



Intraoperative Ultrasonography for Definition of Less Invasive Surgical Technique in Patients with Chiari Type I Malformation

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INTRODUCTION: Chiari malformation type I (CM) is the main congenital malformation disease of the craniovertebral junction. The ideal surgical treatment is still controversial. Invasive procedures inside the cerebrospinal fluid (CSF) space and associated with dural repair are considered the gold standard; however, less invasive surgery with isolated bone decompression without dural opening may be possible in selected patients. Our study evaluates the efficacy of intraoperative CSF flow measurement with ultrasonography (USG) as a determining parameter in the selection of these patients.

METHODS: We analyzed prospectively 49 patients with CM operated on at the Hospital das Clínicas, College of Medicine, University of São Paulo. Patients underwent decompressive surgery with or without opening of the dura mater after intraoperative USG measuring flow rate. A value of 3 cm/second was considered a cutoff. Quality of life before and after surgery and the improvement of neck pain and headache were evaluated.

RESULTS: Among 49 patients enrolled, 36 patients (73%) had CSF flow >3 cm/second and did not undergo duraplasty. In 13 patients (27%) with initial flow <3 cm/second, dural opening was performed together with duraplasty. All patients improved when preoperative and postoperative scores were compared, and all clinical parameters evaluated did not differ between both surgical groups. Patients submitted to bone decompression alone had a lower complication rate.

CONCLUSIONS: Intraoperative USG with measurement of CSF allows the proper selection of patients with CM for less invasive surgery with bone decompression without duraplasty.

INTRODUCTION

Chiari malformation type I (CM) is a congenital disease characterized by herniation of the cerebellar tonsils 5 mm below the foramen magnum. The usual prevalence is approximately 8 per 1000 live births. CM may also occur secondary to increased intracranial pressure in the presence of hydrocephalus, space-occupying lesions in the posterior fossa, and downward movement of brain structures caused by decreased intracranial pressure resulting from a cerebrospinal fluid (CSF) fistula.¹⁻¹⁰

Many patients are asymptomatic, but some may experience progressive neurologic symptoms of muscle weakness, impairments in balance, coordination, and sensitivity, and difficulty swallowing, as well as headache and disabling neck pain, especially on exertion. These symptoms are caused by direct compression of the cerebellum, spinal cord, and brainstem structures, which often leads to the formation of a syringomyelic cavity with involvement of the spinal tracts and cell bodies of neurons in the grey columns of the spinal cord. The pathophysiology is related to CSF flow changes at the craniovertebral junction (CVJ).¹¹ The presence of these symptoms impairs quality of life and requires surgical treatment.¹²⁻³⁴

Key words

- Chiari malformation
- Posterior fossa decompression
- Ultrasonography

Abbreviations and Acronyms

- BOD:** Bone-only decompression
- CM:** Chiari malformation type I
- CN:** Cranial nerve
- CSF:** Cerebrospinal fluid
- CVJ:** Craniovertebral junction
- DD:** Dural opening with duraplasty
- HCFMUSP:** Hospital das Clínicas, University of São Paulo School of Medicine
- PFD:** Posterior fossa decompression
- SF-36:** Short-Form 36

Vf: Cerebrospinal fluid flow rate

USG: Ultrasonography

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The recommended treatment for symptomatic patients consists of posterior fossa decompression (PFD) and reestablishment of adequate CSF flow. However, the optimal technique is still controversial. The extent of bone decompression, dural opening, resection of arachnoid adhesions, coagulation, and even resection of the cerebellar tonsils remain subjects of controversy among specialists. Recently, less invasive methods with bone decompression alone have been reported,³⁴⁻⁴⁶ with good results in selected patients. In most cases, the choice of technique depends on the surgeon's preference and evaluation, because there is no definition in the literature about which patients would benefit from less invasive surgery.³⁴⁻⁴⁶

The advent of intraoperative ultrasonography (USG) has allowed identification of CVJ anatomy and CSF dynamics and CVJ structures with real-time images.^{9,10,30,34,35,46} Within this context, our study aims to evaluate the usefulness of intraoperative USG in patients with CM as a method for selection of candidates for PFD with bone removal alone.

METHODS

This study was approved by the research ethics committee of Hospital das Clínicas, University of São Paulo School of Medicine (HCFMUSP), on September 16, 2009 (opinion number 0903/09).

Using a prospective design, we collected demographic, clinical, and sonographic data from 49 consecutive adult patients who had received a diagnosis of CM at HCFMUSP and underwent decompressive surgery of the posterior fossa at the HCFMUSP Department of Neurosurgery between 2008 and 2012. All procedures were performed by the same surgeon, following a pre-established technique.

Inclusion Criteria

The diagnosis of CM is based on clinical and radiologic criteria (herniation of the cerebellar tonsils 5 mm below the foramen magnum).

Pharmacologic methods (including opioids, tricyclic antidepressants, neuroleptics, and serotonin reuptake inhibitors) were used to treat headache and neck pain. Treatment failure was defined as worsening or absence of improvement in symptoms after at least 6 months of effective pharmacotherapy. Deterioration of neurologic signs and symptoms refractory to medical treatment was considered a definite indication for PFD.

Exclusion Criteria

Patients with other neurologic diseases and those with basilar invagination type A¹⁴ and indication for anterior decompression surgery were excluded from the study protocol. Similarly, patients with rapidly progressive neurologic deterioration were excluded from analysis, being submitted to PFD with dural opening.

Clinical Evaluation

Patients were evaluated for the presence or absence of headache; neurologic symptoms, following the standard neurologic examination protocol of the HCFMUSP Department of Neurology; and quality of life, as assessed by the validated Portuguese-language version of the Short-Form 36 (SF-36).⁷ Each patient's subjective impression regarding their preoperative and postoperative clinical status was also assessed.

Surgical Technique

The procedure is performed under general anesthesia and tracheal intubation, with the patient in the prone position, with the head slightly bent and held in a skull clamp. A midline skin incision is made extending from the external occipital protuberance to the height of the fourth or fifth spinous process of the cervical vertebra. If needed, in patients lacking proper CSF flow, the incision is extended 3 cm to harvest pericranium grafts for duraplasty. This procedure is followed by a suboccipital craniectomy 3–4 cm in diameter with removal of the posterior arc of C1 (Figure 1).

Intraoperative USG is then performed to determine the retrocerebellar space, cisterna magna, and CSF flow through the foramen magnum in the retrocerebellar space. At our facility, a two-dimensional USG system (MicroMaxx Sonosite, Bothell, Washington, USA) with high-frequency and low-frequency transducers (13–6 MHz and 04.08 MHz, respectively) is used.

USG allows identification of the anatomic structures of the CVJ (Figure 2). With the system in B-scan mode, the craniocaudal (Figure 3A), anteroposterior (Figure 3B), and lateral (Figure 3C) dimensions of the cisterna magna are measured. Three different images are obtained in each plane, and the mean of the 3 resulting measurements is taken into account for analysis. CSF flow measurement is performed with the system in Doppler mode, with the transducer placed longitudinally in a window parallel to CSF flow at the level of the foramen magnum. The average speed is determined by the formula $(V_{\max} + 2 \cdot V_{\min})/3$ (Figure 4). Three measurements are obtained, and the weighted average of the 3 is defined as the CSF flow rate (Vf) through the foramen magnum.

All sonographic examinations were performed by physicians from the HCFMUSP Department of Neurosonology with particular expertise in neuroimaging and intraoperative CSF flow measurement.

A Vf value of 3 cm/second was adopted as the threshold for opening the dura and performing duraplasty. This was the lowest Vf value found in the literature for patients who achieved a good outcome after PFD.³³ After bone decompression and dural delamination, CSF flow was measured, and if Vf was equal to or greater than 3 cm/second, the procedure was completed without dural opening. If the Vf was less than 3 cm/second, then a Y-shaped dural opening was fashioned and, under microscopy, arachnoid membrane adhesions were dissected and the foramen of Magendie opened to communicate the subdural space to the fourth ventricle (Figure 5). No tonsillar coagulation was performed in the present sample.

Postoperative Evaluation

Postoperative complications were recorded for each patient. After discharge, all subjects were followed as outpatients and evaluated 1 year after surgery for presence and intensity of headache and neck pain, subjective perception of improvement (defined as better, same, or worse), and quality of life according to the SF-36 questionnaire.

Statistical Analysis

Groups were compared for the mean values obtained for each item before and after surgery, and improvement was quantified by the

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