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Chiari type I anomalies in children and adolescents: minimally invasive management in a series of 53 cases

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Abstract The authors studied the role of the sole posterior fossa bony decompression in the management of symptomatic children affected by Chiari type I anomalies. The series in the pediatric literature on this subject were reviewed and compared with that presented in this article. From May 1994 to December 1998, 53 patients (3 months to 26 years) were observed. They were divided into: asymptomatic patients (27), who received no surgical treatment and were only subject to clinical observation; symptomatic patients (brain stem compression 16, syringomyelia 10, including 7 with holocord). All the symptomatic patients were treated with the same surgical approach: bony decompression of posterior fossa with removal of the posterior arch of C-1 and the outer layer of the dura without dural opening. In all 16 (100%) of the 16 patients with brain stem compression the symptoms resolved or improved; in patients with syringomyelia the symptoms were resolved or improved in 94.4% of cases. Two children required further surgery after 13 and 24 months, respectively. This series seems to demonstrate that even a simple extradural surgical approach, with a lower rate of postoperative complications and short stay in hospital, is sufficient to arrest the disease and to improve the symptomatology in a high percentage of cases (97.2%), which is comparable to that achieved with other, more aggressive, procedures.

Keywords Syringomyelia · Chiari I malformation · Foramen magnum decompression · Children · Scoliosis · MRI

Introduction

In 1891 and 1896, Chiari [17, 18, 19, 20] described a series of anomalies of the caudal cerebellum and brain stem on the basis of autopsy observations. The type I Chiari anomaly was "Elongation of the tonsils and medial part of the inferior lobes of the cerebellum which go along the medulla into the cervical canal." There were no anomalies of the brain stem. The IV ventricle was always in a normal position.

Long before Chiari's descriptions, in 1546, Estienne had described a cystic dilatation of the spinal cord, and in 1824 Ollivier d'Angers 69] used the term syringomyelia. The association of hydromyelia and Chiari anomalies was noted by Russel and Donald in 1935 [76]. Williams [90, 92] divided these cysts into those that communicate with CSF pathways and those that do not. Noncommunicating syringomyelia may be related to neoplasms, trauma, vascular accidents and spinal arachnoiditis. Barnett [7] in 1973 classified syringomyelia into: (1) communicating: from congenital or acquired anomalies; (2) posttraumatic; (3) from spinal arachnoiditis; (4) neoplastic; (5) idiopathic.

There are many hypotheses about the pathogenesis of the Chiari "malformation," and the surgical treatment varieswidely. We studied the role of bony decompression as the sole therapy in the management of this disease in a pediatric population.

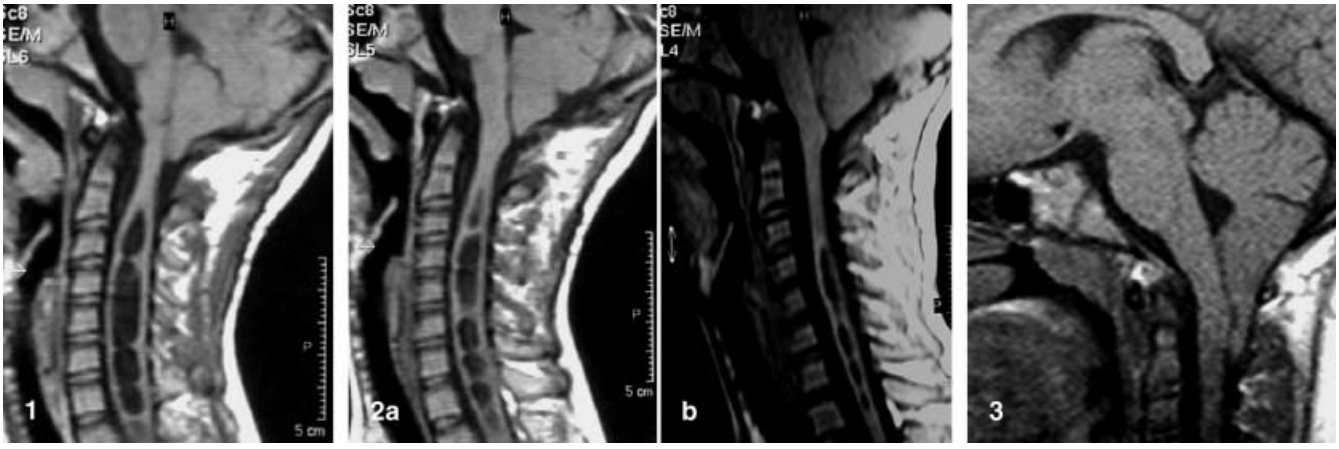

Fig. 1 MR sagittal T1-weighted image with typical Chiari type I with a large syringomyelia

Fig. 2 **a** Preoperative MR imaging (sagittal, T1-weighted) of the 10-year-old girl with scoliosis, showing a Chiari type I malformation with a huge syringomyelia. Note the very small subarachnoid space around the inferior cerebellum, medulla and upper cervical cord. **b** Postoperative (3 months) MR imaging (sagittal, T1-weighted) of the same child after posterior fossa (PF) bony decompression as the sole treatment. Note the increased size of the subarachnoid space, anteriorly and posteriorly, and the reduction in size of the intramedullary cavities

Fig. 3 Chiari type I malformation with "overcrowding" in the PF

Pathogenesis

The Chiari anomalies are associated with syringomyelia in 20–75% of cases [50, 71, 77, 89, 91], (Fig. 1). For this frequent association, Gardner [34, 35, 36, 37], in 1965, worked out the hydrodynamic theory, supposing that the lack of perforation of the rhomboencephalic roof and the persistence of patency between the IV ventricle and the central canal of the spinal cord could be the cause of the malformation. This theory does not explain the lack of patency between the IV ventricle and the central canal, which was the case in 20% of the cases in the University of Iowa series [59], and the existence of spinal cord cysts associated with tumors and trauma or lumbo-peritoneal shunts.

In 1980, Williams [92] proposed that the CSF obstruction at the level of the foramen magnum produced a dissociation of pressure between the cranial and spinal cerebrospinal fluid compartments, which contributes to the syrinx formation. The Valsalva maneuver causes the CSF pressure to increase in the spinal compartment following congestion of epidural veins. The subarachnoidal spinal space is constricted, and the pressure wave rises toward the intracranial cavity. If this balance is modified by scars or tissues inside the foramen magnum, a pressure difference is created between the intracranial and the intraspinal subarachnoidal space, causing alternative pathways either through the obex, if patent, or following the descent of the hindbrain. The syringomyelia develops from craniospinal fluid pressure dissociation, with enlargement of the cavity as a result of a valve-like mechanism produced by anomalies around the foramen magnum [93].

Welch et al. [88] showed a descent of the cerebellar tonsils in patients who underwent lumbo-peritoneal shunting, and therefore assumed that difficulty in balancing intracranial and intraspinal pressure could cause such anomalies; these authors stressed the possibility of a secondary, and not only of congenital, origin of Chiari type I disease. Pathological features of Chiari type I anomalies could be the result of an anomalous pressure gradient. Williams found a high incidence of birth trauma in such patients, which could cause a subarachnoidal hemorrhage with subsequent arachnoidal adhesions, causing an alteration of CSF pressure balance. CSF pressure measurements in the intracranial and intraspinal spaces showed that in many patients the reduction phase of intracranial pressure was clearly prolonged: in severely affected patients a pressure gradient was present even in resting conditions. As reported by many authors, a prolapse of the cerebellar tonsils through the foramen magnum is observed in children with ventriculo-peritoneal or lumbo-peritoneal shunts [21, 41, 43, 67]. Fisher described syringomyelia in a patient with a lumbo-ureteral shunt [32]. A suggestive theory explaining the build-up of a syringomyelic cavity, based on cine-MRI and intraoperative ultrasonography, was described by Oldfield [68]. In his opinion, the Chiari anomalies could induce a piston-like motion affecting the cerebellar tonsils, producing a systolic wave in the CSF flow, acting on the spinal cord and inducing CSF leakage through interstitial and perivascular spaces. According to this author, posterior fossa decompression alone could be successful (Fig. 2a, b). Some authors [2, 4, 8, 63, 79, 83, 87, 97], studying the morphology of posterior fossa, have shown that occipital hypoplasia with a decrease in posterior fossa volume results in overcrowding of posterior fossa struc-

tures and induces herniation of lower hindbrain contents through the foramen magnum. A posterior fossa hypoplasia could be responsible for Chiari disease, with a disproportional growth of the posterior fossa contents. For this reason, Sahuquillo proposed a surgical technique for posterior fossa reconstruction in the management of this disease: a large craniectomy with dural graft saving the arachnoid, which led to the formation of an artificial cisterna magna [78]. A few familial cases seem to ssupport the idea of this etiology [3]: "An evaluation of posterior fossa morphology in these patients suggests that occipital dysplasia and overcrowding of posterior fossa contents may be the substrate for both familial and sporadic case of Chiari type I malformation and suggests a unifying concept of origin." A review of the literature suggests that these cases are rare but underreported [18, 23, 82]. The external compression upon the cerebellum and brain stem could play a role in the difficulties of CSF circulation through the foramen magnum. Nishikawa [63] analyzed the posterior fossa morphology in 50 control cases and in 30 sporadic cases of Chiari type I anomalies. The author demonstrated an underdevelopment of the occipital bone with compression of cerebellum and brain stem that is herniated through the foramen magnum. The measurement of the angle between the Twining line and the tentorium of cerebellum performed by the author showed increased values in patients affected by Chiari disease. We have found the same results in our series of children, with a mean angle of 40.2° (Fig. 3).

For this reason it seems to be reasonable to assert that occipital hypoplasia, posterior fossa overcrowding, small posterior fossa, underdevelopment of occipital bone, vascular malformations, posterior fossa masses, hydrocephalus, lumbo-peritoneal shunts, craniofacial and posterior cranial base malformations in association with a craniospinal pressure dissociation all contribute to the pathogenesis of Chiari type I anomalies [97].

Patients and methods

We studied 53 children (21 girls, 32 boys) observed from May 1994 to December 1998 in the Division of Pediatric Neurosurgery at the Children's Hospital of Turin (O.I.R.M.). The average age at the time of diagnosis was 8 years, ranging from 3 months to 26 years (Tables 1: all patients, 2: symptomatic patients, 3: asymptomatic patients).

It is very important to note that the rate of very young children is quite substantially higher (44.4% vs 15.4%) in the asymptomatic than in the symptomatic group. This suggests there is an evolution between a "morphological" Chiari, without symptoms, and a clinical Chiari with symptoms and signs. All patients underwent spinal MRI.

We classified the patients into three groups (Table 4). We chose this classification to evaluate whether it was possible to correlate the clinical evolution to prognosis.

Group I patients did not undergo surgery but were subject to clinical observation (Table 5) during a period ranging from 1 to 12 months.

As shown in Table 5, Chiari type I anomalies were associated with other pathologic conditions in 70.4% of children of this group. Nine children had a shunted hydrocephalus with various VP shunts. No lumbo-peritoneal shunt was performed. Chiari type I anomalies were found in 5 children with cranio-facial syndromes (Crouzon, etc.) with a huge disproportion in posterior fossa volume with median bony spur (Fig. 4 a, b). All these children were operated on with various cranio-facial techniques, without any posterior procedure. Chiari type I anomalies were found in 4 epileptic children during a routine MR study. One child with a closed spinal dysraphism and lipomeningomyelocele showed MRI features of Chiari type I anomalies.

None of these children developed symptoms or signs related to compression of brain stem or syringomyelia; in any case the follow-up period of this group of patients is too short to make any prognostic judgement.

Table 1 All patients

Age	0–2 years	3–5 years	6–10 years	11–15 years	>15 years
n=53	16	8	9	9	11
%	30.2%	15.1%	17%	17%	20.7%

Table 2 Symptomatic patients

Age	0–2 years	3–5 years	6–10 years	11–15 years	>15 years
n=26	4	5	5	6	6
%	15.4%	19.3%	19.3%	23%	23%

Table 3 Asymptomatic patients

Age	0–2 years	3–5 years	6–10 years	11–15 years	>15 years
n=27	12	2	5	3	5
%	44.4%	7.5%	18.5%	11.1%	18.5%

Table 4 Classification

Group I	Asymptomatic	27	50.9%
Group II	Symptoms of brain stem compression at diagnosis	16	30.2%
Group III	Presence of syringomyelia at MRI	10	18.9%

Table 5 Features of asymptomatic patients (27 cases)

Average age (range)	7.4 years (3 months to 17 years)
Sex	12 Female 15 Male
Associated conditions: 19 cases (70.4%	Hydrocephalus: 9 Cranio-facial syndromes: 5 Epilepsy: 4 Occult spinal dysraphism: 1

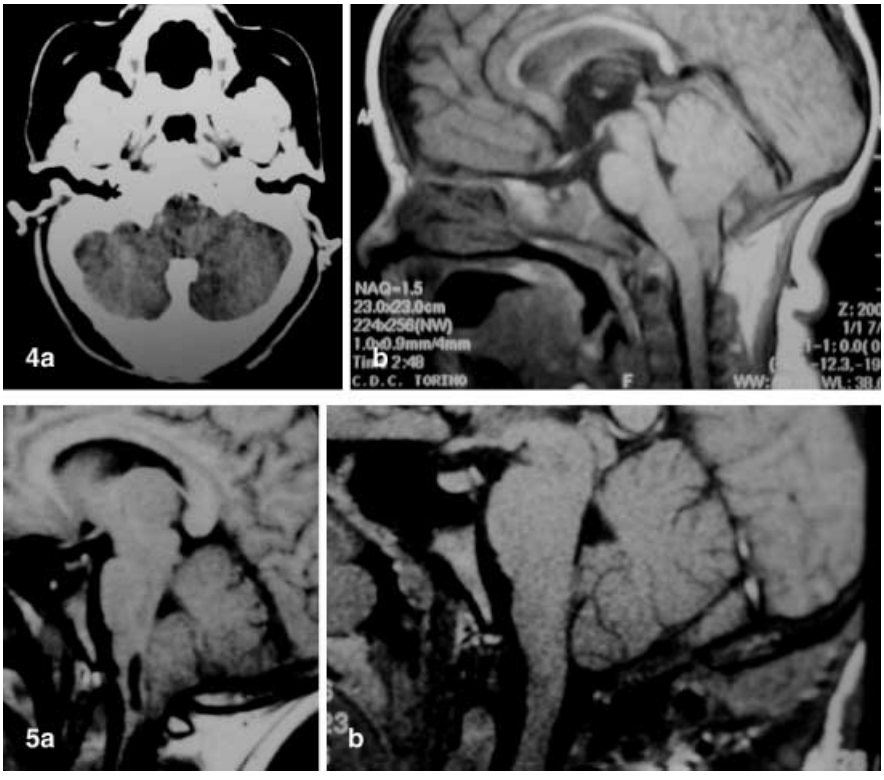

Fig. 4 **a** CT scan of a patient affected by Crouzon syndrome, with a median bony spur in the PF. **b** Sagittal MR of the same child with cranio-facial malformation and volume decrease in the PF

Fig. 5 **a** Preoperative MR with a cavity into the medulla in a 12-year-old boy. **b** Postoperative (1 year) MR imaging of the same child with complete disappearance of the cavity

Table 6 Features of symptomatic patients (26 cases)

Average age (range)	11.4 years (7 months to 26 years)
Sex	10 Female 16 Male
Associated conditions: 11 cases (40.3%)	Cranio-facial syndromes: 6 Hydrocephalus: 3 Precocious puberty: 2

Symptomatic patients showed the characteristics reported in Table 6.

Symptoms related to brain stem compression in group II patients are reported in Table 7. Neck pain, followed by vertigo and headache turned out to be the most frequent symptoms. There were no cases of scoliosis.

The clinical conditions of the patients with syringomyelia are reported in Table 8. The syringomyelic cavity involved the entire spine in 7 cases. The levels affected in the other 3 patients were C2–4, C3–7 and C3–T4.

Group III patients more frequently had a deficit in sensitive pathways, such as paresthesia and sensory loss, than did those in group II. Scoliosis and hypotrophy of various muscular groups were present in 30% of cases. The rate of headache and neck pain were similar in both groups.

The average duration of symptoms differed markedly between groups II and III, as reported in Table 9.

It is evident that children presenting with syringomyelia at diagnosis have a longer duration of symptoms and, of course, a longer evolution of the disease than children with symptoms of brain stem compression. Consequently, it seems to be very important that these symptoms are recognized early to avoid the slow formation of intramedullary cavities.

Table 7 Group II, brain stem compression (16 cases)

Symptoms	*n*	%
Neck pain	7	43.8%
Vertigo	5	31.3%
Headache	5	31.3%
Numbness	4	25%
Swallowing difficulties	3	18.8%
Apnea	3	18.8%
Opisthotonos	3	18.8%
Brachialgia	2	12.5%
Hyposthenia	1	6.3%
Spasticity	1	6.3%
Hemifacial pain	1	6.3%

Table 8 Group III, syringomyelia (10 cases)

Symptoms	*n*	%
Numbness	5	50%
Sensory loss	4	40%
Neck pain	4	40%
Vertigo	4	40%
Scoliosis	3	30%
Hyposthenia	3	30%
Headache	3	30%
Muscle hypotrophy	3	30%
Swallowing difficulties	2	20%
Limb pain	1	10%

Table 9 Duration of symptoms in group II and III

	Minimum	Maximum	Average
Group II (brain stem compression)	1 month	4 years	14.3 months
Group III (syringomyelia)	3 months	12 years	43.2 months
All	1 month	12 years	28.7 months

All symptomatic children were operated on in the prone position with the same procedure: bony decompression of the posterior fossa without dural opening. After a midline incision and muscular dissection, the occipital bone, the atlo-occipital membrane and the posterior arch of C-1 were exposed. No dissections were performed into the C-2 muscles, to avoid later problems in vertebral dynamics. Then we performed a 3×3 cm suboccipital craniectomy for wide opening of the foramen magnum. The posterior arch of C-1 was removed. The fibrous band corresponding to the thickened atlo-occipital ligament was completely removed together with the outer layer of the dura. No dural opening and no dural patches were used. Closure was assured by reabsorbable stitches. The median hospital stay was 3 days (Fig. 5 a, b).

No mortality and no serious morbidity were observed in this series of children. We observed 2 wound infections without meningitis or bony involvements, which needed only minor local treatment.

Results

Patients in group I (asymptomatic patients) were observed over a follow-up period ranging from 1 to 12 months. None developed symptoms or signs related to compression of brain stem or syringomyelia. Group II and III patients were operated on. The average follow-up after surgery was 21.6 months (6–60 months). There were no serious postoperative complications. The procedure entailed 3 days in hospital.

All patients in group II improved soon after surgical posterior fossa decompression.

Clinical results are reported in Table 10. The results show complete resolution of the symptoms in almost all patients. Only in 3 cases was total recovery not achieved, and some improvement was seen in each of these patients.

Patients suffering from syringomyelia (group III) completely recovered from vertigo; headache, weakness and muscle hypotrophy improved in 100% of cases, paresthesia in 80%. Only in 25% of the patients were their problems of sensory loss resolved completely. Scoliosis improved in 66.6% of cases (Table 11).

Intramedullary cavities disappeared in 80% of the patients within a few months after surgery (Fig. 6 a, b). In this group of patients, 2 children required further surgery related to the persistence of syringomyelia in spite of initial clinical improvement. Surgery was carried out after 13 and 24 months, respectively. During the second surgical approach the dura mater was opened, the cerebellar tonsils coagulated and a duraplasty was made. No sign of bone regrowth was observed at the previous site of craniectomy. Later neuroradiological controls showed a progressive reduction of the size of the syringomyelic cavities, although the did not disappear. The same patients showed an improvement in swallowing and in muscular strength.

Table 10 Clinical results (group II, brain stem compression)

Symptoms	*n*	Resolved	Improved	Unchanged
Neck pain	7	6 (85.7%)	1	–
Vertigo	5	5 (100%)	–	–
Headache	5	4 (80%)	1	–
Numbness	4	4 (100%)	–	–
Swallowing difficulty	3	3 (100%)	–	–
Apnea	3	3 (100%)	–	–
Opisthotonos	3	3 (100%)	–	–
Brachialgia	2	2 (100%)	–	–
Hypostenia	1	1 (100%)	–	–
Spasticity	1	–	1	–
Hemifacial pain	1	1	–	–

Table 11 Clinical results (group III, syringomyelia)

Symptoms	*n*	Resolved	Improved	Unchanged
Numbness	5	3 (60%)	1	1
Sensory loss	4	1 (25%)	3	–
Neck pain	4	3 (75%)	1	–
Vertigo	4	4 (100%)	–	–
Scoliosis	3	–	2	1
Hyposthenia	3	2 (66.6%)	1	–
Headache	3	1 (33.3%)	2	–
Muscle hypotrophy	3	2 (66.6%)	1	–
Swallowing difficulties	2	1 (50%)	1	–
Limb pain	1	–	1	–

Discussion

There are many surgical techniques for the treatment of Chiari type I anomalies, and also many controversies. The modern surgical procedures all share the aim of decompressing the inferior cerebellum and cervico-medullary region [15, 16] in order to restore a normal CSF circulation at the level of the foramen magnum [33, 74, 94]. The Pediatric Section of the American Association of Neurological Surgeons laid down the following principles in 1988 [39]: surgical decompression was indicated for brain stem or cranial nerve dysfunction, and otherwise there was no indication for prophylactic decompression in asymptomatic children. There was some disagreement about the patients with isolated headache and a few or mild symptoms.

Posterior fossa decompression and cervical laminectomy make up the basic treatment, but there are many other controversial procedures after the bone decompres-

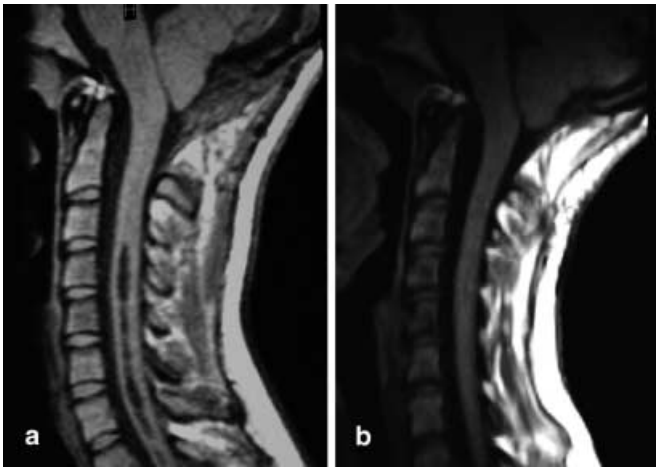

Fig. 6 a Preoperative MR: few Chiari type I anomalies, but cervical syringomyelia. **b** Postoperative MR (3 years): complete disappearance of syringomyelia

sion. The dura can be opened by a Y-shaped incision, and a dural graft could be recommended to maintain the decompression. Many intradural procedures have been described: dissection of the arachnoid overlying the tonsils [25, 27, 58]; coagulation of herniated cerebellar tonsils that respect the integrity of the pia and arachnoid [94]; resection of cerebellar tonsils [31, 91] with a subpial approach when there are very highly gliotic tonsils not reduced by coagulation alone [9, 70]; and obex occlusion with a piece of muscle [7, 11, 22, 32, 34, 35, 37, 41, 43, 42, 55, 56, 59, 66, 72].

Some authors consider it useful to place a shunt between the IV ventricle and the subarachnoidal spaces [22, 32, 58, 70, 75]; others prefer to perform a syringostomy by way of a myelotomy [31, 75]. Abbe and Coley, in 1892 [1], were the first to propose myelotomy for the surgical management of syringomyelia. Syringo-subarachnoid shunting [45, 46, 85, 86] has also been described, especially in the Japanese literature. Some authors have considered a syringo-peritoneal or pleural shunt [6] more effective, owing to a higher differential pressure compared with the subarachnoidal space.

Among other treatments described in the literature we have found section of the filum terminale, as reported by Filizzolo [30] in a comparison with PF decompression, and percutaneous aspiration of the cyst when previous surgical procedures have failed [80].

The extreme variability of the procedures applied confirms the lack of agreement on surgical policies. The advent of MRI has allowed the anatomical features of the Chiari type I anomalies to be much more readily identified. MRI has also been helpful in showing associated syringomyelia. The involvement of MRI in diagnosis modifies the approach to this pathology, allowing minimally invasive diagnosis and identifying cases at an early stage and lowering the age at diagnosis of the disease [28, 29, 51, 60, 73, 84]. Milhorat asserts that with MRI criteria it is possible to classify intramedullary cavities into communicating, noncommunicating and atrophic types, with the possibility of different treatments [60].

Recently cine-MRI has added a significant improvement, especially in the evaluation of results in terms of pulsatility of CSF through the posterior cerebellum [97] (Fig. 7 a, b).

There is now general agreement on the importance of decompressing the cranio-cervical junction [9, 13, 14, 27, 73]. The goal of surgical treatment is to restore CSF flow, thus re-establishing a pressure balance between the intracranial and intraspinal subarachnoidal spaces.

The foramen magnum decompression with removal of the outer dural layer only as treatment for syringomyelia occurring with Chiari type I anomalies was first described by Isu in 1993 [47]. This author reports seven adult patients treated with bony posterior fossa decompression alone without opening of the dura, with good clinical results throughout a postoperative follow-up of 2 years. In 1981 Logue recommended “simple decompression with preservation of the arachnoid membrane, combined with syringostomy in certain cases. The occlusion of the central canal appears to have no greater influence on the progression of the disease” [53]. Yundt, in 1996, said that “in young children under 2 years of age severe arachnoidal adhesion is exceedingly rare and moreover there is no evidence that lysis of the arachnoidal adhesion adds significantly to decompression of the hind brain” [96]. Recently Krieger [49] has reported a series of 31 children who underwent bony decompression of the posterior fossa with dural opening but without any intradural procedures. The dura was left open and over-

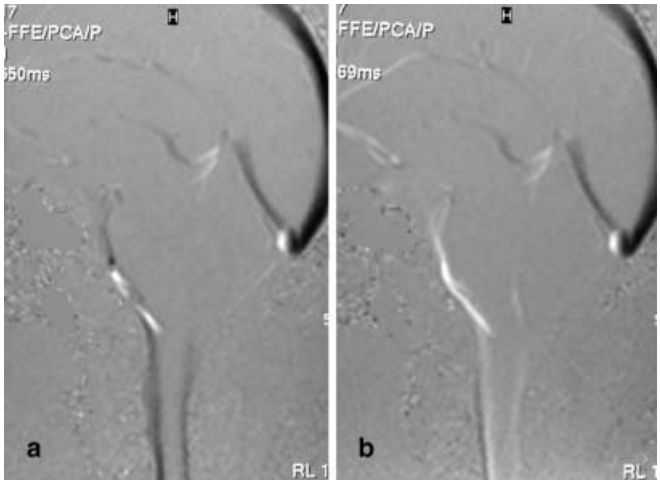

Fig. 7 a, b Postoperative cine-MR (follow-up 1 year) showing restoration of CSF flow from intracranial to perimedullary subarachnoid spaces

laid with oxidized cellulose. The follow-up period ranged from 15 to 93 months. In 88% of children a significant reduction in the size of the syrinx was obtained.

In spite of these good clinical results, the author reports a high incidence of postoperative complications: 25% of patients with headaches, 16% with nausea and vomiting, and 10% with postoperative CSF leak.

Zerah [97], in 1999, reported the largest series of pediatric patients with Chiari type I anomalies and syringomyelia with a long period of follow-up. In this article the author stresses the importance of a simpler operative procedure in spite of the same quality of results.

All children were operated on by a posterior fossa decompression with various intradural procedures in a period ranging from 1985 to 1993. Since 1994 a simpler technique was employed in 79 children (PF decompression without intradural procedures, with an extra-arachnoidal dural opening). The author reports a 95% clinical improvement or stabilization rate among these children.

These results are comparable with those in the modern literature and with those published in the present paper. However, there is a very high rate of complications, originating especially from the dural opening, with aseptic meningitis in 22%, pseudomeningocele in 4%, fistula in 2%, and acute hydrocephalus in 1%.

A simpler posterior fossa decompression without dura opening and without duraplasty has the advantage of being easy and of requiring a very short hospital stay with the same results as reported in this paper. This technique prevents excessive venous bleeding at the opening of the dura [96], pseudomeningocele [10, 97], fluid collection at the operative wound [53, 97], aseptic meningitis [48, 97], spinal cord injury after insertion of the catheter in the cavity of the spinal cord [46, 47], late arachnoid adhesions [9, 55, 57] and hydrocephalus [97]. The enlargement of the posterior fossa frees the cerebellar tonsils and helps to restore normal cerebrospinal fluid dynamics [40].

The analysis of our data indicates very encouraging results in patients with brain stem compression symptoms: 94.3% of pre-existing symptoms disappear, and any remaining symptoms improve. A very positive fact in patients affected by syringomyelia is that progression of the disease has never been observed. In this group, 53.1% of symptoms disappeared, 40.6% improved and 6.3% did not change. The volume of the cavities decreased until their disappearance in 80% of children operated on with this procedure. Syringomyelia and scoliosis are closely correlated, especially in childhood. As reported in the literature [12, 38, 45, 52, 65, 95, 97], it is possible to stop the progression of the vertebral disease, improving symptoms related to syringomyelia. PF decompression seems to be more effective than shunting procedures in this field [64, 97]. Consequently, an MRI examination of the entire spinal cord and posterior fossa is mandatory in cases of juvenile or idiopathic scoliosis [5, 64, 81, 97].

An analysis of the literature on pediatric patients only is reported in Table 12.

Hoffman (1987) presented a series of 47 cases of hydrosyringomyelia: this included 12 patients affected by Chiari type I anomalies. The author noted that posterior fossa decompression with dural grafting alone was not sufficient. Patients improved with plugging of the obex. The Gardner procedure (DPF, plugging obex) was the one most commonly used in his group of patients and resulted in improvement in over 70% of patients [43].

Dyste (1988), who analyzed 16 patients with Chiari anomalies (11 with Chiari I), said that “the group of children who obtained an asymptomatic result had several features: their preoperative deficits were less extensive (none had scoliosis, ataxia, nystagmus or atrophy). In addition there was a significant difference in both the duration of symptoms and the age of these patients.” The author thinks that a child with symptomatic Chiari should undergo immediate surgery in the hope of optimizing the outcome [25].

Isu presented a series of 17 cases of hydrosyringomyelia in children and adolescents, 16 of whom were treated by placement of syringo-subarachnoid shunts. In 88% of the patients the neurological symptoms improved. He thought that such a surgical treatment was effective [45].

Muhonen (1992), in his article on scoliosis in pediatric Chiari anomalies, noted that the preoperative presence of hydrosyringomyelia did not impair postoperative resolution of the scoliosis [61].

Nagib [62] (1994), who studied 16 cases of Chiari type I, stated that the more significant prognostic indicators are the actual clinical signs and symptoms, with scoliosis (<30°), headache and cervical pain and sleep apnea having the most favorable response to the decompressive procedure. Motor deficits and sensory deficits are unlikely to improve.

Yundt [96] (1996) presented seven cases of posterior fossa decompression without duraplasty. All patients improved. He affirms that before 2 years of age severe arachnoidal adhesion are rare. The dura mater along the posterior rim of the foramen magnum was abnormally thick. Incision of the thickened outer layer of the dura may result in visible expansion of the dural sac.

Park [70] treated 68 consecutive patients with Chiari type I anomalies, utilizing a posterior fossa decompression in association with an intradural procedure. He demonstrated that standard bony and dural decompression of the foramen magnum with various intradural procedures restores CSF circulation across the posterior cerebellum. In 93% of cases there was a clear improvement of symptoms.

On review of the literature it is evident that more pronounced clinical improvements can be achieved in pediatric than in adult series, with more than 80% of pediatric patients improving after surgery. Dyste [25] stressed the importance of the duration of symptoms: the longer the clinical history, the worse the prognosis.

Table 12 Pediatric literature

Reference	Patients	Duration of symptoms	Technique	Syrinx	Morbidity	Results
[41]	47 cases 6 months to 18 years 12 Chiari I (5–17 years)	20 months (1 day to 36 months)	Suboccipital craniectomy and plugging obex: 5 Suboccipital craniectomy and dural grafting: 2 Shunt syrinx: 3 Gardner procedure and syrinx shunt: 1 Suboccipital craniectomy and syrinx shunt: 1	12	No	Follow-up: minimal 1 year 83.3% improved
[25]	16 cases 1–19 years 11 Chiari I 3 Chiari II 2 Chiari III		Suboccipital craniectomy + Dural grafting + C1 laminectomy + Dissection arachnoid adhesions + Plugging obex (if central canal patent) 15 Transoral odontoid resection: 3 Cystosubarachnoid shunt: 2 Occipito cervical fusion: 4	16	No	Follow-up: 43 months Resolved 37.5% Improved 50% Stable 12.5%
[24]	7 cases	<12 years	Suboccipital craniectomy + Dural grafting + C1 laminectomy + Syringosubarachnoid shunt: 7			100% improved or unchanged
[46, 47]	17 cases: <20 years	6 years 1 month to 6 years	Syringosubarachnoid shunt: 16 Suboccipital craniectomy without plugging obex: 1 Soc + syringostomy: 1 Syringostomy: 1 VP shunt: 1 (3 patients each had 2 operations)	17	1 shunt failure 1 VP shunt for secondary hydrocephalus	Follow-up: 4 years 88% improved
[61]	11 cases: 2–15 years 9 Chiari I 2 Chiari II 5/11: basilar invagination	1–5 years	Suboccipital craniectomy + Dissection of arachnoid adhesions: 11 Plugging obex: 6 Shunt V4-subarachnoid space: 11 Ventral – transoral decompression: 5 + posterior fusion: 3	8 87.5% resolved	No	Follow-up: 35 months (20–68 months) Symptoms: Resolution: 64% Improved/stable: 36% Scoliosis: Improved: 73% Stable: 9% Worsened: 18%
[62]	16 cases: 4–14 years	3 months to 2 years	Suboccipital craniectomy + C1(C2) laminectomy + Arachnoidal retraction + Duraplasty + Syringo-subarachnoid shunt: 16	5 100% resolved	1 CSF leak	Improved: Scoliosis: 60% Headaches: 86% Sleep apnea: 100% Dysphonia : 100%

Table 12 (continued)

Reference	Patients	Duration of symptoms	Technique	Syrinx	Morbidity	Results
[96]	7 cases: 3 Chiari I 2 Chiari II 2 achondroplasia	3 months to 2.5 years	Suboccipital cramiectomy + C1 laminectomy: 7	0	No	Follow-up: 22 months (4.5 months to 4 years) 100% improved
[70]	68 cases: 17 months to 21 years		Suboccipital craniectomy + C1 laminectomy + Duraplasty: 68 Coagulation of cerebellar tonsils: 40 Shunt V4-cervical subarachnoid space 32 Plugging obex: 23	40 80% resolved	Subdural hygroma: 1% Nausea, vomiting: 22% Headache: 10%	Follow-up: 6–70 months 100% improved
[49]	31 cases: 6 months to 18 years		PF decompression with dural opening: 31	26 88% resolved	Hydrocephalus: 3% Headache: 26%	Follow-up: 46 months
[97]	188 cases: 7 months to 15 years	6 months (0–24 months)	1985–1993: PF decompression + dural opening + stent + patch: 105 1994–1999: PF decompression + extra-arachnoidal dural opening: 79	188 Improvement: 69% Stabilization: 28% Worsening: 3%	Aseptic meningitis: 22% Wound pb.: 6% Pseudo-meningocele: 4% Fistula: 2% Meningitis: 1% Hydrocephalus: 1%	Follow-up: 5.4 years 95% improved
Our series	26 cases: 3 months to 26 years	29 months (1 month to 12 years)	PF decompression without dural opening: 26	10 80% resolved	Wound pb.: 7.7%	Follow-up: 22 months (6–60 months) 97.2% improved

A similar outcome was also obtained in our patients. In group III, carrying a less favorable prognosis, the symptoms lasted longer. According to Nagib [62], sensory deficits were scarcely influenced by treatment, while in contrast to his series, 66% of our patients achieved motor improvement.

In conclusion, our results and the analysis of modern literature indicate that an even simpler treatment of Chiari type I anomalies is possible in children. Posterior fossa decompression, with C-1 laminectomy without opening of the dura, without duraplasty and without any intradural procedures, seems to be effective, with a lower incidence of postoperative complications and a comparable percentage of clinical improvement.

A longer period of follow-up is necessary to confirm the complete efficacy of this approach.

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