

Our experience in surgical treatment of Chiari Type 1 malformations

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Abstract

Introduction: There are four types of Chiari malformations described in the literature. Due to the fact that it is a common finding in the general population (true Chiari being present in 0,75% of the population), Chiari 1 malformation was also called Chiari anomaly.

Material and methods: In the last 5 years 17 patients with Chiari I malformation have been treated in our clinic. There were 12 women and 5 men. The mean age was 43 years (between 21 and 60 years). The symptoms were grouped in 6 syndromes: brain stem and bulbar palsy syndrome, cerebellar syndrome, central cord syndrome, paroxysmal intracranial hypertension, scoliosis, pyramidal syndrome.

Surgical treatment: According to recent literature patients respond best when operated within 2 years from the onset of symptoms. We recommend early surgery for symptomatic patients. Surgical treatment of Chiari I malformation should accomplish several goals. First of all, there is the obvious need to decompress the lower part of the cerebellum. Chiari I malformation being related to a small posterior fossa, the surgical treatment should realise enlargement of the total volume of the posterior fossa(7, 12). In the author's opinion the key point in surgical

treatment of Chiari 1 malformation should be to reestablish the CSF flow at the level of the foramen of Magendie and foramen magnum. Various approaches were used in the last five years in our clinic for the treatment of symptomatic patients: only osseous decompression in 2 patients; osseous decompression with dural grafting and intradural dissection of adhesions in 4 patients; osseous decompression with dural grafting, intradural dissection, and tonsillar coagulation in 11 patients

Results: The long-term (6 months postoperative) surgery-related result was considered excellent if symptoms resolved (7 patients). The result was considered good if the patient experienced significant improvement but also residual symptoms (8 patients). A poor result indicated no change in symptoms (2 patients). As for the surgical technique used, the vast majority of patients with good or excellent outcome at 6 months underwent osseous decompression with dural grafting, intradural dissection, and tonsillar coagulation

Conclusions: Regarding Chiari I malformations, the author considers that a proper patient selection is critical to prevent unnecessary procedures and maximize the outcome. In light of this study results and recent literature, the author considers that the surgical gold standard consists in three

key steps: posterior fossa craniectomy followed by durotomy and subarachnoid decompression of CSF flow and last duroplasty.

Key words: small posterior fossa, osseous decompression, dural graft, syringomyelia

Introduction

Chiari malformations are perhaps one of the most controversial topics in neurosurgery today. There is a lack of agreement as to what defines these malformations, their symptoms and their natural history. If treatment is necessary, a wide variety of techniques have been proposed (9, 12). There are four types of Chiari malformations described in the literature. Due to the fact that it is a common finding in the general population (true Chiari being present in 0.75% of the population), Chiari 1 malformation was also called Chiari anomaly while Chiari 2 malformation is usually associated with myelomeningocele and is found only in this population (23, 29). Chiari 3 and 4 malformations are extremely rare pathologies and are usually fatal until the age of two.

Epidemiology

In a large series of patients, the mean age of onset of Chiari I malformation is reported to be 25 years, and women account for 75% of patients. Average duration of symptoms clearly related to Chiari I is 3.1 years (range 1 month – 20 years)(9).

Material and methods

In the last 5 years 17 patients with Chiari I malformation have been treated in our clinic. There were 12 women and 5 men.

The mean age was 43 years (between 21 and 60 years).

The symptoms were grouped in 6 syndromes:

1. Brain stem and bulbar palsy syndrome: caused by brainstem compression or syringobulbia ,including variable involvement of the cranial nerves, lower brain stem.

Symptoms include “down-beating” nystagmus (1 patient), sleep apnea (2 patients), vocal cord dismotility (2 patients), extraocular eye movement abnormality (2 patients), hearing loss (2 patients), tinnitus (2 patients), dysphagia (5 patients) , headache (9 patients), neck pain (6 patients).

2. Cerebellar syndrome:

Ataxia of the limbs or trunk (3 patients), nystagmus (2 patients), dizziness (6 patients) and dysarthria (3 patients)

3. Central cord syndrome: caused by canal compression or syringomyelia, including pain (frequently “ burning”) (7 patients), dissociated and posterior column sensory loss (12 patients).

4. Paroxysmal intracranial hypertension: exertional headache and nausea, vomiting, and dizziness associated with a headache episode (in 2 patients)

5. Pyramidal syndrome: stiffness and/or spasticity and hyperreflexia (10 patients), motor weakness and variable long tract signs (8 patients), atrophy (3 patients).

6. Scoliosis (3 patients).

Mri evaluation

Magnetic resonance imaging of the cranio-spinal junction represents the gold standard diagnostic tool. A descend of tonsils of 5 mm or more below the foramen magnum is considered a pathological

tonsillar ectopia (18, 20), but this is still under debate (figure 1).

In our study we classified the tonsils ectopia into two major groups: tonsils descended to C-1 (12 patients) and tonsils descended to C-2 or lower (5 patients). We also realised head CT scans which revealed osseous anomalies such as small posterior fossa in 3 patients and platybasia in one patient. We have also studied the presence of syrinx associated with Chiari type 1 malformation (figure 2).

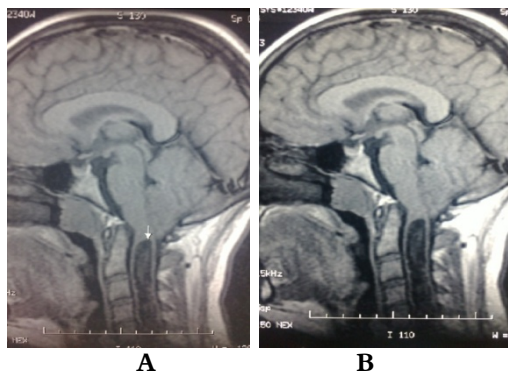


Figure 1 A: Sagittal T1-weighted MR image of the brain showing herniation of cerebellar tonsils and a low-lying obex characteristic of Chiari malformation Type I. **B:** Sagittal MR images of the cervicothoracicspine revealing syringomyelia



Figure 2 T2-weighted MR images of the spinal cord showing a holocord syrinx

Indications for surgery

According to recent literature patients respond best when operated within 2 years from the onset of symptoms. We recommend early surgery for symptomatic patients. Asymptomatic patients may be followed and operated upon if and when they become symptomatic (6, 8, 13).

For patients who have been symptomatic and stable for years, we consider that observation is the correct management, surgery being indicated only if signs of deterioration appear.

There are several strong indications for posterior fossa decompression such as drop attacks, dysphagia with aspiration, apnea and the presence of syrinx (1, 2).

Headache represents a controversial indication for surgery when it is the only symptom. We recommend using imagistic studies such as Cine MRI to establish CSF flow impairment at the foramen magnum. If positive, surgery should be considered (4).

Objectives of surgery

In the author’s opinion, surgical treatment of Chiari I malformation should accomplish several goals. First of all, there is the obvious need to decompress the lower part of the cerebellum. Chiari I malformation being related to a small posterior fossa, the surgical treatment should realise enlargement of the total volume of the posterior fossa (7, 12).

In the author’s opinion the key point in surgery treatment of Chiari 1 malformation should be to reestablish the CSF flow at the level of the foramen of Magendie and foramen magnum.

Although numerous techniques of surgery have been proposed since Chiari first described the pathology, all of them have been reported with advantages and

disadvantages, and none of them managed to fully accomplish the desired goals of surgery.

Techniques of surgery

Various approaches have been used for the treatment of symptomatic patients in our clinic in the last five years: only osseous decompression in 2 patients; osseous decompression with dural grafting and intradural dissection of adhesions in 4 patients; osseous decompression with dural grafting, intradural dissection, and tonsillar coagulation in 11 patients. We perform a more aggressive decompression and intradural dissection when a syrinx is present.

I. Osseus decompression (cranio-vertebral decompression/ laminectomy)

First of all, the craniectomy should not be larger than 3x3 cm in order to avoid cerebellar sag (11, 16, 24). The author recommends C1 laminectomy in most cases, but extension to C2 laminectomy is also possible this being directly related to caudal displacement of the cerebellar tonsils (figure 3).

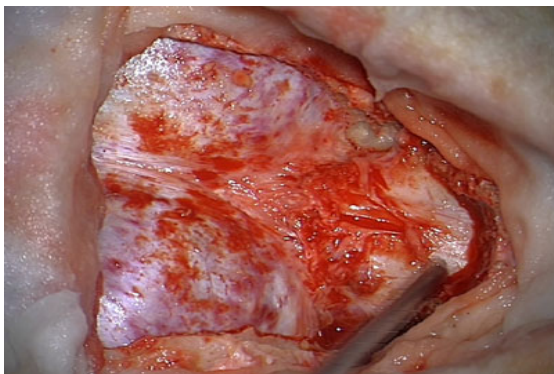


Figure 3 Suboccipital craniectomy and C1 laminectomy

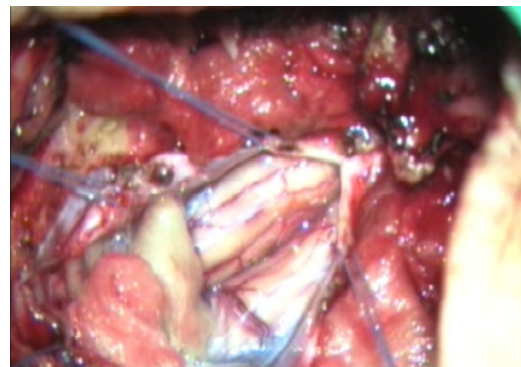
II. Opening of the dura + intra-arachnoid dissection

Opening of the dura and intracranial

dissection of the scarring are, in the author's opinion, the essential steps in Chiari I surgery. The author strongly recommends them because osseous decompression only, does not achieve one of the main objectives of surgery, increasing the global volume of the posterior fossa. The dura is opened through a Y shaped incision (figure 4 A). The next step is represented by intracranial dissection of the arachnoid scarring, reestablishing the normal CSF flow.

Visualization of the choroid plexus of the 4th ventricle and free flow of CSF into the subarachnoid space consist in our opinion the proof of adequate decompression (figure 4 B). The author prefers not to aggressively resect the herniated tonsils; instead he realises a controlled and intended tonsillar ischaemia with bipolar coagulation of PICA tonsillar branches. The author also tries to maintain the flow of the CSF through the foramen of Magendie, by lateral suspension of the medial part of the tonsils (figure 4 C).

The author prefers to reserve syringomyelic drainage for patients who fail to respond (clinically, not radiologically) to initial posterior fossa decompression. Avoiding the risks and complications associated with shunting procedures is desirable.



A

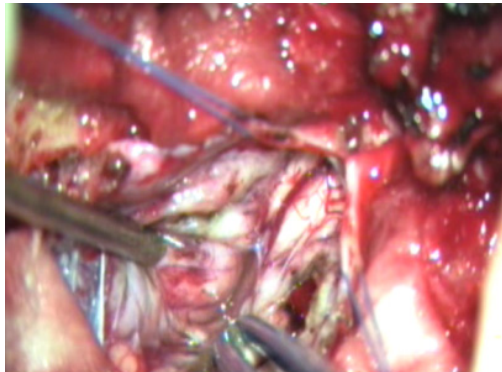
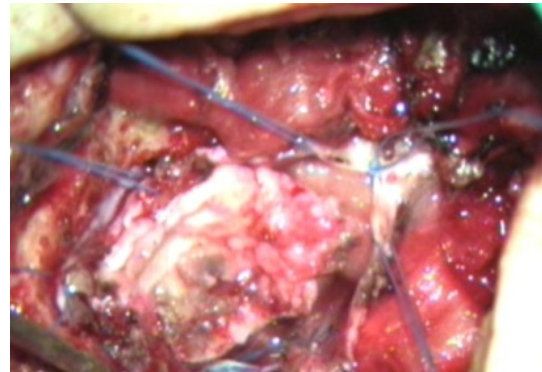
**B****C**

Figure 4 **A** Y shaped durotomy; **B** Internal debulking and coagulation of tonsils; **C** Lateral suspension of the medial part of the tonsils

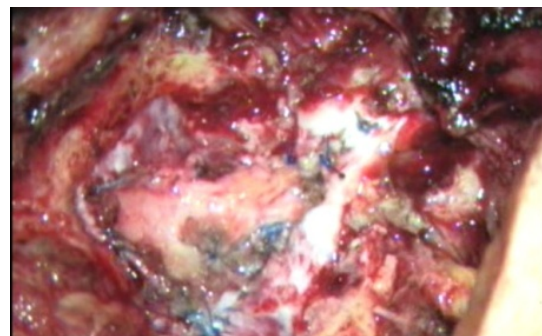
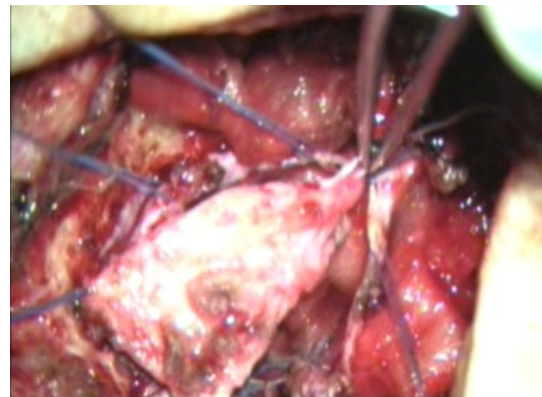


Figure 5 Duroplasty with pericranium

III. Dural graft

Dura mater being inextensible, the author considers that in order to achieve one of the goals of treatment (enlargement of the posterior fossa), it needs to realise a dural graft. The author uses autologous graft – pericranium – in order to achieve a tight closure of the dura without CSF leak (figure 5).

Tight closure of the dura is mandatory in our opinion because it prevents blood leakage from the extradural to the intradural space thus preventing subdural/intra-arachnoid scarring. Also, it prevents CSF leakage and pseudomeningocele (30, 33).

Typical postoperative care involved the patient staying overnight in an intensive care unit. Patients were discharged from the hospital between 4 and 10 days postoperatively.

Results

Postoperatively, the condition of the patients was reassessed at the follow-up visits (1 month, 3 months, 6 months, 1 year) according to: symptoms resolution; signs and symptoms improvement; no change; signs and symptoms worsening.

Those associated with poor outcome include, signs or symptoms suggestive of syringohydromyelia; atrophy suggest a

similarly poor prognosis, whereas weakness in the presence of atrophy is bad response to surgery, as might be expected with the loss of alpha motor neurons. Sensory loss has been widely recognized as largely unresponsive to surgery (figure 6).

Headache specifically, sleep apnea, and cervical pain in general appear to respond

best. Weakness in the absence of atrophy tends to respond well, while mild scoliosis, seems to respond reasonably well to surgery (figure 7).

Patients with signs of cerebellar syndrome and paroxysmal intracranial hypertension respond best to surgical management, followed by patients with pyramidal and brain stem syndromes.

Postoperative MR imaging assessment of the operative site in all patients demonstrates that the level of cerebellar tonsils was normal, the cisternal space at the level of the foramen magnum was indeed enlarged, and the width of CSF column around the spinal cord was enlarged. Postoperative spinal MR images obtained to assess the syringomyelia revealed the disappearance of the syrinx in 5 cases, and a significant decrease in syrinx size in 7 patients (figure 8).

The long-term (6 months postoperative) surgery-related result was considered excellent if symptoms resolved (7 patients).

Figure 6 Poor outcome symptoms and signs

Figure 7 Good outcome signs and symptoms

Figure 8 Preoperative/postoperative MRI showing the reduction of syrinx

The result was considered good if the patient experienced significant improvement but also residual symptoms (8 patients). A poor result indicated no change in symptoms (2 patients).

As for the surgery technique used, the vast majority of patients with good or excellent outcome at 6 months underwent

osseous decompression with dural grafting, intradural dissection, and tonsillar coagulation (Figure 9).

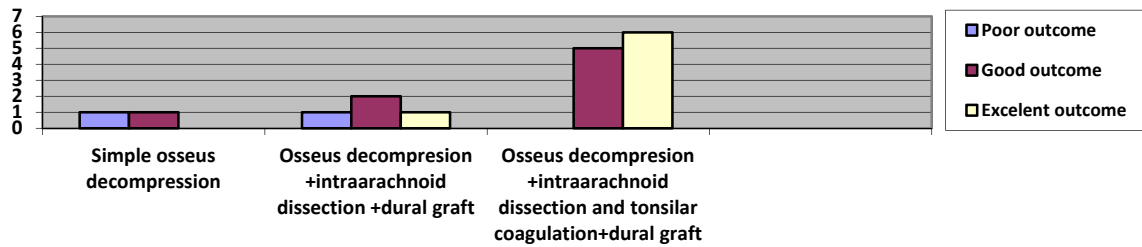


Figure 9 Long-term outcome in relation to the surgery technique used

Complications

There are several types of complications. We distinguish between complications related to conservative surgery (inadequate surgery) and actual postoperative complications (9, 11).

In our study the complications were mostly postoperative complications and included CSF leakage (1 patient), aseptic meningitis (2 patients), intraoperative changes in cardiovascular parameters (1 patient), and pseudomeningocele (1 patient).

Postsurgical moderate headaches, neck pain and fever represent the so-called "aseptic" meningitis and were regarded as adverse events rather than outright complications. Analysis of a CSF sample did not reveal evidence of infection and were successfully treated with steroid and analgesic medications. The symptoms resolved within a postoperative period no longer than 3 weeks and had no influence on the final treatment result.

The CSF leak required wound revision and lumbar drainage.

At later follow up, a pseudomeningocele was clinically and radiologically

demonstrated in one patient. No treatment was required.

We had one recurrence due to conservative surgery (incomplete decompression). In this case only an extradural decompression was realised, so we performed dural grafting, intradural dissection, and tonsillar manipulation and coagulation on the second surgery.

Discussion

In our clinic's series, surgical treatment of Chiari I malformation consisted of suboccipital craniectomy only (2 cases), arachnoid dissection of scarring (4 patients), reestablishing the CSF flow at the foramen of Magendie by shrinkage of the tonsils and duraplasty with autologous material (pericranium) (11 patients).

The most important question that remains to be answered is whether good results could be achieved by leaving the arachnoid or dura intact. According to some authors, leaving the dura and the arachnoid intact can result in lower complication rates, but this data is counterbalanced by high rates of recurrence of symptoms and the need for revision surgeries. Also, leaving the dura and the arachnoid intact results in

lower rates of syrinx reduction (7, 14, 22).

The outcome data resulted from McGirt et al study (21) (which used a simple decompressive craniectomy in 116 cases or craniectomy+duraplasty without arachnoid dissection in 140 cases) was significantly worse than the data provided by Kleklamp et al (14) in their study (craniectomy, opening the arachnoid, opening the foramen of Magendie and duraplasty in 371 cases). In our clinic's series one of the two patients who underwent a simple suboccipital craniectomy experienced recurrence of symptoms, and needed a second surgery for intrarachnoid dissection and tonsils coagulation with duraplasty. Given the low morbidity rates of these studies and the much better long term results in the literature for surgery techniques that involve at least duraplasty, the autor does not support decompression without duraplasty (2, 14, 22, 24).

As for arachnoid opening and dissection there is also no unanimous protocol. The lower morbidity and complication rates reported by some authors are in favor of leaving the arachnoid intact (21). In his study, Kleklamp revealed a correlation between arachnoid scarring and preoperative clinical symptoms and neurological outcome, and recommended opening the arachnoid in every decompression in order to restore CSF pathways (27). In light of these studies, the author also recommends opening the arachnoid and dissection of scarring in order reestablish the CSF flow at the level of foramen of Magendie.

Because of high rates of morbidity related to syrinx drainage, the author does not recommend this maneuver as first choice (2, 8, 27).

Conclusions

Regarding Chiari I malformations, the author considers that a proper patient selection is critical to prevent unnecessary procedures and maximize outcomes. The author believes that preoperative longtime neurological deficit is a predictor of poorer outcome, making early surgery, mandatory. The key point of surgery in Chiari I malformation is to allow a CSF flow at the level of foramen of Magendie.

In light of this study results and recent literature, the author considers that the surgical gold standard consists of three key steps: posterior fossa craniectomy followed by durotomy and subarachnoid decompression of CSF flow and last, duroplasty.

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