Original Investigation

The Role of Cine Flow Magnetic Resonance Imaging in Patients with Chiari 0 Malformation

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ABSTRACT

AIM: To define the role of phase-contrast cine magnetic resonance imaging (MRI) in deciding the therapeutic strategy and underlying pathophysiology resulting in syrinx formation in patients with Chiari type 0 malformation.

MATERIAL and METHODS: Seven patients who were admitted to our clinic with the diagnosis of Chiari 0 malformation from January 2005 to July 2016 were enrolled in the study. All patients underwent a detailed preoperative neurological examination. Entire neuroaxis MRI and phase-contrast cine MRI were obtained preoperatively and postoperatively.

RESULTS: Seven patients (5 female and 2 male) with Chiari type 0 malformation fulfilled the inclusion criteria. All of the patients had absent cine flow at the craniovertebral junction except two patients. These five patients underwent surgical interventions; suboccipital decompression and duraplasty. All of them showed both clinical and radiological improvement in the postoperative period.

CONCLUSION: Cine flow MRI appears to be a useful tool in the management of patients with Chiari 0 malformation. There was a good correlation between the clinical presentation and cine flow preoperatively, and between clinical improvement and cine flow in the postoperative period.

KEYWORDS: Chiari 0 malformation, Cine flow magnetic resonance imaging, Decompression, Duraplasty

■ INTRODUCTION

The term "Chiari malformation" has been used to define conditions with ectopia of cerebellar tonsils below the level of the posterior edge of the foramen magnum for several decades. Since the late 1990s, the "Chiari type 0 malformation" is defined as syringomyelia without tonsillar herniation that responds to posterior fossa decompression (23-25). The increasing awareness of this malformation has led to widespread use of magnetic resonance imaging (MRI). The pathophysiology resulting in syrinx formation in this group of patients is poorly understood and there is no consensus on the management of the patients. The responses to surgery suggest that a "Chiari-like" pathophysiology may be present in the absence of tonsillar herniation.

Phase-contrast cine flow MRI is a non-invasive method of studying cerebrospinal fluid (CSF) circulation in the brain and the craniocervical junction and also enables understanding the pathophysiology of the Chiari malformation. All the theories relating to the cause of Chiari type 0 malformation with an associated syrinx implicate abnormal CSF flow.

In the present study, we investigated the role of phase-contrast cine MRI in deciding the therapeutic strategy and the underlying pathophysiology resulting in syrinx formation in patients with Chiari type 0 malformation.

■ MATERIAL and METHODS

The medical records of the patients who were admitted to our clinic with the diagnosis of Chiari 0 malformation from

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January 2005 to July 2016 were reviewed. Seven patients (5 female and 2 male) fulfilled the inclusion criteria, which included Chiari type 0 malformation (cervical syringomyelia without cerebellar tonsils ectopia) without other causes of a syrinx. Five cases were operated, and suboccipital decompression and duraplasty were performed. All patients underwent entire neuroaxis MRI (GE 1.5T) with or without contrast enhancement (Figure 1A, B). The phase-contrast sequence was obtained in the midsagittal plane, gated with the cardiac cycle using the specific software of GE for this test. Phasic and cine flow images were obtained and studied pre- and postoperatively at 3 months.

Several variables were analyzed including age at presentation, duration of symptoms, main symptomatology, and management. MRI studies were reviewed and the following variables were recorded: presence or absence of cine flow at the craniovertebral junction and diameter of the syrinx cavity.

■ RESULTS

The mean age of the patients at presentation was 36 years. Six patients were symptomatic for Chiari type 0 malformation, showing a wide variation as severe suboccipital headache, cervical pain, vertigo, dizziness and motor/sensory deficits (Table I). The mean duration of symptoms was 20 months. Two patients were diagnosed incidentally as a result of MRI performed for other medical reasons. Five patients with Chiari type 0 malformation underwent surgical procedures; suboccipital decompression and duraplasty were performed. Mean clinical follow-up period was 23 months. All 5 patients showed both clinical and radiological improvements in the postoperative period. One month later, postoperative MRI showed mega cisterna manga, but the syrinx cavity showed no change in diameter. Syrinx cavity diameter had decreased at the sixth month after surgery (Figure 2A, B). All patients with Chiari type

0 malformation and absent cine flow preoperatively improved after suboccipital decompression and duraplasty 3 months later (Figures 3, 4). One month after surgery, the presenting symptoms of all patients had improved. There was a good correlation between the clinical presentation and the radiological findings in pre- and postoperative periods.

The patients with normal cine flow at the craniovertebral junction in MRI were followed without surgical intervention for approximately two years. Cranial MRI and phase-contrast cine flow MRI were performed at six-months periods. There was progression of clinical and radiological findings or new findings in the follow-up periods.

■ DISCUSSION

The Chiari malformations are a group of congenital hindbrain abnormalities which are observed in conjunction with spinal cord malformations. The term "Chiari malformation" has been used to define conditions with ectopia of cerebellar tonsils below the level of the posterior edge of the foramen magnum for several decades. The first published reports were described by Hans Chiari in 1891 (7). They are categorized into four traditional types, with Types I and II accounting for almost 99% of the clinical cases. Chiari I malformation is characterized by underdevelopment and overcrowding of the posterior cranial fossa and the associated neurological dysfunction secondary to hindbrain compression and syringomyelia. Caudal herniation of cerebellar tonsils more than 5 mm below the foramen magnum has been described as "Chiari I malformation". Since the late 1990s, there is increasing evidence that many patients with an overcrowded posterior cranial fossa and typical Chiari malformation type I signs, such as syringomyelia, do not show tonsillar descent; many authors refer to these cases as Chiari malformation type 0 (23-26).
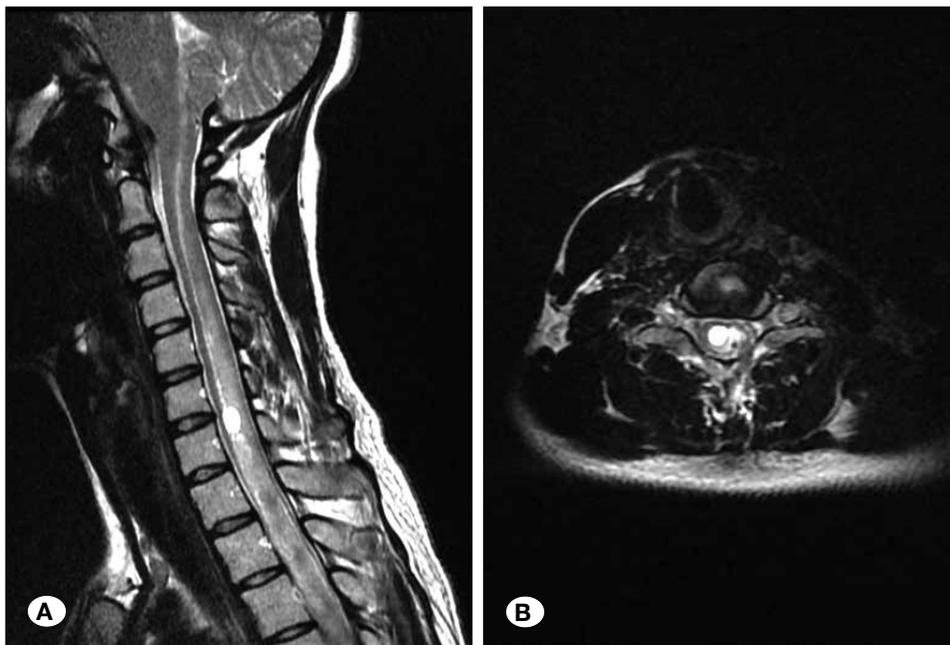
Figure 1: Preoperative **A)** sagittal and **B)** axial cervical MRI revealing cervicothoracic syringomyelia without tonsillar herniation (Chiari type 0 malformation) (syrinx diameter is 9 mm).

Iskandar et al. first described 5 patients with syringomyelia and no evidence of tonsillar herniation (14). MRI of the entire neuroaxis ruled out other causes of a syrinx. Ultimately, abnormal CSF flow at the posterior fossa or foramen magnum may be a possible cause of syrinx. All five patients underwent a posterior fossa decompression and duraplasty without direct fenestration or management of the syrinx. Significant syrinx resolution and symptom improvement were observed in all patients. Their improvements after surgery suggest that "Chiari-like" pathophysiology may be present in the absence of tonsillar herniation, which may possibly be intermittent. This contention is supported by the identification of crowded

Table I: Chiari 0 Patients' Data and Preoperative and Postoperative Radiological Findings

Patient	Age (years)/ Sex	Symptoms	Phase-contrast cine MRI	Management	Preoperative syrinx diameter	Postoperative/ Follow-up syrinx diameter (6th month)
1	30/F	Asymptomatic	Positive cine flow	Follow-up	4 mm	4 mm
2	40/F	Neck pain	Positive cine flow	Follow-up	8 mm	8 mm
3	35/F	Quadriparesis	Absent cine flow	Suboccipital decompression and duraplasty	13 mm	7 mm
4	36/M	Numbness and pain in the arms	Absent cine flow	Suboccipital decompression and duraplasty	7 mm	3 mm
5	32/M	Neck pain, headache and vertigo	Absent cine flow	Suboccipital decompression and duraplasty	9 mm	3 mm
6	33/F	Neck pain and quadriparesis	Absent cine flow	Suboccipital decompression and duraplasty	8 mm	4 mm
7	43/F	Neck pain, numbness and pain in the arms	Absent cine flow	Suboccipital decompression and duraplasty	9 mm	4 mm

M: Male, F: Female, MRI: Magnetic resonance imaging.
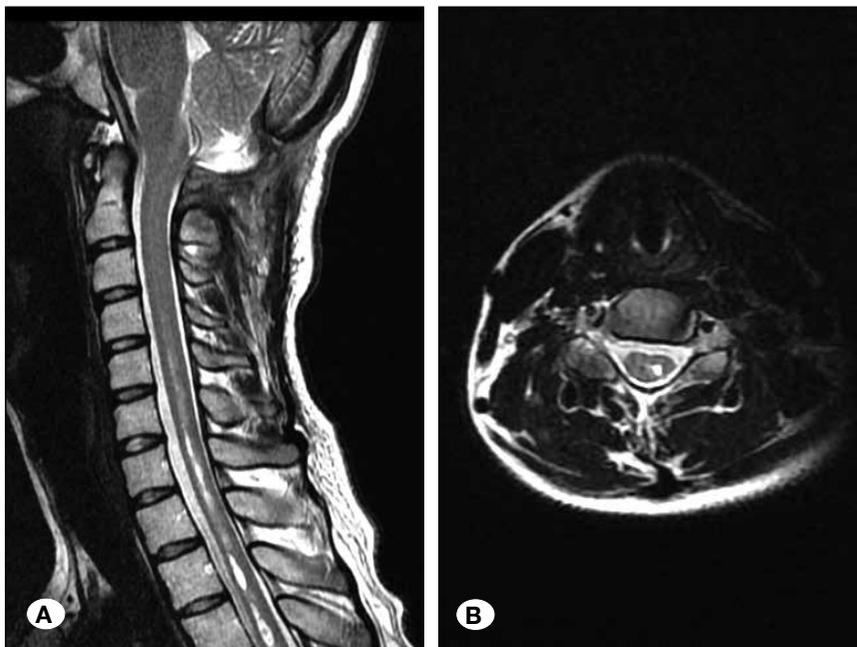
Figure 2: Postoperative **A)** sagittal and **B)** axial cervical MRI of the same patient revealing the regression of syringomyelia (postoperative syrinx diameter is 4 mm).
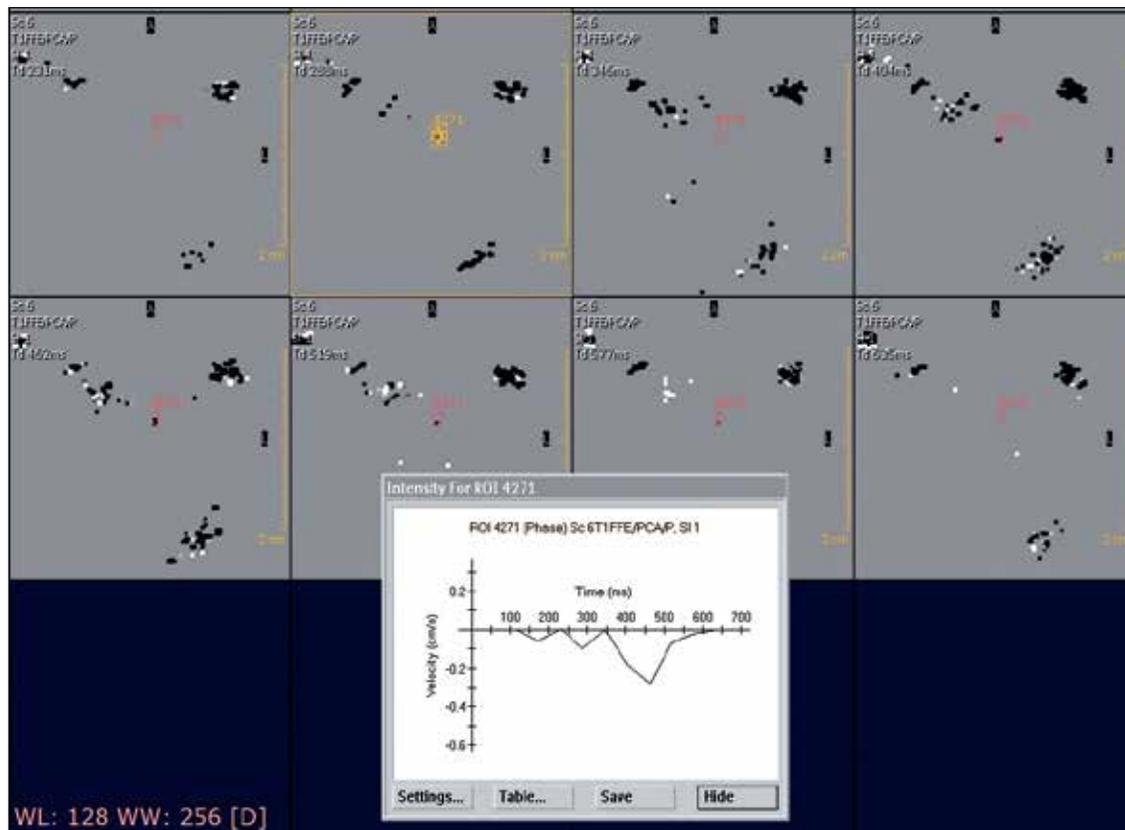
Figure 3: Preoperative CSF velocity-time graph obtained at the foramen magnum level demonstrates absence of flow.
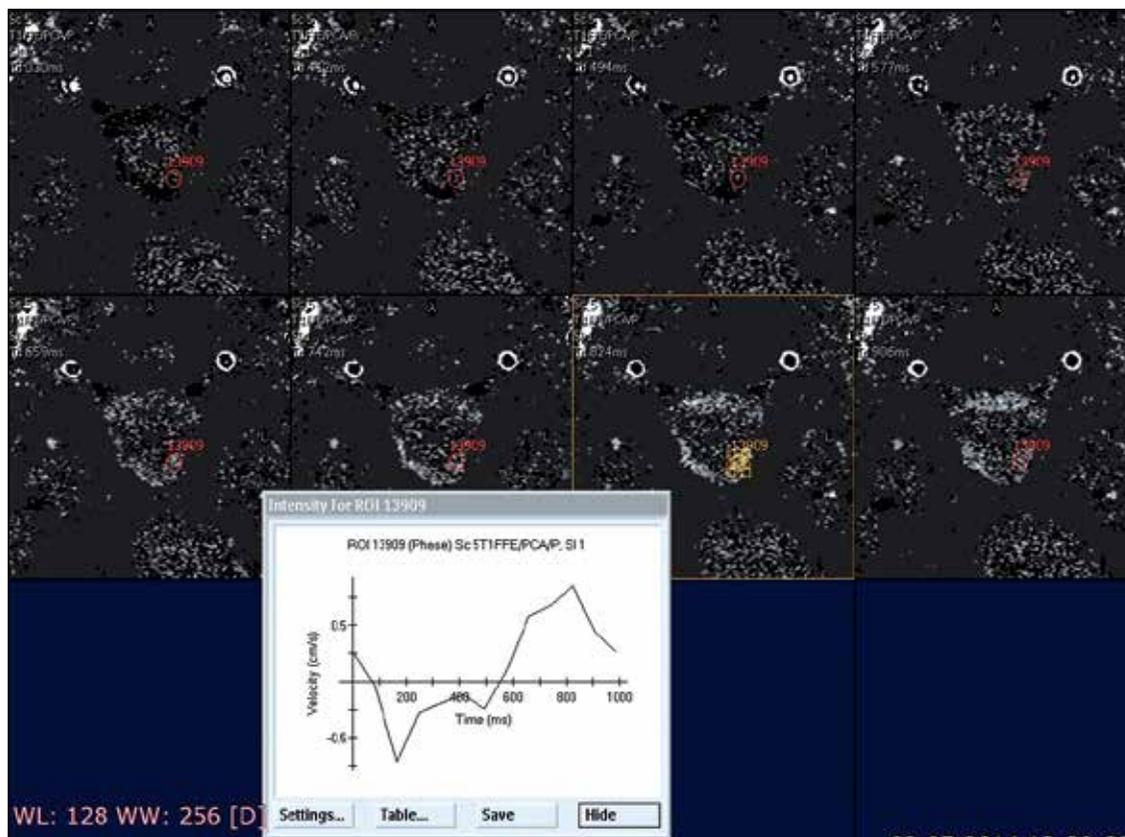
Figure 4: Postoperative graph of the same patient at the same level demonstrates normal sinusoidal flow pattern.

foramen magnum in two patients, multiple arachnoid adhesions in two others, and a fourth ventricular arachnoid veil in one. Each of these findings can alter CSF flow, although the pathophysiology resulting in syrinx formation in this group of patients is poorly understood (25).

MRI is an excellent non-invasive method of appreciating the anatomy of this malformation and the associated anomalies, not only as part of the preoperative work-up, but also in the follow-up of surgically treated patients. The incidence of Chiari 0 malformation has increased dramatically over the last decade. This finding is attributed mainly to the widespread use of MRI in clinical practice. Chiari type 0 malformation is also diagnosed incidentally as a result of MRIs performed for other reasons as in our 2 cases.

On MRI, posterior fossa volume tends to be smaller in Chiari malformation when compared with normal individuals, even though this is not always the case (26). Morphometric studies in Chiari malformation patients collaborated the overcrowding of a normally developed hindbrain within a hypoplastic posterior cranial fossa (26). Nishikawa et al. suggested calculating the "volume ratio," defined as brain volume divided by the cranial volume of posterior fossa (evaluated by means of MRI and computed tomography respectively) (19). Nishikawa et al. also reported that posterior fossa volume is smaller in the Chiari malformation (19).

The clinical manifestations of all types of Chiari malformations are classically attributed to the crowding of the posterior fossa structures at the foramen magnum leading to obliteration of the CSF circulation at the craniovertebral junction (27). Phase-contrast cine flow MRI provides useful additional information on this particular aspect. During the cardiac cycle, there is bi-directional craniospinal displacement of CSF. Greitz et al. demonstrated this pulsatile motion using phase contrast MRI (12). During systole, there is an increase in cerebral blood volume and CSF is displaced caudally through the foramen magnum as a compensatory mechanism. During diastole, the elastic recoil of the spinal dura mater propels the CSF in a caudocranial direction towards the intracranial compartment (4,9,10,16). Phase-contrast cine flow MRI studies have demonstrated that Chiari type I malformation can have pathological CSF flow at the level of foramen magnum (1,5,16,21,22). Following posterior fossa decompression, abnormal CSF flow reverts to normal and parallels symptomatic improvement (1,5,16,21,22). In the present study, we first investigated the significance of the abnormal CSF circulation at the craniocervical junction in Chiari type 0 patients for understanding the pathophysiology of syrinx development. Our six cases have an obstruction in cine flow MRI at the craniovertebral junction and this finding improved after suboccipital decompression and duraplasty. There is no literature knowledge about altered CSF dynamics at the craniovertebral junction correlated with clinical symptomatology in patients with Chiari 0 malformation.

The clinical significance of these radiological findings remains questionable for patients who were diagnosed incidentally. In these cases, cine flow MRI studies may be performed to evaluate CSF flow at the craniovertebral junction.

Syringomyelia was demonstrated in 50-70% of all types of patients with Chiari malformation (6,17,20,28). Their more frequent location is cervical and thoracic levels, even though C1 is almost never affected; in about 20% of the cases the syrinx extends to the whole spinal cord (6). In our cases, syringomyelia was located at the lower cervical levels.

The management algorithm of Chiari type 0 syndrome is still controversial. There are only few reports in which the authors published their treatment modalities and postoperative results for these patients. We reported 7 cases with Chiari type 0 syndrome and reviewed the literature to determine the differences between the treatment options of Chiari type 0 with syringomyelia.

Patients with Chiari I malformation and associated syringomyelia usually show improvement after suboccipital decompression and duraplasty (18,27). However, these patients do not improve after suboccipital decompression alone (3,15,18). The degree of hindbrain CSF flow obstruction correlated with presenting symptomatology and predicted the response to the surgery. McGirt et al. also reported that normal preoperative hindbrain CSF flow was an independent risk factor for treatment failure after decompression for Chiari type I malformation regardless of the degree of tonsillar ectopia (16). Based on these observations, it seems that augmentation duraplasty contributes significantly to the restoration of CSF flow at the craniovertebral junction (3,13). Therefore, our symptomatic Chiari type 0 patients were treated surgically by posterior fossa decompression and duraplasty. As such, a thorough neuroaxis evaluation is necessary to exclude other causes of syrinx before consideration of posterior fossa decompression (25).

The management of asymptomatic patients that was incidentally found during routine MRI studies may be problematic. In this case, cine flow MRI studies may be performed to evaluate cine flow at the craniovertebral junction. If cine flow MRI studies are normal, asymptomatic patients would be followed without surgical intervention. This condition may be progressive and it is recommended that patients with Chiari 0 malformations found incidentally should undergo careful clinical and radiological follow-up as in our cases. Despite these encouraging outcomes, the number of cases in this study is not large enough to draw a solid conclusion.

Resolution of clinical symptoms after hindbrain decompression in patients with Chiari malformation associated syrinx ranges from 35% to 100% in different series (2,8,11). In fact, apart from clinical improvement, the success of posterior fossa decompression should be integrated by neuroimaging studies. The reduction in size of a syrinx in MRI can be utilized as an objective neuroradiological sign of improvement. McGirt et al. reported that marked reduction and resolution of syringomyelia in 65% of patients who were followed by MRI (16). Stabilization in syrinx size was observed in the remaining 35% of patients. Many patients with symptomatic Chiari type I malformation presented with recurrent symptoms after surgical decompression alone. In these cases, McGirt et al. reported that phase-contrast cine flow MRI may be a valuable tool (16). Phase-contrast cine flow MRI can identify patients

who are less likely to respond to surgical decompression. Our symptomatic Chiari type 0 patients with abnormal cine flow MRI studies showed both clinical and radiological improvements after surgical intervention. Complete preoperative CSF flow obstruction predicted long-term resolution of symptoms.

■ **CONCLUSION**

Cine flow MRI appears to be helpful in the diagnosis and management of patients with Chiari 0 malformations. CSF flow turns to normal ranges after surgical treatment, which correlated radiologically with syrinx resolution or reduction, and symptomatic improvement.

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