. The subjective hearing difficulties improved in 1 of the 2 patients with this finding. The single patient with tongue atrophy did not experience any improvement. The 2 patients who subsequently expired were Chiari type II patients (see section: Operative morbidity and mortality).

Discussion

The need for treatment of the symptomatic Chiari malformation has been the subject of extensive review. Whereas infants who present with significant symptoms of the Chiari malformation at birth may not benefit from operative intervention owing to malformation and absence of cranial nuclei [8], there is abundant literature to substantiate that the older child with clinical progression may benefit from surgery. This is felt to be due to a continual change in the relationship of the hindbrain to the cranial base, as a consequence of each individual child's cranial base, cervical canal and CSF dynamics [17]. As the ongoing changes create direct compression of the hindbrain, symptoms present and progress [17].

The operative interventions advocated have varied according to the surgeons' choice and the presence or absence of syringomyelia/hydromyelia. Some advocate for cervical decompression without any decompression of the posterior fossa [9, 14]. With this a cervical durotomy to the level of the foramen magnum has also been recommended [9, 14]. Other variations that have been implemented are suboccipital and cervical bone decompression with durotomy [13, 17], limited occipital decompression with durotomy and C1 laminectomy [12], lysis of arachnoidal adhesions when present, exploration of the fourth ventricle [16, 21], and plugging of the obex if hydromyelia is present [9, 14, 21]. Other authors have advocated that in addition to occipital craniectomy, C1 laminectomy, durotomy and duroplasty should also be performed [13], that durotomy should be carried out without duroplasty [12], and that coagulation of the cerebellar tonsils should also be performed [13, 22]. Some advise that the information obtained from the neuroimaging may be useful in selection of the operative intervention to be performed [16, 21]. Others suggest that the age at the time of presentation may be the most useful indicator of what type of operative intervention should be done [9, 13, 16]. The importance of duroplasty following the bone decompression, for adequate expansion of the posterior fossa, has been emphasized by others [11, 18]. It has also been suggested that the extent of the spinal subarachnoid space (by spinal ultrasonography at the time of craniectomy) may be of importance [13]. However, this approach was recommended only for infants [13]. "Dural thinning" to permit the dura to stretch and expand the posterior fossa following the bone decompression, has also been recommended [11]. The postoperative results in terms of neurological improvement reported in the literature vary extensively. Some authors report 52% long-term improvement [14], while others report 90% or more [9, 11, 13, 18, 21].

Limited information is available on the length of hospital stay after surgery, duration of the operative intervention, length of the postoperative intubation, neurological complications, CSF fistula, pseudomeningocele formation, infection, and postoperative hydrocephalus in the operative treatment of Chiari malformation. This is probably because such issues are expected to be a part of such a complicated disease process.

The lack of scientific evidence that might indicate the correct selection of the type of operative intervention for each patient is further compounded by the lack of consistent long-term outcome in patients who do undergo an operation.

The decision to perform the suboccipital and cervical decompressive procedure without durotomy was based on the fact that the dura of children and adolescents retains a certain degree of elasticity that should allow for the hindbrain to be properly decompressed without durotomy. In addition, significant arachnoiditis may be generated by opening the dura and exploring the area. This in turn could lead to subsequent sclerosis of the subarachnoid space and subsequent scarring, and recurrence of symptoms may occur following the use of Silastic dural substitutes [2]. This was seen in 1 of our patients, who had to be treated for recurrent symptoms of deteriorating Chiari malformation. Ten years earlier, he had had a cervical decompression, durotomy, and Silastic duroplasty. At operation, upon removal of the Silastic graft, we encountered severe and extensive sclerosis of the subarachnoid space.

It may be argued that in patients with associated syringomyelia decompression without durotomy may be an inadequate procedure, and that a more extensive operation may avoid additional progression and/or a syringoperitoneal shunt such as was performed in some of the patients in this series [12]. In the series of Krieger et al. [12], 23 of the 26 patients had a resolution or a reduction in the size of the syrinx. It must be noted that despite the durotomy, 2 patients required a shunt procedure to treat the syringomyelia [12]. Since syringomyelia may spontaneously resolve despite no operative intervention [19, 20], the "correct" way to address persistent syringomyelia needs to be further studied in a larger series of patients.

Conclusion

We suggest that occipital craniectomy, with cervical laminectomy to the lowest level of tonsillar herniation and without durotomy, should be considered an alternative procedure of choice in pediatric patients and young adults with symptoms attributable to the Chiari malformation. A larger series is needed to document this preliminary study.

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