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Cranio-Cervical Decompression for Chiari Type I-Malformation, Adding Extreme Lateral Foramen Magnum Opening and Expansile Duroplasty with Arachnoid Preservation. Technique and Long-Term Functional Results in 44 Consecutive Adult Cases – Comparison with Literature Data

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Summary

Background. Posterior cranio-cervical decompression by opening at least foramen magnum and C1-lamina usually with corresponding dural and arachnoid opening, is the procedure most currently used for treating Chiari I malformation (alone or in association with syringomyelia). To optimize decompressive effects together with reducing risks, a procedure was developed which consists of a suboccipital craniectomy and a C1 (or C1/C2) laminectomy, plus an extreme lateral Foramen Magnum opening, a "Y" shaped dural incision with preservation of the arachnoid membrane, and an expansile duroplasty employing autogenous periosteum.

The purpose of the article is:

1. to report the long-term functional results in a consecutive series of 44 adult patients affected by symptomatic Chiari Malformation type I (CM) using the procedure described.
2. and to compare this technical modality with the other modalities reported in the literature.

Method. This series includes 44 patients harboring CM type I and operated on between 1990 and 2000. 15 patients had CM with syringomyelia (34%) and 29 CM alone (66%). Functional status was evaluated by using the Karnofsky disability scale. Before surgery 37 patients (84.1%) were independent (of whom 13 had syringomyelia) and 7 patients (15.9%) were dependent – i.e., they required assistance – (of whom 2 had syringomyelia). Outcomes were analized with follow-up ranging from 1 to 10 years (4 years on average).

Findings. There was no operative mortality, and surgery did not provoke any neurological aggravation. After surgery all the patients were independent. For the patients with CM only, the averaged Karnofsky score was 90 at latest follow up, versus 76 before surgery. For the patients with syringomyelia, the averaged latest Karnofsky score was 89 after surgery, versus 74 before.

Interpretation. The presented technique was compared with the other surgical modalities reported in the literature. This comparative study shows that cranio-cervical decompression with extreme lateral resection of the posterior rim of Foramen Magnum out to the level of the occipital condyles on either side, associated with an enlargement duroplasty with preservation of the arachnoid membrane, achieved the best results with minimal complications and side-effects.

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

Introduction

Chiari Type I-Malformation consists of herniation of the cerebellar tonsils through Foramen Magnum into the cervical spinal canal without descent of the brainstem. Syringomyelia is frequently associated with this malformation. Posterior decompression of the cervico-occipital junction is the commonly admitted standard surgical treatment. The procedure aims at suppressing compression of the neural structures and at re-establishing CSF circulation within Cisterna Magna, which seems the most logical means counteracting pathophysiology of syringomyelia.

Decompression can be performed using a variety of technical modalities. It may be limited to a simple suboccipital craniectomy associated with laminectomy of C1 (and more or less of C2) [15]. Simple bone resection may lead to insufficient decompression, therefore craniectomy and laminectomy can be completed with enlargement of the dura of the posterior fossa [2, 3, 6, 8, 11, 16, 19, 20, 21, 22]. An option is to only incise the external layer of the dura, the internal layer being respected [9, 12, 13]. Another option is to open both layers of the dural sheath, along a vertical line [4] or in a Y-shaped manner [2, 6, 21] and to patch the enlarged opening with a duroplasty of periosteum [2, 22, 25, 26], fascia lata [4, 14] or artificial dura [21] rather than leaving it open [27].

Dural incision may be accompanied by a large arachnoid opening to explore the Foramen of Magendie [5, 14, 17, 28, 29] so as to ascertain its patency, especially in cases with hydrocephalus; but arachnoid opening predisposes to pseudomeningocoele and/or CSF fistulas. Bilateral resection of tonsils has been advocated by some authors to achieve an optimal decompression of the cervico-occipital junction [7, 10, 18].

When performing dural incision and duroplasty, arachnoid may be opened; but one may prefer to preserve the arachnoid membrane, which avoids CSF leakage [12, 22]. This is this later option which we prefered for treating our referred cases over the last ten years.

The purpose of this study was to check the efficiency, as well as the inocuity, of Foramen Magnum decompression adding extreme lateral bone resection and using an expansile duroplasty with an attempt at arachnoid preservation. This article reports our experience with this method and compares its results with the various other techniques published in the literature; which comparison to our knowledge, has not been done so far in a detailed manner.

Table 1. *Presenting Symptoms (A) and Signs (B) in Chiari Type I Malformation, Without or with Syringomyelia, Listed in Decreasing Order*

A – Symptoms	
Pain:	86,4%
– Neck (sub-occipital, cervical)	38,7%
– Trigeminal neuralgia	20,5%
– Brachial neuralgia	15,9%
– Headaches	6,8%
– Back pain	4,5%
Paresthesias	29,5%
Dizziness	18,2%
Unsteadiness	4,5%
Diplopia	2,3%
Hypo-acousia	2,3%
Difficulty in swallowing	2,3%
Vomiting	2,3%
Miction imperiosa	2,3%
Attacks of unconsciousness	2,3%
B – Signs	
Pinprick altered sensation	27,3%
Thermal altered sensation	15,9%
Upper extremity weakness	13,6%
Hand atrophy	9,1%
Hyperactive lower extremity reflexes	6,8%
Gait disturbances	6,8%
Hyperactive upper extremity reflexes	4,5%
Miction imperiosa	4,5%
Dysmetry	4,5%
Dysarthria	4,5%
Facial hemispasm	2,7%
Dysphonia	2,3%
Hypopallesthesia	2,3%
Lower extremity weakness	2,3%
Babinski sign	2,3%
Urinary incontinence	2,3%

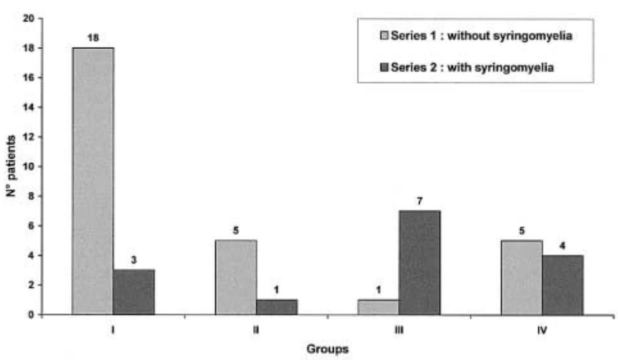


Fig. 1. Regrouping of patients affected with Chiari malformation type I without syringomyelia (series 1) and with syringomyelia (series 2), according to neurological presentation (see description of the various groups in paragraph on Neurological status)

Clinical Material

In the last ten years (January 1990 to January 2000), 44 adult patients with symptomatic Chiari Malformation type I (CM) were referred for surgery to our Neurosurgical Department. 15 patients had CM with signs of syringomyelia on MRI (34%) and 29 had CM without any significant abnormal neuro-imaging of associated syringomyelia (66%). 13 (29%) were males and 31 (71%) females. Age ranged from 14 to 63 years, 40 years on average. Mean duration of symptoms from origin to surgery amounted to 3 years and 6 months, with extremes of 3 months and 15 years. The low rate of Chiari malformations associated with syringomyelia in this series (34% compared to the classical rate in the literature: 60 to 80%) is probably related to the fact that our series started only in 1990 with an easy access to MRI examination.

Preoperative Clinical Status

Neurological Status. All cases underwent full neurological examination. Presenting symptoms and signs – whether there was syringomyelia associated to the CM or not – are listed in Table 1.

According to neurological presentation, the patients were classified into one of the four following groups (Fig. 1). *Group I* (21 patients, 47%) corresponded to patients who had subjective neurological symptoms (neck and/or shoulder pain, brachialgias, paresthesias, dizziness . . .) without observable signs; *Group II* (6 patients, 14%), to patients with objective clinical signs of cranial nerve deficits and/or cerebellar disturbances, suspected to be in relation with posterior fossa dysfunction; *Group III* (8 patients, 18%), to cases with motor and/or sensory changes attaining the extremities and presumed to be due to spinal cord dysfunction; *Group IV* (9 patients, 21%), to patients with mixed disturbances of the three previous groups.

Table 2. *Karnofsky Scale*

Score	
100	normal; no complaints, no evidence of disease
90	able to carry on normal activity; minor symptoms
80	normal activity with effort; some symptoms
70	cares for self; unable to carry on normal activity
60	requires occasional assistance; cares for most needs
50	requires considerable assistance and frequent care
40	disabled; requires special care and assistance
30	severely disabled; hospitalised, death not imminent
20	very sick; active supportive care needed
10	moribund; fatal processes are progressing rapidly

Functional Status. Functional status was evaluated by using the Karnofsky Disability scale (Table 2). 37 patients (84.1%) had score above 70, that is they were independent; of these, 24 had no syringomyelia, whilst 13 had. 7 patients (15.9%) had a score below 70 i.e., they were dependent, in other words they required, at least occasional, assistance; 5 of them had no syringomyelia, whilst 2 had. Details on Karnofsky scores in the group without syringomyelia and the group with syringomyelia are given in Tables 3A and 3B, respectively.

Neuro-Imaging

Conventional MRI Scan of the brain, as well as of the entire spinal cord, with T1 and T2 sequences, were performed in all patients, in order to diagnose tonsilar herniation and search for associated syringomyelia down to the conus medullaris. A syrinx was found in 15 of the 44 patients. It is important to mention that only one patient had hydrocephalus in our series. In this patient, hydrocephalus was presumed to be the consequence of tonsilar herniation, as ventricular size partly decreased after Foramen Magnum decompression. No tethered cord was found in our series.

Tonsilar herniations were classified into four grades according to level of descent through the Foramen Magnum (Fig. 2). Grade I (T1) corresponded to level of tonsils descent between Foramen Magnum and C1, II (T2): at C1 level, III (T3): between C1 and C2 and IV (T4): at C2 level. 31 patients (71%) had herniation grade I, 8 (18%) grade II, 3 (7%) grade III and 2 (4%) grade IV.

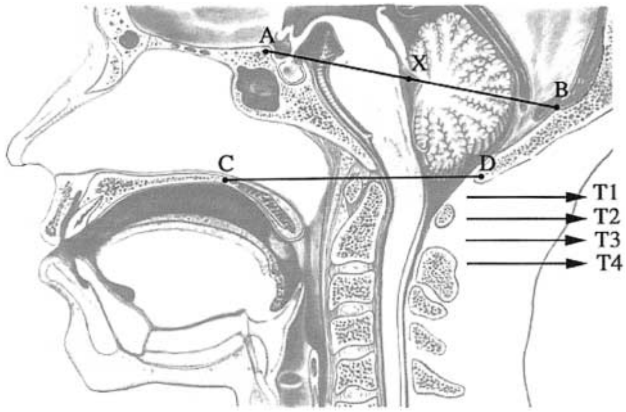


Fig. 2. Drawing of cervico-occipital junction with mention of the various degrees of tonsilar herniation (as found on preoperative MRI). *A–B*: The Twining line goes from the sellar tubercle to the internal occipital protuberance. The middle of the line (×) corresponds to the midpoint of the fourth ventricle. *C–D*: The Chamberlain line goes from the posterior border of the bony palate to the posterior margin of Foramen Magnum. The summit of the dens stays slightly below Chamberlain's line. Our classification of tonsilar herniations is as follows: *T1* level of tonsils descent between Foramen Magnum and C1, *T2* at C1 level, *T3* between C1 and C2, *T4* at C2 level

Of the 15 patients with associated syringomyelia, the syrinx was cervical in 4 (27%), thoracic in 3 (20%), cervicothoracic in 2 (13%) and panmedullary in 6 (40%).

Cine-MRI to assess CSF velocity/flow, was not performed in this study.

Surgical Procedure

All patients in this series were operated on under general anesthesia with endotracheal intubation, in the sitting position. All patients were equiped with a right atrial central venous catheter and dopler ultrasonography to monitor the possible occurrence of air embolism.

Table 3. *Pre- and Post-Operative Karnofsky Scores (with 4 Years Follow-up on Average) in the Group of Patients with Chiari Malformation Type I Without (A) and with Syringomyelia (B)*

A – Chiari type I without Syringomyelia (N 29)							
		Karnofsky: pre-op.	Karnofsky: post-op.				
Group	N(%)		2 weeks	2 months	6 months	1 year	updated
I	18(62)	79	83	87	89	88	91
II	5(17)	72	76	79	86	86	84
III	1(4)	70	80	90	90	90	90
IV	5(17)	68	76	80	82	84	92
Total	29(100)	76	80	85	87	87	90
B – Chiari type I with Syringomyelia (N 15)							
		Karnofsky: pre-op.	Karnofsky: post-op.				
Group	N(%)		2 weeks	2 months	6 months	1 year	updated
I	3(20)	90	93	90	97	97	100
II	1(6)	70	80	80	80	80	100
III	7(47)	73	77	81	86	89	89
IV	4(27)	65	70	73	75	78	78
Total	15(100)	74	79	80	85	87	89

The strategy of surgery was based on increasing the capacity of the occipito-cervical junction in its two axial-plane diameters: the antero-posterior and the transverse, so as to achieve an optimal decompression of the neural structures and get free cisternal spaces with good CSF circulation in Cisterna Magna, as well as around brainstem and cervico-occipital junction.

Operative Steps

The main operative steps are illustrated in drawings of Fig. 3. A midline occipito-cervical skin incision was made, from 5 cm above the external occipital protuberance (to expose the supra-occipital periosteum) down to the level of the C-4 spinous process. The periosteum was preserved in the supra-occipital region so that it could be harvested to patch the dural opening at closure. Then posterior cervical muscles were elevated to expose the suboccipital skull and Foramen Magnum, as well as the posterior arch of C-1 in all cases and more or less of C-2, according to the degree of tonsilar herniation.

The vertebral arteries at occipito-cervical level were identified on both sides and protected. A low occipital craniectomy, made by turning a small bone flap of an approximately triangular shape

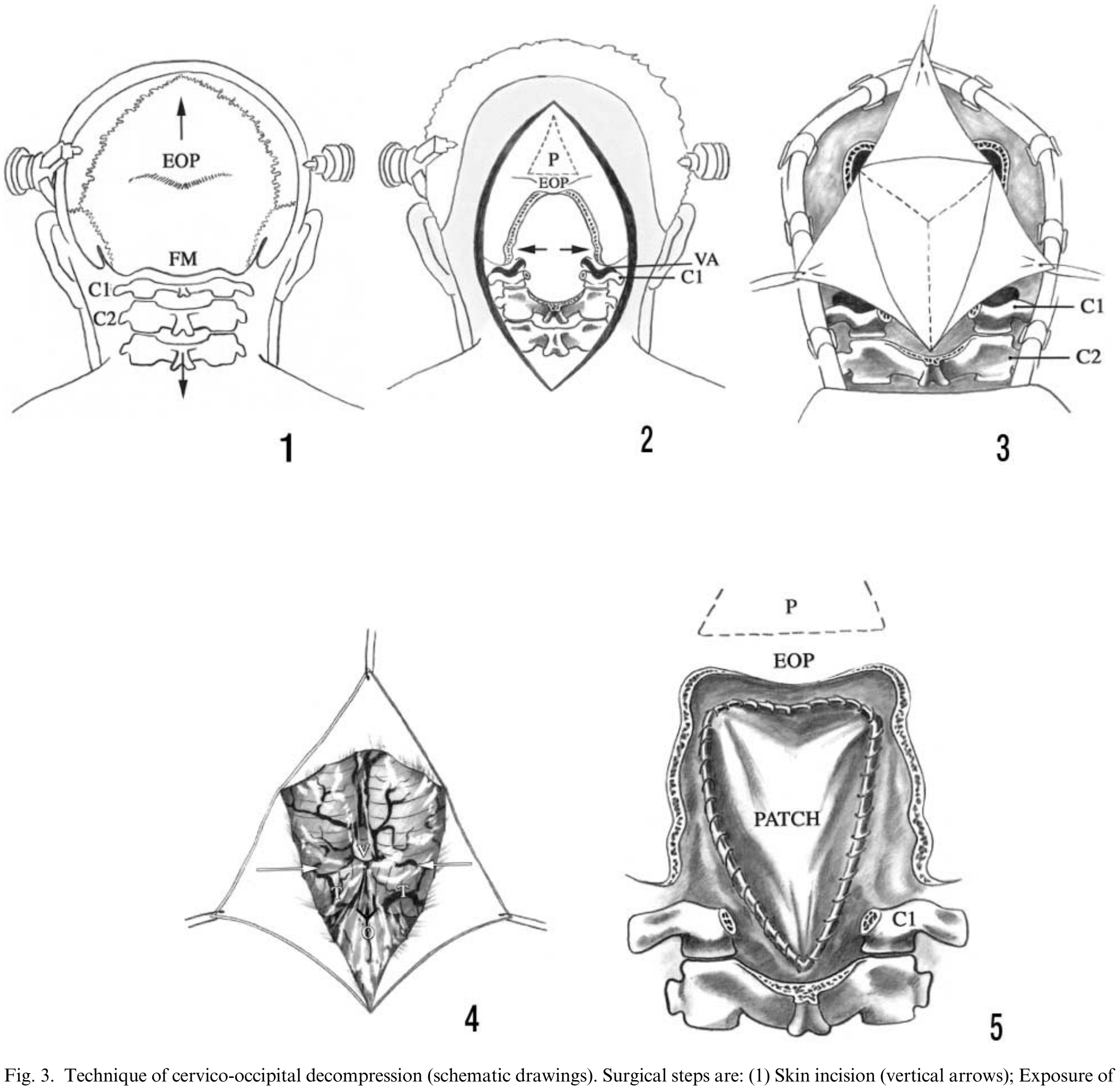


Fig. 3. Technique of cervico-occipital decompression (schematic drawings). Surgical steps are: (1) Skin incision (vertical arrows); Exposure of bony structures: external occipital process (*EOP*), Foramen Magnum (*FM*), C1 and C2 laminae. (2) Bone opening: sub-occipital craniotomy, with extreme lateral opening of Foramen Magnum (horizontal arrows); Vertebral artery exposure (VA) on both sides; C1 laminectomy. (3) Y-shaped dural opening, with arachnoid preservation. (4) Exposure of herniated tonsils with marked engrooving by the posterior margin of Foramen Magnum (arrows); Obex (*O*), Vermis (*V*), Tonsils (*T*). (5) Enlargement duroplasty with periosteal patch (*PATCH*): Periosteum for patching, harvested (*P*) above the external occipital process (*EOP*)

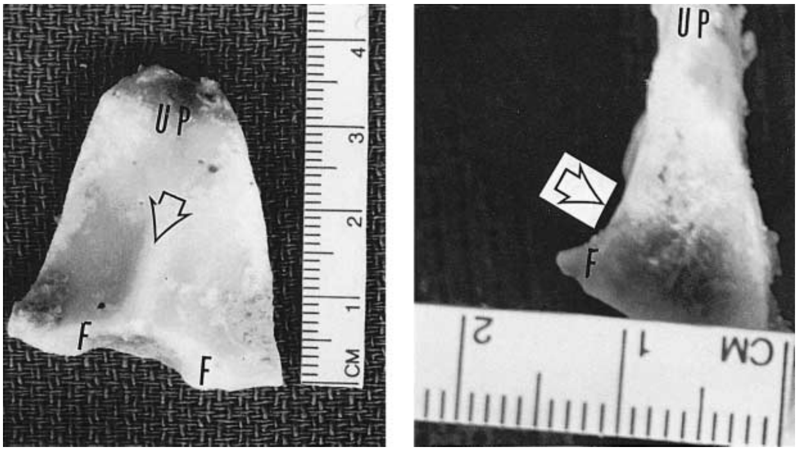


Fig. 4. Photography of the piece of bone from suboccipital craniotomy. [Left: inner (endocranial) view; right: lateral view]. Notice in this example – as in many other cases – the importance of the medial suboccipital crest (arrow) (1.5 cm in width), that markedly reduced the antero-posterior diameter of Foramen Magnum, and contributing to cervico-medullary compression in addition to Chiari malformation. Upper part (UP); lower part: posterior margin of Foramen Magnum (F)

Fig. 5. Post-operative CT-scan. *Left*: axial section, showing the extent of the Foramen Magnum (*FM*) opening. Bone resection includes, not only the posterior edge of FM, but also the lateral edges anteriorly to the gleno-condylar atlo-occipital joint on both sides. *Right*: midline sagital section, showing (small) sub-occipital craniectomy, C1-laminectomy and partial resection of the upper part of C2 spinous process.

with an inferior base (Fig. 4), was performed to decompress the medullary-spinal cord junction and the tonsils, posteriorly. Then craniectomy was completed with an extreme lateral opening of the rim of the Foramen Magnum by bone resection with fine rongeurs out to the level of the occipital condyles on either side – i.e., condyle to condyle – with the aim to achieve a good decompression of both tonsils, not only posteriorly but also laterally (Fig. 5). After laminectomy of C-1 (and C-2 if necessary) was performed, a "Y"-shaped opening of the dura-mater was made to enlarge Cisterna Magna. This was done with careful hemostasis of the occipital venous sinuses.

The arachnoid membrane was preserved to avoid CSF leakage and consequently prevent the brain from collapse. Then a duroplasty was made with periosteum patch, harvested from the supra-occipital region, to enlarge the posterior fossa and restore a spacious Cisterna Magna. The dural patch was tighted with three running sutures, in a triangular shaped manner with the base superior.

Finally, the wound was meticulously closed, each layer at a time: muscles, fascia, subcutaneous tissue and skin, with interrupted sutures. Special care was taken not to injure the occipital nerve on both sides.

Post-Operative Care

After the surgery, the patient was placed in the Intensive Care Unit for one night. A cervical collar was offered to the patient to keep his neck comfortable with a physiological lordosis and to decrease pain. Analgesics and anticoagulation were systematically given. The patient was discharged on the 10th day, on average. Then, at one month postoperatively, physiotherapy with particular mobilization of both upper limbs was undertaken to avoid frozen shoulders. At 2 months, an outpatient clinic visit was organized before deciding the patient returns to work and – hopefully – resumes normal life.

Post-Operative Assessment

Post-operative neurological and functional status were regularly assessed; the latter was quantified by using the Karnofsky Disability scale. Evaluation took place by direct examination on discharge approximataly two weeks after surgery and at the first outpatient clinic visit two months after discharge; then by mail or telephone questionnaire, at 6 months and one year or more, according to follow-up. Depending on patients, the latest follow-up ranged from 13 months to 10 years (4 years and 2 months on average).

All the 15 patients with syringomyelia had MRI control, of both brain and entire spinal cord. In patients with CM alone, a post-operative cranio-cervical MRI was performed only when disabling clinical manifestations persisted at the six month follow-up check, that is in 13 patients.

Results

Surgical Events

Accidental "pin point" opening without real penetration of the arachnoid, but with some transient droplets of CSF but with spontaneous cessation within a few seconds, occurred in 13 cases. A large accidental opening of the arachnoid causing post-operative CSF fistula happened in 2 cases.

Morbidity and Mortality

There was no surgical mortality, and no neurological aggravation postoperatively. There were two cases of CSF leak, without any neurological consequences. The leak ceased spontaneously in one case, but required external lumbar drainage for two weeks in the other. Healing was delayed in 5 cases, particularly at the top of the wound due to superficial skin necrosis (of the order of one cm^2); none required re-operation for surgical repair, none necessitated postponement of the patient's discharge. There was one case with short transient laryngeal edema and one case with pneumonia who rapidly recovered under antibiotics.

Outcome

Neurological Outcome. Outcomes in time, according to neurological presentations, are illustrated in Fig. 6.

Functional Outcome. Evaluation of functional results was the purpose of this study. Functional outcome was rated excellent when the Karnofsky score was above 90, good when between 70 and 90, fair when between 50 and 70, and poor when score was below 50.

In the group of the 29 patients with CM only, outcome at latest follow-up was excellent in 79% and good in 21%. In the 15 patients with syringomyelia associated with CM, outcome was excellent in 26% and good in 74%.

Benefit from Surgery. Prior to surgery, 15.9% of the patients needed at least occasional assistance, i.e., they were dependent. After surgery, all the patients were independent (i.e., with a Karnofsky score above to 70).

In the population of patients with CM only ($N = 29$), the averaged Karnofsky score was 90 at latest follow-up, versus 76 before surgery (Table 3A).

For the patients with syringomyelia associated to CM ($N = 15$), the latest averaged Karnofsky score was 89 after surgery, versus 74 before (Table 3B). Of these patients, 12 improved. The 3 patients who did not experience amelioration, belonged to clinical group IV; it is important to note that all these 3 patients had pansyringomyelia.

Briefly and from a practical point of view, 36 patients of the overall 44 cases of the series (i.e., 82%) could resume a normal life, whilst 8 patients (18%) did not return to full work and/or a completely normal life. Details on these 8 patients are provided in Table 4.

Results on Imaging

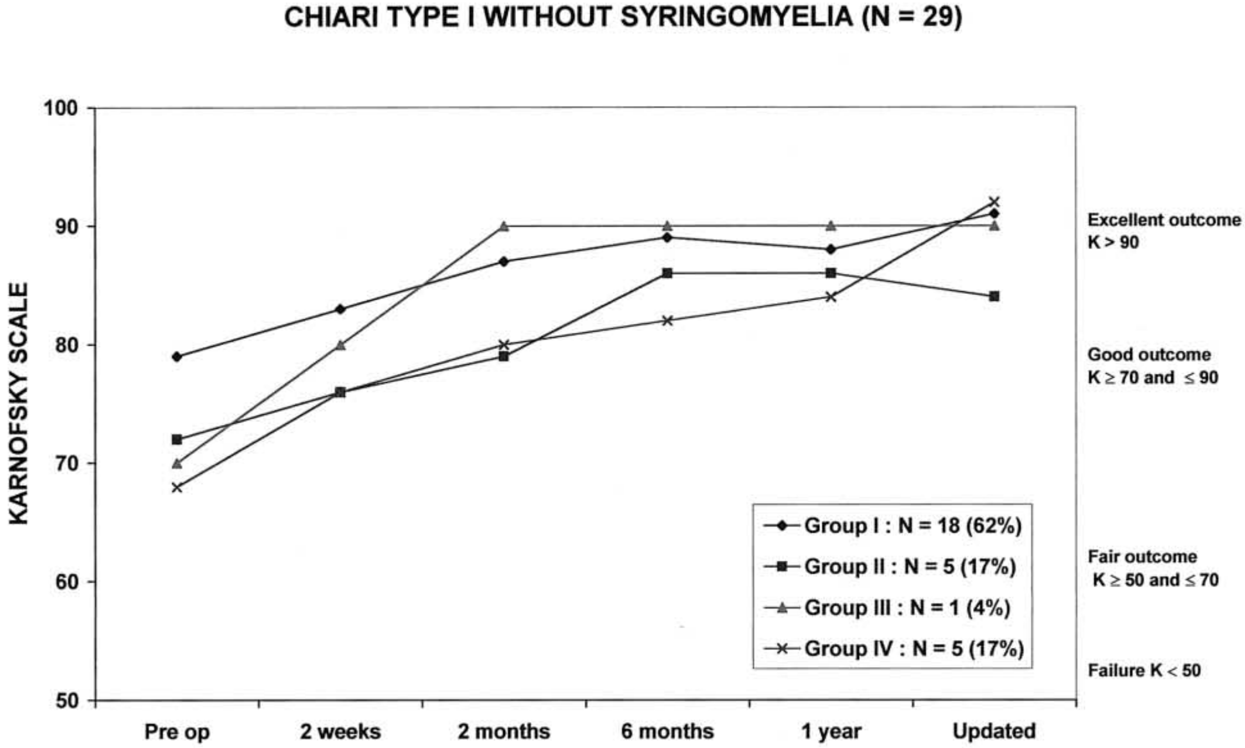
MRI control was performed within the range of 6 months to 4 years after surgery (8 months on average). Illustrated examples are presented in Fig. 7.

The occipito-cervical junction was considered totally decompressed with a "new" spacious Cisterna Magna in all the 28 patients in whom a post-operative MRI control was done (i.e., all 15 who had syringomyelia associated with CM and the 13 checked patients with CM only).

In the patients with syringomyelia ($N = 15$), images of the syrinx were as follows. For the 4 patients with cervical syrinx, 3 were improved and 1 unchanged; for the 3 patients with thoracic syrinx, 1 was improved and 2 unchanged; for the 2 patients with cervico-thoracic syrinx, the 2 were improved; and for the 6 patients with a panmedullary syrinx, 3 were improved and 3 unchanged. In summary, MRI images of syringomyelia were improved in 9 of the 15 patients (60%) and stabilized in 6 (40%); none had aggravation.

Discussion

This study shows that a low suboccipital craniectomy and C1-laminectomy, with extreme lateral Foramen Magnum decompression – together with



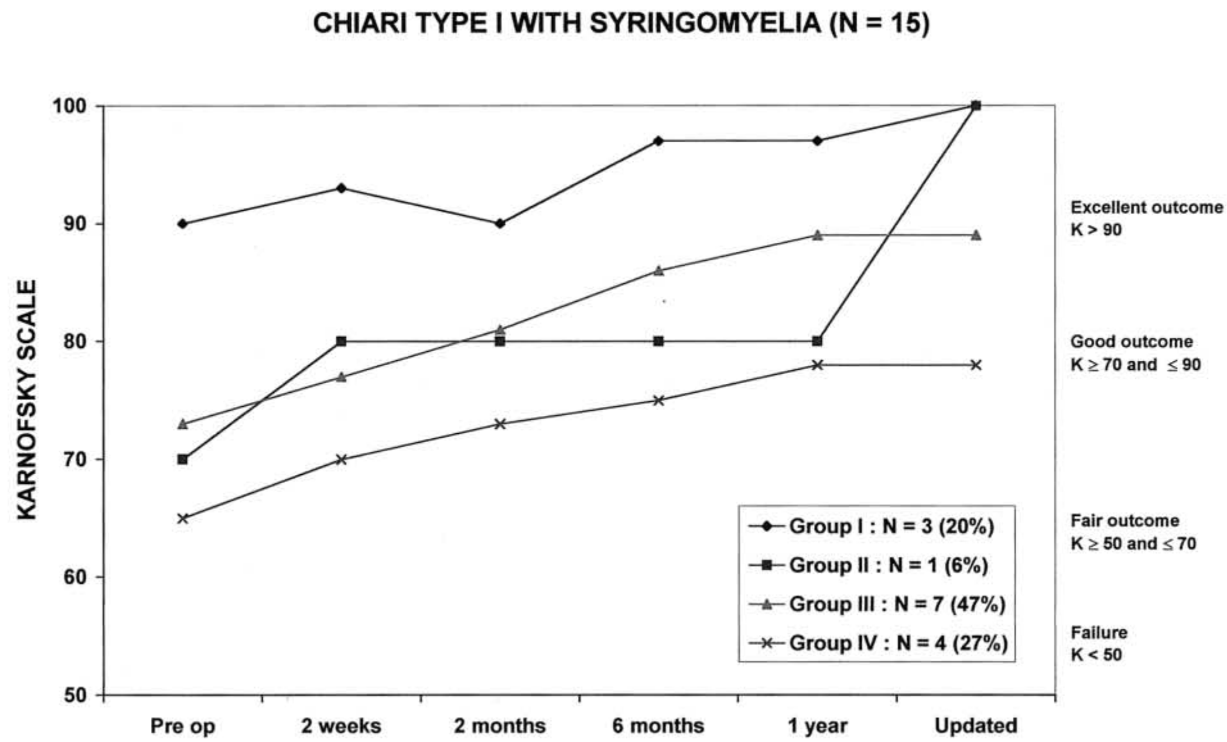


Fig. 6. Averaged outcome (evaluated using Karnofsky scale = K) according to clinical groups, (i.e., neurological presentations), along follow-up, in Chiari type I malformation without (N = 29) and with syringomyelia (N = 15). Outcome was considered excellent when $K > 90$, good when $K \geq 70$ and ≤ 90, fair when $K \geq 50$ and ≤ 70; outcome was considered as failure when $K < 50$

Table 4. *Patients who did not Return to Normal Life: Hypothesized Causes of Failure. K1 (Pre-Operative) and K2 (Post-Operative) Karnofsky Scores*

Patient	Age	Syrinx	K − 1	K − 2	Hypothesized cause of failure to resume normal life
1	57	(−)	80	70	multiple retinian lesions
2	71	(−)	70	70	ankylosing spondylitis
3	70	(−)	60	70	hydrocephalus
4	56	+	60	70	panmedullary syrinx
5	62	+	60	70	panmedullary syrinx, not improved by kysto-subarachnoid shunt
6	69	+	70	70	large cervicothoracic syrinx
7	63	(−)	70	70	associated atypical trigeminal neuralgia
8	61	(−)	70	80	associated atypical trigeminal neuralgia

K − 1 Preoperative Karnofsky score, *K − 2* post-operative Karnosky score.

dural opening but with preservation of arachnoid, followed by enlargement duroplasty – was able to obtain improvement in 83% of patients with CM alone and in 80% of patients with CM associated with syringomyelia. This technique did not provoke any neurological or any severe or lasting associated complication. This technical modality has to be compared with the other varieties of cranio-cervical decompression techniques.

Literature Review and Comparison Between Technical Modalities

On the occasion of this study, we reviewed the 119 articles published during the past ten years, given by the MEDLINE system. Only the reports with information on the techniques used and their corresponding long-term outcomes, sufficiently detailed to allow comparaison between series, were retained. They are listed in Table 5 for CM alone and Table 6 for CM with syringomyelia.

In order to evaluate the effectiveness and risks of each of the various techniques, the literature series [or cases from series] which corresponded to the same technical modality were regrouped together. Such an inventory, made separately for CM alone and CM associated with syringomyelia, is presented in Table 7. Histograms representing averaged outcome values corresponding to the various technical modalities, are presented in Fig. 8 for CM alone and Fig. 9 for CM associated with syringomyelia.

Chiari Malformation Without Syringomyelia (Fig. 8). Craniocervical bone resection without dural opening (technique N°1) was performed in only 4 cases; only 3 patients (75%) were improved. FM decompression with dural opening but with preservation of the arachnoid membrane and duroplasty to enlarge Cisterna Magna (Technique N°2) was performed in 38 cases; an improvement was obtained in 87%, with very few complications as shown in Table 7. Systematic opening of the arachnoid in addition to dural opening (Technique N°3) was performed in 78 cases. It did not reveal any superiority compared to the procedure without arachnoid opening; improvement rate was 81% (versus 87%) and there was 2% of aggravation (versus none). Statistical analysis using the method of Confidence Interval (95% CI), was carried out to compare techniques N°2 and N°3 (Technique N°1, applied to only 4 patients, was not included in the statistical study). Although not statistically significant, the better results achieved with technique N°2 tend to show that dural opening with preservation of arachnoid membrane, is preferable as complications due to arachnoid opening were frequent and sometimes severe, as shown in Table 7.

Chiari Malformation with Syringomyelia (Fig. 9). Results with the various reported techniques are summarized in Fig. 9. Statistical analysis (Confidence Interval 95% CI) showed that Foramen Magnum decompression with simple incision of the dural outer layer (modality N°2) or complete dural opening followed by duroplasty (modalities N°3, 5 or 7), were significantly better than simple bone resection (modality N°1) or sole shunt placement (modality N°8) ($p < 0.05$). Modality N°6, i.e., dural together with arachnoid opening, but without duroplasty, was the worst ($p < 0.05$).

Interestingly enough, in a study comparing the effects of two varieties of craniectomy: one small and one large (both associated with dural opening, arachnoid opening and duroplasty) Klekamp *et al.* observed better results with the smaller bone resection. As a matter of fact, syrinx was decreased in 87%, unchanged in 11% and increased in 2% in the group with small craniectomy, versus 72%, 6% and 22%, respec-

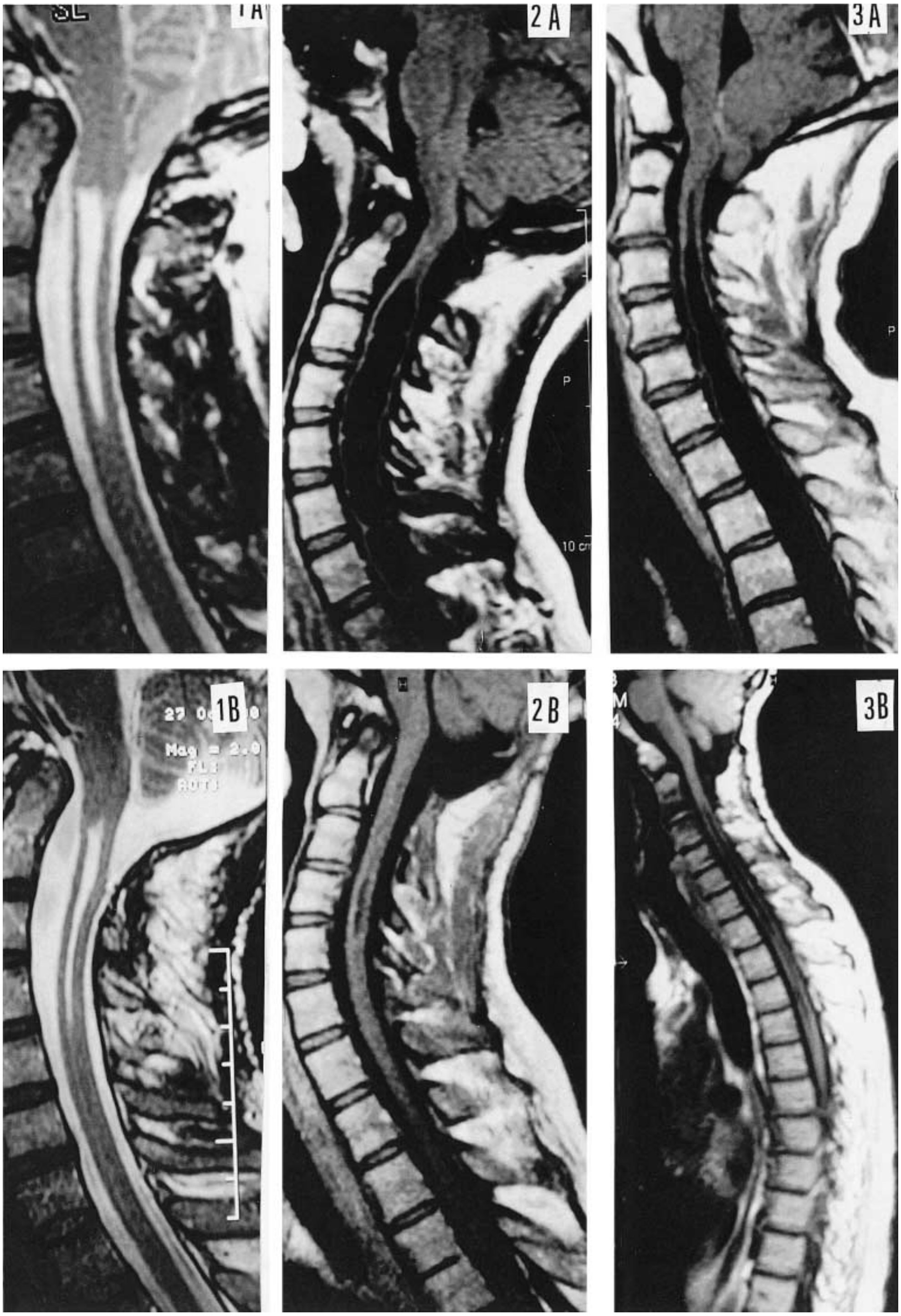


Fig. 7. *Case N°1:* Chiari I malformation with a cervico-bulbar syrinx before (1A) and 2 years after (1B) Foramen Magnum Decompression. On the post-operative MRI (1B), Cisterna Magna is patent and syrinx has decreased in size [i.e., its anterior – posterior (AP) diameter at the C2–C3 level passed from 55% to 32% of the total AP diameter of the spinal cord. *Case N°2:* Chiari I malformation with a cervical-thoracic syrinx before (2A) and 6 months after (2B) Foramen Magnum decompression. On the post-operative MRI (2B), Cisterna Magna is patent and the syrinx has mostly disappeared. *Case N°3:* Chiari I malformation with a pan-medullary syringomyelia before (3A) and 9 months after (3B) Foramen Magnum decompression. On the post-operative MRI (3B), Cisterna Magna is patent and syringomyelia has markedly receded (Index of the syrinx decreased from 70% to 30%)

Table 5. *Summary of Literature Results in Chiari-Type I Malformation Alone, after Surgical Decompression*

Chiari malformation alone					
Series	Technical modalities	N	Clinical outcome: N (%)		
			IMP	STAB	AGG
Pillay, 1991 (17)	FMD + DO + AO + DU	14	12(86)	2(14)	0(0)
Bindal, 1995 (3)	FMD + DO + DU	9	9(100)	0(0)	0(0)
Fisher, 1995 (7)	FMD + DO + AO + RT + DU	3	2(67)	1(33)	0(0)
Klekamp, 1996 (14)	FMD + DO + AO + DU	36	*		
Blagodatsky, 1999 (4)	FMD + DO + AO + DU	16	14(88)	1(6)	1(6)
Munshi, 2000 (15)	FMD + DO + AO + DU	11	10(91)	1(9)	0(0)
	FMD	4	3(75)	1(25)	0(0)
Sindou, 2001	FMD + DO + DU	29	24(83)	5(17)	0(0)

IMP Improved, *STAB* Stabilized, *AGG* Aggravated, *FMD* Foramen Magnum Decompression, *DO* Dural Opening, *AO* Arachnoid Opening, *DU* Duroplasty, *RT* Resection of Tonsils.
* In Klekamp *et al.*'s series [14] average Karnofsky score passed from 68 (pre operatively) to 77 (at one year post-operatively) with large craniectomy and from 77 (pre op.) to 83 (at one year post op.) with small craniectomy. Kaplan-Meier analysis demonstrated that 9 of the 36 patients without a syrinx showed progressive worsening of at least some of their symptoms and signs after a mean follow-up of 39 ± 52 months.

tively, in the large craniectomy group [14]. FM decompression with dural opening and enlargement duroplasty but without opening the arachnoid (modality N°3) was the most effective and at the same time the less dangerous modality. As a matter of fact, with this technique – which was performed in 78 cases – 85% of the patients were improved, whereas only 2% were made worse by the procedure and/or deteriorated in spite of surgery. Further, complication rate was minimal, as shown in Table 7.

Bone decompression with dural incision of only the outer layer (modality N°2) gave only slightly inferior results compared to complete dural opening. This technique was performed in 27 cases: 85% of the patients were improved and 4% deteriorated. FM decompression without any dural opening at all (modality N°1) was less effective. With this technique, performed in only 7 cases, improvement rate was 71%. In both techniques, complications were nil, as shown in Table 7.

Whether the dura was reconstructed (modalities N°4 and N°5) or left open (modality N°6), opening of the arachnoid was followed by a number of complications. These complications were particularly severe when the dura was not closed, as shown in Table 7. The 7 patients who were reported as having died from complication in the literature [1, 4, 10, 14], had large arachnoid openings. Opening of the arachnoïd did not guarantee better effectiveness as shown in Table 7. On the contrary, with technical modality N°4, only 5% of the 591 patients were improved and as many as 12% were made worse. Outcome was worst with modality N°6 (15 patients reported); there was improvement in only 27% and aggravation in as much as 33%.

Plugging the obex in addition to arachnoid opening that we found reported in 106 patients in the literature [4, 17, 22, 24], did not bring any positive effect. Improvement rate was 66% and aggravation 9% (not shown in the Table).

It appears from literature data, that complementary tonsilar resection (modality N°5) did not add much, compared to FM decompression with enlargement duroplasty only; improvement was obtained in 85% of the 97 patients and there was aggravation in 5% (versus 85% and 2% with modality N°3 for example). The same holds true for shunting in addition to FM decompression with duroplasty (modality N°7); improvement was achieved in 88% of the 16 patients and aggravation occurred in 6%.

Treatment of syringomyelia associated with Chiari I malformation with shunting only, whatever the method used, (technical modality N°8) has been widely done during the past years [1, 12, 23, 27]. Shunting was followed by improvement in 60% of the 95 patients and by deterioration in 16%, which are significantly the worst results compared to the ones achieved with FM decompression with duroplastic enlargement.

Conclusion

According to present experience and review of data from the literature, the following can be concluded.

Table 6. *Summary of Literature Results in Chiari-Type I Malformation with Syringomyelia, after Surgical Decompression and/or Shunt. Clinical Outcome and Evolution of Syrinx on Post-Operative MRI*

Chiari malformation with syringomyelia								
Series	Technical modalities	N	Clinical outcome			Syrinx on post-op MRI		
			IMP	STAB	AGG	IMP	STAB	AGG
Vaquero, 1990 (27)	FMD + DO + AO	15	4(27)	6(40)	5(33)	14(93)	1(7)	0(0)
	Shunt	15	10(67)	2(13)	3(20)	15(100)	0(0)	0(0)
Pillay, 1991 (17)	FMD + DO + AO + DU	17	9(53)	6(35)	2(12)			
Fujii, 1991 (8)	FMD + DO + DU	5	4(80)	0(0)	1(20)	3(60)	1(20)	1(20)
	FMD + DO + DU + SH	8	8(100)	0(0)	0(0)	6(75)	2(25)	0(0)
Isu, 1993 (13)	FMD + DO (ext. Layer)	7	6(86)	1(14)	0(0)	7(100)	0(0)	0(0)
Tognetti, 1993 (23)	FMD + DO + AO + DU	17	14(82)	2(12)	1(6)	12(100)	0(0)	0(0)
	Shunt	12	4(33)	5(42)	3(25)	12(100)	0(0)	0(0)
Raftopoulos, 1993 (18)	FMD + DO + AO + RT + DU	8	8(100)	0(0)	0(0)	8(100)	0(0)	0(0)
Van Velthoven, 1993 (24)	FMD + DO + AO + DU	25	10(40)	9(36)	6(24)			
Versari, 1993 (28)	FMD + DO + AO + DU	40	25(63)	9(22)	6(15)	28(70)	12(30)	0(0)
Oldfield, 1994 (16)	FMD + DO + DU	7	5(71)	2(29)	0(0)	7(100)	0(0)	0(0)
Sahuquillo, 1994 (21)	FMD + DO + DU	10	8(80)	2(20)	0(0)			
	FMD + DO + AO + DU	10	2(20)	5(50)	3(30)			
Bindal, 1995 (3)	FMD + DO + AO + DU	12	7(58)	5(42)	0(0)			
Fisher, 1995 (7)	FMD + DO + AO + RT + DU	16	9(56)	7(44)	0(0)	14(93)	1(7)	
Hida, 1995 (12)	FMD + DO (ext. Layer)	12	(82)			30(94)		
	FMD + DO + DU	21						
	Shunt	37	36(97)			37(100)	0(0)	0(0)
Klekamp, 1996 (14)	FMD + DO + AO + DU	88	*					
Vanaclocha, 1997 (25)	FMD + DO + AO + DU (+BR)	28	20(73)	8(27)	0(0)	11(39)		
Gambardella, 1998 (9)	FMD + DO (ext. Layer)	8	7(87)	0(0)	1(13)	7(87)		
Guyotat, 1998 (10)	FMD + DO + DU	50	18(36)	14(28)	18(36)	14(58)		
	FMD + DO + AO + RT + DU	8	7(87)	1(13)	0(0)			
	FMD + DO + DU + SH	8	6(75)	1(13)	1(12)			
Aghakhani, 1999 (1)	FMD + DO + AO + DU	242	92(38)	121(50)	29(12)	36(15)		
	Shunt	31	0(0)	8(71)	3(29)	(7)		
Blagodatsky, 1999 (4)	FMD + DO + AO + DU	44	34(78)	8(18)	2(5)			
	FMD + DO + AO + RT + DU	11	7(64)	3(27)	1(9)			
Sakamoto, 1999 (22)	FMD + DO + DU + BR	20	20(100)	0(0)	0(0)	20(100)	0(0)	0(0)
	FMD + DO + AO + DU + BR	20	17(85)	3(15)	0(0)	20(100)	0(0)	0(0)
	FMD + DO + AO + DU + BR	4	3(75)	1(25)	0(0)	3(75)	1(25)	0(0)
Munshi, 2000 (15)	FMD + DO + AO + DU	12	10(84)	2(16)	0(0)	9(100)	0(0)	0(0)
	FMD	7	5(71)	2(29)	0(0)	3(50)	0(0)	3(50)
Sindou, 2001	FMD + DO + DU	15	12(80)	3(20)	0(0)	9(60)	6(40)	0(0)

IMP Improved, *STAB* Stabilized, *AGG* Aggravated, *FMD* Foramen Magnum Decompression, *DO* Dural Opening, *AO* Arachnoid Opening, *DU* Duroplasty, *SH* Shunt, *RT* Resection of Tonsils, *BR* Bone Reconstructed.

* In Klekamp *et al.*'s series [14], Karnofsky score passed from 68 (pre op.) to 74 (at one year post op.) with large craniectomy and from 71 (pre op.) to 77 (at one year post op.) with small craniectomy. Kaplan-Meier analysis demonstrated that 16 of the 88 patients with a syrinx showed progressive worsening with time, but much less with a small craniectomy (14%) than with a large craniectomy (62%).

In Klekamp *et al.*'s study [14], there were actually two groups according to the size of craniectomy. In the group of patients with a small craniectomy the size of the syrinx decreased in 87% was unchanged in 11% and increased in only 2% (versus 72%, 6% and 22%, respectively, in the large craniectomy group).

– F.M. decompression with extreme lateral rim resection, followed by dural enlargement, revealed the most effective treatment for CM whether associated with syringomyelia or not. As pointed out by Klekamp *et al.* [14], small craniectomy offers better results than large craniectomy.

– Complete opening of the dura achieved (slightly) better results than simple incision of only the outer dural layer.

– Preservation of the arachnoid membrane, whenever there is no evidence of obstruction of the Foramen of Magendie and/or arachnoiditis, decreases com-

Table 7. *Summary of Results in the Literature, According to the Technical Modality Used. Clinical Outcome [N number; (%)]*

			Clinical outcome			Complications	
			IMP	STAB	AGG	Local	Neuro
1. FMD without DO							
Munshi, 2000 (15)	CM	4	3(75)	1(25)	0(0)	wound infection 1(9)	0
Munshi, 2000 (15)	S	7	5(71)	2(29)	0(0)		
Total	CM	4	3(75)	1(25)	0(0)		
	S	7	5(71)	2(29)	0(0)		
2. FMD with DO (outer layer)							
Gambardella, 1998 (9)	S	8	7(88)	0(0)	1(12)		
Hida, 1995 (12)	S	12	10(82)	2(18)	0(0)		
Isu, 1993 (13)	S	7	6(86)	1(14)	0(0)		
Total	S	27	23(85)	3(11)	1(4)		
3. FMD + DO + DU (arachnoid integra)							
Bindal, 1995 (3)	CM	9	9(100)	0(0)	0(0)		
Fujii, 1991 (8)	S	5	4(80)	0(0)	1(20)		
Hida, 1995 (12)	S	21	17(82)	3(17)	1(1)		motor deterioration (13)
Oldfield, 1994 (16)	S	7	5(71)	2(29)	0(0)	0	0
Sahuquillo, 1994 (21)	S	10	8(80)	2(20)	0(0)	aseptic meningitis 1(5)	
Sakamoto, 1999 (22)	S	20	20(100)	0(0)	0(0)	0	0
Sindou, 2001	CM	29	24(83)	5(17)	0(0)	delayed wound healing 5(12)	0
	S	15	12(80)	3(20)	0(0)		0
Total	CM	38	33(87)	5(13)	0(0)		
	S	78	66(85)	10(13)	2(2)		
4. FMD + DO + AO + DU							
Aghakhani, 1999 (1)	S	242	92(38)	121(50)	29(12)	wound hematoma 17(6), meningitis 17(6)	
Bindal, 1995 (3)	S	12	7(58)	5(42)	0(0)		
Blagodatsky, 1999 (4)	CM	16	14(88)	1(6)	1(6)		
	S	44	34(78)	8(18)	2(5)		
Guyotat, 1998 (10)	S	50	18(36)	14(28)	18(36)	meningitis 1(1)	
Klekamp, 1996 (14)*	CM	36	27	0	9	infection 2(5), aseptic meningitis 13(35), CSF leak 14(38)	hydrocephalus 1(3), cerebellar 1(3) and posterior cerebral 1(3) infarction, apnea 2(5), mesencephalic disturbances 2(5) swallowing dysfunction 3(8)
	S	88	72	0	16	infection 3(4), CSF-leak 1(1)	
Munshi, 2000 (15)	CM	11	10(91)	1(9)	0(0)	CSF leak 2(9), aseptic meningitis 1(4), subgaleal CSF 4(17), infection 3(13), occipital nerve pain 1(4)	
	S	12	10(84)	2(16)	0(0)		
Pillay, 1991 (17)	CM	14	12(86)	2(14)	0(0)		
	S	17	9(53)	6(35)	2(12)		
Sahuquillo, 1994 (21)	S	10	2(20)	5(50)	3(30)		hydrocephalus 1(10)
Sakamoto, 1999 (22)	S	24	20(83)	4(17)	0(0)	aseptic meningitis 1(4)	
Tognetti, 1993 (23)	S	17	14(82)	2(12)	1(6)		
Vanaclocha, 1997 (25)	S	28	20(73)	8(27)	0(0)		
Van Velthoven, 1993 (24)	S	25	10(40)	9(36)	6(24)		
Versari, 1993 (28)	S	40	25(63)	9(22)	6(15)		
Total	CM	78	63(81)	4(10)	1(2)		
	S	591	300(51)	193(37)	68(12)		
5. FMD + DO + AO + RT + DU							
Blagodatsky, 1999 (4)	S	11	7(64)	3(27)	1(9)		
Fisher, 1995 (7)	S	16	15(94)	1(6)	0(0)	aseptic meningitis 2(11), kyphotic deformity 1(5), mild hearing loss 1(5)	
Guyotat, 1998 (10)	S	8	7(87)	1(13)	0(0)	meningitis 1(1)	
Raftopoulos, 1993 (18)	S	8	8(100)	0(0)	0(0)		
Williams, 1993 (29)	S	54	45(83)	5(9)	4(8)		
Total	S	97	82(85)	10(10)	5(5)		

Table 7. *Continued*

			Clinical outcome			Complications	
			IMP	STAB	AGG	Local	Neuro
6. FMD + DO + AO							
Vaquero, 1990 (27)	S	15	4(27)	6(40)	5(33)	cephalalgias 7(47), meningitis 2(13)	Neuralgia Vth 1(7)
Total	S	15	4(27)	6(40)	5(33)		
7. FMD + DO + DU + SH							
Fujii, 1991 (8)	S	8	8(100)	0(0)	0(0)		
Guyotat, 1998 (10)	S	8	6(75)	1(13)	1(12)		
Total	S	16	14(88)	1(6)	1(6)		
8. Shunt							
Aghakhani, 1999 (1)	S	31	7(23)	15(48)	9(29)		
Hida, 1995 (12)	S	37	36(97)	1(3)	0(0)	shunt malfunction 4(11)	
Tognetti, 1993 (23)	S	12	4(33)	5(42)	3(25)		Leg dysesthesia 1(8)
Vaquero, 1990 (27)	S	15	10(67)	2(13)	3(20)		Leg dysesthesia 8(53)
Total	S	95	57(60)	23(24)	15(16)		

IMP Improved, *STAB* Stabilized, *AGG* Aggravated, *Neuro* Neurological, *CM* Chiari type I Malformation alone, *S* Chiari Malformation with Syringomyelia, *FMD* Foramen Magnum Decompression, *DO* Dural Opening, *AO* Arachnoid Opening, *DU* Duroplasty, *SH* Shunt, *RT* Resection of Tonsils.
* For Klekamp *et al.*'s series see legends of Tables 5 and 6 for interpretation.

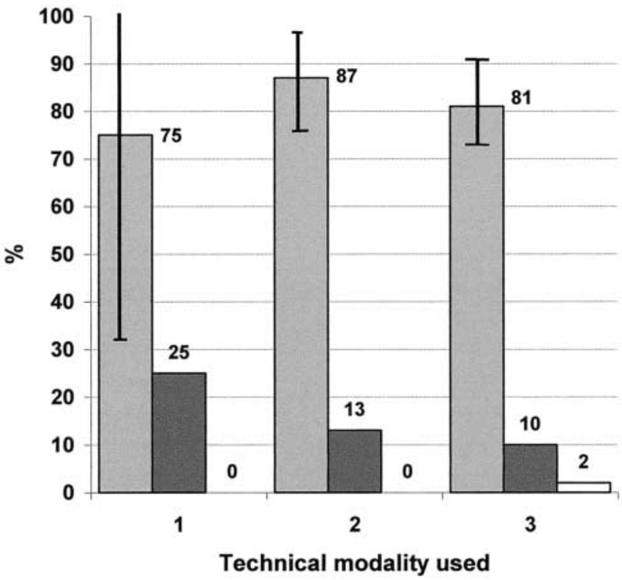


Fig. 8. Clinical outcome according to the various technical modalities used for *Chiari Malformation type I alone*. *FMD* Foramen Magnum Decompression, *DO* Dural Opening, *DU* Duroplasty, *AO* Arachnoid Opening, *RT* Resection of Tonsils, *SH* Shunt.
□ Improved, ■ Stabilized, □ Aggravated
1 FMD WITHOUT DO, *2* FMD + DO + DU (arachnoid integra), *3* FMD + DO + AO + DU
Best results were achieved with technical modality N°2

plication risks. In other words, the arachnoid should be dissected in cases of Chiari with syringomyelia and/or hydrocephalus whenever simple decompression would not appear sufficient to re-establish a good CSF flow, i.e., when there is evidence of "blocking" arachnoiditis.

– Tonsilar resection does not seem to add much, provided FM decompression is performed not only posteriorly but also laterally, i.e., condyle to condyle.
– Large opening of the arachnoid, together with exploration of the Foramen of Magendie, as well as complementary subpial resection of tonsils, could be kept as a secondary option in the rare cases in whom CSF circulation at the cervico-occipital junction is considered to remain insufficient.
– Plugging the obex not only does not bring any positive benefit, but entails significant additional risks.
– Shunting does not seem anymore an appropiate method of treatment for syringomyelia considered in relation to a dysfunctioning circulation due to Chiari I Malformation.

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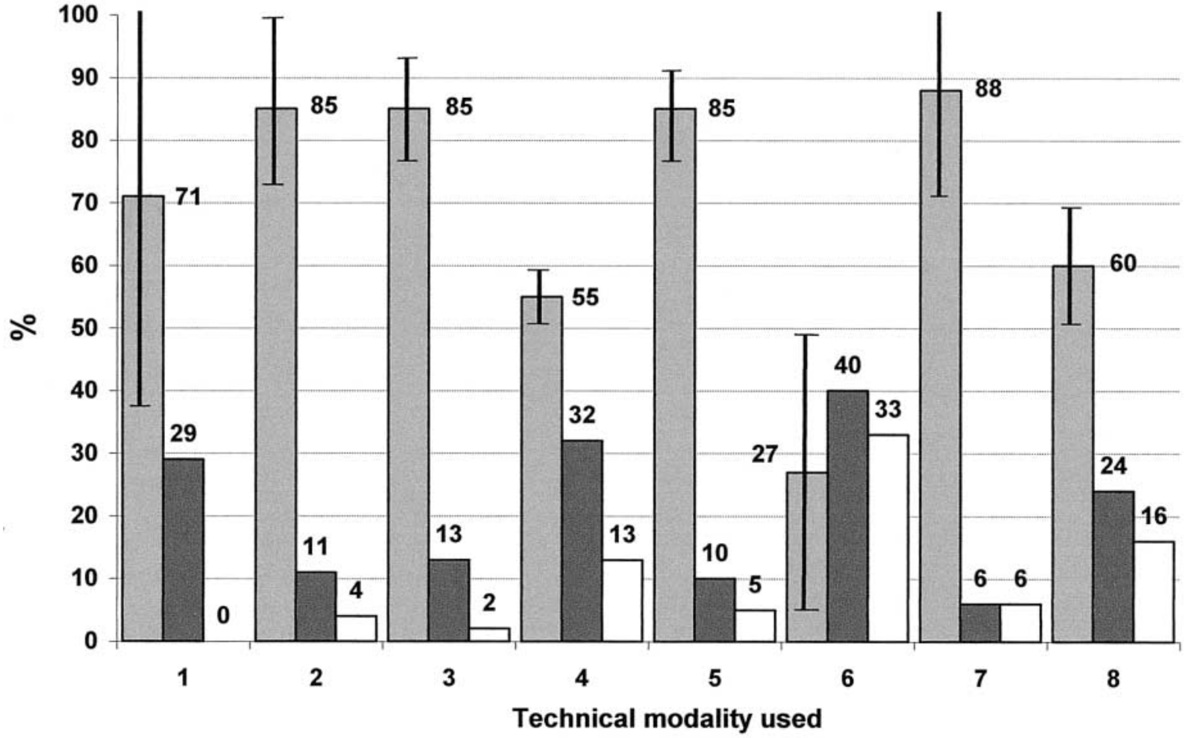

Fig. 9. Clinical outcome according to the various technical modalities used for *Chiari Malformation type I associated with syringomyelia*. *FMD* Foramen Magnum Decompression, *DO* Dural Opening, *DU* Duroplasty, *AO* Arachnoid Opening, *RT* Resection of Tonsils, *SH* Shunt.
■ Improved, ■ Stabilized, □ Aggravated
1 FMD WITHOUT DO, *2* FMD WITH DO (outer layer), *3* FMD + DO + DU (arachnoid integra), *4* FMD + DO + AO + DU, *5* FMD + DO + AO + RT + DU, *6* FMD + DO + AO, *7* FMD + DO + DU + SH, *8* SHUNT
Best results were achieved with technical modality N°3, worst results were when using techniques N°4 or 6

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