ORIGINAL ARTICLE

The use of magnetic resonance phase-contrast cine in Chiari malformation with syringomyelia

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ABSTRACT

Introduction: Chiari malformation (CM) is a disorder of mesodermal origin and is commonly associated with syringomyelia. Foramen magnum decompression is the first-line of standard treatment in symptomatic patients with a confirmed radiographic diagnosis. Magnetic resonance (MR) cine allows accurate evaluation of cerebrospinal fluid (CSF) physiology at the craniovertebral junction but often this is under-utilised in Malaysia.

Methods: In this series, we looked into nine cases of CM with syringomyelia from clinical and radiological perspective before and after surgery. The radiological parameters were herniated tonsillar length, syrinx: cord ratio, syrinx length and diameter. Flow velocity and morphologic changes in Chiari were illustrated.

Results: Seven patients showed either reduction in syrinx length, syrinx: cord ratio or both postoperatively. Clinical recovery somewhat varied in motor and sensory symptoms. Four patients gained better functional grade in modified Rankin scale (MRS) while the rest remained similar. The study highlighted the advantage of CSF flow dynamics information over MR anatomical radiographic improvement in addressing the neurologic and functional recovery. We also discussed the practicality of cine sequence in preoperative patient selection, syrinx analysis and postoperative flow evaluation in anticipation of clinical outcome.

Conclusion: Phase-contrast cine MRI is a useful tool dictated by resource availability. We recommend its routine use in preoperative analysis and subsequent observational follow-up after surgery.

KEYWORDS:
Chiari malformation, syringomyelia, MRI, CINE, CSF flow dynamics

INTRODUCTION

Chiari malformations (CM) was first described by John Cleland and later classified by Hans Chiari in 1891.[1] The disease is a disorder of mesodermal origin with mixed neuroectodermal component. It is often associated with syringomyelia, up to 85% had been reported in Chiari I malformation.[2] Syringomyelia is a condition of fluid containing cavities in the parenchyma of the spinal cord.[3] For a long time the hypotheses evolved from Gardner's hydrodynamic theory, William's craniospinal pressure dissociation, to the recent Oldfield or Ball and Dayan's Virchow Robin Space theory. Many more theories were described and yet none was able to wholly reconcile with the clinical findings.[1,3] Posterior fossa foramen magnum decompression remains as the standard surgery to restore CSF flow at the craniovertebral junction. However, there is no consensus about the best operative technique especially in the context of the extent of suboccipital bony decompression.[4] An inadequately small decompression unlikely to resolve the Chiari symptoms; likewise, excessive large decompression will lead to cerebellar ptosis. Thus, the surgical outcome is variable and unpredictable. Those patients who experience poor clinical outcomes require more surgeries but often this is detected late.[5] In this study, we emphasise the use of phase-contrast cine as a routine sequence to analyse CSF flow dynamics, therefore to improve patient selection, evaluate the extent of surgical decompression, and correlate CSF physiology to clinical recovery.

MATERIALS AND METHODS

Patient selection

In this clinico-radiological cross-sectional study, nine patients were diagnosed symptomatic CM type I with syringomyelia, six were females and three were males. They were treated in Hospital Universiti Sains Malaysia (HUSM). Radiographic criteria of inclusion were defined as tonsillar herniation of at least 4mm below the foramen magnum (McRae's line) on mid-sagittal T1-weighted Magnetic resonance (MR) images. Those patients with craniovertebral junction abnormalities, hydrocephalus, and syndromic craniosynostosis were excluded. All the nine cases had posterior fossa foramen magnum decompression, C1 laminectomy and durofascioplasty.

Neurological Assessment

Preoperative clinical examinations of patients were conducted, and functional grade was determined using a modified Rankin scale (MRS). The preoperative symptoms

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and signs were summarised in Table I. The patients were followed-up and reassessed in the neurosurgical clinic postoperatively at six and twelve months. Patients were then classified as having had improvement, were the same or had worsening of clinical symptoms. Improvement was defined as complete or partial clinical recovery of motor, sensory deficits or both.

Radiological Imaging

We used 3.0 Tesla MRI T2-weighted sequence to measure the lengths of tonsillar herniation, syrinx lengths and its thickest diameter, cord diameter, syrinx: cord diameter ratio at mid-sagittal plane. Pre and postoperative radiological parameters were analysed by using measurement tools available on Picture Archiving and Communication System (PACS). Postoperative MRI were obtained between six and twelve months after surgery. MR phase-contrast cine CSF flow study was included in pre and postoperative studies for all the cases. The CSF flow velocity was measured at the level of foramen magnum. The sagittal scout view sequences were used as localisers to determine the anatomic levels. The anterior and posterior subarachnoid spaces were selected as region of interest (ROI) and applied to each of the multiple phases acquired in phase-contrast MR. The peak positive (systolic flow) and negative (diastolic flow) velocities were charted and plotted for each voxel in each phase. CSF flow was synchronised with cardiac cycle via peripheral gating of R wave, divided into 16 images at a velocity gradient of 5cm/sec.

Surgery

All patients had posterior fossa foramen magnum decompression and C1 laminectomy. Patients were placed in prone position and head fixed to a Mayfield clamp. A posterior midline incision was made from inion and extended to the C2 spinous process. Subperiosteal dissection to expose suboccipital bone and posterior arch of C1. A standard suboccipital craniectomy was performed, 30mm x 30mm (height x width) from the posterior lip of foramen magnum. Lamina of C1 posterior arch was rongeured. Atlanto-occipital band was released. Dura was opened in "Y" shape manner. The arachnoid layer was kept intact to prevent CSF leak or pseudomeningocoele. Tonsils were not resected. Durofascioplasty was performed and wound closed in layers.

Statistical Analysis

IBM SPSS version 24 was utilised to analyse the data to appraise the relationship between radiological signs and clinical symptoms or functions (MRS) to evaluate the adequacy of surgical decompression. P-value of less than 0.05 was considered significant.

RESULTS

Demographic data

The age of patients ranged from 15 to 41 years old, and the five were adolescents, 55.6%. Six cases were presented with either headache or neck pain which was localised to the occipital region or nape of neck. The headache was precipitated by straining or cough. The onset of symptoms was gradual and most of the patients were not able to accurately tell when the symptoms began. The mean duration of symptoms was 24 months. Their symptoms progressed and involved motor weakness or sensory deficit. Four patients had quadriparesis, 3 had bilateral upper limb weakness and 2 had upper limb monoparesis. Two of the quadriparetic patients were severely disabled with poor functional grade (MRS 5). Otherwise, most of the cases (n=5) had moderate disability. We encountered 2 cases with atypical presentation. One patient presented with acute type 2 respiratory failure, which required ventilatory support. Another patient presented with chest and neck pain, associated with progressive scoliosis. (Table I).

Preoperative radiological findings and clinical symptoms correlation

Our Chiari cases had a mean tonsillar herniation of 9.9mm. The syrinx of the patients was extensive in overall. It spanned at a mean length of 12±5.24 vertebral level across cervico-thoracic region. Syrinx were thickest at cervical region, predominantly C5 to C6 bony level. The mean syrinx: cord ratio was 0.74±0.14. All the cases demonstrated CSF flow obstruction at craniovertebral junction, and none showed decreased fourth ventricular outlet and aqueductal flow. Spearman's rank-order correlation showed statistically significant strong correlations between motor weakness, syrinx diameter (r=-0.858, p=0.003), tonsillar herniation (r=-0.702, p=0.035), and syrinx: cord diameter ratio (r=-0.687, p=0.041). Similarly, there was a strong correlation between ordinal preoperative MRS and syrinx: cord diameter ratio, which was statistically significant (r=-0.719, p=0.029).

Postoperative MR cine, radiological findings and neurologic recovery

Seven patients (77.0%) showed radiological improvement after the surgery. They either showed reduction in syrinx length (n=2), syrinx: cord diameter ratio (n=1), or both (n=4). The syrinx length reduction ranged from complete resolution to 6 vertebral body levels. Overall, the mean syrinx length was reduced by two vertebral levels and syrinx: cord ratio improved approximately 20% by 0.14. The syrinx: cord diameter ratio showed up to 75% of significant reduction, ranged from 0.20 to 0.69. Wilcoxon signed-rank test elicited statistically significant improvement in syrinx: cord ratio and syrinx length after the surgery (p-value <0.05) (Table II). Mean suboccipital bony decompression diameter was 33.8mm and C1 posterior arch diameter of 14.8mm. There were no major postoperative complications (massive blood loss, cerebellar ptosis, CSF leak or meningitis) that hindered neurologic recovery.

Although seven patients showed the radiographic improvement, functionally, there were only 4 patients with grade 3 to 4 MRS recovered to grade 2. These patients were able to ambulate and carry out daily living without assistance despite some disability. The condition of the other 5 patients remained unchanged despite surgery, in which two patients were bedridden and fully dependent. Neurologically, five out of nine patients had motor or sensory improvements after surgery and the other four patients showed static deficits. Six patients had complete resolve of their headache or neck pain (Table III).

Table I: Demographic, clinical and radiological data

Case No.	Age (yrs), Sex	Duration of symptoms (mos)	Headache/ Neck pain	Motor deficit	Sensory loss	MRS (Pre-op)	Radioimaging (MRI)			
							Tonsillar herniation	Syrinx length (Total level)	Syrinx thickest level	Syrinx: Cord ratio
1	24, F	66	Y	Quadriparesis	C4-S4 (Both)	5	10.3mm	C2-T10 (16)	C6	0.68
2	15, M	36	N	UL weakness (L)	C5-C8 (L)	3	11.6mm	C1-T12 (19)	C6-C7	0.88
3	52, F	240	Y	Quadriparesis	C5-S1 (Both)	4	11.0mm	C4-T10 (14)	T3	0.48
4	41, F	24	Y	UL weakness (Both)	C6-T1 (Both)	2	5.1mm	C2-C6 (5)	C5	0.83
5	19, F	2	N	Quadriparesis	C2-L5 (Both)	5	9.0mm	C2-T11 (17)	T2-T3	0.61
6	23, F	20	Y	UL weakness (Both)	C8-T1 (Both)	3	11.3mm	C1-T8 (15)	C5-C6	0.91
7	15, M	21	Y	Hemiparesis (R)	C4-T4 (R)	3	4.8mm	C2-T8 (14)	T3-T4	0.68
8	20, M	6	N	Quadriparesis	C5-S1 (Both)	3	16.0mm	C1-C6 (6)	C4-C5	0.75
9	16, F	24	Y	UL weakness (Both)	C4-T1 (Both)	3	10.0mm	C3-L3 (20)	C6-C7	0.83

*Y =Yes, N = No, UL = Upper limb, L = Left, R = Right

Table II: Pre and post op radiological parameters

		Mean (SD)	Median	z	p-value
Syrinx: cord ratio	Pre-op, n=9	0.74 (0.14)	0.75	-2.255	0.024
	Post-op, n=9	0.58 (0.22)	0.58		
Syrinx length in vertebral level	Pre-op, n=9	14.00 (5.24)	15.00	-2.023	0.043
	Post-op, n=9	12.00 (6.56)	13.00		

Table III: Surgical outcome in Chiari series

Case	Pre Op	Post Op							
		Functional grade/ Clinical symptoms				Radiological (MRI and CT)			
	MRS	MRS	Headache	Motor	Sensory	CINE CSF flow study	Syrinx length	Syrinx: Cord ratio	Decompression (cranial & C1 laminectomy)
1	5	5	C	S	S	No	Same	0.58 (Reduced)	B35mm, C17mm
2	3	3	NA	I	S	Return	Same	0.82	B30mm, C16mm
3	4	2	C	I	R	No	Reduced	0.32 (Reduced)	B34mm, C15mm
4	2	2	C	S	S	No	Resolved	0.20 (Reduced)	B36mm, C13mm
5	5	5	NA	S	S	No	Reduced	0.67	B33mm, C16mm
6	3	2	C	I	R	Return	Reduced	0.69 (Reduced)	B40mm, C14mm
7	3	2	C	R	S	Return	Same	0.57 (Reduced)	B35mm, C13mm
8	3	2	NA	I	R	Return	Reduced	0.45 (Reduced)	B35mm, C13mm
9	3	3	C	S	I	Return	Same	0.88	B31mm, C15mm

**NA = Not applicable, C = completely resolved, S = Similar, R = Recovered, I = Improved

Subgroup analysis was performed to appraise the relationship between radiographic and neurologic findings. Spearman signed-rank test showed positive moderate correlation between ordinal post-op MRS, syrinx: cord diameter ratio (r=0.553, p=0.122), level of syrinx (r=0.505, p=0.166) and diameter of bony decompression (r=-0.452, p=0.222). But, the results not statistically significant.

In the MR cine analysis, all the patients were categorised into two groups based on return of CSF flow. We found those patients with good return of CSF flow at anterior and posterior portion of craniovertebral junction showed clinical improvement (Figure 1). This was statistically significant in the Fisher's exact test (p=0.048). Peak velocity CSF flow waveforms at foramen magnum showed typical normal bi-directional sinusoidal waveform. There was an increase in the systolic magnitude with shorter duration. Longer duration of diastolic flow was seen as well. On the other hand, those with only radiological improvement had restricted or no CSF flow at the level of foramen magnum. The peak velocity CSF flow showed multiple short upward and downward pulsations indicating spatial inhomogeneity in velocities without sinusoidal pattern. Some of the cases showed low peak velocity with unchanged uni-directional

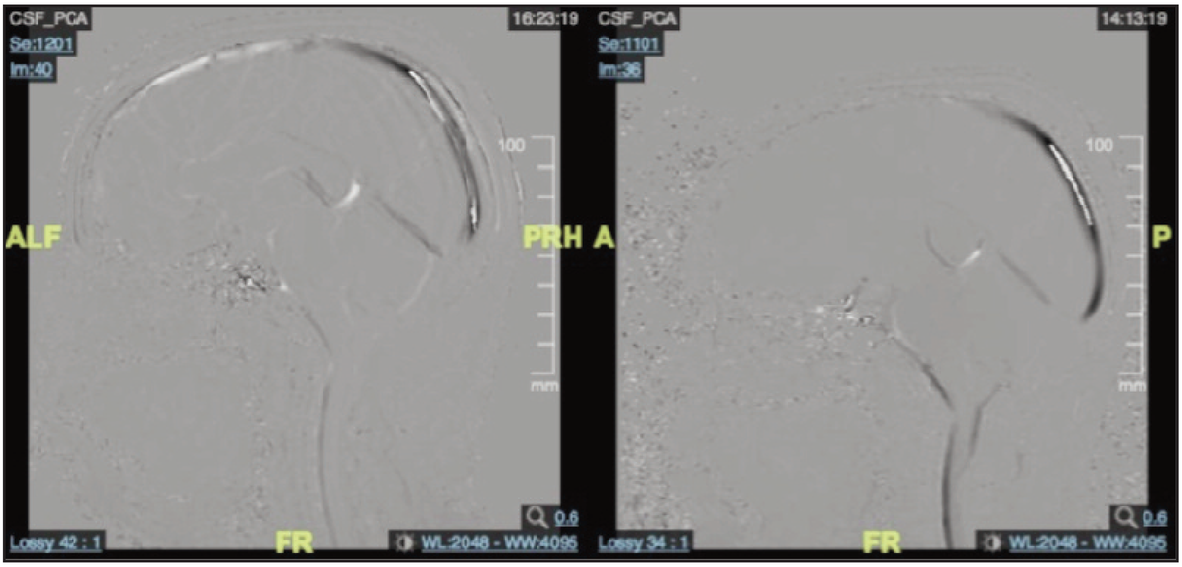

Fig. 1: MRI CINE showed pre (Left) and postoperative (Right) CSF flow at posterior foramen magnum.

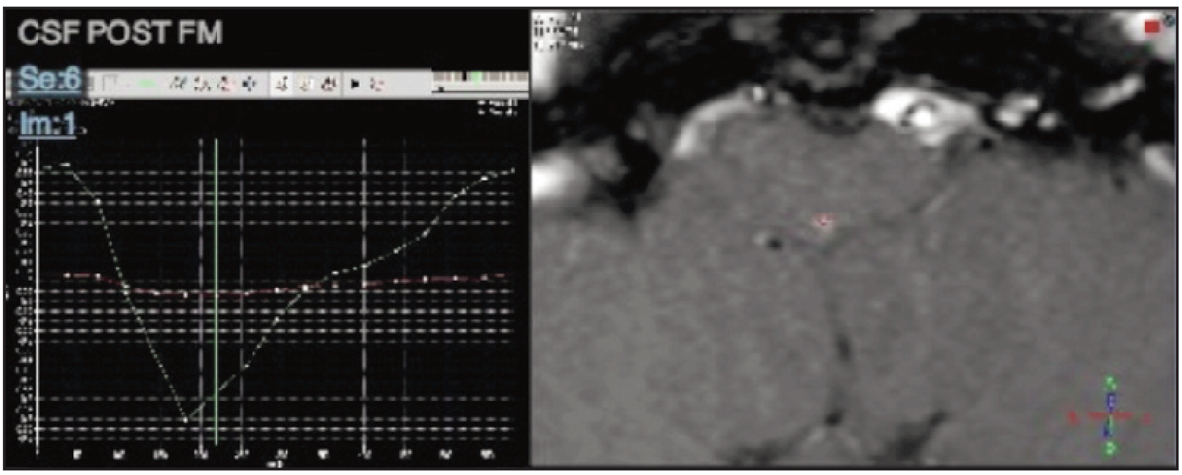

Fig. 2: Peak velocity CSF flow waveforms at foramen magnum shows normal bidirectional sinusoidal waveform.

waveform either in positive or negative flow. There were a substantial net cranial or caudal flows within voxels during each cardiac cycle.

DISCUSSION

Literature on Chiari decompression is fundamentally sound. Good clinical outcomes of CM are seen with standard decompression in most patients. Although the surgical criteria are based mainly on presenting symptoms and tonsillar herniation, some patients do not improve or even fare worse after the surgery. Such are the cohort of patients who require further surgical treatment, but early recognition of this state is somewhat difficult. While periodic clinical evaluation and assessment are time-consuming, it is inadequate to rely solely on MR anatomic changes. The use of MR cine provides valuable analysis from physiological perspective. First, the degree of CSF flow obstruction enables better selection of patients who are most responsive to surgery.[6,7] Secondly, changes in CSF velocity profile following surgery is practical in anticipation of symptomatic improvement.[8]

Cine phase-contrast MRI is a non-invasive imaging technique sensitive to the dynamics of CSF flows. This is not a novel method and its capacity to assess the CSF spaces beyond the bony margins is distinctive.[9] It works via mechanical coupling of cerebral blood and CSF flows in coordinated temporal sequence throughout cardiac cycle. Therefore, it detects the flow in three cardinal directions along which bipolar encoding gradients are applied. Over the past two decades, the technique has been utilised in many neurological conditions, including Chiari malformation; assessment and functionality of shunt in hydrocephalus; confirmation of aqueductal stenosis; and determining patency of a third ventriculostomy.[9-11]

In pulsatile flow theory, during systole, increase in cerebral blood volume and CSF is displaced caudally via dorsal subarachnoid space of foramen magnum. When diastole comes, the elastic recoil of spinal dura propels the CSF in a reverse caudocranial direction towards the intracranial compartment via ventral subarachnoid spaces. The CSF flow study is typically viewed in sagittal and axial manner. It captures the intracranial blood and CSF flow in a typical flush and fill manner, giving an alternating black and white motion effect signals along the CSF and venous pathway. The visualisation of such motion effect in dynamic loop denotes bidirectional physiological flow and can be plotted in a graph format. The line graph will display different time courses of CSF flow in dual axis. Y-axis indicates the magnitude of flow in positive and negative directions, in centimetre per second; while the X-axis shows time as a decimal fraction of the cardiac cycle, in milliseconds. The change in the direction of the CSF flow annotates transition between the systolic and diastolic phase. It will appear as an asymmetrical sinusoidal pattern along the time course. Duration of CSF flow in the systolic phase is shorter but has greater magnitude than that in the diastolic phase.

Nevertheless, this modality is dictated by resource availability in local setup. It is often under-utilised and unfamiliar to the end-user, resulting in unavailability. CM cases will only receive the conventional spin-echo, inversion recovery and diffusion-weighted sequences for MR imaging. In HUSM, we had the providential opportunity to collaborate with the radiology team to include cine sequence in Chiari cases. This had given us the additional advantage to evaluate preoperative CSF flow patency, velocity and waveform pattern at the level of foramen magnum. Although selection criteria for surgery are mainly based on clinical symptoms and structural abnormalities, we took into consideration of CSF flow obstruction at craniovertebral junction, both ventrally and dorsally. Postoperatively, we analysed the impact and extent of decompression on the overall CSF flow through its morphologic changes and correlated with clinical assessment. Thus, early re-intervention could be planned ahead of the conventional watchful observation.

Although our study had demonstrated radiographic shrinkage of syrinx in most patients after the standard foramen magnum decompression and durofascioplasty, only half had functional or clinical recovery. Such discrepancy between radiographic and neurologic findings was not able to be entirely explained even with correlation showed in the Spearman test. However, further analysis from CSF flow physiology perspective had rationalised the gap. Those Chiari cases with improved neurologic or functional grade had return of CSF flow and sinusoidal pattern. These findings paralleled improvement or resolution of clinical symptoms. Results were consistent and could be noticed as early as six months (Figure 2). On the other hand, the remaining patients who did not show restoration of CSF flow at foramen magnum had either unresolved clinical symptoms or static functional recovery.

This study had suggested that the radiologic improvement in syrinx and craniovertebral junction could only partially relate to resolution of symptoms. This was similar to other radiologic features like tonsillar ectopia and posterior fossa volume.[12,13] Clinical outcomes were better illustrated with additional information about CSF flow dynamic study, although the small sample size was likely a limiting factor. Nevertheless, it was also less likely to achieve large series in single centre due to rarity of the disease. Various studies showed that clinical symptoms were correlated well with abnormal cine flow imaging as compared to anatomical changes.[14,15] Another study showed that symptomatology was more accurately related to hindbrain CSF flow obstruction rather than degree to tonsils ectopia.[16] Chiari patients with obstructed CSF flow at foramen magnum preoperatively had better clinical response after surgery than those without.[17] One study showed that 40% of patients with no CSF flow pathology experienced treatment failures, while only 5% of patients with CSF flow obstruction before surgery.[18] MR cine indirectly highlighted the adequacy of standard decompression and emphasized the physiological abnormalities element in this complex structural disorder.[9,18,19] It can provide a direct evaluation for the follow-up and the postoperative surgery in patients with syringomyelia.[20] This could be noticed in our three cases that had similar surgery but did not achieve return of CSF flow. This might either mean that the standard decompression was not optimal to facilitate complete restoration of CSF flow for those cases, or other technical modalities were needed to be considered in second-look surgery. Information from MR cine would encourage earlier decision to intervene. However, we did not further intervene as two out of the three were very ill with poor functional grade. Our third patient opted for conservative measures.

CONCLUSIONS

Chiari malformation with syringomyelia is still a challenging disease in neurosurgery, and much regarding its pathogenesis remains debatable. MR cine is a sensitive method to determine CSF flow dynamics for a better selection of patients responding to the standard surgery preoperatively. Improvement of CSF flow dynamics following surgery is practical in anticipation of clinical and functional recovery. However, its usage, even as an adjunct in local setup is still sub-optimum. Effort should be made to include the sequence as a routine in MRI study of Chiari cases. Timely evaluation of CSF physiology and subsequent early intervention is imperative.

REFERENCES

1. Schijman E. History, anatomic forms, and pathogenesis of Chiari I malformations. Childs Nerv Syst 2004; 20: 323-8.
2. Bejjani GK; Cockerham KP. Adult Chiari malformation. Contemp Neurosurg 2001; 23: 1-7.
3. Chang HS, Nakagawa H. Hypothesis on the pathophysiology of syringomyelia based on simulation of cerebrospinal fluid dynamics. J Neurol Neurosurg Psychiatry 2003; 74: 344-7.
4. Alamar M, Teixidor P, Colet S, Muñoz J, Cladellas JM, Hostalot C. [Comparison (corrected) or Chiari I Malformation treatment using suboccipital craniectomy and posterior arch of C1 resection with or without dural graft]. Neurocirugia (Astur) 2008; 19: 233-41.
5. Noudel R, Gomis P, Sotoares G, Bazin A, Pierot L, Pruvo JP, et al. Posterior fossa volume increase after surgery for Chiari malformation Type IL a quantitative assessment using magnetic resonance imaging and correlations with the treatment response. J Neurosurgery 2011; 115: 647-58.
6. Bhadelia R, Frederick E, Patz S, Dubey P. Cough-associated headache in patients with Chiari I malformation: CSF flow analysis by means of cine phase-contrast MR imaging. J Neuroradiol 2011; 32:739-42.
7. Iskandar J, Quigley M, Haughton M. Foramen magnum cerebrospinal fluid flow characteristics in children with Chiari I malformation before and after craniocervical decompression. J Neurosurg Pediatr 2004; 01:169-78.

8. Battal B, Kocaoglu M, Bulakbasi N, Husmen G, Tuba Sanal H, Tayfun C. Cerebrospinal fluid flow imaging by using phase-contrast MR technique. Br J Radiol 2011; 84:758–65.
9. Bond AE, Jane JA Sr, Liu KC, Oldfield EH. Changes in cerebrospinal fluid flow assessed using intraoperative MRI during posterior fossa decompression for Chiari malformation. J Neurosurg 2015; 122(5): 1068-75.
10. Mbonane S, Andronikou S. Interpretation and value of MR CSF flow studies for paediatric neurosurgery. S Afr J Rad 2013; 17(1):26-9.
11. Panigrahi M, Reddy BP, Reddy AK, Reddy JJ. CSF flow study in Chiari I malformation. Childs Nerv Syst 2004; 20:336–40.
12. 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; 44:1005-17.
13. Nishikawa M, Sakamoto H, Hakuba A, Nakanishi N, Inoue Y: Pathogenesis of Chiari malformation: a morphometric study of the posterior cranial fossa. J Neurosurg 1997; 86:40-7.
14. Quon JL, Grant RA, DiLuna ML. Multimodal evaluation of CSF dynamics allowing extradural decompression for Chiari malformation Type I. J Neurosurg Spine 2015; 22: 622-30.
15. Enzmann DR, Pelc NJ. Normal flow patterns of intracranial and spinal cerebrospinal fluid defined with phase-contrast cine MR imaging. Radiology 1991; 178: 467-74.
16. Ventureyra EG, Aziz HA, Vassilyadi M: The role of cine MRI in children with Chiari I malformation. Childs Nerv Syst 2003; 19: 109-13.
17. Caldarelli M, Di Rocco C. Diagnosis of Chiari I malformation and related syringomyelia radiological and neurophysiological studies. Childs Nerv Syst 2004; 20:332–5.
18. McGirt MJ, Atiba A, Attenello FJ, Wasserman BA, Datoo G, Gathinji M, et al. Correlation of hindbrain CSF flow and outcome after surgical decompression for Chiari I malformation. Childs Nerv Syst 2008; 24:833–40.
19. Menezes AH. Craniovertebral junction abnormalities with hindbrain herniation and syringomyelia: Regression of syringomyelia after removal of ventral craniovertebral junction compression. Clinical article. J Neurosurg 2012; 116:301–9.
20. McGirt MJ, Nimjee SM, Fuchs HE, George TM. Relationship of cine phase-contrast magnetic resonance imaging with outcome after decompression for Chiari I malformations. Neurosurgery. 2006; 59(1):140-6.