

## Surgical Outcome of Chiari-I Malformation

NAQIB ULLAH ACHAKZAI, IRFAN ADIL, SALEEM KHAN

Department of Neurosurgery, Bolan Medical College/  
Sandeman Provincial Teaching Hospital, Quetta 87300 Pakistan

### ABSTRACT

**Objective:** To determine outcome in patients operated for Chiari-I Malformation.

**Study:** Descriptive case series.

**Materials and Methods:** Patients admitted in the Department of Neurosurgery, Bolan Medical College/ Hospital Quetta Pakistan from January 2013 to June 2017 with Chiari-I Malformation diagnosed on MRI and candidates for surgery based on consistent symptoms. MRI findings regarding cerebellar tonsillar termination in mm were recorded. Surgical Intervention consisted of suboccipitalcraniectomy, C1+/- C2 laminectomy(ies), reduction/ resection of cerebellar tonsil and expansion/augmentation duroplasty. Patients were followed-up after 6 months and outcome was determined by Chicago Outcome Scale for Chiari Malformation.

**Data Analysis:** SPSS version 20 software was used.

**Results:** Out of total of 39 patients 30 (77%) were females and 9 (23%) were males with Female to Male ratio of 3.3:1. Age ranged from 21 to 42 years. 35 (90%) patients presented with Valsalva headache, 34 (87%) with neckache, 25 (65%) with ataxia, 25 (65%) with paresthesia/numbness, 17 (44%) with sleep apnea, 16 (40%) with dysphagia, 15 (39%) with weakness, 12 (31%) with dizziness, 11 (27%) with visual disturbances, 9 (24%) with nausea/vomiting while 7 (18%) with tinnitus. 34 (88%) patients had tonsillar herniation more than 5mm while 5 (12%) had tonsillar herniation more than 10mm. Out of total of 39 patients 30 (77%) had good outcome while 9 (23%) had worse outcome.

**Conclusions:** Surgical decompression in Chiari-I Malformation should be offered to only those patients who are consistently symptomatic and their symptoms correlate with CM1. Surgery has favorable outcome in patients without significant cord damage and less severe neurological deficit before surgery.

**Key Words:** Chiari malformation, Tonsillar herniation, Foramen magnum decompression, Duroplasty, Chicago Chiari Outcome Scale (CCOS).

### INTRODUCTION

Chiari malformations are a group of disorders that comprises the downward herniation of the contents of the posterior fossa into the upper part of the cervical spinal canal through the foramen magnum.<sup>1,2</sup> These malformations are of different clinico-pathological entities having different etiology, pathophysiology and clinical signs and symptoms. Chiari malformations represent different degrees of posterior fossa contents herniation. Professor Hans Chiari classify these entities by using four tier scheme.<sup>3,4</sup> The most common Chiari malformation is type I (CM1) which is the

downward displacement of cerebellar tonsil into upper cervical spinal canal and it has incidence of 1 in 1000 births with slight female predominance and female to male ratio is 1.3:1. The other type Chiari-II is associated with neural tube defects in around 100% of cases particularly myelomeningocele.<sup>5</sup> Many theories have been proposed for the pathogenesis of this clinical entity.<sup>6</sup> Sarnat suggested that there is a primary defects in the genetic programming of hindbrain segmentation and of growth of associated bones and other cranial structures that results in Chiari malformations.<sup>7</sup> Another theory (hydrodynamic pulsion theory) suggests

that Chiari malformations are caused by early progressive hydrocephalus in fetus pushing down on the cerebellum and brainstem.<sup>8</sup> Some researchers believe that during early fetal development, defective closure of the neural tube results in the leakage of cerebrospinal fluid (CSF), which leads to insufficient distention of the fetal ventricular system and results in a small posterior fossa and cerebral disorganization.<sup>9</sup> Recently, Kong et al presented a new explanation called “evolutional mismatch theory.”<sup>10</sup> Most common symptom is occipital headaches that occurs in 73% to 98% of patients.<sup>2,11,12</sup> These headaches usually exacerbated by Valsalva maneuvers, such as coughing, sneezing, laughing etc. Other common symptoms include pain in the neck and shoulders, diplopia, dizziness, tinnitus, ear pain, weakness of upper extremities, abnormal sensations, dysphagia, apnea during sleep, poor fine skilled motor movements and sometimes occasional nausea and vomiting. Physical signs include nystagmus, diminished palate elevation, diminished gag reflex. Sometimes other signs such as dysmetria, hyperreflexia and ataxia may be present. Syringomyelia is associated with CMI in 24% to 75% of cases. If present then it may results in classic cape-like of upper extremity nociception sensory loss and also as atrophy of the intrinsic muscles of the hand. Scoliosis is seen in nearly 25% of patients with CMI.<sup>2,11,14,15</sup> To diagnose Chiari type I malformation, neuroimaging techniques are used whether patients having symptoms or no symptoms. The recommended technique for diagnosis is magnetic resonance imaging (MRI). MRI also gives details of the of the posterior fossa volume and flow dynamics of the cerebrospinal fluid. The diagnosis of associated syringomyelia should be made by MRI of the entire spine along with MRI of brain.<sup>16</sup> To determine which patient need surgery, MRI of whole craniospinal axis along with history and physical examination are the most important and effective tools. Patients having consistent symptoms with herniation of cerebellar tonsils below the foramen magnum detected on MRI are usually a candidate of surgery and surgical decompression is offered.<sup>17</sup> Patients of Chiari type I malformation who are asymptomatic and without syringomyelia should not be considered for surgery.<sup>13</sup> Decompression of the posterior fossa is the most common surgical approach done for CMI.<sup>18</sup>

Surgery is performed with duroplasty or without.<sup>19-22</sup> previously outcomes of surgery have been very difficult to assess because there was no measurement system that was standardized. Many attempts have been made and many different reporting schemes

were used in the past that include a scale know as gestalt impression of overall postoperative performance, that reports in terms of good, and fair etc. Out of these schemes, only the Chicago Chiari Outcome Scale (CCOS) is thought to standardized and validation has been done which is based quantitative measurement of signs and symptoms.<sup>23,24,25</sup> In this study we determine outcome of Chiari-I decompression surgery. The aim of this study is to determine the outcome of surgery of Chiari-I malformation in our department so that we can share our experience with other healthcare professionals so that they come to know the importance of surgery in Chiari-I malformation when recommended.

## MATERIALS AND METHODS

All patients admitted with Chiari-I malformation diagnosed on the MRI brain basis, who are candidates for surgery on the basis of MRI brain finding and clinical findings in the Department of Neurosurgery at Sandeman Provincial Teaching Hospital and Bolan Medical Complex Hospital Quetta from January 2013 to June 2017.

The basic demographic information were recorded like name, age, sex, address and hospital registration number. Clinical findings of patients, presented with one or more of the following; Valsalva headaches, pain in the neck, walking difficulties/ ataxia, paresthesias/numbness, apnea during sleep, dysphagia, limb weakness, dizziness, disturbances in vision, nausea/vomiting if present and tinnitus were recorded. MRI findings regarding cerebellar tonsillar herniation in mm (Fig-1) were recorded. All surgeries were performed by neurosurgeons and consisted of sub occipital craniectomy, C1+/- C2 laminectomy(ies), reduction/resection of the tonsils of the cerebellum and expansion/augmentation duraplasty were performed as usual by microsurgery. The tip(s) of the cerebellar tonsil(s) were coagulated, and/ or resected, depending upon findings and to ensure adequate decompression and patency of fourth ventricle (Fig. 2). Fourth ventricle carefully inspected to ensure the free flow of CSF fluid from the fourth ventricle laterally and also flow into the subarachnoid space. Patients without complications were discharged on 6<sup>th</sup> – 7<sup>th</sup> postoperative day. Patients were followed at 6 months and outcome was determined by Chicago Chiari Outcome Scale (CCOS). Signs and symptoms were measured in numerical values ranges from 1 to 4 in four different categories: first category is pain symptoms, second is non-pain

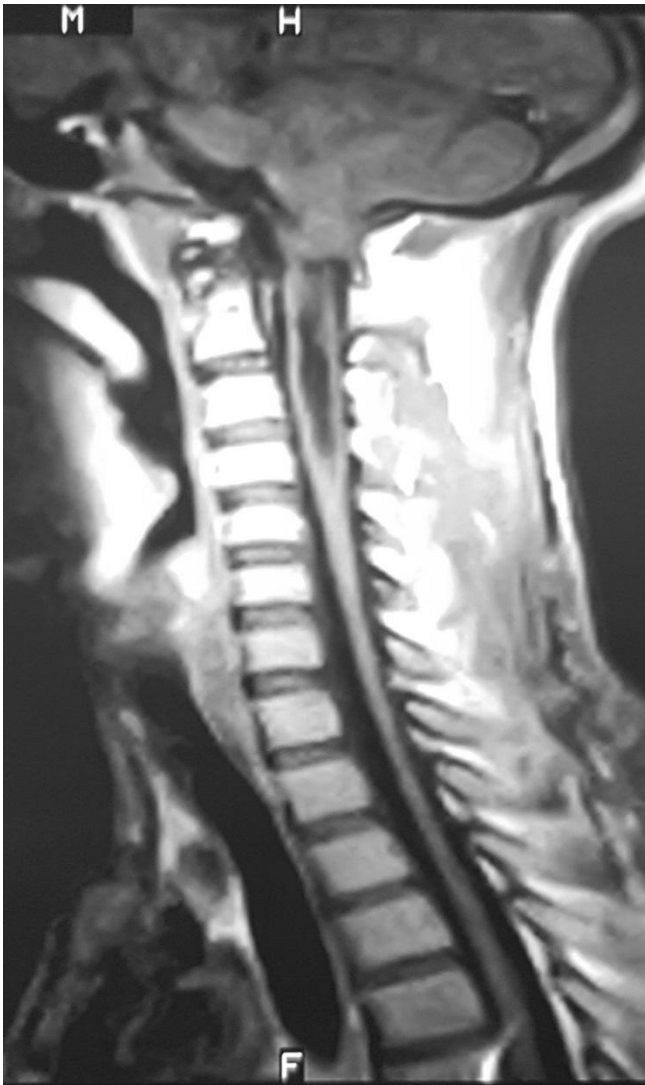


Fig. 1:

symptoms, third category is functionality and fourth category is complications. If there was a score of four in a category it shows complete resolution of symptoms of pain and symptoms other than pain, having full activity as before disease and without having any complication. If score was three then there is some improvement in preoperative pain and symptoms other than pain, and postoperative pain and symptoms other than pain controlled by nonsurgical methods. If score was two, it shows no change in preoperative pain, no change in symptoms other than pain, physical function impairment moderately or a complication that became prolonged but improved with nonsurgical modality. A score of one shows worsening of preoperative pain, worsening of symptoms other than pain, no functional improvement and a complication that is permanent and



Fig. 1a:

not responding to nonsurgical therapies. By summing score from each category, it resulted in a total score that ranges from 4 to 16. The score of 4 indicates complete post-surgery incapacitation. A score of 16 indicate excellent recovery. We use a cut-off score of 11 to denote a good or worse outcome. Using these criteria a score of 11 or above denotes good outcome while score less than 11 is considered as worse outcome.

### Data Analysis

SPSS software was used. Frequencies of variables like age, Valsalva headaches, pain in neck, ataxia, paresthesias, apnea during sleep, dysphagia, limb weakness, dizziness, disturbed vision, nausea/vomiting, and tinnitus, cerebellar herniation, good outcome, worse out-

come were described. Male to female ratio (M:F) was calculated.

**RESULTS**

39 patients were treated by surgery by decompression between 2012 and 2017. Out of 39, 30 (77%) patients were females and 9 (23%) were male with mean age at surgery of  $32 \pm 9$  years (Fig. 3). Female to male ratio was 3.3: 1. Age ranged from 23 to 41 years. 35 (90%) of patients presented with Valsalva headaches, 34 (87%) with pain in neck, 25 (65%) with ataxia, 25 (65%) with paresthesias/ numbness, 17 (44%) with apnea during sleep, 16 (40%) with dysphagia, 15 (39%) with weakness, 12 (31%) with dizziness, 11 (27%) with disturbed vision , 9 (24%) with nausea/ vomiting and 7 (18%) with tinnitus (Table 1). 34 (88%) patients had tonsillar herniation more than 5mm while 5 (12%) had tonsillar herniation more than 10 mm (Fig. 4). 30 (77%) patients had good outcome while 9 (23%) patients had worse outcome (Fig.5).



Fig. 2:

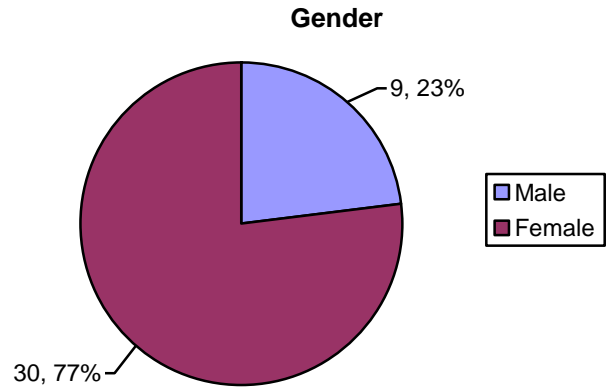


Fig. 3:

Table 1:

Clinical Presentation	Yes (%)	No (%)
Valsalva Headaches	35 (90%)	04 (10%)
Pain in neck	34 (87%)	05 (13%)
Ataxia	25 (65%)	14 (35%)
Paresthesias/Numbness	25 (65%)	14 (35%)
Apnea during sleep	17 (44%)	22 (56%)
Dysphagia	16 (40%)	23 (60%)
Weakness	15 (39%)	24 (61%)
Dizziness	12 (31%)	27 (69%)
Disturbance of vision	11 (27%)	28 (73%)
Nausea/vomiting	09 (24%)	30 (76%)
Tinnitus	07 (18%)	32 (82%)

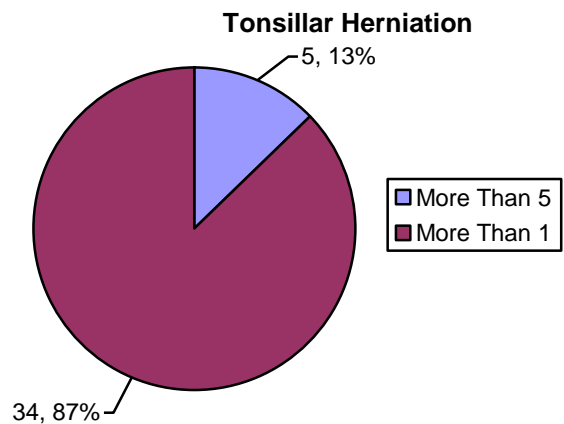


Fig. 4:

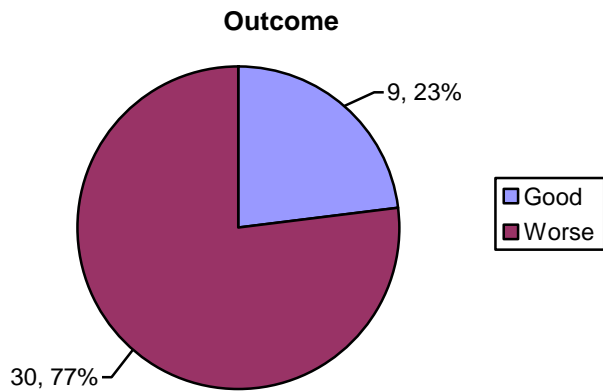


Fig. 5:

## DISCUSSION

Chiari Malformation comprises a complex syndrome that involves caudal herniation of the tonsils of the cerebellum through foramen magnum. Herniation should be at least 3–5 mm. In 1883, the first case of CMI was described by Cleland.<sup>13</sup> However, 1891 Chiari made a more detailed description. by the mid of 1970s, the syndrome was again named after Chiari. CMI sometime associated with an elongation of the fourth ventricle. It usually originate from mesoderm, but other origins, such as neuroectoderm and has also be in reportes.<sup>26</sup>

Chiari type I malformation along with syringomyelia are noteworthy among other many craniovertebral junction malformations. Their prevalence and the complexity of their symptoms made this entity very important. If a patient is symptomatic then surgical treatment should be offered. Literature emphasized that surgical treatment is given only if the patient is symptomatic with CM1.<sup>27</sup>

In our study there were 30 females and 9 males which shows female predominance and the mean age at presentation is 32 years which are consistent with previous studies.<sup>11, 28-31</sup>

Valsalva headache which is present in 35 of patients and is the very common clinical presentation in our series which is in agreement with other studies.<sup>13,17</sup> In our study the other most common presentations were neck pain in 34 patients, 25 with ataxia, 25 with paresthesias/numbness, 17 with apnea during sleep or snoring, 16 with dysphagia, 15 with limb weakness, 12 with dizziness, 11 with disturbed vision, 9 with nausea/vomiting, and 7 with tinnitus which are near to other international studies.<sup>1,17,21,22</sup> Similarly, 16 CM1 cases studied by Nagib and he concluded that clinical

features such as headache, pain in the cervical region, apnea during sleep, scoliosis are the significant prognostic factors and if they are present then patient shows favorable response after posterior fossa decompressive surgery.<sup>32</sup>

In this study, the preoperative symptoms were classified according to Chicago Chiari Outcome Scale into pain symptoms, symptoms other than pain and functionality.<sup>24</sup> In our study pain symptoms are more than non-pain symptoms which are in consistent with previous studies.<sup>17</sup>

Our study shows more than 5mm of cerebellar tonsillar herniation in 34 patients, and more than 10mm of herniation in 5 patients. These displaced tonsils are considered to be responsible for alterations of CSF dynamics at the Cervicomedullary junction.<sup>33-36</sup>

Outcome measurements were according to the validated numerical CCOS system which shows good out come in 30 patients and worse outcome in 9 patients. Our result correlates with other studies.<sup>13,17</sup>

## CONCLUSIONS

The Chiari malformations are a group of central nervous system defects that involve the posterior cranial fossa.

Valsalva (occipital) headache, pain in neck, ataxia, paresthesias/numbness, apnea during sleep, dysphagia, dizziness, limb weakness, disturbed vision, nausea/vomiting, and tinnitus are the main presenting symptoms of patients with CM1 Malformation.

Surgical decompression should be offered to only those patients who had constant symptoms and these symptoms correlate clinically with the CM1.

Surgery has favourable outcome when performed in consistently symptomatic patients without significant cord damage and less severe neurological deficit before surgery.

## Disclosure

The authors have no financial conflicts of interest.

Address for Correspondence:

Dr. IrfanAdil

Department of Neurosurgery, Bolan Medical College

at Sandeman Provincial Teaching Hospital

Quetta 87300 Pakistan

E-mail: doctorirfan777@yahoo.com

## REFERENCES

1. McVige JW, Leonardo J. Neuroimaging and the clinical manifestations of Chiari malformation type I (CMI). *Curr Pain Headache Rep.* 2015; 19: 18.
2. Fischbein R, Saling JR, Marty P, et al. Patient-reported Chiari malformation type I symptoms and diagnostic experiences: a report from the national Conquer Chiari Patient Registry database. *Neurol Sci.* 2015; 36: 1617–24.
3. Chiari H. ÜberVeränderungen des kleinhirns, des pons und der medulla oblongata infolge von congenitalerhydrocephalie des grosshirns. *Deutsche Medizinische-schrift.* 1891; 17: 1172–1175.
4. Chiari H. Überveränderungen des kleinhirns, des pons, und der medulla oblongata infolge von congenitalerhydrocephalie des grosshirns. *Denkschriften der Akademie der Wissenschaften Wien.* 1986; 63: 71–116.
5. Hidalgo J, Dulebohn S. Arnold Chiari Malformation. 2017 Jun. [Medline]. [Full Text].
6. Cesmebasi A, Loukas M, Hogan E, et al. The Chiari malformations: a review with emphasis on anatomical traits. *Clin Anat.* 2015; 28: 184–94.
7. Sarnat HB. Regional ependymal upregulation of vimentin in Chiari II malformation, aqueductal stenosis, and hydromyelia. *PediatrDevPathol.* 2004; 7: 48–60.
8. Royo-Salvador MB, Solé-Llenas J, Doménech JM, et al. Results of the section of the filumterminale in 20 patients with syringomyelia, scoliosis and Chiari malformation. *ActaNeurochir (Wien),* 2005; 147: 515–23. Discussion 523.
9. McLone DG, Knepper PA. The cause of Chiari II malformation: a unified theory. *PediatrNeurosci.* 1989; 15: 1–2.
10. Kong X, Yang Y, Gao J, et al. A new pathogenetic explanation of human chiari malformations. *J Pak Med Assoc.* 2015; 65: 804–5.
11. Milhorat TH, Chou MW, Trinidad EM, Kula RW, Mandell M, Wolpert C, et al. Chiari I malformation redefined: clinical and radiographic findings for 364 symptomatic patients. *Neurosurgery,* 1999; 44: 1005-1017.
12. Mueller DM, Oro JJ. Prospective analysis of presenting symptoms among 265 patients with radiographic evidence of Chiari malformation type I with or without syringomyelia. *J Am Acad Nurse Pract.* 2004; 16: 134-138.
13. Fernández AA, Guerrero AI, Martínez MI, Vázquez ME, Fernández JB, Octavio EC, et al. Malformations of the craniocervical junction (Chiari type I and syringomyelia: classification, diagnosis and treatment). *BMC Musculoskelet Discord.* 2009; 10 (Suppl. 1): S1.
14. Arnautovic A, Splavski B, Boop FA, Arnautovic KI. Pediatric and adult Chiari malformation type I surgical series, 1965-2013: a review of demographics, operative treatment, and outcomes. *J Neurosurg Pediatr.* 2015; 15: 161-177.
15. Tubbs RS, McGirt MJ, Oakes WJ. Surgical experience in 130 pediatric patients with Chiari I malformations. *J Neurosurg.* 2003; 99: 291-296.
16. Ventureyra EC, Azíz HA, Vassilyadi M. The role of cine flow MRI in children with Chiari I malformation. *Childs Nerv Syst.* 2003; 19: 109–13.
17. Gilmer HS, Xi M, Young SH. Surgical Decompression for Chiari Malformation Type I: An Age-Based Outcomes Study Based on the Chicago Chiari Outcome Scale. *World Neurosurg.* 2017 Nov; 107: 285-290. Doi: 10.1016/j.wneu.2017.07.162. Epub 2017 Aug 5.
18. Chavez A, Roguski M, Killeen A, Heilman C, Hwang S. Comparison of operative and nonoperative outcomes based on surgical selection criteria for patients with Chiari I malformations. *J ClinNeurosci.* 2014; 21: 2201-2206.
19. Hankinson T, Tubbs RS, Wellons JC. Duraplasty or not? An evidence-based review of the pediatric Chiari I malformation. *Childs Nerv Syst.* 2011; 27: 35-40.
20. Lee A, Yarbrough CK, Greenberg JK, Barber J, Limbrick DD, Smyth MD. Comparison of posterior fossa decompression with or without duraplasty in children with type I Chiari malformation. *Childs Nerv Syst.* 2014; 30: 1419-1424.
21. Gurbuz MS, Karaaslan N, Caliskan T, et al. Comparison of the surgical results for foramen magnum decompression with and without duraplasty in Chiari malformation type I. *Turk Neurosurg.* 2015; 25: 419–24.
22. Chotai S, Medhkour A. Surgical outcomes after posterior fossa decompression with and without duraplasty in Chiari malformation-I. *Clin Neurol Neurosurg.* 2014; 125: 182–8.
23. Greenberg JK, Milner E, Yarbrough CK, Lipsey K, Piccirillo JF, Smyth MD, et al. Outcome methods used in clinical studies of Chiari malformation type I: a systematic review. *J Neurosurg.* 2015; 122: 262-272.
24. Aliaga L, Hekman KE, Yassari R, Straus D, Luther G, Chen J, et al. A novel scoring system for assessing Chiari malformation type I treatment outcomes. *Neurosurgery,* 2012; 70: 656-664 [discussion: 664-665].
25. Yarbrough CK, Greenberg JK, Smyth MD, Leonard JR, Park TS, Limbrick DD Jr. External validation of the Chicago Chiari Outcome Scale. *J Neurosurg Pediatr.* 2014; 13: 679-684.
26. Schijman E. History, anatomic forms, and pathogenesis of Chiari I malformations. *Child's Nervous System,* 2004; 20 (5): 323–328.
27. Nash et al. in their review article support that surgical treatment is indicated only in symptomatic CM-I patients with radiographic evidence of hindbrain abnormalities.
28. Saez RJ, Onofrio BM, Yanagihara T. Experience with Arnold-Chiari malformation, 1960 to 1970. *J Neurosurg.* 1976; 45: 416–22. Doi: 10.3171/jns.1976.45.4.0416.

29. Dyste GN, Menezes AH, VanGilder JC. Symptomatic Chiari malformations. *J Neurosurg.* 1989; 71: 159–68. Doi: 10.3171/jns.1989.71.2.0159.
30. Bindal AK, Dunsker SB, Tew JM. Chiari I malformation: classification and management. *Neurosurgery,* 1995; 37: 1069–74. Doi: 10.1097/00006123-199512000-00005.
31. Pillay PK, Awad IA, Little JR, Hahn JF. Symptomatic Chiari malformation in adults: a new classification based on MRI with clinical and prognostic significance. *Neurosurgery,* 1991; 28: 639–45. Doi: 10.1097/00006123-199105000-00001.
32. Nagib MG. An approach to symptomatic children (ages 4–14 years) with Chiari type I malformation. *Pediatric Neurosurgery,* 1994; 21 (1): 31–35.
33. Bunck AC, Kroeger JR, Juettner A, Brentrup A, Fiedler B, Crelrier GR, Martin BA, Heindel W, Maintz D, Schwindt W, Niederstadt T. Magnetic resonance 4D flow analysis of cerebrospinal fluid dynamics in Chiari I malformation with and without syringomyelia. *European Radiology,* 2012; 22: 1860–1870.
34. Clarke EC, Stoodley MA, Bilston LE. Changes in temporal flow characteristics of CSF in Chiari malformation Type I with and without syringomyelia: implications for theory of syrinx development Clinical article. *Journal of Neurosurgery,* 2013; 118: 1135–1140.
35. Hofmann E, Warmuth-Metz M, Bendszus M, Solymosi L. Phase-contrast MR imaging of the cervical CSF and spinal cord: volumetric motion analysis in patients with Chiari I malformation. *AJNR Am J Neuroradiol.* 2000; 21: 151–158.
36. Oldfield EH, Muraszko K, Shawker TH, Patronas NJ. Pathophysiology of syringomyelia associated with Chiari I malformation of the cerebellar tonsils. Implications for diagnosis and treatment. *J Neurosurg.* 1994; 80: 3–15.

**AUTHORS DATA**

Name	Post	Institution	E-mail	Role of Authors
Dr. NaqibUllahAchakzai		Department of Neurosurgery, Bolan Medical College/ Sandeman Provincial Teaching Hospital, Quetta 87300 Pakistan	achakzai_66@hotmail.com	Writing of the Paper
Dr. IrfanAdil			doctorirfan777@yahoo.com	Data Collection
Dr. Saleem Khan				Data Analysis

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