Surgical Management of Chiari Malformations: Preliminary Results of Surgery According to the Mechanisms of Ptosis of the Brain Stem and Cerebellum

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Abstract

Introduction We classified Chiari malformation type I (CM-I) according to the mechanism of ptosis of the brain stem and cerebellum, based on a morphometric study of the posterior cranial fossa (PCF) and craniovertebral junction (CVJ). Surgery was performed to manage the mechanism of the hindbrain ptosis.

Materials and Methods We calculated the volume of the PCF (VPCF) and the area surrounding the foramen magnum (VSFM) and measured the axial length of the enchondral parts of the occipital bone (occipital bone size) and the hindbrain. According to these measures, we classified CM-I into type A (normal VPCF, normal VSFM, and normal occipital bone size), type B (normal VPCF, small VSFM, and small occipital bone size), and type C (small VPCF, small VSFM, and small occipital bone size). Foramen magnum decompression (FMD) (280 cases) was performed on CM-I types A and B. Expansive suboccipital cranioplasty (ESCP) was performed on CM-I type C. Posterior craniocervical fixation (CCF) was performed in cases with CVJ instability. Lysis of the adhesion and/or sectioning of the filum terminale were performed on cases with tethered cord syndrome.

Results Both ESCP and FMD had a high rate of improvement of neurological symptoms (87%) and recovery rate. There was only small number of complications. CCF had a high rate of improvement of neurological symptoms (88%) and joint stabilization.

Conclusion In the management of Chiari malformation, appropriate surgical methods that address ptosis of the hindbrain should be chosen. Each surgical approach resulted in a good improvement of neurological symptoms.

Keywords
- Chiari malformation
- morphometric study
- posterior cranial fossa
- craniovertebral junction
- surgical management
- outcome

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Background and Purpose

Previously, we reported that patients with a Chiari malformation have an underdeveloped occipital bone, so that the posterior cranial fossa (PCF) becomes shallow.[1,2] This shallowness results in the brain stem and cerebellum sagging into the spinal canal, that is, the pathogenesis of Chiari malformation is insufficiency of the para-axial mesoderm, which is the origin of the occipital bone.[1–7] In this case, foramen magnum decompression (FMD) to enlarge the space of the foramen magnum has been performed widely. Other mechanisms of ptosis of the brain stem and cerebellum, that is, hypermobility and instability of the craniovertebral junction (CVJ), traction by tethering, and pressure coning, have also been reported.[8–13] In those cases, FMD does not resolve the underlying mechanism. Misunderstandings of the mechanism of ptosis of the brain stem and cerebellum in Chiari malformation have resulted in confusion when choosing a surgical approach, and the use of inappropriate treatment has resulted in the recurrence of neurological symptoms.

Since 2006, we have performed morphometric studies of the PCF using magnetic resonance imaging (MRI) and computed tomography (CT) reconstructed images, and have conducted multiple analyses on cases with a Chiari malformation. In addition, we have also examined the mechanism of ptosis of the brain stem and cerebellum using morphometric analysis and have redefined and classified Chiari malformation type I (CM-I) into three independent subgroups: CM-I types A, B, and C.[14] We have performed surgical procedures to treat the mechanism of ptosis of the brain stem and cerebellum (►**Tables 1** and **2**).[14] We also present the preliminary results of treatments for ptosis of the brain stem and cerebellum in cases with CM-I.

Materials

We recruited 100 healthy control volunteers with no neurological symptoms or abnormalities in the neural axis (16–69 years old, mean: 38.7 years, 40 males, 60 females) under approval from the Institutional Review Boards of Koudoukai Health System (Osaka City University Graduate School of Medicine, Osaka, Japan) and North Shore University Hospital–Long Island Jewish Health System (New York, New York, United States). For the cases, 500 subjects with CM-I (cerebellar tonsil herniation $\geq$ 5 mm from the McRae line, i.e., between the basion and opisthion) (16–69 years old, mean: 37.7 years, 237 males, 263 females) and 50 subjects with an absence of CM-I (defined as cerebellar tonsil herniation $<$ 5 mm from the McRae line, but having brain stem symptoms and/or myelopathy due to associated syringomyelia) (17–55 years old, mean: 35.4 years, 18 males, 32 females) were examined. The distribution and mean of age and sex were not significantly different between the normal controls and CM-I group. CM-I was associated with syringomyelia in 221 cases, hereditary disorders in connective tissue in 187 cases, basilar invagination in 23 cases, and other bony anomalies in the CVJ in 57 cases. Patients who had syringomyelia due to other etiology were excluded. These patients were diagnosed and treated from April 2006 to March 2017.

Table 1 Subtypes of CM-I classified based on morphometric analyses and surgical indication

	CM-I type A	CM-I type B	CM-I type C	CM-absence
	187 cases	178 cases	155 cases	50 cases
PCFV	Normal	Normal	Small	Normal
VAFM	Normal	Small	Small	Small
PFBV/PFCV	Normal	Large	Large	Normal
Occipital bone size	Normal	Small	Small	Normal
Surgery	Others and FMD	FMD	ESCP	FMD

Abbreviations: CM-absence, cases which have neurological brain stem symptoms but tonsillar herniation less than 5 mm; CM-I, Chiari malformation type I; ESCP, expansive suboccipital cranioplasty; FMD, foramen magnum decompression; PCFV, posterior fossa cranial volume; PFBV, the volume of brain in posterior cranial fossa; VAFM, the volume of area of foramen magnum.

Table 2 Other mechanisms of ptosis of the brain stem and cerebellum, and surgical indications

	CVI	Traction (tethering)	Others
	50 cases	20 cases	17 cases
PCFV	Normal	Normal	Normal
VAFM	Normal	Normal	Normal
PFBV / PCFV	Normal	Normal	
Brain stem and cerebellum	Normal	Elongation and/or downward displacement	
Surgery	CCF	Untethering/SFT	VPS and/or others

Abbreviations: CCF, craniocervical fixation; CVI, craniovertebral instability; PCFV, posterior fossa cranial volume; PFBV, the volume of brain in posterior cranial fossa; SFT, section of filum terminale; VAFM, the volume of area of foramen magnum; VPS, ventriculoperitoneal shunt.

Methods: Morphometric Analyses and Surgical Indication

Morphometric Analyses and Classification of CM-I (►Table 1)

By MRI, two-dimensional (2D) and three-dimensional CT reconstructed images using OsiriX software (free access) were used to calculate the volume of the PCF (VPCF), brain volume in the PCF (VPCB), and volume of the area surrounding the foramen magnum (VSFM). The axial length of the basiocciput, exocciput, and supraocciput of the enchondral parts of the occipital bone (occipital bone size), axial length of the brain stem, cerebellum (excluding herniated tonsils), and the position of the fourth ventricle were also measured. By multiple analyses of the results, CM-I was classified into three independent groups: CM-I types A (167 cases) (normal VPCF, normal VSFM, and normal occipital bone size), B (178 cases) (normal VPCF, small VSFM, and small occipital bone

size), and C (155 cases) (small VPCF, small VSFM, and small occipital bone size).[1,2,13]

The volume of the brain stem and cerebellum and morphometric analyses (axial length of the brain stem and cerebellum) indicated there was no significant difference between the cases and healthy controls. Of the measured items, the following variables were significantly different in the cases compared with the controls: VPCF, the ratio of VPCB to VPCF (VPCB: VPCF), occipital bone size (axial length of the basiocciput and exocciput), and VSFM. In CM-I type A (167 cases), there was no significant difference in VPCF, VSFM, and occipital bone size compared with normal controls (►**Table 1**). In CM-I type B (178 cases), there was no significant difference in VPCF compared with normal controls, but VSFM and occipital bone size were significantly smaller than in normal controls. In this group, the volume above Twining's line (the line between the tubercle sellar and internal occipital protuberance) (VPCF-ATL) was significantly smaller than in the other groups, and the volume below Twining's line (VPCF-BTL) was also significantly smaller than in the other groups (►**Table 1**). In CM-I type C (155 cases), VPCF, VSFM, and occipital bone size were significantly smaller than in normal controls. In this group, VPCF-ATL and VPCF-BTL were significantly smaller than in the other groups (►**Table 1**). Moreover, the axial length of the brain stem was significantly longer than in the other groups, suggesting elongation of the brain stem in CM-I type C.

In CM-II (30 cases), VPCF and occipital bone size were significantly smaller than in normal controls, while the brain stem was longer, similar to the pattern observed in CM-I type C. However, VSFM was significantly larger than in normal controls (►**Table 1**). In addition, VSFM was significantly larger in CM-I type B than in CM-I type A.

The VSFM of patients with CM-I type A who had tethered cord syndrome was larger than in normal controls; however, VSFM was not significantly different between patients with CM-I type A who had a tumor and normal controls.

Hypermobility and Instability at the Craniovertebral Junction and Tethered Cord Syndrome (►Table 2)

Diagnosis of instability at the CVJ was confirmed by a morphometric study and a craniocervical traction test, using morphometric analyses described by the authors and Goel et al.[11–13] The first author (M.N.: neurosurgeon), second author (P.B.: neurosurgeon), and fourth author (H.I.) performed the craniocervical traction test and measurements. For the craniocervical traction test, a tong was attached to the skull under intravenous anesthesia in the supine position and morphometric measurements were taken. Then, the patient was placed in an upright position and craniocervical instability was revealed as neurological symptoms (checked by the third author, R.K.: neurologist), and when morphometric measurements were taken, displacement at the occipitoatlantoaxial joints (>1 standard deviation [SD]) was observed. Craniocervical traction of 10 to 15 kg was then applied and the neurological symptoms resolved (checked by the fourth author, R.K.: neurologist) and a reduction of the craniocervical junction was observed.

Tethered cord syndrome was diagnosed by neurological symptoms (e.g., motor weakness at both lower extremities, sensory loss, neurogenic bladder, pes equinus, and conus medullaris lower than the L2 vertebral body and/or diameter of the filum terminale ≥ 2 mm).[15] The third author (R.K.: neurologist) diagnosed tethered cord syndrome.

Surgical Indications and Procedures (►Table 3)

According to the mechanism of ptosis of the brain stem and cerebellum, a surgical procedure was selected. The indications for surgery were the presence of myelopathy, upper cervical cord symptoms, brain stem symptoms, and <14

Table 3 Subtypes (CM-I types A, B, and C), surgical indication, and associated anomalies

Total	550 cases	Male/ female	Age	Mean (SD)
		255/295	16–69 y	37.7 y
Subtypes				
CM-I type A	167 cases	80/87 cases	16–69 y	39.6 y (10.1)
CM-I type B	178 cases	83/95 cases	17–64 y	37.8 y (10.8)
CM-I type C	155 cases	74/81 cases	16–59 y	33.4 y (10.3)
CM-absence	50 cases	18/32 cases	17–55 y	35.4 y (10.4)
Surgery	585 surgeries			
FMD	280 cases	112/168 cases	16–69 y	37.6 y (10.5)
ESCP	150 cases	69/81 cases	16–59 y	33.4 y (10.3)
+ OCF	52 cases			
CCF	110 cases	52 / 58 cases	16–51 y	30.4 y (10.7)
OCF	64 cases			
C1/2 PLF	46 cases			
SFT	25 cases	74/81	16–23 y	21.8 y (7.8)
VPS	20 cases	8/12 cases	16–22 y	20.4 y (4.4)
Associated anomalies				
Syringomyelia	221 cases (55%)			
HDCT	187 cases (34%)			
Basilar invagination	23 cases (4.2%)			
Other bony anomalies	57 cases (10%)			

Abbreviations: CCF, craniocervical fixation; CM-absence, cases which have neurological brain stem symptoms and/or myelopathy but tonsillar herniation less than 5 mm; CM-I, Chiari malformation type I; ESCP, expansive suboccipital cranioplasty; FMD, foramen magnum decompression, HDCT, hereditary disorders of connective tissue; OCF, occipitocervical fixation; PLF, posterior lateral fixation; SD, standard deviation; SFT, section of the filum terminale; VPS, ventriculoperitoneal shunt.

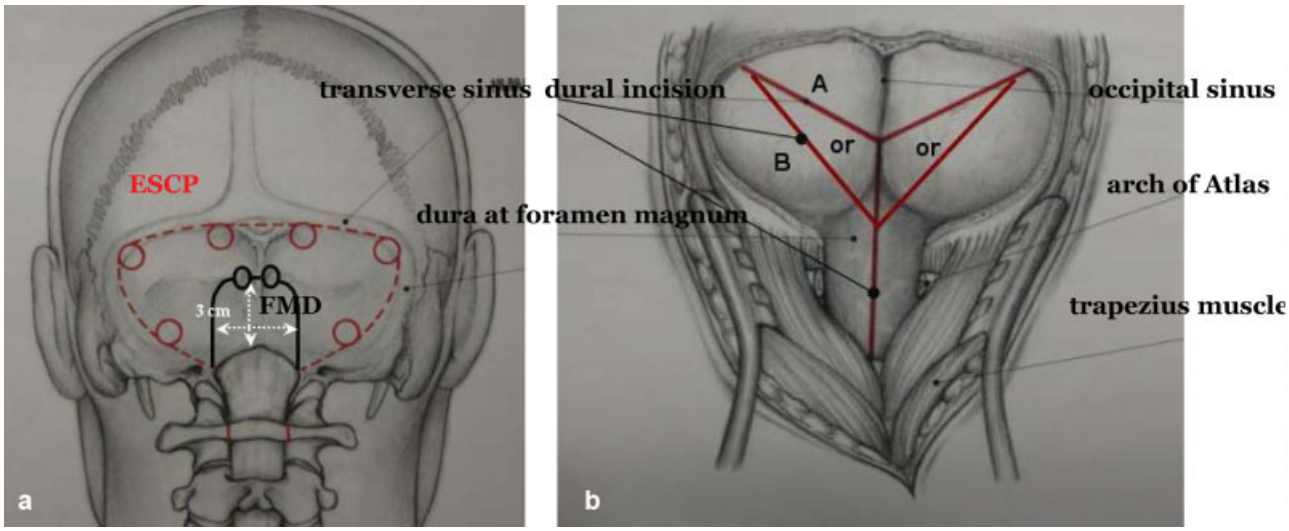

Fig. 1 Craniotomy with expansive suboccipital cranioplasty (ESCP) and foramen magnum decompression (FMD). (**Left**) Craniotomy with ESCP along the transverse and sigmoid sinuses, and craniotomy with FMD of 2 to 3 cm^2. In FMD, the suboccipital muscle group is preserved. (**Right**) Dural incision in both procedures.

points in the Japanese Orthopedics Association Cervical Myelopathy Evaluation Questionnaire (JOACMEQ).[15]

FMD consists of craniectomy to decompress the surrounding area (2–3 cm^2).[16] Expansive suboccipital cranioplasty (ESCP), which was described by Sakamoto et al and Nishikawa and Ohata, can also be used for extensive decompression (along the transverse and sigmoid sinuses) and osteoplasty and dural plasty (►**Figs. 1** and **2**).[17,18] CM-I types A and B (280 cases) underwent FMD to expand the area surrounding the foramen magnum and major cistern and resolve compression of the brain stem and cerebellum (►**Figs. 1** and **2**).[15] CM-I type C (150 cases) underwent ESCP to expand the area surrounding the foramen magnum and the PCF (►**Figs. 1** and **2**).[16,17] CM-absence (30 cases) underwent FMD.[15] C1 laminectomy and dural plasty were performed in all cases. For the cases with instability at the craniocervical junction, posterior craniocervical fixation (CCF) was performed.[18] In the cases who received ESCP and FMD, CCF was performed on 52 subjects.[10–12] CCF was used in a total of 110 cases, of which CCF was performed on 64 cases and posterior atlantoaxial fixation was utilized in 46 cases. For tethered cord syndrome, untethering (lysis of the arachnoid adhesion and/or sectioning of the filum terminale [SFT]) was administered to 25 cases.[9,13] For the cases with increased intracranial pressure and/or hydrocephalus, a ventriculoperitoneal shunt was performed in 20 cases. The other 10 cases had a mass lesion of the PCF, which was removed; 7 cases had an arachnoid cyst, for which fenestration of the cyst was performed.

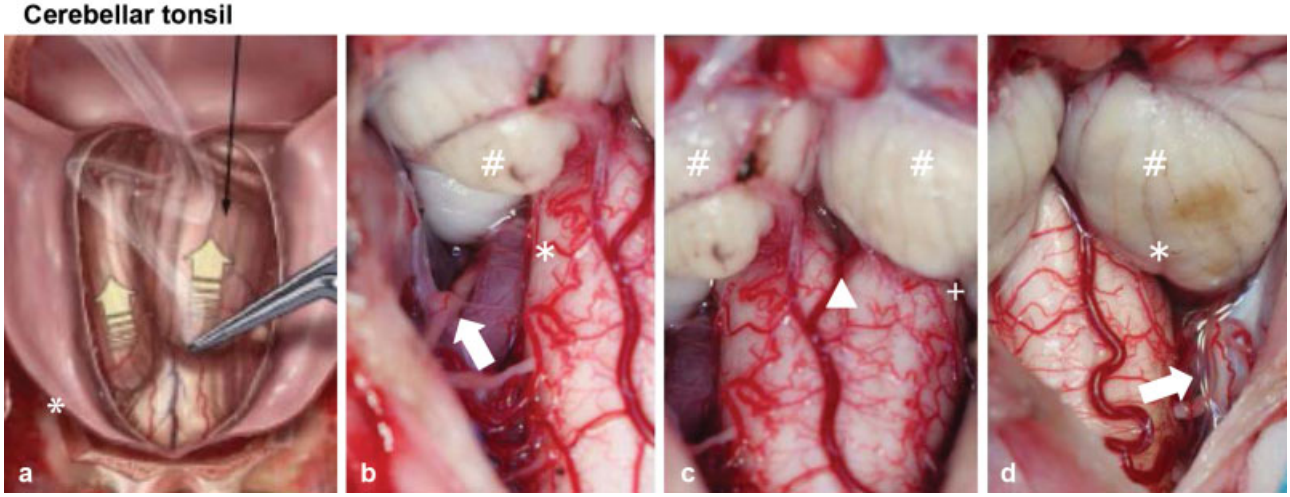

Fig. 2 Operative images, tonsillar burning, and the foramina of Magendie and Luschka. (**a**) Burning of the cerebellar tonsils. (**b**) Left foramen of Luschka (*), burned and shrunken tonsil (#), accessory nerve (arrow), and vertebral artery (+). (**c**) Foramen of Magendie (*), burned and shrunken tonsils (#), and obex (arrowhead). (**d**) Right foramen of Luschka (*), burned and shrunken tonsil (#), and accessory nerves (arrow). If the tonsils are large and cerebrospinal fluid (CSF) flow is normal after craniotomy and opening the dura mater is not confirmed, the tonsils should be burned to shrink them. Confirmation of CSF flow from the foramina of Magendie and Luschka (*) is important after shrinking the tonsils (#); CSF flowed from the foramen of Luschka and decompression was performed for accessory nerves (arrows) and the vertebral artery (+), and the obex was observed (arrowhead).

Examination of Cerebrospinal Fluid Space and Flow

After bony decompression, before opening the arachnoid membrane and dura mater, and after opening the dura mater and dural plasty, color Doppler ultrasonography (CDU) was performed to observe the dynamics of cerebrospinal fluid (CSF) flow from the foramina of Magendie and Luschka, and the volume of the major cistern and CSF dynamics were estimated (►**Fig. 3**).[19–21] Using this approach, Milhorat and Bolognese reported that, as a final goal, the CSF space of the major cistern should be ≥8 mL and the maximum CSF flow velocity should be ≥5 cm/s.[19] If the final goal is not achieved, enlargement of the bony decompression should be performed initially; however, if the final goal is still not achieved, burning and shrinking of the cerebellar tonsils should be conducted.[19]

Follow-up and Postoperative Examinations and Determination of Joint Fixation

Postsurgery, neurological symptoms, activities of daily living (JOACMEQ score), recovery rate of the JOACMEQ score (JOACMEQ score RR), as calculated by (postsurgery points − presurgery points/full points [17] − presurgery points) × 100%,[15] and neuroradiological findings (dynamic X-ray, 2D CT, and MRI of the cervical spine) were examined every 3 months. Joint fixation was examined to certify that there was no joint instability and/or continuity between bones by cervical spine dynamic X-ray and 2D CT imaging.

Statistical Analysis

For the comparison of means between two groups, the Mann–Whitney's test was used. For the comparison of more than two groups, the Kruskal–Wallis' test was used. A p-value < 0.01 was used to determine significance. Outcome was assessed using chi-square and Fisher's tests. A pathological condition was defined when VPCF, VSFM, and occipital bone size were <2 SD.

The first author (M.N.: neurosurgeon), second author (P.B.: neurosurgeon), third author (R.K.: neurologist), and fourth author (H.I.: neuroradiologist) performed measurements; M.N., P.B., and fifth author (T.T.) performed surgery; M.N. and R.K. performed statistical analyses; and the last author (K.O.) and T.M. conducted and supported this study.

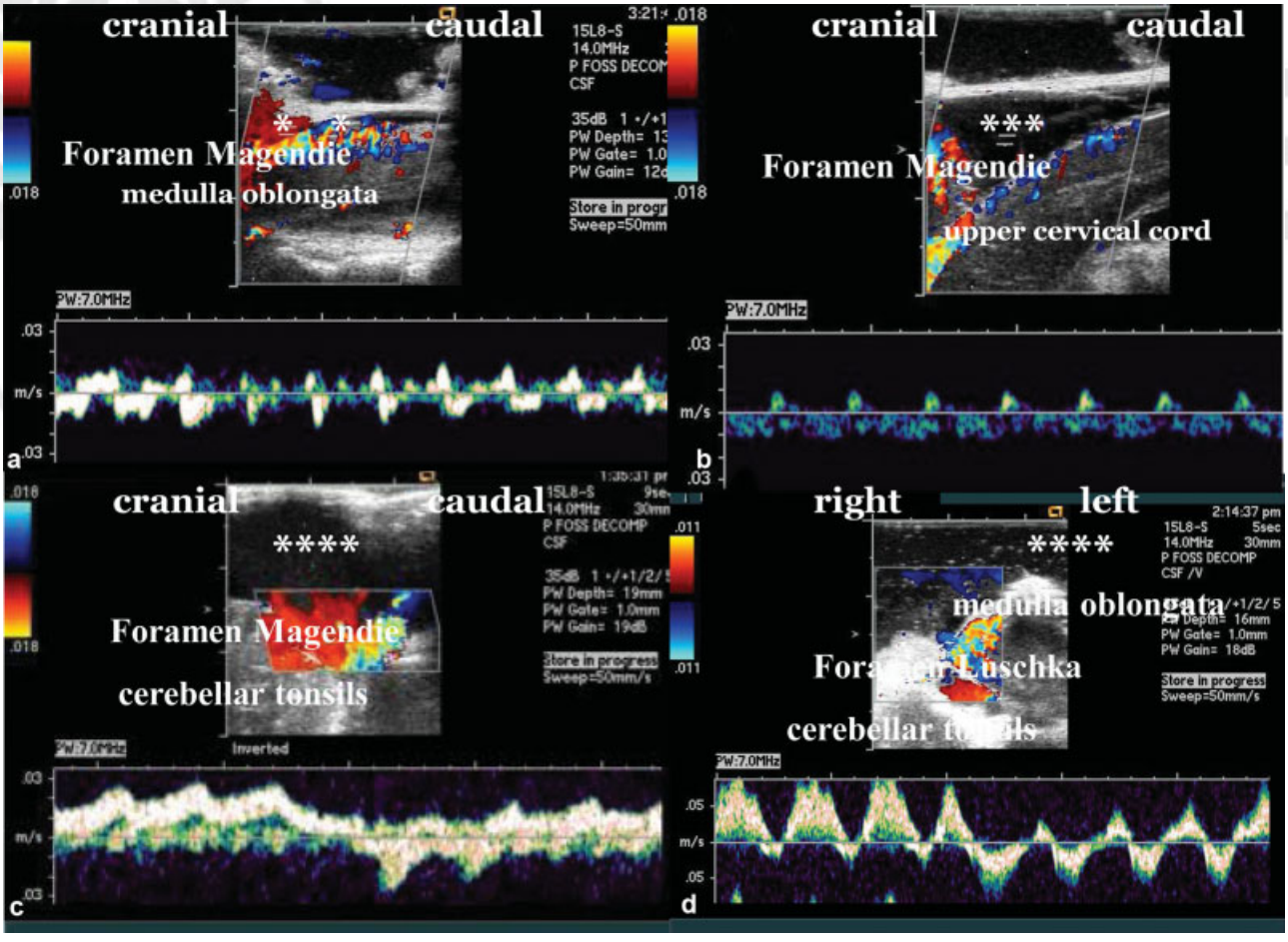

Fig. 3 Color Doppler ultrasonography (CDU). (**a**) CDU at the foramen of Magendie after craniotomy. The cerebrospinal fluid (CSF) space of the cisterna magna was very small. A small amount of CSF flow was observed. (**b**) CDU at the foramen of Magendie after cutting the external layer of the dura mater. Although CSF space was slightly increased, there was no change in CSF flow, with only slight flow observed. (**c**) CDU at the foramen of Magendie after dural plasty. (**d**) CDU at the foramen of Luschka after dural plasty. In this case, after craniotomy, CDU was performed, and the space of the cisterna magna was found to be small with insufficient CSF flow, and so dural plasty was performed. After dural plasty, CDU was performed again. A large amount of CSF flow from the foramina of Magendie and Luschka was confirmed. Suitable space of the cisterna magna: 12 mL. Maximum CSF flow velocity: 3 cm/s at the foramen of Magendie and 10 cm/s at the foramen of Luschka. *, CSF space.

Table 4 Overall results: FMD and ESCP

	Improved	JOACMEQS RR (%)	Stabilization	Deteriorated
FMD: 280 cases (for CM-I types A, B, and CM-absence)	247 (88%)[a]	66.7 ± 10.2	25 (8.9%)	8 (2.9%)
Without OAM: 145	126 (87%)[a]	66.9 ± 10.5	15 (10%)	
With OAM: 135	121 (90%)[a]	66.4 ± 8.7	10 (7.4%)	
ESCP 150 cases (for CM-type C)	131 (87%)[a]	66.7 ± 10.1	16 (11%)	3 (2.0%)
Without OAM: 72	63 (88%)[a]	70.5 ± 9.6	8 (11%)	1 (1.4%)
With OAM: 78	68 (87%) [a]	64.5 ± 10.7	8 (11%)	2 (2.6%)
Complication: wound infection: 3 (1.1%) in FMD				
Wound infection: 2 (1.3%) and cerebellar slumping: 10.7%) in ESCP				

Abbreviations: CM-absence, cases which have neurological brain stem and/or myelopathy but tonsillar herniation less than 5 mm; CM-I, Chiari malformation type I; ESCP, expansive suboccipital cranioplasty; FMD, foramen magnum decompression; JOACMEQS RR, recovery rate of Japanese Orthopedics Association Cervical Myelopathy Evaluation Questionnaire Score; OAM, opening of arachnoid membrane.
Note: Follow-up period: 18 to 130 months, mean: 40.5 months.
[a]Significantly high ($p < 0.001$).

Results

Outcome of Surgical Treatment

The follow-up period was 18 to 130 months (mean: 40.5 months). Outcome was estimated using the most recent data. Twenty-eight cases dropped out of the study at more than 3 years after surgery, in whom the results of the final examination were estimated. Only 12 cases were missed during follow-up.

FMD was performed in cases with CM-I type A, CM-I type B, and CM-absence. The JOACMEQ score RR for FMD was 66.7%, while 97% of cases showed an improvement or stabilization of their neurological symptoms. There was no significant difference in the JOACMEQ score RR between the cases in which the arachnoid membrane was or was not opened. In eight cases (2.9%), their neurological symptoms deteriorated later (►**Table 4**). In 7 of 207 cases, syringomyelia remained.

ESCP was performed for CM-I type C. The JOACMEQ score RR of ESCP was 66.7%, and 98% of cases had an improvement or stabilization of their neurological symptoms. There was no significant difference in the JOACMEQ score RR between the cases in which the arachnoid membrane was or was not opened. In three cases (2.0%), their neurological symptoms deteriorated later. In all 110 cases, syringomyelia vanished.

There was no significant difference between the FMD and ESCP groups for the improvement or stabilization of neurological symptoms and the JOACMEQ score RR. Complications were observed in three cases (1.1%) with FMD, and two cases (1.3%) with ESCP had a wound infection. In MRI, cerebellar slugging without neurological symptoms occurred with ESCP. In both the FMD and ESCP groups, there was no mortality and no permanent morbidity (►**Table 4**).

Of the cases who underwent CCF, 97 (88%) had an improvement of their neurological symptoms and the JOACMEQ score RR was 76.9%. Sixty-eight cases (62%) had complete bony fusion, while 33 cases (30%) were stabilized. Nine cases (8%) still had incomplete stabilization. Complications consisted of a transient swallowing disturbance in one case (0.9%), injury to the vertebral artery without neurological symptoms in one case (0.9%), and wound infection in one case (0.9%). There was no permanent morbidity (►**Table 5**).

Untethering was performed in 25 cases with tethered cord syndrome; 9 cases (36%) demonstrated an improvement of their neurological symptoms, while the other 16 cases (64%)

Table 5 Overall results: CCF

Clinical symptoms	Improved	JOACMEQS RR (%)	Unchanged
110 cases	97 (88%)	76.9 ± 13.2	13 (12%)
Fixation and stabilization of joints	Complete fusion	Stabilization	Incomplete
110 cases	68 (62%)	33 (30%)	9 (8%)
Complication: transient swallowing disturbance: 1 (0.9%)			
Wound infection: 1 (0.9%)			
Vertebral artery injury: 1 (0.9%)			
Permanent complication: none	Morbidity: 0		

Abbreviations: CCF, craniocervical fixation; JOACMEQS RR, recovery rate of Japanese Orthopedics Association, Cervical Myelopathy Evaluation Questionnaire Score.
Note: Follow-up period: 18 to 130 months, mean: 40.5 months.

had no neurological improvement, but rather deteriorated, and so FMD was added and their neurological symptoms improved. In the cases with increased intracranial pressure and/or hydrocephalus, a ventriculoperitoneal shunt was performed in 20 cases; 10 cases (50%) demonstrated an improvement of their neurological symptoms. The other 10 cases (50%) had no neurological improvement, so FMD was added and their neurological symptoms improved.

Reoperation Cases (►Table 6)

There were 22 cases (5.1%) who had deterioration of their neurological symptoms and/or the syrinx was not reduced: CM-I type A, 11 cases (8.5%); CM-I type B, 8 cases (6.0%); CM-I type C, 1 case (0.7%); and CM-absence: 2 cases (6.7%). There were significantly more of these cases in the CM-I type A, CM-I type B, and CM-absence groups than in the CM-I type C group. The most common reason for a second operation was instability at the CVJ in 12 cases (55%), with 11 of these cases in the CM-I type A, CM-I type B, and CM-absence groups, while inappropriate decompression was the cause in 7 cases (32%) in the CM-I type A and B groups. Two cases (9%) in the CM-I type A group developed tethered cord syndrome and SFT was added. One case (4.5%) in the CM-I type B group developed an arachnoid adhesion due to blood flow into the subarachnoid space during the initial operation, and lysis of the adhesion and untethering were performed. Only one case of cerebellar slumping (downward displacement of the cerebellum into the enlarged major cistern and displacement of the brain stem and cerebellar hemisphere) was observed in the ESCP group.

Discussion

Mechanisms: Pathogenesis of Ptosis of the Brain Stem and Cerebellum, and Surgical Indication and Outcome in CM-I type B, CM-I type C, and CM-absence

In these cases, ptosis of the brain stem and cerebellum was caused by a small VSFM and underdevelopment of the occipital bone. From the embryological viewpoint, the occipital bone is formed from enchondral bone that originates from occipital somites. Therefore, the pathogenesis of CM-I type B, CM-I type C, and CM-absence was insufficiency of the para-axial mesoderm, as the source of the occipital bone.[1,2]

In the management of Chiari malformation, appropriate surgical methods that can treat ptosis of the brain stem and cerebellum should be chosen. On the basis of this idea, we selected FMD for CM-I type B and CM-absence because VSFM and occipital bone size were small.[15] We chose ESCP for CM-I type C because FMD was not considered appropriate under the setting of a small VSFM, small VPCF, and small occipital bone size.[16] ESCP can normalize CSF flow, but it is also possible that the enlarged space created by ESCP could result in displacement of the brain stem and cerebellum into the PCF.

Both ESCP and FMD had good surgical outcomes. These outcomes were better than those reported previously in cases receiving only FMD, and morbidity and complications were also less than in previous reports.[16,17,19,22–26] This suggested that the choice of ESCP or FMD according to classification by morphometric analyses was appropriate.

Mechanisms of Ptosis of the Brain Stem and Cerebellum, and Surgical Indication in CM-I type A

In CM-I type A, which has normal VPCF, normal VSFM, and normal occipital bone size, there was no significant difference in the VPCB/VPCF ratio compared with the normal control group. Therefore, the mechanism underlying ptosis of the brain stem and cerebellum was not the narrowness of the PCF, but due to other mechanisms. Milhorat et al and the authors of the present study have reported the phenomenon of functional cranial settling, in which the cranium (occipital bone) falls into the CVJ. This functional cranial settling could cause ptosis of the brain stem and cerebellum.[8] In addition, Milhorat et al and the authors of the present study have indicated that tethering could cause ptosis of the brain stem and cerebellum by a traction effect.[9]

In CM-I type A, various mechanisms could cause ptosis of the brain stem and cerebellum. Other surgical methods that can treat ptosis of the brain stem and cerebellum must be chosen. CCF should be chosen for cases with craniocervical instability. Untethering and/or SFT should be chosen for cases with a lesion of traction and/or tethering.

Table 6 Reoperation cases

	CM-I type A	CM-I type B	CM-I type C	CM-absence
	FMD	FMD	ESCP	FMD
22/430 cases	11/117	8/133	1/150	2/30 (6.7%)[a]
5.1%	8.5%[a]	6.0%[a]	0.7%[a]	6.7%[a]
Other mechanism of ptosis of the brain stem and cerebellum				
CVJ instability	5	4	1	2
Incomplete decompression	4	3		
Tethered cord	2			
Arachnoid adhesion		1		

Abbreviations: CM-absence, cases which have neurological brain stem symptom and/or myelopathy but tonsillar herniation less than 5 mm; CM-I, Chiari malformation type I; CVJ, craniovertebral junction; ESCP, expansive suboccipital cranioplasty; FMD, foramen magnum decompression.
Note: Follow-up period: 18 to 130 months, mean: 40.5 months.
[a]Significantly high compared with CM-I type C group ($p < 0.001$).

In five cases who needed CCF after FMD or ESCP, CCF using the diploic screw technique, which we have described previously, was effective.[27]

Color Doppler Ultrasonography

The examination of CSF flow using CDU was very important.[19] In the present study, there was no difference in outcome between opening or not opening the arachnoid membrane. We were not concerned about preserving the arachnoid membrane at the major cistern, and confirming that CSF flowed out from the foramina of Magendie and Luschka was much more important. In all cases, we examined the dynamics of CSF flow at the major cistern using CDU (►**Fig. 3**). At the point of bony decompression, the CSF flow dynamics data never reached their previous levels. In the early stage (ESCP: five cases; FMD: five cases) at the point of incision of the outer membrane, CSF flow dynamics never reached their previous levels; therefore, dural plasty was added to all cases. Then, only outer membrane resection was not performed.

Preliminary Results of CCF

CCF led in a significant improvement of neurological symptoms and had a high rate of joint fixation. These results suggested that CCF was an effective surgical approach for cranial settling due to craniovertebral hypermobility or instability (►**Table 5**). To investigate hypermobility and instability at the occipitoatlantoaxial joints, a craniocervical traction test should be performed.[8]

Preliminary Results of Lysis, SFT, and Ventriculoperitoneal Shunt

Less than half of the cases with both untethering (lysis and/or SFT) and a ventriculoperitoneal shunt demonstrated an improvement of their neurological symptoms. Most of these cases were followed by FMD/ESCP and other surgical procedures, leading to an improvement of their symptoms. Therefore, only lysis and/or SFT and a ventriculoperitoneal shunt are not fundamental treatments for cases with CM-I and CM-absence (►**Table 5**).

Reoperation Cases (►Table 6)

Craniocervical instability was the most common reason for reoperation, and was observed significantly more often in CM-I type A, CM-I type B, and CM-absence than in CM-I type C.

Cases with CM-I type A, CM-I type B, and CM-absence should undergo a craniocervical traction test and examination of hypermobility and instability at the occipitoatlantoaxial joints. CCF had a good surgical outcome with regard to the improvement of neurological symptoms and fixation. In cases with CM-I types A and B, the FMD examination should include CDU to measure CSF flow and decompression of the brain stem and cerebellum.

Conclusion

Morphometric analyses of the PCF and CVJ are required to determine the mechanism and treatment of ptosis of the brain stem and cerebellum.

According to the mechanism of ptosis of the brain stem and cerebellum in the three types of CM-I, which were suggested by VPCF and morphometric analyses, a surgical procedure was selected. Each surgical treatment resulted in a good improvement of symptoms and safety course.

In the management of Chiari malformation, appropriate surgical methods that address ptosis of the brain stem and cerebellum should be chosen. It is important to perform appropriate decompression, while examination of CSF dynamics using CDU during surgery is recommended.

Note

This study has been approved by Institutional Review Boards of Koudoukai Health System, Osaka City University Graduate School of Medicine, Osaka, Japan, and North Shore University Hospital–Long Island Jewish Health System, New York, United States.

Conflict of Interest

None declared.

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