Clinico-Radiological Outcome of CHIARI I Malformation with Syringomyelia after Surgery

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ABSTRACT

BACKGROUND

Chari I malformation Type I represents a complex anatomical and / or a distended syrinx cavity. It is characterized by caudal descent of cerebellar tonsil. We wanted to analyse age and sex distribution of patients having CHAIRI I malformation with syringomyelia. We also wanted to analyse the clinical and radiological outcome of surgery.

METHODS

All patients who are surgically treated at the Department of Neurosurgery, SCB Medical College and Hospital, Cuttack, with a diagnosis of Chari-1 malformation with syringomyelia, reviewed prospectively from October 2015 - February 2018 were included in the study and this included minimum 6 weeks of follow up. Data regarding demographics, clinical symptoms and signs at the time of follow up was collected. Age groups of 2 - 65 years with CIM and syringomyelia were included.

RESULTS

32 cases were included in the study. Out of which 19 were males (59.37%) and 13 were females (40.62%). Male to female ratio is 1:1.46. The mean age of the patients was 29.5 years. Clinical characteristics showed four common presentations. Sensory loss 62.5%, headache and neck pain 53.1% and lower limb weakness with hyperreflexia in 50%. In this study, all the patients underwent MRI. Patients with syringomyelia were taken in account. The surgical treatment is always individualized for each patient. Sub-occipital craniotomy along with removal of posterior arch of atlas and augmentation of duraplasty is the most commonly performed procedure. The risk for complications for duraplasty was minimum in our series. A favourable clinical outcome resulted from relief of spinal cord distention as the syrinx becomes smaller in size.

CONCLUSIONS

The average age of the patient in the study was 29.5 years with a range of 2 - 65 years. Female to male ratio was 1:1.46. Hence, Chari 1 malformation was traditionally viewed as the problem of young adults. Surgery gave a good result and on long term follow up no problems related to cervical spine instability was noted.

KEYWORDS

CHARI I Malformation, Syringomyelia.

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DOI:10.18410/jebmh/2020/335

How to Cite This Article:
Bhujabal SN, Gouda AK, Dhir MK, et al. Clinico-radiological outcome of CHARI I malformation with syringomyelia after surgery. J Evid Based Med Healthc 2020; 7(32), 1596-1601. DOI:10.18410/jebmh/2020/335

Submission 09-05-2020,
Peer Review 15-05-2020,
Acceptance 25-06-2020,
Published 10-08-2020.

BACKGROUND

Chiari I malformation (CIM) is characterized by caudal descent of cerebellar tonsils and may or may not be associated with the presence of a syrinx. Herniation of the tonsils more than 5 mm below the foramen magnum on MRI (Magnetic Resonance Imaging) is considered diagnostic in adults.[1] Mikulis DJ. et al.

Criteria for ectopia of the cerebellar tonsils: 1st decade of life, 6 mm; 2nd and 3rd decades, 5 mm; 4th to 8th decades, 4 mm; and 9th decade, 3 mm.[2] It is more common in women, with a female-to-male ratio variably reported as 3:1 to 1:1.[3,4] If only patients with CIM associated with syringomyelia are included, the female-to-male ratio ranges from 1:1 to 1.3.[5] The reported prevalence of tonsillar herniation extending more than 5 mm below the foramen magnum on MRI of the head is 0.78%.[6] Some patients with this malformation are completely asymptomatic, and the diagnosis is established incidentally when MRI is performed for some other reason. The most common presenting symptom is pain (60% to 70%),[1,6] usually occipital and upper cervical in location.[7] It is described as a heavy, crushing, or pressure-like sensation at the back of the head that radiates to the vertex, behind the eyes, to the neck, shoulders and often induced by Valsalva manoeuvre such as laughing, sneezing, and coughing. Most patients also experience ocular disturbances, such as blurred vision, photophobia, diplopia, and visual field deficits. However, the neuro-ophthalmological examination in these patients usually is normal. Ten to 21% paediatric patients present with a lower cranial nerve palsy.[8,9,10] the most common symptoms in this group are dysphagia, sleep apnoea, dysarthria, hoarseness. Incidence of syringomyelia in CIM ranges from 30 to 70%.[11,12,13,4,14]

Patients with syringomyelia more frequently suffered motor deficits than those without syringomyelia (ratio 7:1). Sensory symptoms were also more frequent in this group (ratio 4:1).[9] The diagnosis of CIM is suspected when analysing the clinical course and physical examination, but it must be confirmed with imaging studies. MRI is considered the most important study for establishing the diagnosis and planning the surgical treatment. A T1-weighted sagittal view of the CVJ (Cranio vertebral junction) usually shows both the tonsillar herniation and syringomyelia, but in patients with small spinal cord cavities, T2- weighted imaging also can be very helpful. MRI also is useful for identifying other related anomalies, such as tumours and CVJ problems. Dynamic MRI helps demonstrate abnormalities in CSF (Cerebro spinal fluid) flow at the foramen magnum and the benefits of decompressive surgery in patients with the CIM.[15] USG (Ultrasonography) is an useful method in CIM with mild tonsillar herniation (rostral to C1), but it is used only during surgery to identify when the CSF circulation has been re-established during the procedure. The management strategy follows "top down" rule.[8,16,17] This implies that one proceeds from the cranial to caudal direction. If there is hydrocephalus it is dealt with first by ventriculoperitoneal shunt or other appropriate shunting technique. If the shunting does not ameliorate the symptoms then foramen magnum decompression with or without lax expansile duraplasty is done to deal with the impaction of tonsils into the upper cervical canal. The expansile duraplasty allows more room around the foramen magnum and opens the subarachnoid pathways. If with this manoeuvre the syrinx cavity resolves, or flow disappears then there is no need for further intervention. With the natural history known incompletely, poorest prognosis is seen in patients with central cord signs; the best prognosis is found in patients with paroxysmal intracranial hypertension.[18] Saez et al. classify patients into preoperative prognostic categories revealed that more than 80% of patients with paroxysmal intracranial hypertension or cerebellar dysfunction achieved a favourable outcome, that 65% of patients with foramen magnum compression improved, and that only 33% of patients with central cord disturbance improved.

METHODS

All patients who were surgically treated at the Department of Neurosurgery, SCB Medical College and Hospital, Cuttack, with the diagnosis of CIM with syringomyelia, reviewed prospectively for the period from October 2015 to February 2018 and this includes minimum 6 weeks follow up. Data were collected regarding demographics, clinical symptoms and signs at the time of admission, MRI features, surgery and postoperative complications. Post-operative follow up at 6 weeks, 6, 12, months was done to evaluate resolution of signs and symptoms, and imaging features. Maximum diameter of syrinx was measured preoperatively. Postoperatively the diameter was measured at the same level for comparison. Our results were compared to data in previously mentioned literature.

Inclusion Criteria

1. Age group 2-65 yrs.
2. All cases of CIM with syringomyelia.

Exclusion Criteria

1. Patient with secondary causes, such as a history of meningitis, encephalitis, hydrocephalus, intracranial lesions, occipito-cervical anomaly (including basilar invasion), brain or spine surgery and head trauma.
2. Patients who were treated by other methods like ventriculoperitoneal or other forms of shunt.

RESULTS

The results obtained from the study are expressed in the following section in tabular format. Numerical, continuous data are expressed with mean.

Demographics

There were 19 males (59.37%) and 13 females (40.62%), with female to male ratio of 1:1.46. The mean age of the patients was 29.5 years, with a range of 2 to 65 years.

	Male	Female
0 - 20 Years	2	1
21 - 40 Years	14	9
41 - 65 Years	3	3

Table 1. Age and Sex Distribution of Study Population

Clinical Characteristics

The 4 common presentations were sensory loss (segmental/dissociated) 62.5% (n = 20), headache, neck pain 59.3% (n=19), upper limb pain/numbness/weakness 53.1% (n = 17) and lower limb weakness, hyperreflexia in 50% (n = 16). Other presentations were cerebellar signs 34.3% (n=11), gait ataxia 28%, urinary incontinence 15.6%, lower cranial nerve dysfunction 9.3%, facial numbness, drop attack, scoliosis 6.25% each, chronic emesis 3.1%.

Clinical Characteristics	No. of Patients
Sensory loss	20
Headache/neck pain	19
UE pain/weakness/numbness	17
LE weakness/numbness	16
Cerebellar signs	11
Gait ataxia	9
Urinary incontinence	5
Lower CN palsy	3
Facial numbness	2
Drop attack	2

Table 2. Clinical Characteristics

Magnetic Resonance Imaging

All the patients were evaluated with magnetic resonance imaging to look for Tonsillar descent, presence and extent of syrinx, scoliosis. Tonsillar descent was present in all the patients, of which in 27 (range: 5 - 22 mm; mean 10.88 mm) the extent of tonsillar herniation was measured and mentioned in case records (Fig. 16). In the rest 5 patients, only the relation of the inferior border of cerebellar tonsils to C1, C2 was documented. Syrinx was present in all patients. Most commonly involved segment in syrinx was cervico-dorsal 71.8%, only cervical in 12.5%, only thoracic segment 6.25%, holocord syrinx 9.3%. Scoliosis was present in 6.25% (n=2), one toward right side and other toward left side.

Location	No. of Patients
Cervical	4
Cervico-dorsal	23
Dorsal	2
Holocord	3

Table 3. Syrinx Distribution

Surgical Procedure and Intraoperative Findings

Of the 32 patients with CIM, 59.37% (n = 19) underwent foramen magnum decompression with lax duraplasty, 40.62% (n=13) treated with foramen magnum decompression alone.

Type of Operative Procedure	No. of Patients
FMD	13
FMD with lax duraplasty	19

Table 4. Surgery

Patients who underwent posterior cranial fossa decompression with lax duraplasty dura was augmented using autologous pericranium (n=12), and fascia lata (n=7). The posterior arch of C-1 was bifid in 10% of patients. After removing the posterior arch of C-1, a dense constrictive band causing compression was observed in 58% of patients. The dura mater of the craniocervical junction was thickened, based on subjective assessment, in 40% of patients. Other anomalies noted at operation included thickened midline keels (internal occipital crests) and arachnoid adhesions within the fourth ventricle.

Complications

Out of 32 patients two patients died due to post-operative meningitis. CSF leak occurred in 6 patients, pseudo-meningocele developed in 3 patients, donor site hematoma occurred in patients in whom fascia lata was used as dural graft in 1 patient. All these complications occurred in patients in whom foramen magnum decompression with lax duraplasty was done.

Complications	No. of Patients
CSF leak	6
Pseudo meningocele	3
Donor site hematoma	1
Death	2

Table 5. Complications

Outcomes

Patients were assessed clinically at the time of discharge and at follow up. There were two deaths (post-operative meningitis) and two patients were lost to follow up.

Outcome Profile	No. (%)
Same as admission	22 (68.75%)
Improved	10 (31.25%)

Table 6. Outcome Profile at Discharge

Outcome Profile	No. (%)
Symptomatic improvement	23 (71.8%)
Stabilized	5 (15.62%)
Progression of features	0
Lost to follow up	2 (6.25%)
Death	2 (6.25%)

Table 7. Outcome Profile at 12 Months Follow Up

At Discharge

At discharge 31.25% of patients had definite clinical improvement (Table 6).

Follow-Up MRI

At 6 months follow up MRI was available for 25 patients (78%). Syrinx was diminished in size or resolved in 50% patients and remaining same for 28.12%. Mean syrinx diameter decreased from 7.34 ± 2.8 mm on the preoperative

MRI to 5.3 mm at 6 months follow up (Table 10). Six months after surgery pointed tonsils become rounded, cervicomedullary protuberance disappeared and mean tonsillar ectopia diminished.

Syrinx	No. of Patients
Diminished in size/resolved	16
Remaining same	9
Imaging not available	7

Table 8. Follow Up MRI (Syrinx) at 6 Months

At discharge, 31.25% of our patients had clinical improvement; but during follow-up, this progressively increased to 71.8% and 15.62% of patients become stabilized. Our results showed that patients who had a duraplasty had more morbidity compared to excision outer fibrous layer of dura alone. At 12 months follow up, sensory loss (segmental/dissociated) improved or stabilized in 45% of cases, headache, neck pain improved in 89.47% of cases, upper limb pain/numbness/weakness improved in 88.23% of cases, lower limb weakness, hyperreflexia improved in 81.25% of cases, cerebellar signs improved in 36.36% of cases, gait ataxia improved in 55.5% of cases. At 6 months follow up MRI was available for 25 patients (78%). Syrinx was diminished in size or resolved in 50% of patients and remaining same for 28.12% of patients. Mean syrinx diameter decreased from 7.34 ± 2.8 mm on the preoperative MRI to 5.3 mm at 6 month follow up .6 months after surgery pointed tonsils become rounded, cervicomedullary protuberance disappeared and mean tonsillar ectopia diminished. Nicholas M. Wetjen, John D. Heiss, and Edward H. Oldfield (2008) in their series reported that mean syrinx diameter decreased from 6.9 ± 2.1 mm on the preoperative MRI to 1.2 ± 1.5 mm at 3 to 6 months.

Dyste et al.[8] (1989) 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. There was no improvement in sensory deficits in 61% of patients and 43% had no change in cranial nerve deficits.

Cristante et al. (1994) published their results in which cephalgias disappeared in 17 out of 20 patients, 8 patients with lower cranial nerve deficits improved, and 7 patients stabilized. They also found that 60% had improvement in motor weakness, and 50% of patients had stabilization of sensory deficits with posterior column loss showing a tendency to recover.

Batzdorf et al. (1988) reported that headaches improved in almost all, motor features in the majority, and sensory deficits only a few of his patients.

Dones et al. (2003) concluded that the main benefit of surgery for CIM was to arrest the progression of the disease.

Asgari et al. (2003) 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 MRI.

M. J. McGirt et al.[14] (2003) reported that posterior fossa decompression relieves headache/neck/back pain in 80% of cases and 83% relieve in preoperative signs and symptoms. Symptomatic improvement in present study is comparable to earlier studies.

DISCUSSION

CIM is characterized by caudal descent of cerebellar tonsils and may or may not be associated with the presence of a syrinx, a degree of medullary descent and buckling of the lower medulla may also be present. A tonsillar herniation of more than 5 mm is widely considered pathological in adults. In children, cerebellar and neocortical growth causes a physiological herniation of the cerebellar tonsils, whereas in old age atrophy of the brain may lead to tonsillar ascent. Symptoms of CIM are related to age in infancy, signs of brainstem compression predominate with apnoea spells, cyanosis attacks, and swallowing problems, whereas in later childhood scoliosis becomes the most common presenting sign. The typical clinical features of occipital headache, gait ataxia, sensory disturbances, and motor weakness are observed predominantly in adults.

Demographics

The average age of the patients in the study was 29.5 years, with a range of 2 to 65 years, female to male ratio is 1:1.46. CIM have been traditionally viewed as a problem of young adults. Attal et al. (2004) reported more males in their series, all aged above 22 years.[19] Our study is in concordance with the study by R. S. Tubbs et al. and Cahan and Bentson et al in view of sex distribution. Average age of patients is higher in our study (29.5 years) as compared to previous studies (11 years) as there is combination of paediatric and adult age group.

Clinical Characteristics

Clinical presentation in patients with CIM is related to the compression of neural structures by the inferiorly herniated tonsils or related to the associated syringomyelia.[20] These may be motor, sensory, cerebellar or lower cranial nerve involvement in various combinations. In present study sensory features in the form of dissociated and suspended sensory loss were the most common presentation, being seen in 62.5%, while posterior column impairment in the form of ataxia was seen in 28%. Only a few studies had predominant sensory features as presenting complaints. Attal et al. (2004) concluded that the duration of sensory deficits is the best predictive factor for the efficacy of surgery.[19] In present study 59.3% patients presented with headache/neck pain. Headache and neck pain are the most common complaints in both children and adults,[8,20] but

James and Brant et al. (2002) reported headache only in 33% of their patient.

In a series by R. S. Tubbs et al. (2011) headache/neck pain was the presentation in 40% of patients. In a series by M. J. McGirt et al. (2003) 42.3% patients presented with headache/neck pain. Presentation as headache is in concordance with earlier published studies.

Motor symptoms, including long tract signs like brisk reflexes, were seen in half of our patients. In present study upper limb pain/numbness/weakness in 53.1% of patients and lower limb weakness, hyperreflexia in 50% of patients. In a study by Dones et al. (2003) half of the patients presented with weakness, especially involving the upper limbs. Other studies also reported motor weakness as a predominant feature. In present study cerebellar signs were present in 34.3% patients but other studies shows a much lower incidence. Gait ataxia present in 28% of patients. In a series by R. S. Tubbs et al. (2011) ataxia was the present in 3.8%. M. J. McGirt et al. (2003) observed that ataxia was present in 9.2% of patients. Presentation as ataxia varied significantly in different series. Urinary involvement was found in 15.6% of patients. Goel and Desai (2000) in their series demonstrated that 55 out of 163 patients (33.74%) presented with urinary complaints. In the present study only 6.25% of patients had scoliosis. Scoliosis was described by many series as a very common finding. In a series by R. S. Tubbs et al. (2011) scoliosis was present in 18%. In a series by M. J. McGirt et al. (2003)17.7% of patients had scoliosis.

Magnetic Resonance Imaging

In our study all patients underwent MRI. Presently, MRI is the investigation of choice for diagnosis. It is noninvasive, correlates well clinically, and dynamic imaging can be done. Tonsillar descent was present in all the patients, of which in 27 the extent of tonsillar herniation was measured and mentioned in case records. In the rest 5 patients, only the relation of the inferior border of cerebellar tonsils to C 1 and C 2 was documented.

Tonsillar Descent

5 - 10 mm: 13 patients,
11 - 20 mm: 13 patients,
>20 mm: 1 patient

In other 5 patients lower limit of tonsil was between foramen magnum and C1 in 3 patients, up to C2 level in 2 patients. In a series by M. J. McGirt et al. (2003) the inferior border of the cerebellar tonsil to be situated between the foramen magnum and C1 in 20% patients, at the level of C1 in 46.9% patients, at the level of C2 in 32.3% patients and at the level of C3 in 0.7% patients. In present study, all the patients had syringomyelia. The most commonly involved segment in spinal cord was cervico-dorsal 71.8%, only cervical in 12.5%, only thoracic in 6.25%, holocord syrinx 9.3%. In a series by R. S. Tubbs et al. (2011) syringomyelia was present in 57% patients. Syringomyelia involving holocord was most common seen in 39.3%, followed by cervicothoracic in 24.9%, cervical in 15.4%, thoracic in 14.7%, lumbar in 3.5% and syringobulbia in 2.1%. M. J. McGirt et al. (2003) demonstrated that syringes were present in 58% of patients. Syringomyelia involving holocord was most commonly seen in 44%, followed by cervical in 21.3%, thoracic in 16%, cervicothoracic in 12%, syringobulbia in 4% and lumbar in 2.7%. The most commonly involved segments in present study are cervicodorsal followed by cervical. In earlier studies syringomyelia was present in holocord followed by cervico-thoracic. Syringobulbia and syrinx involving the lumbar region are not seen in present study.

Surgical Procedure

In present study 59.37% (n=19) underwent foramen magnum decompression with lax duraplasty, 40.62% (n = 13) treated with foramen magnum decompression alone. In present study autologous pericranium in 12 cases and fascia lata in 7 cases was used as graft for duraplasty. In all cases the posterior arch of C 1 was removed and a dense constrictive band causing compression was observed in 58% of patients.

Heiss et al. (2012) reported successful surgical outcome (100% normalization of hindbrain herniation) was without arachnoid opening.

Dones et al. (2003) suggested that the main benefit of surgical management in CIM was to arrest the progression of the disease. In a series by R. S. Tubbs et al. (2011) all patients except one, underwent foramen magnum decompression and lax duraplasty at first operation with cadaveric pericardium in 8% of cases, autologous pericranium 91% of cases. One patient underwent lax duraplasty at second operation due to persistent symptoms. Posterior element of C 2 removed in 0.4% patients, tonsillar coagulation in 9.8% patients and craniospinal fusion in 2.4%. In a series by M. J. McGirt et al. (2003) all patients except one, underwent foramen magnum decompression and lax duraplasty at first operation. One patient underwent lax duraplasty at second operation due to persistent symptoms. Posterior element of C 2 removed in 0.8% patients, tonsillar coagulation in 16.9%. Foramen magnum decompression with lax duraplasty is most commonly done surgery in all studies.

Complications

In the present study 18.75% patients developed CSF leak, 9.37% patient developed pseudo meningocele (in one case primary closure, in two cases conservative management), one patient developed donor site hematoma (in whom fascia lata was used as dural graft), two patients died due to post-operative meningitis.

In a series by M. J. McGirt et al. (2003) complication occurred in 2.3%. 1.5% developed subdural extra axial collection which was managed with external ventricular drainage. 0.8% developed severe life-threatening signs of

brainstem compression following surgery managed with a transoral odontoidectomy and occipitocervical fusion. No infection, CSF leak, arterial injury or any death.

CONCLUSIONS

Chiari malformation Type I represents a complex anatomical and clinical entity. Patients with a symptomatic Chiari malformation and/or a distended syrinx cavity are ideal candidates for surgery where the main goal of surgery is to arrest the progression of neurological deficits and normalization of CSF flow. The role of surgery in patients with asymptomatic Chiari malformation continues to be a matter of debate. The surgical treatment should be individualized for each patient. Sub occipital craniectomy along with atlas's posterior arch removal and augmentation duraplasty seems to be the most commonly performed procedure. Simply performing foramen magnum decompression with removing outer layer of dura is a good option. The risk of complications from duraplasty was minimal in our series. A favorable clinical outcome resulted from relief of spinal cord distension as the syrinx became smaller and did not require complete disappearance of syrinx fluid on MRI. Symptoms usually improved incompletely after surgery; residual signs and symptoms did not signal a failure of therapy, but rather confirmed that the syrinx permanently injured the spinal cord before surgery. On long term follow up, no problems related to cervical spine instability was noted following removal of posterior arch of C1. Proper patient selection is necessary for achieving appropriate postoperative results.

Financial or Other Competing Interests: None.

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