

Chiari I malformations: an Indian hospital experience

Ramnarayan R, Praharaj M S, Jayakumar P N

ABSTRACT

Introduction: A consensus on the ideal surgical procedure for Chiari I malformation has not been achieved. The purposes of the study were to report on the experience of treating this condition in an Indian hospital, and to look into the management of this not too uncommon condition with a view to improve the treatment and prognosis.

Methods: A retrospective case note study of all cases of Chiari I malformations operated over a ten-year period from 1989 to 1999 and followed-up for another two years, were analysed for clinical and radiological features. All patients who underwent posterior fossa decompression were included. Patients who underwent other procedures like shunts were excluded. The data obtained included clinical and radiological features.

Results: A total of 51 cases were analysed. All patients had posterior fossa decompression with duroplasty in the majority. Many patients had a delayed improvement noticed during the follow-up period. Headache, neck pain and motor features showed an improvement in the majority of the patients but sensory deficits did not.

Conclusion: This study showed the effectiveness of posterior fossa decompression with duroplasty as a procedure, which benefited most patients with Chiari I malformations.

Keywords: cerebellar tonsils, Chiari I malformations, duroplasty, hindbrain herniation, posterior fossa decompression, syringomyelia

Singapore Med J 2008;49(12): 1029-1034

INTRODUCTION

The Chiari malformations collectively describe increasing degrees of hindbrain herniation through the foramen magnum.⁽¹⁾ The type I malformation is defined as herniation of the cerebellar tonsils below the plane of the foramen magnum.⁽²⁾ Syringomyelia is associated with this condition

Table I. Age and gender profile of patients.

Age (years)	Gender (no.)		Total
	Male	Female	
< 20	6	3	9
21–40	23	10	33
> 40	4	5	9
Total	33	18	

in up to 75% of cases.⁽³⁾ Clinical presentations include neck pain, motor and sensory deficits or cerebellar features. Magnetic resonance (MR) imaging has now emerged the investigation of choice, though previously myelography and later intrathecal contrast computed tomography (CT) were used.⁽⁴⁾ Many treatment modalities have been proposed, including posterior fossa decompression, with or without duroplasty, and different types of shunts.⁽⁵⁾ The aim of this study was to ascertain the postoperative course of events in patients with Chiari I malformations and to compare the treatment modalities of durotomy and duroplasty after posterior fossa decompression.

METHODS

This was a retrospective case note analysis of all cases of type I Chiari malformation treated at The National Institute of Mental Health and Neurosciences (NIMHANS) in a ten-year period, 1989–1999. Cases were identified from the medical records library. The information that was taken into consideration included the clinical presentations of these patients, the radiological findings and the condition of the patients at discharge and at follow-up. This information was used for analysis. All cases of Chiari I malformations only, with or without syringomyelia, were included in this study. Excluded were patients with other types of Chiari malformations, patients with other associated malformations like basilar invagination or other craniovertebral anomalies, and patients who were treated by other methods like ventriculoperitoneal or other forms of shunts.

RESULTS

The case records of 51 patients that satisfied the criteria were obtained. The patients were categorised by age and gender (Table I). There were more males, and more than 60% of patients were in the third and fourth decades of life. About half of the patients presented with limb weakness and

Department of
Neurosurgery,
The National
Institute of Mental
Health and
Neurosciences,
Bangalore 560029,
Karnataka,
India

Ramnarayan R, MCh
Associate Professor
and Consultant

Praharaj MS, MCh
Additional Professor

Department of
Neuroimaging

Jayakumar PN, MD
Professor and Head

Correspondence to:
Dr Ramnarayan
Ramachandran
Malar Hospital,
Chennai,
India
Tel: (91) 44 2249 3265
Email: r_ramnarayan
@yahoo.com

Table II. Clinical and radiological profiles.

Profile	No. (%)
Clinical profile	
Headache including cough headache	18 (35)
Neck pain	15 (29)
Urinary complaints	3 (6)
Neck tilt	2 (4)
Gait disturbances	19 (37)
Nystagmus	21 (40)
Impaired corneal reflex	7 (14)
Muscle wasting	19 (37)
Reduced power	25 (48)
Sensory loss (segmental and dissociated)	32 (62)
Posterior column loss	16 (31)
Brisk deep tendon reflex	29 (56)
Cerebellar signs	17 (33)
Radiological profile	
Myelogram	1 (2)
Intrathecal contrast CT	26 (51)
MR imaging	24 (47)
With syrxinx	15 (29)
Without syrxinx	36 (71)

Table III. Operative profile.

Operative profile	No. (%)
Posterior fossa decompression with duroplasty	47 (92)
Posterior fossa decompression only	4 (8)

sensory complaints, and about a third had gait problems. Urinary involvement was seen only in three cases, whereas two patients came to the hospital for neck tilt. Clinical examination showed sensory involvement in the form of dissociated and segmental sensation, and posterior column loss as the most common sign. Motor signs in the form of wasting and weakness were seen in up to 50% and cerebellar signs in about a third of cases (Table II). The first patient in this series had a myelogram to identify the pathology. The diagnosis in half of the patients was confirmed with intrathecal contrast CT (Table II), especially the delayed scan done 24 hours later as described by Li and Chui.⁽⁴⁾ All patients in the latter part of the series (47%) had MR imaging to confirm the diagnosis. The radiological findings noted were the extent of the the descent of the tonsils and the presence and extent of the syrinx. More than 70% of patients did have an associated syringomyelia.

All patients were treated operatively with posterior fossa decompression (Table III). Of these, 47 patients had duroplasty done to increase the volume of the posterior fossa. Four patients had the posterior fossa decompression; the dura was opened but not closed (durotomy). Five patients had cerebrospinal fluid (CSF) leak, and this was controlled with drainage lumbar punctures. It was interesting to note that of these five patients, four were those who did not have duroplasty. Pseudomeningocele was treated conservatively in five patients. One of our patients

Table IV. Outcome profile.

Outcome profile	No. (%)		
At discharge			
Same as admission	29 (58)		
Improved	22 (42)		
Follow-up			
		Syrinx	No syrinx
Symptomatic improvement	38 (75)	8	30
Stabilised	6 (12)	3	3
Progression of features	4 (8)	3	1
Lost to follow-up	2 (3)	0	2
Death	1 (2)	1	0

Table V. Postoperative clinical improvement profile.

Feature	At discharge (%)	Follow-up (%)
Neck pain and headache	86	100
Gait disturbances	62	89
Cranial nerve deficits	33	76
Motor features	28	78
Sensory features	6	12
Cerebellar signs	30	39
Urinary complaints	0	11

with diabetes mellitus developed postoperative meningitis and, in spite of appropriate antibiotics, could not be saved.

The patients were assessed clinically at the time of discharge and at follow-up (Table IV). At discharge, 42% of patients had definite clinical improvement. We had a follow-up of one year in 46 (92%) of our patients, and 33 (66%) patients were followed-up for two years. During the follow-up period, improvement was noted in 38 patients (76%), six patients (12%) had stabilisation of the disease, and four patients (7%) continued to deteriorate. There was one death (postoperative meningitis) and two patients were lost to follow-up. In patients with syringomyelia, only 21% improved; whereas in those without syringomyelia, 79% improved. We then compared the surgical procedures with the outcome at discharge and at follow-up. There was no difference in the outcome between the procedures at discharge. But all the patients with the dura left open had CSF leak and had to undergo repeated lumbar punctures, leading to an increase in morbidity. During follow-up, it was noticed that all patients who had duroplasty had some improvement or stabilisation of the disease process. However, in the other group, no patient improved and one patient (2%) had non-progression of the disease. However, this finding may not be very relevant as the numbers of cases in the groups were disproportionate.

The clinical features at discharge and at follow-up were compared with the preoperative status (Table V). At discharge, neck pain and gait disturbances improved in 86%; and cranial nerve deficits, motor signs and cerebellar signs improved in a third. However, sensory features and urinary involvement did not improve much. During follow-

Table VI. Comparison of age, duration of illness and outcome.

Variable	No. (%) with outcome at follow-up			χ^2	p-value
	Improved	Same	Deteriorated		
Age (years)				5.79	NS
< 20	7 (88)	1 (12)	0		
20–40	27 (84)	3 (10)	2 (6)		
> 40	4 (50)	2 (25)	2 (25)		
Duration of illness (years)				12.66	0.00
≤ 5	34 (70)	3 (6)	1 (2)		
> 5	4 (8)	3 (6)	3 (6)		

NS: not significant

up, headache and neck pain disappeared in all patients and gait improved in the majority. Cranial nerve deficits, especially nystagmus, improved in 76% of cases. Motor features like motor power and brisk reflexes improved in 78% of cases. However, impaired corneal reflex and muscle wasting did not change in most patients. Sensory features, except posterior column signs, cerebellar signs and urinary involvement, did not make any progress during the follow-up period. In those cases with syringomyelia, the improvement of sensory features and bladder involvement was very little. Comparison was made between the age of presentation, the duration of symptoms before being operated on, and the postoperative outcome (Table VI). It was noted that age was inversely proportional to improvement but this correlation was not significant (chi-square value 5.79). Similarly, when the duration of illness was more than five years, the patients who improved were only a fraction of those with a duration of illness of five years or less. This finding was statistically significant (chi-square value 12.66, $p = 0.000$).

DISCUSSION

Chiari I malformation constitutes a group of entities of congenital or acquired aetiology that have, in common, descent of the cerebellar tonsils into the cervical spinal canal.⁽⁵⁾ This is associated with syringomyelia in 30%–75% of cases.^(4,6) Chiari I malformations have been traditionally viewed as a problem of young adults. In the series by Cahan and Bentson,⁽⁶⁾ there was a slight male preponderance. Majority of the patients were above 21 years of age. There were more females in the study by Panigrahi et al,⁽⁷⁾ and only four of the subjects were below 21 years of age. James and Brant had more females in their series, with ages ranging from 2 to 28 years.⁽⁸⁾ However, with the introduction of MR imaging, more and more young children are being diagnosed.^(3,9,10) Navarro et al described 96 patients, with ages ranging from 0.5 to 18 years.⁽¹¹⁾ Cheng et al described a female-to-male ratio of 3:2.⁽¹⁾ Attal et al had more males in their series, all aged above 22 years.⁽¹²⁾ In this series, males were also a majority, with most patients being young adults in their third and fourth decades of life.

Clinical presentation in patients with Chiari malformations is related to the compression of neural structures by the inferiorly-herniated tonsils or related to the associated syringomyelia.⁽¹⁰⁾ These may be motor, sensory, cerebellar or lower cranial nerve involvement in various combinations. Headache and neck pain is the most common complaints in both children and adults,^(9,10) but only a third of James and Brant's patients had headache.⁽⁸⁾ Ataxia as another feature was described by many authors.^(2,6,13) Headache and/or neck pain was seen in 64% of patients in this series, and 37% had ataxia as the presenting feature. Cranial nerve involvement was manifested by nystagmus and impaired corneal reflex in some patients. Nystagmus has been described by many authors,^(2,14,15) but no mention of impaired corneal reflex was found in the literature. Motor symptoms, including long tract signs like brisk reflexes, were seen in half of our patients. In the study by Dones et al,⁽¹⁴⁾ half of the patients presented with weakness, especially involving the upper limbs. Other studies also reported motor weakness as a predominant feature.^(2,9,15)

Sensory features in the form of dissociated and suspended sensory loss were the most common presentation, being seen in 62%, while posterior column impairment in the form of ataxia was seen in 31% in this series. Only a few studies had predominant sensory features as presenting complaints.^(6,15) Attal et al concluded that the duration of sensory deficits is the best predictive factor for the efficacy of surgery.⁽¹²⁾ Cerebellar signs were seen in 33% of our patients, but a much lower incidence had been being quoted in other studies.⁽³⁾ Three of our patients had upper motor type of urinary involvement. In the series of Goel and Desai, 55 out of 163 patients presented with urinary complaints.⁽¹³⁾ Two of our patients presented only with neck tilt; however, we did not find any other such reported finding. Scoliosis was described by many series as a very common finding,^(2,6) but not in this series.

In this study, one patient had only myelography; 50% of patients were diagnosed with intrathecal CT with delayed images, and the rest had MR imaging performed. Presently, MR imaging is the investigation of choice for

diagnosis. It is noninvasive, correlates well clinically, and dynamic imaging can be done.⁽¹⁶⁾ Pillay et al published a MR imaging classification of adult Chiari malformation based on objective anatomical criteria with clinical and prognostic relevance.⁽¹⁷⁾ Milhorat et al classified intramedullary cavities into anatomical and pathological types having different treatment requirements based on MR imaging criteria.⁽¹⁸⁾ Recently, Panigrahi et al concluded that cardiac-gated phase-contrast cine MR imaging flow study was an effective tool for deciding the type of surgery to be performed and also for postoperative monitoring.⁽⁷⁾ In fact, with the widespread use of MR imaging in diagnosis, more incidental and asymptomatic Chiari malformation are being detected. Nishizawa et al showed the benign nature of these lesions.⁽¹⁹⁾ Previously, myelography and CT with intrathecal contrast, especially with delayed images, were used for diagnosis.^(4,20)

The management of Chiari I malformations, with or without an associated syringomyelia, is controversial. Many types of procedures have been described, such as posterior fossa decompression, syringostomy, terminal ventriculostomy, and percutaneous aspiration of the cyst.⁽³⁾ However, most series have either done foramen magnum decompression or syringosubarachnoid shunting, or both. Cahan and Bentson noted that results of surgery for Chiari malformations were good, but at least one half of patients with syringomyelia showed continued progression of symptoms.⁽⁶⁾ Schijman and Steinbok conducted an international survey on the treatment of Chiari I malformation with syringomyelia and found much variation in the management.⁽⁵⁾ However, there was a consensus that no operation should be carried out in asymptomatic patients. Suboccipital decompression was the standard surgical procedure, and the majority favoured routine dural opening and closure with a patch. Batzdorf studied five patients and concluded that suboccipital craniectomy with duroplasty showed progressive collapse of the syringomyelic cavity taking place over several weeks.⁽²¹⁾

Milhorat et al based their study on MR imaging criteria and developed an algorithm for the treatment of syringomyelia from various causes. They advised decompression or use of shunts in Chiari malformation.⁽¹⁸⁾ Munshi et al compared posterior fossa decompression with and without duroplasty. Eight out of 11 patients who had only posterior fossa decompression, showed symptomatic improvement, but only 50% showed reduction in hydromyelia. With duroplasty, 20 out of 23 patients showed improvement in symptoms and 100% showed a decrease of hydromyelia.⁽²²⁾ Isu et al described a method of foramen magnum decompression with the removal of the outer layer of dura in seven patients, and showed improvement in six

over a period of two years. Of these, pain and numbness improved within a week, sensory improvement was noted in six patients, and motor improvement in four patients by one month.⁽²³⁾ Navarro et al also described a similar method of resecting the thickened dural band, meticulous scoring of the dura with preservation of the inner layer and avoiding dural laceration. They compared this method with duroplasty, and with duroplasty and tonsillar manipulation, and reported that dural scoring had a much better result. They also noted that patients without hydromyelia fared better.⁽¹¹⁾

Sindou et al did a literature comparison, and noted very good improvement with foramen magnum decompression with dural opening, but with preservation of arachnoid membrane and duroplasty. Opening the arachnoid also had similar results but much higher complication rates. Tonsillar resection did not show good results.⁽²⁴⁾ Klekamp et al showed the effectiveness of a small craniectomy rather than a large one. They suggested decompression of the foramen magnum with a small craniectomy, establishment of CSF flow and dural grafting as the surgical procedure of choice. They also commented that syrinx shunting has no place in Chiari I malformation, not even in patients with failed foramen magnum decompression.⁽²⁵⁾ Bindal et al showed that symptoms secondary to brain stem compression seemed reversible with decompressive surgery, whereas results with syringomyelia were less dramatic.⁽²⁾ Goel and Desai divided their patients into three groups based on aetiological factors and treatment considerations, and concluded that group I patients should be offered syringosubarachnoid shunting and group II foramen magnum decompression. Shunting only without decompression in group II patients was found to produce a poor outcome.⁽¹³⁾ In this group, none of the 22 patients showed any improvement and 12 cases actually worsened clinically in spite of collapse of the syrinx in postoperative imaging. In 15 patients, foramen magnum decompression was done subsequently, resulting in significant amelioration of symptoms. Vaquero et al studied 30 patients and concluded that both posterior fossa decompression and syringosubarachnoid shunting were equally useful in inducing syrinx collapse, and obex plugging was not necessary for syrinx collapse.⁽²⁶⁾

Hida et al compared foramen magnum decompression with syringosubarachnoid shunting. They concluded that shunting may be superior to decompression as an initial treatment for syringomyelia with Chiari I malformation especially with large syringes.⁽¹⁵⁾ But this study had a selection bias in that patients with symptoms of Chiari I malformation and/or a small syrinx underwent foramen magnum decompression, but patients with large syringes underwent syringosubarachnoid shunting. James and

Brant argued that craniocervical decompression without durotomy may be a suitable treatment modality for symptomatic Chiari malformation in children and young adults.⁽⁸⁾ Depreitere et al studied 22 patients who underwent posterior fossa decompression. Syringoperitoneal shunting was the treatment of choice in the unit in Belgium before 1989, but posterior fossa decompression was reintroduced. This was due to the fact that in cases which underwent only shunt surgery, the clinical outcome was less favourable when compared to the radiological outcome.⁽²⁷⁾ Ergun et al favoured posterior fossa decompression and syringosubarachnoid shunt in the same sitting as the most rational procedure in treatment of Chiari malformations.⁽¹⁶⁾ Dones et al suggested that the main benefit of surgical management in Chiari I malformation was to arrest the progression of the disease.⁽¹⁴⁾ Krieger et al presented 31 children who underwent limited occipital craniectomy and dural opening. The dura was left open and overlain with oxidised cellulose. They concluded that this is a very safe, effective and simple procedure in children.⁽²⁸⁾ 47 of our patients underwent foramen magnum decompression with duroplasty. Four patients had the dura left open. It was seen that the postoperative complications were more in this group of patients.

At discharge, 42% of our patients had clinical improvement; but during follow-up, this progressively increased to 76%. In 12% of our patients, the clinical features stabilised during the follow-up period of up to two years. Batzdorf⁽²¹⁾ and Dones et al⁽¹⁴⁾ had commented on this delay in improvement following surgery. Our results showed that patients who had a duroplasty had less morbidity compared to durotomy alone with marginally better improvement during follow-up. Neck pain and headache improved very well in our patients. At discharge, 86%, and during follow-up, 100% of patients were relieved symptomatically. Similarly, the gait also improved in 89% at follow-up. Cranial nerve deficits, especially nystagmus and motor features like tone and brisk reflexes, showed minimal improvement (28%) only at discharge, but during follow-up, these features improved in 78% of patients. Cerebellar signs and urinary problems had marginal improvements (9% and 11%, respectively) in a few patients but sensory involvement did not make much improvement, even during follow-up.

Dyste et al reported that 20% of their patients became asymptomatic, 66% improved, and 8% stabilised. Comparing the symptomatology, they reported that 81.5% of cases had improvement in pain, 70% of patients with their motor features improved, and 21% regained normal strength. 61% of patients had no improvement in sensory

deficits and 43% had no change in cranial nerve deficits.⁽⁹⁾ Cristante et al published their results in which cephalgias disappeared in 17 out of 20 patients, eight patients with cranial nerve deficits improved, and seven stabilised. They also found that 60% had improvement in motor weakness, and 50% of patients had stabilisation of sensory deficits with posterior column loss showing a tendency to recover.⁽²⁹⁾ Logue and Edwards suggested that neck pain and cough headache improved well, motor symptoms (30%) and posterior column loss (23%) improved moderately, and sensory symptoms very poorly (5%).⁽³⁰⁾ Pillay et al noted that foramen magnum symptoms and cerebellar signs improved in their patients but not features suggestive of central cord syndromes.⁽¹⁷⁾ Batzdorf reported that headaches improved in almost all, motor features in the majority, and sensory deficits only a few of his patients.⁽²¹⁾ Depreitere et al presented a retrospective analysis of 22 patients in whom best recovery was seen for weakness in the upper and lower limbs.⁽²⁷⁾ Attal et al studied the effects of surgery on sensory deficits, and noted that the effect of surgery on thermal deficits correlated with duration of sensory symptoms and vibration sense detection thresholds improved significantly.⁽¹²⁾ Dones et al concluded that the main benefit of surgery for Chiari I malformation was to arrest the progression of the disease.⁽¹⁴⁾

Asgari et al studied 31 patients with hindbrain herniation, with a mean follow-up of 35 months. They concluded that there was no correlation between clinical outcome and age, and between clinical outcome and duration of postoperative symptoms. But they noticed a good correlation between clinical outcome and result of postoperative MR imaging.⁽³¹⁾ Stevens et al analysed 141 patients retrospectively, and concluded that patient age and duration of clinical symptoms had no significant association with operative outcome.⁽³²⁾ Bindal et al studied 27 adults, and concluded that early diagnosis and treatment are critical in obtaining the best outcome especially in progressive neurological disease.⁽²⁾ Arora et al looked at seven factors, including the duration of symptoms and the extent of tonsillar descent, and concluded that the significant factors predicting good clinical outcome were the presence of basilar invagination, duration of symptoms and respiratory distress.⁽³³⁾ In this series, it was also noted that age was not related to outcome statistically, but duration of illness, especially if more than five years, was a significant factor affecting outcome.

In this retrospective study, it was very clear that posterior fossa decompression with durotomy alone does have a higher morbidity compared to posterior fossa decompression with duroplasty even though the numbers

are small. Munshi et al also had similar results.⁽²²⁾ Other studies also support posterior fossa decompression with duroplasty as the best method of treating Chiari I malformations.^(5,24,25,27) Depreitere et al concluded that decompression of the posterior fossa is a safe procedure with a considerable chance of clinical improvement.⁽²⁷⁾ Although total syrinx collapse is not as frequently seen as in syrinx shunting procedures, the clinical outcome may be better. There seemed to be no unequivocal correlation between clinical outcome and postoperative syrinx size. Goel and Desai commented that group II cases treated by a posterior decompression had a better long-term outcome when compared with group I cases treated by syringosubarachnoid shunt alone.⁽¹³⁾ In conclusion, posterior fossa decompression with duroplasty should probably be the procedure of choice in cases of Chiari I malformations with or without an associated syrinx. Durotomy alone may be associated with a higher morbidity.

REFERENCES

- Cheng JS, Nash J, Meyer GA. Chiari type I malformation revisited: diagnosis and treatment. *Neurologist* 2002; 8:357-62.
- Bindal AK, Dunsker SB, Tew JM Jr. Chiari I malformation: classification and management. *Neurosurgery* 1995; 37:1069-74.
- Wisoff JH. Chiari malformations and hydromyelia. In: Tindall GT, Cooper PR, Barrow D, eds. *The Practice of Neurosurgery*. Baltimore: Williams & Wilkins, 1995: 1875-954.
- Li KC, Chui MC. Conventional and CT metrizamide myelography in Arnold-Chiari I malformation and syringomyelia. *Am J Neuroradiol* 1987; 8:11-7.
- Schijman E, Steinbok P. International survey on the management of Chiari I malformation and syringomyelia. *Childs Nerv Syst* 2004; 20:341-8.
- Cahan LD, Bentson JR. Considerations in the diagnosis and treatment of syringomyelia and Chiari malformation. *J Neurosurg* 1982; 57:24-31.
- Panigrahi M, Reddy BP, Reddy AK, Reddy JJ. CSF flow study in Chiari I malformation. *Childs Nerv Syst* 2004; 20:336-40.
- James HE, Brant A. Treatment of Chiari malformation with bone decompression without durotomy in children and young adults. *Child's Nerv Syst* 2002; 18:202-6.
- Dyste GN, Menezes AH, VanGilder JC. Symptomatic Chiari malformations. An analysis of presentation, management and long term outcome. *J Neurosurg* 1989; 71:159-68.
- Steinbok P. Clinical features of Chiari I malformations. *Childs Nerv Syst* 2004; 20:329-31.
- Navarro R, Olavarria G, Seshadri R, et al. Surgical results of posterior fossa decompression for patients with Chiari I malformation. *Childs Nerv Syst* 2004; 20:349-56.
- Attal N, Parker F, Tadie M, Aghakani N, Bouhassira D. Effects of surgery on the sensory deficits of syringomyelia and predictors of outcome: a long term prospective study. *J Neurol Neurosurg Psychiatry* 2004; 75:1025-30.
- Goel A, Desai K. Surgery for syringomyelia: an analysis based on 163 surgical cases. *Acta Neurochir (Wien)* 2000; 142:293-302.
- Dones J, de Jesus O, Colen CB, Toledo MM, Delgado M. Clinical outcome in patients with Chiari I malformations: a review of 27 cases. *Surg Neurol* 2003; 60:142-8.
- Hida K, Iwasaki Y, Koyanagi I, Sawamura Y, Abe H. Surgical indications and results of foramen magnum decompression versus syringosubarachnoid shunting for syringomyelia associated with Chiari I malformation. *Neurosurgery* 1995; 37:673-9.
- Ergun R, Akdemir G, Gezici AR, et al. Surgical management of syringomyelia-Chiari complex. *Eur Spine J* 2000; 9:553-8.
- Pillay PK, Awad IA, Little JR, Hahn JF. Symptomatic Chiari malformation in adults: a new classification based on magnetic resonance imaging with clinical and prognostic significance. *Neurosurgery* 1991; 28:639-45.
- Milhorat TH, Johnson WD, Miller JI, Bergland RM. Surgical treatment of syringomyelia based on magnetic resonance imaging criteria. *Neurosurgery* 1992; 31:231-45.
- Nishizawa S, Yokoyama T, Yokota N, Tokuyama T, Ohta S. Incidentally identified syringomyelia associated with Chiari I malformations: Is early interventional surgery necessary? *Neurosurgery* 2001; 49:637-41.
- Woosley RE, Whaley RA. Use of metrizamide in computerized tomography to diagnose the Chiari I malformation. *J Neurosurg* 1982; 56:373-6.
- Batzdorf U. Chiari I malformation with syringomyelia. Evaluation of surgical therapy by magnetic resonance imaging. *J Neurosurg* 1988; 68:726-30.
- Munshi I, Frim D, Sline-Reyes R, et al. Effects of posterior fossa decompression with and without duroplasty on Chiari malformation associated hydromyelia. *Neurosurgery* 2000; 46:1384-90.
- Isu T, Sasaki H, Takamura H, Kobayashi N. Foramen magnum decompression with removal of the outer layer of the dura as treatment for syringomyelia occurring with Chiari I malformation. *Neurosurgery* 1993; 33:844-50.
- Sindou M, Chavez-Machuca J, Hashish H. Craniocervical decompression for Chiari type I malformation, adding extreme lateral foramen magnum opening and expansile duroplasty with arachnoid preservation technique and long term functional results in 44 consecutive adult cases – comparison with literature data. *Acta Neurochir* 2002; 144:1005-19.
- Klekamp J, Batzdorf U, Samii M, Bothe HW. The surgical treatment of Chiari I malformations. *Acta Neurochir (Wein)* 1996; 138:788-801.
- Vaquero S, Martinez R, Arias A. Syringomyelia-Chiari complex: Magnetic resonance imaging and clinical evaluation of surgical treatment. *J Neurosurg* 1990; 73:64-8.
- Depreitere B, Van Calenburgh F, van Loon J, Goffin J, Plets C. Posterior fossa decompression in syringomyelia associated with a Chiari malformation: a retrospective analysis of 22 patients. *Clin Neurol Neurosurg* 2000; 102:91-6.
- Krieger MD, McComb JG, Levy ML. Toward a simpler surgical management of Chiari I malformation in a pediatric population. *Pediatr Neurosurg* 1999; 30:113-21.
- Cristante L, Westphal M, Herrmann HD. Craniocervical decompression for Chiari I malformation. A retrospective evaluation of functional outcome with particular attention to motor deficits. *Acta Neurochir (Wien)* 1994; 130:94-100.
- Logue V, Edwards MR. Syringomyelia and its surgical treatment – an analysis of 75 patients. *J Neurol Neurosurg Psychiatry* 1981; 44:273-84.
- Asgari S, Engelhorn T, Bschor M, Sandalcioğlu IE, Stolke D. Surgical prognosis in hindbrain related syringomyelia. *Acta Neurol Scand* 2003; 107:12-21.
- Stevens JM, Serva WA, Kendall BE, Valentine AR, Ponsford JR. Chiari Malformations in adults: relation of morphological aspects to clinical features and operative outcome. *J Neurol Neurosurg Psychiatry* 1993; 56:1072-7.
- Arora P, Behari S, Banerji D, Chhabra DK, Jain VK. Factors influencing the outcome in symptomatic Chiari I malformation. *Neurol India* 2004; 52:470-4.