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Transverse Microincisions of the Outer Layer of the Dura mater Combined with Foramen Magnum Decompression as Treatment for Syringomyelia with Chiari I Malformation

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Summary

Numerous surgical procedures have been proposed for treatment of syringomyelia associated with Chiari I malformation, but the optimal treatment has not yet been uniformly standardised. The main aim of the surgical treatment of syringomyelia/Chiari I complex is directed toward restoration of physiological cerebrospinal fluid dynamic at the craniovertebral junction. We report the surgical results of eight patients, affected by syringomyelia and Chiari I malformation, age range from 18 to 62 years, treated by bony foramen magnum decompression combined with transverse microincisions of the outer layer of the dura mater. In an average postoperative follow-up period of two years neurological symptoms and signs improved in seven patients. Postoperative Magnetic Resonance showed a decrease in size of the syrinx in seven patients. These results suggest that foramen magnum decompression combined with transverse microincisions of the outer layer of the dura 1) is an effective and safe treatment option for syringomyelia and Chiari I malformation, 2) corrects the circulatory disturbances of cerebrospinal fluid dynamic, 3) leads to a decrease in size of the syrinx and to a significant improvement in neurological signs and symptoms, 4) avoids complications of intradural approaches and syringosubarachnoid shunting.

Keywords: Chiari I malformation; foramen magnum decompression; syringomyelia.

Introduction

Syringomyelia, a syndrome of craniospinal pressure dissociation and disturbance of cerebrospinal flow [20], is frequently associated with Arnold-Chiari I malformation [2, 25]. In the literature different pathomechanisms have been proposed for explain this association. It has been postulated that obstruction of the outlets of the fourth ventricle, with diversion of the CSF pulse wave into the central canal [8, 9], or of the foramen magnum may causes a pressure gradient between the intracranial and intraspinal compartments drawing fluid from the fourth ventricle into the central canal [33, 34].

Numerous surgical procedures have been advocated for curing syringomyelia associated with Chirari I malformation and there is still considerable controversy about the optimal method of surgical treatment. Foramen magnum decompression with or without obex plugging [4–6, 8, 10, 12, 18, 23, 26] syringosubarachnoid shunting [5, 10, 25, 27, 29], syringoperitoneal shunting [3, 5, 22, 28], syringopleural shunting [35], thecoperitoneal shunting [30], and syringocisternostomy [19] are the most frequent surgical procedures generally adopted. In 1993 Isu *et al.* proposed an extradural approach by enlarging the cisterna magna through foramen magnum decompression combined with removal of the outer layer of the dura. This treatment option was characterised by a significant recovery of pre-operative neurological symptoms and signs combined with avoidance of post-operative complications [11].

At the Neurosurgical Clinic, University of Messina, Italy, we have performed in eight patients a more conservative surgical treatment for syringomyelia and Chiari I malformation through an extradural approach performed by oblique transverse microincisions of the outer layer of the dura mater combined with foramen magnum decompression. The aim of this report is to describe this surgical option, in the light of the more recent literature upon the theories of syringomyelia associated with Chiari I malformation origin and the surgical procedures advocated in the past.

Cases

Eight patients with syringomyelia associated with Chiari I malformation were surgically treated. The patients were six men and two women, ranging in age from 18 to 62 years (mean 42 yr.).

Five of the eight patients presented with paresthaesias, hyporeflexia and weakness and numbness in the upper extremities. The average duration of symptoms was 5 years. In these patients, on admission, neurological examination revealed muscular atrophy of the upper extremities, sensory loss of the dissociated type and spastic paraparesis; moreover in two cases ataxo-spastic gait and sphincteric disturbances were present. The remaining three patients were affected by loss of pain and temperature sensation, associated with progressive weakness of the upper limbs; seven patients presented also neck pain radiating to the shoulder and the arm, exacerbated by coughing and exertion (Fig. 1). The youngest patient was affected by von Recklinghausen's disease too.

The diagnosis of syringomyelia was made when a low-signal intensity area within the spinal cord was observed on T1-weighted Magnetic Resonance Imaging (MRI). The position of the cerebellar tonsils was located between the foramen magnum and the C1 level in all eight patients, thus suggesting the concomitant occurrence of Chiari I malformation (Figs. 2 and 3). T2-weighted gradient echo images showed a narrowed cisterna magna in all patients. The syrinx was visualised from the cervical to the thoracic level in five patients, only at cervical level in the remaining three cases.

Surgical Technique

The operations were performed in all patients in the prone position, with a skin incision extending from the external occipital protuberance to the spinous process of C3. A suboccipital craniectomy with an opening of the foramen magnum and a removal of the arch of C1 and a C2 laminectomy was performed. In the suboccipital craniectomy the occipital bone was removed far laterally and anteriorly along the foramen magnum. Exposure of the dura mater of the cervico-occipital junction, combined with careful preservation of dural vascularization, was made. When a complete exposure of the dura mater had been achieved, three microincisions were made

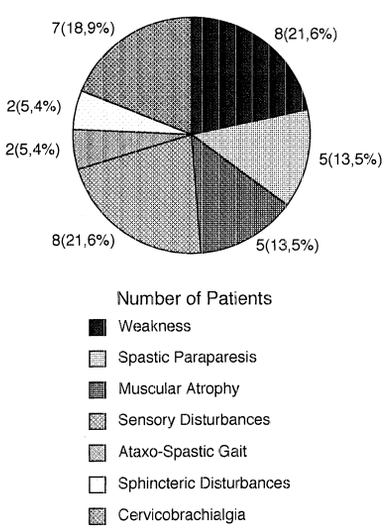

Fig. 1. Pre-operative neurological signs. Presenting signs in 8 patients with syringomyelia/Chiari I complex

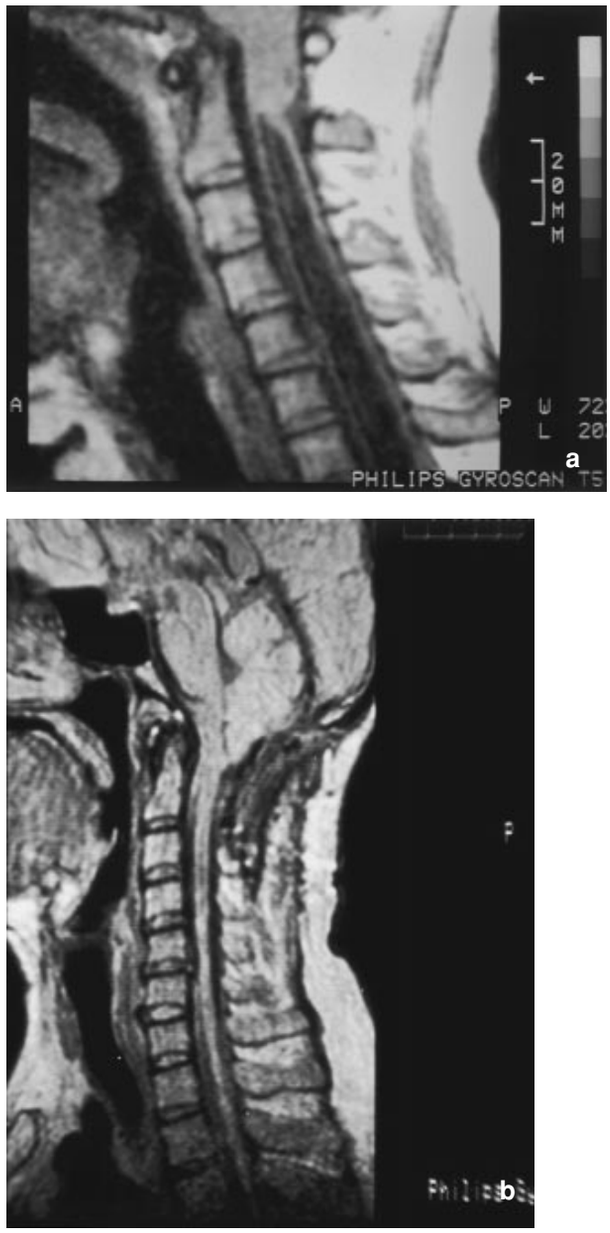

Fig. 2. (a) Pre-operative T1 weighted MRI scan in a 45-year-old man showing a syringomyelia cavity extending from C1 to T1 and a Chiari I malformation. (b) Postoperative MRI scan demonstrating significant reduction of the syrinx cavity

in the external layer of the dura leaving intact the inner layers on each side. The microincisions converge in their caudal direction and progressively decrease in length. The microincisions on the right side begin 4 mm more cranially than on the contralateral side and cross the midline for 1.5 mm without joining. The first two microincisions, 6 mm long, are performed on the occpital dura reaching the midline at the level of the foramen magnum; the second two microincisions, 4.5 mm long are made at the level of the posterior arch of the atlas; the remaining two microincisions, about 4 mm long, are performed at the level of the cranial part of the spinous

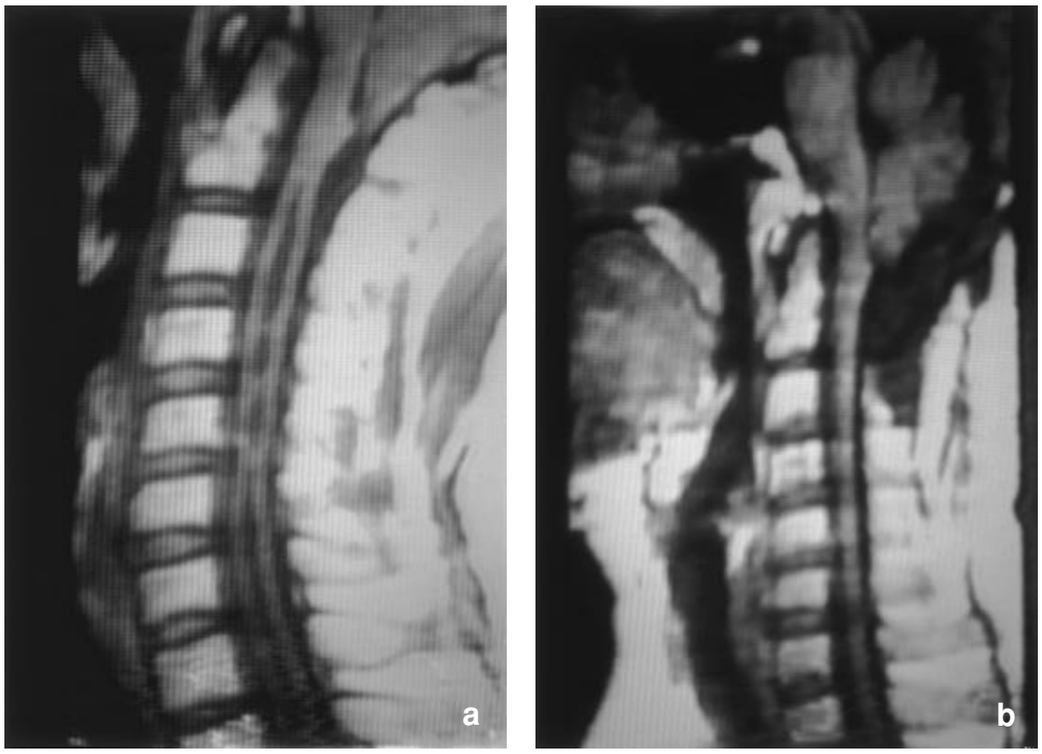

Fig. 3. (a) Pre-operative T1-weighted MRI scan in a 60-year-old man. It is visible the presence of a large syringomyelia cavity extending from C1 to C7. (b) Postoperative MRI scan one year from surgical treatment demonstrating the decrease in size of the syrinx

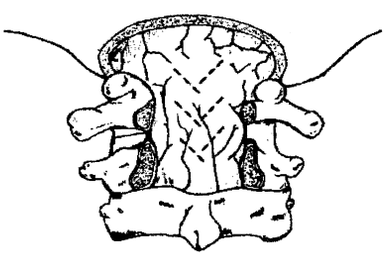

Fig. 4. Illustration showing the six oblique transverse microincisions in the outer layer of the dura mater after suboccipital craniectomy and removal of the arch of C1 and C2 laminectomy

process of C2 (Fig. 4). We used Beaver mini-blades (Mod. 6700, R Beaver Inc., Waltham, MA 02154, USA) the angular arm of which is sharp only in its inner upper side, allowing easy dural splitting with reduced risk of inner dural layer laceration. Careful avoidance of accidental arachnoidal lining laceration is essential in order to prevent the risk of extradural fluid collections. The microincisions were oriented thus in order to be able to observe the occurrence of a visible bulging of the inner layer of the dura together with significant return of pulsation of the cerebellar tonsils without any damage to the dura mater itself and of its structural capacities.

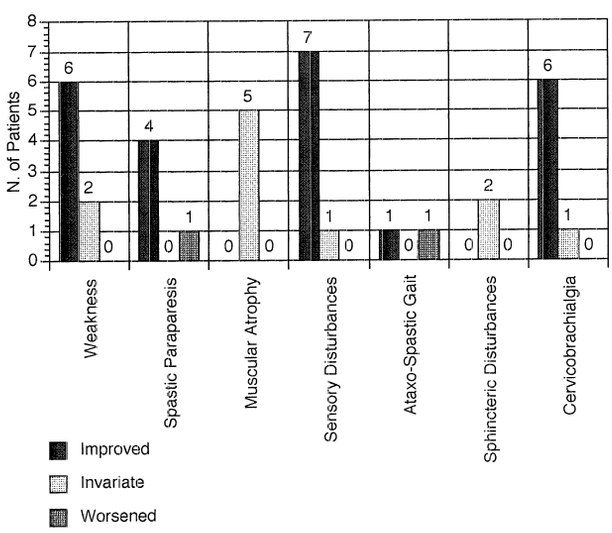

Fig. 5. Post-operative neurological signs. Long-term outcome in 8 patients with syringomyelia/Chiari I complex two years after transverse microincisions of the outer layer of the dura mater

Results

The postoperative course was satisfactory in seven patients. Neurological symptoms and signs markedly improved in seven of eight patients within the first few days (88%) (Fig. 5). Cervical pain improved within one week after surgery in seven patients and, after discharge, between the second and the third week also the weakness of the upper extremities was reduced in seven patients. Within one month in

7 of the 8 patients sensory deficit had significantly decreased.

Postoperative MRI was performed in all patients within one month after surgery. In seven patients MRI showed a significant reduction in syrinx size and T2 weighted MRI scans demonstrated an expanded cisterna magna (Figs. 2 b and 3 b). The average postoperative follow-up period was two years. All treated patients experienced long-term subjective improvement in sensory and motor deficit, together with moderate amelioration of neurological signs. Only one patient showed further progression of deficit during the follow-up period.

Discussion

The pathomechanisms underlying origin, maintenance and propagation of syringomyelia associated with Chiari I malformation are still controversially reported in the literature.

As far as 1965 Gardner *et al.* proposed a "hydrodynamic theory" based upon the existence of a communication between the fourth ventricle and the syrinx caused by a delayed and incomplete embryonic opening of the outlet of the fourth ventricle. This outflow obstruction may cause, during the systolic period, a considerable increase of the cerebro-spinal fluid (CSF) pressure. A pulsatory "water-hammer" effect can be transmitted through the obex to the central canal, causing development and progression of a cord cavitation [7–9]. More recently Williams suggested that foramen magnum obstruction may produce a dissociation pressure between the cranial and the spinal CSF compartments which "sucks" fluid from the fourth ventricle into the central canal [33, 36]. Epidural venous pressure increase may cause itself a pressure wave ascending to the spinal axis and acting on the spinal cord externally. These forces expand the syrinx by asymmetrical pressure and propel the CSF through the syringomyelia cavitation (slosh effect). Both hypothesis are based on dynamic movement of CSF through an opened channel which extends from the obex to the syrinx. However it should be considered that a direct communication between the ventricular system and the syrinx rarely occur [4, 14, 21]. Furthermore it has been demonstrated that in many cases the syrinx cavity may arise at a considerable distance below the foramen magnum and is separated from the fourth ventricle by a long segment of normal spinal cord [21, 31].

In a recent study Marin-Padilla has proved that Chiari I malformation may be caused by a mesodermal insufficiency occurring after closure of the neural folds [15, 16]. According to this hypothesis, a small posterior cranial fossa may be an essential factor in the hindbrain hernia formation. This phenomenon leads to herniation of the tonsils below the foramen magnum. Recently Oldfield *et al.* proposed the so-called "tonsillar piston theory", where the systolic brain expansion, in the presence of a foramen magnum CSF obstruction, plug the cerebellar tonsils posteriorly and downward and acting as a piston on the spinal canal. This phenomenon may be responsible for the origin and maintenance of syringomyelia by the pulsatile pressure waves, forcing CSF into the cord through the perivascular and interstitial spaces [20].

In addition it should be considered that Chiari I malformation is characterised by occurrence of frequent concomitant severe adhesions between the dura-arachnoid and neural tissue. This arachnoid scarring may cause itself clinical symptoms, which are independent from brain stem compression and syringomyelia, acting as an additional factor for impaired CSF-flow [24].

In recent years, in order to examine the pathomechanisms related to syringomyelia associated with Chiari I malformation, it was advocated the use of dynamic MRI and intra-operative ultrasonography. These tools assess the syrinx cavity and spinal cord interface together with the dynamic relationship with the cerebellar tonsils [11, 20].

Controversy surrounding the origin, maintenance and propagation of syringomyelia with Chiari I malformation underlies the development of numerous surgical procedures. The use of drains has a simple and immediate appeal and has been practised for 100 years [1]. Shunting of syringomyelic cavities is based on fluid diversion into either the subarachnoid space or into extracavitary locations (peritoneal and pleural cavities). However the shunting procedures are somewhat disadvantageous in that filling patho-mechanism may not be deactivated, and so "suck" and "slosh" effects may continue to act around the subarachnoid block.

The main aims of surgical treatment in patients with Chiari I malformation with syringomyelia are to relieve the pressure gradient existing through the subarachnoid block, to restore the subarachnoid space, to eliminate the syrinx while relieving the brain stem compression.

Different surgical approaches have been described

in the literature including foramen magnum decompression with plugging of the obex [7, 8], opening of both dura mater layers [14], posterior cranial fossa reconstruction with dural grafting [26]. However, it should be emphasised that none of these approaches are free of complications (i.e., fluid collection in the operative wound, pseudomeningocele, meningitis [4, 11, 12, 17]).

Recently, a number of publications have emerged suggesting to leave the arachnoid layers intact during decompression of the foramen magnum [13, 14]. In 1993 Isu *et al.* proposed a novel surgical approach through foramen magnum decompression with removal of the outer layer of the dura mater. This procedure was demonstrated to produce a significant correction of the circulatory disturbance of CSF, leading to a decrease in the syrinx size and marked improvement in symptoms. The intra-operative use of ultrasound allowed evaluation of the decompressive effect, revealing restoration of good pulsation of the cerebellar tonsils [11].

In eight patients affected by syringomyelia and Chiari I malformation we have performed a modification of the Isu technique where, instead of removing the outer layer of the dura mater, we accomplished a more conservative microsurgical transverse incision of the external layer of the dura mater with foramen magnum decompression.

The rationale of this modification was to obtain an extradural correction of the circulatory disturbance of CSF at the foramen magnum reducing the risk of inadvertent opening of the bulging inner dura mater layer. Although based on a limited number of patients and on a two-year follow-up evaluation, the results of this study revealed a significant decrease in the size of the syrinx, as demonstrated by the follow-up MRI examination, togheter with marked improvement of neurological signs and symptoms lasting for two years. We did not perform an intra-operative ultrasound evaluation of eventual resolution of CSF circulatory disturbances. However, it should be emphasised that the described technique is an extradural approach similar to that of Isu. Therefore, the achievement of a significant long-term neuroradiological and clinical result suggests the occurrence of a reasonable immediate restoration of good pulsation of the cerebellar tonsils together with a return of physiological CSF dynamics. These pitfalls do not allow us to draw definite conclusions in regard to this surgical approach and need further careful confirmation with a larger number of patients and longer follow-up evaluation. However, effective neuroradiological and clinical results combined with the lack of post-operative complications, suggest foramen magnum decompression with microsurgical obliquely transverse outer dura layer incisions as an effective treatment for syringomyelia associated with Chiari I malformation.

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Comment

The authors have submitted their revised manuscript. I would like to add the following comment for publication:

The authors describe their experience with extensive craniectomies of the posterior fossa combined with splitting of the outer dura layer in 8 cases of Chiari I Malformation with syringomyelia. This technique avoids dangers and complications associated with opening of the dura and arachnoid, such as CSF-fistulas or damage to perforating arteries of the brain stem. However, this advantage is somewhat countered by several disadvantages:

The Chiari I Malformation is a heterogenous group of patients. Some patients – possibly the majority – have bony anomalies of the craniocervical juniction resulting in a small posterior fossa. However, as Badie *et al.* (Neurosurgery 37: 214–218, 1995) have shown in their study, a significant proportion of patients have a normal sized posterior fossa. It seems questionable that the surgical method suggested in this report would offer any benefit for this subgroup of cases.

From my experience, three main pathomechanisms have to be considered in patients with Chiari Malformations and syringomyelia: brain stem compression, CSF-flow obstruction and arachnoid scarring, which may tether the cord to cerebellar tonsils and overlying dura and lead to additional CSF-flow obstruction ot the foramen magnum. Even with the use of intra-operative ultrasound it will be very difficult if not impossible to assess the quality of CSF-flow without opening the dura and arachnoid. An obstruction at the level of the Foramen of Magendie by arachnoid adhesions, which is a common finding from my experience, will not be detected. This means that only brain stem compression and – to some degree – CSF-flow obstructions can be corrected with the technique described in this paper.

On "re-operated" patients with failed foramen magnum procedures, severe arachnoid scarring at the level of the foramen magnum was the most prominent intra-operative finding. Arachnoid scarring is of clinical relevance. Therefore, I expect a higher rate of clinical recurrences with time compared to techniques with intradural exploration and opening of the Foramen of Magendie.

A second concern of mine with this paper is the recommended extensive craniectomy. From my experience, it is not necessary to extend the craniectomy to more than 3×3 cm. The craniectomy should be tailored to accommodate the cisterna magna and to decompress the brainstem. The patient needs a foramen magnum and not a posterior fossa decompression. Extensive craniectomies carry the danger of cerebellar herniation towards the spinal canal with consecutive brain stem compression, CSF-flow obstruction and arachnoid scarring.

In summary, the aim of the authors – to do as good or better with less risks for the patient – is honourable. But only time and honest reports on long-term results will tell whether this surgical strategy should be recommended.

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