Chiari I malformation with and without basilar invagination: a comparative study

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OBJECT Chiari I malformation is the most common craniocervical malformation. Its combination with basilar invagination in a significant proportion of patients is well established. This study presents surgical results for patients with Chiari I malformation with and without additional basilar invagination.

METHODS Three hundred twenty-three patients underwent 350 operations between 1985 and 2013 (mean age 43 ± 16 years, mean history of symptoms 64 ± 94 months). The clinical courses were documented with a score system for individual neurological symptoms for short-term results after 3 and 12 months. Long-term outcomes were analyzed with Kaplan-Meier statistics. The mean follow-up was 53 ± 58 months (the means are expressed $\pm$ SD).

RESULTS Patients with ($n = 46$) or without ($n = 277$) basilar invagination in addition to Chiari I malformation were identified. Patients with invagination were separated into groups: those with ($n = 31$) and without ($n = 15$) ventral compression by the odontoid in the foramen magnum. Of the 350 operations, 313 dealt with the craniospinal pathology, 28 surgeries were undertaken for degenerative diseases of the cervical spine, 3 were performed for hydrocephalus, and 6 syring catheters were removed for cord tethering. All craniospinal operations included a foramen magnum decompression with arachnoid dissection, opening of the fourth ventricle, and a duraplasty. In patients without invagination, craniospinal instability was detected in 4 individuals, who required additional craniospinal fusion. In patients with invagination but without ventral compression, no stabilization was added to the decompression. In all patients with ventral compression, craniospinal stabilization was performed with the foramen magnum decompression, except for 4 patients with mild ventral compression early in the series who underwent posterior decompression only. Among those with ventral compression, 9 patients with caudal cranial nerve dysfunctions underwent a combination of transoral decompression with posterior decompression and fusion.

Within the 1st postoperative year, neurological scores improved for all symptoms in each patient group, with the most profound improvement for occipital pain. In the long term, late postoperative deteriorations were related to reobstruction of CSF flow in patients without invagination (18.3% in 10 years), whereas deteriorations in patients with invagination (24.9% in 10 years) were exclusively related either to instabilities becoming manifest after a foramen magnum decompression or to hardware failures. Results for ventral and posterior fusions for degenerative disc diseases in these patients indicated a trend for better long-term results with posterior operations.

CONCLUSIONS The great majority of patients with Chiari I malformations with or without basilar invagination report postoperative improvements with this management algorithm. There were no significant differences in short-term or long-term outcomes between these groups. Chiari I malformations without invagination and those with invaginations but without ventral compression can be managed by foramen magnum decompression alone. The majority of patients with ventral compression can be treated by posterior decompression, realignment, and stabilization, reserving anterior decompressions for patients with profound, symptomatic brainstem compression.

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KEY WORDS basilar invagination; Chiari I malformation; syringomyelia; craniospinal instability; Klippel-Feil syndrome

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THE management of Chiari I malformations and basilar invaginations has been quite controversial in recent years. Issues such as whether the dura mater should be opened, if duraplasties are required, or whether the arachnoid should be opened have been discussed for patients with Chiari I malformations, and the necessity for transoral decompressions in basilar invagination has been questioned, emphasizing that realignment and C1–2 fusion is sufficient. Although most patients with basilar invagination will also demonstrate a Chiari I malformation, almost all studies on basilar invagination concentrate on the bony realignment and fusion, but do not address the management of the associated Chiari I malformation.

In this paper the results of surgical treatment for patients with Chiari I malformations with and without basilar invagination presenting between 1985 and 2013 are documented and compared.

Methods

Between 1985 and 2013 surgery was performed in 323 patients with a Chiari I malformation with or without basilar invagination in the neurosurgical clinics of Nordstadt Krankenhaus in Hannover (1985–2003) and at the Christliches Krankenhaus in Quakenbrück (2004–2013), Germany. In 1991 a spinal cord database was started, and since then information has been entered for all patients with craniocervical and spinal cord pathologies. All patients were followed up and data incorporated into the database continuously up to the present day. Nineteen patients underwent operation between 1985 and 1990 and were incorporated retrospectively. For this publication, all data up to 2013 were analyzed. In this cohort, 277 patients with Chiari I malformation without basilar invagination underwent 300 operations, and 46 patients with basilar invagination in addition to the Chiari I malformation underwent 50 operations.

Apart from general patient data and associated features of this pathology, the neurological examinations before surgery, before discharge from the hospital, after 3 months postoperatively, and yearly thereafter were documented

for individual symptoms according to a scoring system devised by Klekamp and Samii,³¹ adding a score developed subsequently for swallowing function²⁹ (Table 1). This scoring system was used for descriptive purposes only, to illustrate postoperative courses of individual symptoms.

All patients underwent MRI of the head and cervical spine. Basilar invagination was defined as the odontoid crossing the Chamberlain line between the hard palate and posterior rim of the foramen magnum for more than 5 mm.^{10,54} Preoperative CT scans and radiographs of the cervical spine in ante- and retroflexion were obtained in each patient with basilar invagination, and combined with CT angiography in patients with narrow margins for screw placement at C-1 or C-2. Whenever a spinal stabilization was planned, a lateral radiograph in neutral position was obtained for exact intraoperative positioning.

Surgery was recommended for symptomatic patients only. All posterior operations were performed with the patient placed prone and involved a decompression of the foramen magnum with opening of dura and arachnoid, as well as a duraplasty to establish and maintain a physiological CSF flow^{29,30,32} (Fig. 1). Patients undergoing a spinal stabilization were placed in the neutral position with the cervical spine extended under fluoroscopic control according to the preoperative radiograph. The bony decompression consisted of a craniectomy of 3- to 4-cm diameter, which included the foramen magnum and a laminectomy of C-1 (Fig. 1). Care was taken to preserve the spinous process and arch of C-2 to allow secure placement of a bone graft between occiput and C-2 in patients requiring additional fusions (Fig. 2). With exposure of the dura, the atlantooccipital membrane was coagulated and dissected off the dura, which was then incised in a Y shape under the microscope. The arachnoid was incised to spread the tonsils apart for inspection of the foramen of Magendie. In most instances the cerebellar tonsils were coagulated at their tips and medially for this purpose.

If the foramen was found to be patent and no arachnoid adhesions were detectable elsewhere, no further dissections were performed. Whenever the foramen was obstructed, it was opened by sharp dissection. No dissec-

TABLE 1. Neurological scoring system*

Score	Pain	Sensory Dist, Dysesthesias	Motor Weakness	Gait Ataxia	Sphincter Function	Swallowing Function
5	None	Normal	Full power	Normal	Normal	Normal
4	Slight, no medication	Present, not significant	Movement against resistance	Unsteady, no aid	Slight dist, no catheter	Discomfort
3	Good control with medication	Significant, function not restricted	Movement against gravity	Mobile with aid	Residual, no catheter	Nerve deficit compensated
2	Insufficient control with medication	Some restriction of function	Movement without gravity	Few steps with aid	Rarely incontinent	Aspiration, coughing preserved
1	Severe despite medication	Severe restriction of function	Contraction without movement	Standing with aid	Often catheter	Aspiration, coughing insufficient
0	Incapacitating	Incapacitated function	Plegia	Plegia	Permanent catheter	No function

Dist = disturbance.

* Modified from Table 1 in Klekamp J, Samii M: *Syringomyelia: Diagnosis and Treatment*. Berlin: Springer-Verlag, 2002. With kind permission of Springer Science+Business Media.

FIG. 1. **A:** Sagittal MRI study obtained in a 52-year-old woman with a 3-year history of occipital pain and gait ataxia shows a Chiari I malformation with syringomyelia. **B–E:** Intraoperative photographs. After opening of the dura mater the arachnoid appears normal (**B**). After bipolar coagulation of the tonsils (**C**), the foramen of Magendie is inspected and appears partially obstructed (**D**). The dura is closed with a duraplasty (**E**). **F:** The postoperative MRI obtained after 3 months demonstrates a free CSF passage at the foramen magnum and complete resolution of the syrinx. Occipital pain and gait ataxia improved.

tion was performed laterally to avoid injury to perforating vessels of the brainstem or spinal cord. A duraplasty was inserted with a running suture. A variety of materials were used for this purpose: Goretex (W.L. Gore & Associates GmbH), Neuro-Patch (B. Braun Melsungen AG), fascia lata, or galea (Fig. 1). For patients requiring fusion, lateral mass screws have been used for the subaxial cervical spine since 2004. For C-2, pedicle screws or laminar screws were used. Prior to 2004, posterior rods were fixed to cervical laminae by soft wire. Whereas occipital rods applied laterally to the occiput were used up to 2009, occipital plates that can be fixed to the medial and thicker parts of the occipital bone have been used since then. An iliac crest graft was placed between occiput and C-2 and fixed with small screws (Fig. 2). Additionally, bone material was placed into the C1–2 joints. Finally the wound was closed, paying particular attention to tight suturing of the muscular layer, which had to cover all implants completely.^{29,30,32}

For anterior decompressions (i.e., transoral resections of the dens axis), the patients were placed supine for the

operation, which was always followed by a posterior decompression and fusion a few days later. No splitting of the soft or hard palate was performed in these patients. The incision was centered at the level of the anterior arch of C-1. The anterior part of the atlas was removed, and then the odontoid was detached from the body of C-2 at its base, according to intraoperative fluoroscopic guidance, and then removed in toto after resection of the ligaments at the odontoid tip. The wound was closed in a single layer. A gastric tube was left in place and a Philadelphia collar applied until the posterior decompression and fusion had been performed a few days later (Fig. 3).

Postoperatively, all patients were supervised on the intensive care unit for at least 24 hours before being transferred back to the regular ward. Postoperatively, all patients underwent an MRI study before discharge and a CT control study and lateral radiograph if a fusion had been performed. A further MRI study was obtained after 3–6 months. Further scans were obtained in patients with unsatisfactory clinical outcomes or in those with no resolution of an associated syrinx. For patients with posterior

FIG. 2. **A:** Sagittal MRI scan of a 26-year-old man with a 4-year history of occipital pain and gait ataxia demonstrates a Chiari I malformation with syringomyelia and a basilar invagination with ventral compression of the medulla. **B:** Intraoperative photograph. After dura and arachnoid opening, adhesions encasing both posterior inferior cerebellar arteries and obstructing the foramen of Magendie become apparent. **C:** Postoperative sagittal MRI obtained after 3 months shows the realignment achieved by extension and fixation of Oc–C3 and a decrease of the syrinx.

FIG. 3. A: Sagittal MRI scan of a 43-year-old man with a 3-year history of neck pain, gait ataxia, and profound swallowing dysfunctions due to ventral compression by the odontoid and posterior compression by the Chiari I malformation. The patient underwent a transoral and a posterior decompression with Oc–C4 fusion. **B:** Postoperative sagittal MR image shows the decompression of the upper cervical cord and medulla. **C:** Sagittal CT reconstruction reveals the bony fusion Oc–C4. Gait ataxia, swallowing problems, and neck pain improved.

fusions, CT scans were ordered 1 year after the operation to rule out hardware problems.

For statistical tests of significance, Student t-tests and Fisher tests were used. Long-term follow-up was analyzed with Kaplan-Meier²⁶ statistics to determine percentages of patients with and without postoperative clinical recurrences. A clinical recurrence was defined as a new neurological deficit or a worsening of a preoperative deficit after surgery. For all statistical analyses the software package PC-Statistik version 4.0 was used (Hoffmann-Software).

Results

Clinical Presentations and Anatomical Features

Three hundred twenty-three patients with a Chiari I malformation underwent 350 operations between 1985 and 2013, with a follow-up of 53 ± 58 months (mean $\pm$ SD). Preoperative symptoms are presented in Table 2, and patient data in Table 3. There were 16 children and 307 adults in this series.

Patients with Chiari I malformation and basilar invagination were significantly less affected by neuropathic pain and sensory deficits compared with patients with a Chiari I malformation alone, but these patients were more severely compromised by gait ataxia and swallowing dysfunctions. Furthermore, they were significantly younger and demonstrated syringomyelia less often and of smaller size if present.

Looking at additional bony anomalies, patients with invagination were more commonly affected by assimilations of the atlas to the occiput or Klippel-Feil syndrome of the cervical spine. Functional radiographs disclosed

craniocervical instability significantly more often in patients with invagination than in those without (65.2% and 2.7%, respectively).

Despite the younger age for patients with invagination in addition to Chiari I malformation, the average preoperative history was almost identical in both groups, and extended for approximately 5.5 years on average.

Craniocervical Operations

Surgery was offered to all symptomatic patients; 313 operations dealt with the craniocervical pathology—consisting of a foramen magnum decompression, arachnoid dissection, and a duraplasty with or without stabilization. Of these, 56 operations were secondary interventions (47 in patients without invagination, 9 in those with invagination). Intraoperatively, severe arachnoid scarring was evident in 15% of all decompressions, without a significant difference between those with or without additional invagination (16% and 13%, respectively). The foramen of Magendie was found to be closed by a membrane in 36% of all foramen magnum decompressions, and it was opened in these patients (Fig. 1). There was again no difference in this respect between patients with or without additional invagination (34% and 36%, respectively).

Patients without invagination but with evidence of craniocervical instability underwent the decompression with additional craniocervical fixation. Patients with invagination were divided into those with ($n = 31$) or without ($n = 15$) ventral compression by the odontoid in the foramen magnum. Patients with ventral compression were significantly more likely to present with swallowing problems

TABLE 2. Preoperative neurological symptoms

Group	Occipital Pain	Neuropathic Pain	Hypesthesia	Gait	Motor Power	Sphincter Function	Swallowing Function
Chiari I malformation	79%	48%*	70%*	65%*	41%	15%	16%*
Chiari I + invag	86%	34%*	56%*	86%*	34%	10%	32%*
All	80%	46%	68%	68%	40%	14%	18%

Invag = invagination.

* Significant difference ($p < 0.05$) between groups.

TABLE 3. Clinical data and surgical management

Variable	Chiari I	Chiari I + Invag	All
Clinical data			
No. of patients	277	46	323
Age (mean $\pm$ SD)	44 $\pm$ 15 yrs*	38 $\pm$ 18 yrs*	43 $\pm$ 16 yrs
Sx duration (mean $\pm$ SD)	64 $\pm$ 96 mos	66 $\pm$ 88 mos	64 $\pm$ 94 mos
Syringomyelia	223*	31*	254
Craniospinal instability	4*	30*	34
Atlas assimilation	2*	26*	28
Klippel-Feil syndrome	7*	17*	24
Management			
Pst decompression†	264	19	283
Pst decompression + pst fusion	4	17	21
Combined dec + pst fusion†	0	9	9
Syrinx catheter re- moval	6	0	6
VP shunt	3	0	3
Ventral fusion cervical spine	16	4	20
Pst fusion cervical spine	7	1	8
Total	300	50	350

Combined dec = transoral and posterior decompression; pst = posterior; Sx = symptom; VP = ventriculoperitoneal.

* Significant difference ($p < 0.05$) between groups.

† Two patients with ventral compression underwent subsequent stabilizations after a posterior decompression. Two patients required repeat fixations after combined decompressions and fusions due to hardware failures.

FIG. 4. **Left:** Sagittal MRI scan of a 31-year-old woman with sensory deficits and neuropathic pain in her left arm for the past 10 years demonstrates a Chiari I malformation, a basilar invagination without ventral compression, and a scoliosis of her entire spine. A profound syrinx extended from C-3 to T-3. The patient underwent a foramen magnum decompression without fusion. **Right:** Sagittal postoperative MRI shows the free CSF passage at the foramen magnum and a decrease of the syrinx. Her neuropathic pain improved corresponding to a significant reduction of her analgesic medication.

Spine Operations

In the group without invagination, 6 operations were performed to remove syrinx shunt catheters implanted at other institutions, because these had caused symptoms related to cord tethering. Three patients were primarily treated for their hydrocephalus only.

Another group of operations addressed degenerative diseases of the cervical spine after sufficient treatment of the craniospinal pathologies (Fig. 5). These surgeries made up 8% of all operations consisting of posterior or anterior decompressions and fusions (Table 3). Ventral fusions were chosen for patients with radicular symptoms, whereas posterior decompressions and fusions were used for patients presenting with a progressive myelopathy.

Postoperative Outcome for Craniocervical Operations

After foramen magnum decompressions with or without fusions, complications within 30 days were observed in 19.2%, and were significantly more common in patients with than in those without basilar invagination in addition to Chiari I malformation (35.6% and 16.4%, respectively; $p = 0.0006$) (Table 4). Whereas complications in the Chiari group without invagination were predominantly related to the foramen magnum, complications unrelated to the foramen magnum such as pneumonias were more common in the group with additional invagination.

Syringomyelia declined after 74% of all operations and remained stable in 23%. The rate for postoperative increases was very low, at 3%. Due to the smaller syrinx sizes in patients with invagination, the rate of syrinx cavities remaining unchanged was higher in this group, without reaching statistical significance (41% and 21%, respectively).

Looking at short-term results for the 1st postoperative year after decompressions (Table 5), both groups experienced profound improvements for occipital pain with minor improvements for the remaining symptoms. At this

than were those without ventral compression (39% and 12%, respectively).

In the group with basilar invagination, all patients without ventral compression underwent a foramen magnum decompression without additional stabilization (Fig. 4). Another 4 patients with mild anterior compression were also treated without fusion. Among these 4 patients treated without fusion, 3 developed instabilities subsequently. Two of the 3 had segmentation abnormalities, with assimilation of the atlas in 1 and Klippel-Feil syndrome in both of them. The third patient had presented without segmentation abnormalities but with a profound swan neck deformity. Two of these patients underwent stabilizations in a second operation. The third patient refused further surgery and died 7 years later due to respiratory failure.

In the group with basilar invagination and ventral compression, 15 operations consisted of a foramen magnum decompression with fixation (Fig. 2). All fixations included the occiput, with 1 exception; that patient underwent a C1–2 fusion. In another 9 patients with ventral compression and signs of caudal cranial nerve deficits, a transoral resection of the odontoid was performed first, followed by the posterior decompression and fusion (Fig. 3). Of these 9 patients, 2 required refixations at the occiput 2 and 3 years later due to hardware failures early in the series.

FIG. 5. A: Sagittal MRI scan of a 38-year-old woman with a Chiari I malformation and syringomyelia at C4–6. The patient underwent a foramen magnum decompression leading to resolution of the syrinx. Six years later a ventral fusion C5–6 was performed for treatment of radicular symptoms. Another 10 years later the patient presented with signs of a cervical myelopathy. **B:** The sagittal MRI at this time shows the free CSF passage at the foramen magnum, no reappearance of the syrinx, and multiple osteochondroses. **C:** After laminectomies of C3–6 and fusion of C3–7, the sagittal MRI shows the decompression of the cervical spinal cord. Neck pain and gait ataxia improved.

time point, only 4% considered their neurological status to be worsened by the operation, with the rate for postoperative improvements being higher for those with additional invagination than in those without (81.6% and 76.4%, respectively).

The analysis of long-term results revealed no significant difference between patients with or without basilar invagination in addition to Chiari I malformation. Rates for clinical recurrences within 5 and 10 years were not significantly different according to Kaplan-Meier analysis, although a smaller proportion of patients without invagination tended to develop new neurological deficits in the long term (Chiari I: 13.2% and 18.3%; Chiari I with invagination: 24.9% and 24.9% after 5 and 10 years, respectively; log-rank test: not significant) (Fig. 6). Clinical recurrences

in patients with invagination were exclusively related either to instabilities becoming manifest after a foramen magnum decompression (3 patients) or to a hardware failure (2 patients). Postoperative neurological deteriorations for patients without invagination were more evenly spread during follow-up and were predominantly related to reobstructions of the CSF passage at the foramen magnum.

A comparison between primary and secondary decompressions revealed significantly higher clinical recurrence rates in the group without additional invagination for secondary interventions, due to the profound arachnoid pathology in this subgroup (first surgery: 8% and 9.6%; revision surgery: 28.3% and 37.3% after 5 and 10 years, respectively; log-rank test: $p = 0.0005$) (Fig. 7). A similar difference for patients with invagination was not detectable (first surgery: 29.4% and 29.4%; revision surgery: 14.3% and 14.3% after 5 and 10 years, respectively; log-rank test: not significant) (Fig. 8), presumably because instability was the major reason for revision rather than reobstruction of the CSF pathway at the foramen magnum. In absolute numbers, 15 of 230 patients without invagination and 5 of 37 patients with invagination demonstrated a postoperative clinical recurrence after a first decompression, requiring a surgical revision in 4 patients without and 3 patients with invagination.

Postoperative Outcome for Spine Operations

For patients treated for their degenerative cervical disc disease, 81.5% reported a postoperative improvement of their symptoms after surgery, whereas the remainder considered their condition unchanged or worse. Looking at individual symptoms, the postoperative results were very similar to those observed after foramen magnum decompressions. Occipital pain and gait ataxia responded best, with marginal improvements for the remainder of symptoms (Table 5).

The analysis of long-term results for patients treated

TABLE 4. Complications after craniospinal decompressions with and without fusions

Complication	Chiari I	Chiari I + Invag	All
CSF fistula	11	1	12
Wound infection	8	2	10
Hydrocephalus	7	3	10
Hemorrhage	3	0	3
Swallowing dysf	1	1	2
Cerebellar infarction	1	0	1
Instability	0	1	1
UTI	9	2	11
Pneumonia	3	5	8
Thrombophlebitis	1	0	1
Angioneurotic edema	0	1	1
Total	44 (16.4%)	16 (35.6%)	60 (19.2%)

Dysf = dysfunction; UTI = urinary tract infection.

TABLE 5. Neurological course in the 1st postoperative year*

Symptom	Decompressions			All Cervical Fusions
	Chiari I	Chiari I + Invag	All Craniocervical	
Hypesthesia				
Preop	3.7 ± 0.9	4.1 ± 0.9	3.7 ± 1.0	3.7 ± 0.8
Postop	3.8 ± 0.9	4.2 ± 0.8	3.9 ± 0.9	3.8 ± 0.8
3 mos	3.9 ± 0.9	4.3 ± 0.9	4.0 ± 0.9	3.9 ± 0.8
1 yr	4.0 ± 0.9	4.3 ± 0.7	4.0 ± 0.9	4.0 ± 0.8
Neuropathic pain				
Preop	4.2 ± 0.9	4.5 ± 0.8	4.2 ± 0.9	4.4 ± 0.9
Postop	4.3 ± 0.8	4.6 ± 0.7	4.4 ± 0.8	4.5 ± 0.9
3 mos	4.4 ± 0.8	4.6 ± 0.7	4.4 ± 0.8	4.5 ± 0.9
1 yr	4.4 ± 0.8	4.6 ± 0.7	4.4 ± 0.8	4.5 ± 0.9
Occipital pain				
Preop	3.6 ± 0.9	3.4 ± 0.8	3.6 ± 0.9	3.6 ± 0.9
Postop	4.1 ± 0.6	3.8 ± 0.7	4.1 ± 0.7	4.1 ± 0.7
3 mos	4.4 ± 0.7	4.1 ± 0.7	4.3 ± 0.7	4.3 ± 0.6
1 yr	4.4 ± 0.7	4.2 ± 0.8	4.4 ± 0.7	4.2 ± 0.6
Motor weakness				
Preop	4.3 ± 0.9	4.6 ± 0.6	4.3 ± 0.9	4.0 ± 0.9
Postop	4.3 ± 0.9	4.6 ± 0.6	4.4 ± 0.9	4.1 ± 0.8
3 mos	4.4 ± 0.9	4.6 ± 0.6	4.4 ± 0.9	4.3 ± 0.8
1 yr	4.4 ± 0.9	4.7 ± 0.6	4.4 ± 0.9	4.3 ± 0.8
Gait ataxia				
Preop	4.0 ± 1.0	3.8 ± 0.7	4.0 ± 0.9	3.8 ± 1.1
Postop	4.1 ± 1.0	3.9 ± 0.7	4.1 ± 0.9	3.8 ± 1.1
3 mos	4.3 ± 1.0	4.1 ± 0.5	4.3 ± 0.9	4.0 ± 1.0
1 yr	4.3 ± 1.0	4.2 ± 0.6	4.3 ± 0.9	4.1 ± 0.9
Bladder function				
Preop	4.8 ± 0.7	4.8 ± 0.7	4.8 ± 0.7	4.8 ± 0.6
Postop	4.8 ± 0.6	4.9 ± 0.4	4.8 ± 0.6	4.8 ± 0.6
3 mos	4.8 ± 0.6	4.9 ± 0.2	4.8 ± 0.6	4.8 ± 0.5
1 yr	4.8 ± 0.6	4.9 ± 0.2	4.8 ± 0.6	4.8 ± 0.5
Swallowing				
Preop	4.8 ± 0.6	4.5 ± 0.8	4.7 ± 0.6	
Postop	4.9 ± 0.5	4.6 ± 0.7	4.8 ± 0.5	
3 mos	4.9 ± 0.4	4.7 ± 0.5	4.9 ± 0.4	
1 yr	4.9 ± 0.4	4.8 ± 0.5	4.9 ± 0.4	
Overall				
Better	76.4%	81.6%	77.1%	81.5%
Unchanged	19.2%	16.3%	18.9%	14.8%
Worse	4.4%	2.0%	4.0%	3.7%

* Unless otherwise specified, all values are expressed as the mean ± SD.

for their degenerative cervical disc diseases revealed no postoperative clinical recurrences for those with additional invaginations. However, follow-up was limited to 26 months in this subgroup. In the group without invagination, 5 patients reported neurological deteriorations due to adjacent-level disease. In 4 of these, a ventral fusion in 1 or 2 segments had been performed (Fig. 5). Among patients treated by posterior decompression with laminectomy of C3–6 and fusion of C3–6 or C3–7, only 1 clinical recur-

rence was detected. The maximum follow-up in this subgroup was 10 years.

Discussion

History and Diagnosis

Except for 16 children between 12 and 17 years of age, all other patients in this series were adults. Basilar invagination was detected in 14.2% of all patients presenting

FIG. 6. Kaplan-Meier analysis for clinical recurrence rates after foramen magnum decompressions in patients with Chiari I malformation with and without basilar invagination (log-rank test: no significant difference).

with a Chiari I malformation. Among their series of 364 adult patients with Chiari I malformation, Milhorat et al.⁴⁰ reported a comparable rate of 12% for patients with basal invagination.

The clinical history extended over 5.5 years irrespective of the presence or absence of invagination. Symptoms started most commonly with occipital pain or gait ataxia, and these remained the major complaints by the time of presentation. There were some differences between patients with or without invagination, however. Patients with invagination tended to be more affected by caudal cranial nerve deficits and gait disturbances due to the additional ventral compression, whereas patients without invagination were more likely to suffer also from neuropathic pain or sensory disturbances, which was most likely related to higher rates of syringomyelia in patients without invagination. Moreover, syrinx cavities tended to be smaller in the invagination group, which suggests that a high percentage of them were asymptomatic. Nevertheless, the combination of anterior as well as posterior compression with C1–2 instability may lead to very dangerous and rapid clinical deteriorations.³ If the diagnosis has been made and clinical signs of a high cervical myelopathy are present, early surgery should be recommended.

The radiological diagnosis of a Chiari I malformation is almost always made by MRI. In the presence of invagination, however, additional radiological examinations are mandatory to establish all associated features. The diagnostic workup has to include a detailed analysis of the bony and vascular anatomy as well as functional studies apart from an MRI image. Preoperative CT scans with reconstructions in sagittal and coronal planes are extremely helpful for surgical planning to evaluate the bony anatomy of the craniocervical junction (Table 3).²⁰ In combination with a CT angiography study, the exact courses of the vertebral arteries can be demonstrated to avoid vascular injuries during surgery. The CT studies in ante- and retroflexion allow a good evaluation of functional aspects (such as instabilities of C1–2, in particular). Functional radiographs may serve the same purpose. However, the often short necks of these patients may make a good visualization of the C1–2 segment difficult. Segmentation anomalies such

FIG. 7. Kaplan-Meier analysis for clinical recurrence rates after first and secondary foramen magnum decompressions in patients with Chiari I malformation (log-rank test: $p = 0.0005$). Invag. = invagination.

as fusion of the atlas to the occiput or Klippel-Feil syndrome of the subaxial spine were much more common in the invagination group and should always be considered as features predisposing to craniocervical instability in patients with a Chiari I malformation.

Management of the Craniocervical Pathology

Symptomatic patients with Chiari I malformation require a decompression of the foramen magnum. But how should this decompression be designed? If a basilar invagination is present as well, do these patients require a ventral, transoral decompression? Is a sagittal realignment and posterior fusion sufficient for these patients, or should a foramen magnum decompression be performed as well?

The ongoing debate on the optimal foramen magnum decompression cannot be solved with this paper because all patients were treated in the same pattern: they underwent a suboccipital craniectomy enlarging the foramen magnum, a laminectomy C-1, dura and arachnoid opening, opening of the fourth ventricle if obstructed, and a duraplasty (Fig. 1). A detailed analysis of results incorporating data from the Department of Neurosurgery at the University of California Los Angeles had established the superiority of this technique compared with interventions that left the arachnoid intact.^{29,32}

For patients with basilar invagination, some publications suggest that a ventral decompression alone,^{13,37} cervical traction,²⁴ or posterior sagittal realignment and fusion with or without a bony decompression^{17,22,23} may be sufficient. In this series, a foramen magnum decompression was always incorporated into the procedure rather than limiting the operation to posterior alignment and fusion, because the realignment does release the ventral compression by the odontoid but not necessarily the posterior compression by the cerebellar tonsils and the CSF obstruction associated with it (Fig. 2).

The treatment algorithm (Fig. 9) presented here stratifies patients according to the presence or absence of a basilar invagination and distinguishes further between patients with (Fig. 2) or without ventral compression by the odontoid peg (Fig. 4). Patients without ventral compression do

FIG. 8. Kaplan-Meier analysis for clinical recurrence rates after first and secondary foramen magnum decompressions in patients with Chiari I malformation and basilar invagination (log-rank test: no significant difference).

not require a reduction of a ventral component and can be treated by a foramen magnum decompression alone—unless instability is likely to develop due to segmentation anomalies demonstrated on preoperative functional images or detected during surgery, in which case fusion should be added.¹⁸

Preoperative functional radiographs or CT scans may not pick up instabilities, however, because patients may restrict their range of motion according to their biomechanical limitations. Measurements of several angles between different skull base structures and the upper cervical spine have been proposed and used to aid in the determination whether craniocervical instability is present or likely to be so.^{7,54} Such measurements were not used in this series, and whether they would have been useful in deciding if fu-

sion should be performed in patients without ventral compression cannot be evaluated in retrospect. Segmentation anomalies and intraoperative judgment of stability were considered sufficient for identification of patients who require fusion in this subgroup. After induction of general anesthesia and muscle relaxation, one may encounter considerable hypermobilities not apparent on preoperative images. With the head securely fixed in the Mayfield clamp, the competence of C1–2 joints as well as those of the subaxial spine can be evaluated intraoperatively. Even though no objective criteria can be defined to diagnose instability during surgery, it is my experience that the intraoperative evaluation of C1–2 stability is a reasonable adjunct to decide whether a fusion is required.

Patients with basilar invagination without ventral compression, with no pre- or intraoperative evidence of instability but with abnormalities of segmentation in the upper cervical spine carry a considerable risk of postoperative instability if treated by a posterior decompression alone. Two of 7 such patients required a secondary stabilization later on in this series. Among such patients with segmentation anomalies treated at other institutions by posterior decompression without fusion, and who required a surgical revision, all had to undergo posterior fusion. Assessments of patients with secondary stabilizations revealed a considerably worse long-term outcome for occipital pain compared with all other patients. Several of them suffered from chronic pain syndromes requiring permanent analgesic medication.

The most likely explanation appears to be that the detachment of neck muscles from occiput and C-1 may cause problems in compensating their biomechanical handicaps sufficiently after surgery. Therefore, for patients in this subgroup—basilar invagination with no ventral compression but with segmentation anomalies—it is recommended

FIG. 9. Algorithm for surgical management of Chiari I malformation in patients with and without additional basilar invagination. CCN = caudal cranial nerves.

that the foramen magnum decompression be combined with a stabilization procedure, even if no instability can be demonstrated before or during surgery. Patients with ventral compression should undergo a foramen magnum decompression with posterior realignment and fusion⁷ (Fig. 2). This realignment can be achieved by preoperative Crutchfield traction (Jürgen Harms, personal communication, 2001),²⁴ a halo vest,⁵² and distraction of C1–2 by intraarticular spacers¹⁷ or by intraoperative traction under neuromuscular blockade,¹¹ as was done in this series. If C1–2 is mobile, such maneuvers will reduce ventral compressions to such a degree that anterior decompressions are no longer necessary.^{8,17,19,23,27,47} Fusion is then required to secure this corrected position permanently—irrespective of pre- or intraoperative evidence of instability.

If a posterior fusion is required, the occiput can be spared whenever the atlas is not assimilated to the occiput.²² Without abnormalities of the subaxial spine, an Oc–C2 or C1–2 fixation can be sufficient.⁵⁵ Experimental studies have confirmed the biomechanical safety of preserving realignment of the craniocervical junction by posterior distraction and fusion of C1–2 alone.¹² With segmentation abnormalities or degenerative changes below C-2, the fixation can be extended caudally as required.

Can all patients with ventral compression be treated in this fashion? Are transoral decompressions obsolete?²⁰ This is a controversial issue. Basilar invagination may reach a point that a sufficient reduction may no longer be possible by distraction. The younger the patient the more likely it is that extension will reduce an anterior compression. On the other hand, the combined decompressions in this series were particularly successful in improving caudal cranial nerve functions in particular. Similar observations were made by Salunke et al. for pediatric patients.⁵⁰ Nobody can tell in retrospect whether the same results would have been obtained with posterior management alone. Looking at patients from recent years, the number of combined decompressions has gradually decreased in this series, because more cases than previously thought can indeed be managed well without transoral surgery. However, for adult patients with profound anterior compression and caudal cranial nerve deficits, a transoral decompression is still a sensible option (Fig. 3) and may still be required in patients in whom a realignment via the posterior approach was not successful.

Transoral decompressions should always be followed by a posterior fusion. Menezes et al. described a group of patients treated by ventral decompression alone without developing postoperative instability.³⁹ In the literature, some reports describe stable situations after a transoral²⁵ or transnasal resection^{15,51} of the odontoid in basilar invagination. However, the risks of postoperative instability are considerable.^{16,18,42,44} Early in this series, 1 patient without preoperative instability underwent a transoral decompression with no posterior fusion planned. This rendered him severely unstable while still in intensive care, and required posterior fusion immediately. Therefore, I cannot recommend stand-alone transoral decompressions. In a recent review, Menezes recommended posterior fusions after transoral resections of the odontoid in every patient as well.³⁷

Management of the Degenerative Cervical Pathology

Another important aspect for patients with Chiari I malformations is the recognition of degenerative diseases of the cervical spine^{28,53} (Fig. 5). Most patients undergo a foramen magnum procedure at an age before they have developed significant degenerative problems in their cervical spines. However, due to the longstanding cord compression by cerebellar tonsils or the odontoid peg and the cord damage by an associated syrinx, the cervical cords of these patients are very vulnerable to degenerative spine diseases. Most patients will demonstrate some degree of cord atrophy despite a sufficient decompression and reduction of a syrinx. This may suggest that a cervical stenosis or a kyphotic angulation may be irrelevant. The significance of degenerative changes is easily overlooked unless functional studies of the cervical spine are performed. Hypermobilities appear to be particularly harmful to such an atrophic spinal cord. It is my experience that these patients should undergo cervical fusions as soon as clinical signs of a cervical myelopathy become apparent or progress.²⁸ The results presented here suggest that posterior decompressions with laminectomies of C3–6 combined with lateral mass fixations may lead to better long-term results than ventral fusions, which tended to address the 1 or 2 most affected levels—only to be followed by adjacent-level disease soon thereafter.

Complications After Foramen Magnum Decompressions

The complication rate of 19.2% in this study is in the reported range for foramen magnum decompressions that incorporate a duraplasty.^{2,21,34,57} Lower rates are described for decompressions that leave the dura intact.^{36,45} In this study, complication rates were significantly higher for patients with invagination compared with those without (Table 4). Most of this difference was related to higher rates for postoperative pneumonia in the invagination group. The rates for local complications (i.e., CSF fistulas, hemorrhages, and so on), were almost identical. Only 3.2% developed a postoperative ventricular dilation and clinical signs of increased intracranial pressure despite a sufficient decompression and required a ventriculoperitoneal shunt. This is a well-documented but not well-understood complication after foramen magnum decompressions for Chiari I malformations. This complication is not related to arachnoid opening or postoperative formation of subdural hygromas in the posterior fossa,²⁹ but may in some way be related to the altered CSF flow at the foramen magnum, which some patients have problems adapting to in the immediate postoperative phase.

Swallowing dysfunctions and problems of opening the mouth are well-recognized complications after occipitocervical stabilizations,³⁸ and may be aggravated further by short necks and smaller pharynx cavities in patients with invagination. In this series these dysfunctions were transient in 3 patients and did not require any measures apart from logopedic therapy. Such problems can be limited by meticulous positioning of the patient in neutral position.^{5,35,41,56} Care should be taken to avoid too much antelexion, in particular. For that purpose, a preoperative lateral radiograph obtained in the neutral position is extremely helpful to aid correct positioning on the operating table under fluoroscopic control.

Outcome After Foramen Magnum Decompressions

With the type of decompression performed in this study, 74% of patients with a syrinx experienced a sustained postoperative decrease in syrinx size, whereas 23% demonstrated no changes on postoperative MRI. For patients undergoing operation for the first time this figure was 83%, and was considerably higher compared with studies in which the dura was left intact for treatment of a Chiari I malformation.^{9,45} Because syrinx cavities tended to be considerably smaller in patients with additional invagination, a higher percentage showed no postoperative change of syrinx size compared with patients who had Chiari I malformations without invagination.

Looking at clinical results in the 1st postoperative year, the overwhelming majority of patients experienced improvements for almost all symptoms,²³ as outlined in Table 5. Occipital pain responded best according to this analysis.

Contrary to other reports,^{1,4} basilar invagination was not associated with a worse long-term outcome compared with Chiari I malformation without invagination. This may be explained by the additional foramen magnum decompression as performed in this series. Long-term results were very gratifying for patients with invagination, as long as hardware failures could be avoided⁴⁶ and no instability had resulted from the initial management.⁴⁸ For patients without invagination, long-term results depended on the postoperative course of associated arachnoid pathologies.²⁹ If the CSF pathway remained open, the rate for clinical recurrences was very low (Fig. 6). Revision surgeries, on the other hand, carried an increased risk for reobstructions leading to higher clinical recurrence rates due to the often severe arachnoid pathologies in this group^{28,29} (Fig. 7).

Compared with primary decompressions that leave the dura intact or incise only the outer layer, clinical recurrence rates greater than 10% are reported in the literature, with considerably smaller patient numbers and shorter follow-up.^{14,33,43,45,49} Pediatric neurosurgeons in particular favor this technique increasingly due to lower complication rates and shorter hospitalization times. However, none of these studies used Kaplan-Meier analyses to account for the varying follow-up times. If such analyses had been used, even higher clinical recurrence rates would have been the result. In this study, 6.5% of patients experienced a clinical recurrence after a first foramen magnum decompression, corresponding to a recurrence rate of 9.6% in 10 years according to Kaplan-Meier analysis. Therefore, a duraplasty should be part of each foramen magnum decompression.^{6,29} The results of this study demonstrate that the treatment algorithm presented here provides good short- and long-term results for patients with Chiari I malformations with or without an additional basilar invagination.

Summary

Symptomatic patients with a Chiari I malformation should undergo a foramen magnum decompression as outlined in this paper. In the absence of basilar invagination, craniocervical instability is a rare feature of this malformation, so that a posterior fusion is rarely required. If basilar invagination is combined with a Chiari I malformation, however, posterior fusion has to be performed

along with the foramen magnum decompression whenever craniocervical instability is present or realignment of C1–2 has to be maintained to relieve a ventral compression of the medulla. This study also demonstrates that, in a significant number of patients with basilar invagination and no ventral compression by the odontoid, but with segmentation anomalies at Oc–C1 or the subaxial spine, the craniocervical junction will become unstable after a foramen magnum decompression. Therefore, posterior fusion is recommended for these patients as well, irrespective of pre- or intraoperative signs of instability. The traditional method of treating an invagination by transoral odontoid resection followed by posterior fusion can be replaced by such a posterior operation in the great majority of patients. In this study, transoral decompressions were reserved for patients with ventral compression and significant dysfunctions of caudal cranial nerves. With the treatment algorithm as presented here, the great majority of patients reported sustained postoperative improvements, with no significant differences between those with or without additional basilar invagination.

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