Long-Term Outcomes of a New Minimally Invasive Approach in Chiari Type 1 and 1.5 Malformations: Technical Note and Preliminary Results

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■ BACKGROUND: **The treatment options for patients with Chiari malformation type 1 (CM1) and Chiari malformation type 1.5 (CM1.5) have not yet been standardized. In these malformations, the main factors include obstruction at the level of the foramen magnum and dural and ligamentous thickening. Here we present our outcomes of surgery and decompression using a minimally invasive surgery (MIS) technique.**

■ METHODS: **Sixty-one patients admitted to our clinics between 2009 and 2016 due to CM1 or CM1.5 and who had undergone MIS were investigated retrospectively. All patients were followed up for a mean period of 55 months, both clinically and radiologically, and the outcomes were recorded.**

■ RESULTS: **All 61 patients underwent foramen magnum decompression through a 1.5-cm mini-open incision, C1 laminectomy and C2 medial inner side tour, posterior atlanto-occipital membrane removal, external dural delamination, and widening of the internal dura with longitudinal incisions. Fifty-six patients (91.8%) were satisfied with the outcome, 4 patients (6.5%) remained the same, and 1 patient (1.6%) reported a poor outcome. Forty-five percent of the patients with syringomyelia demonstrated resolution within 2 years, and 92% demonstrated resolution in 5 years. Scoliosis was seen in 5 patients (8.1%). The rate of benefit from the surgical procedure was statistically significant ($P = 0.0045$), and no patient required additional surgery because of poor decompression.**

■ CONCLUSIONS: **MIS is effective for uncomplicated cases of CM1 and CM1.5 due to its minimal connective and muscular tissue damage, short surgical duration, short recovery time, early mobilization, effective posterior foramen magnum widening, lack of liquor fistula development, and better clinical and radiologic improvement during long-term follow-up.**

INTRODUCTION

Olivier d'Angers was the first researcher to define syringomyelia, in 1824.[1] Sixty-six years later, Chiari malformation was defined as the downward displacement of the cerebellar tonsils through the foramen magnum into the upper cervical spinal canal.[1] Discussions of its etiology and treatment continue today. The common features of this syndrome (with 4 defined types) are obstruction at the foramen magnum level and dural and posterior atlanto-occipital membrane thickening. The most commonly seen clinical type is Chiari malformation type 1 (CM1).

The incidence of syringomyelia is lower in patients with CM1 compared with those with Chiari malformation type 1.5 (CM1.5).[2-4] CM1.5 is usually observed at earlier ages, and the most common characteristics are hindbrain herniation, an impaired odontoid clivus relationship, occipitalization of the atlas, a retroverted dens, a superomedial tendency of the squamal part of the occipital bone, platybasia, and other congenital abnormalities.[4] Therefore, a certain number of these patients are candidates for complex surgery and may require arthrodesis, whereas some may be undergo repaired via a minimally invasive surgery (MIS) approach, as in CM1 cases. Significant syringomyelia may manifest in these cases. Some authors consider CM1.5 a more advanced form of untreated CM1.[4]

Key words
- Chiari malformation
- Chiari 1.5
- Minimally invasive surgery
- Syrinx

Abbreviations and Acronyms
CFS: Cerebrospinal fluid
CM1: Chiari malformation type-1
CM1.5: Chiari malformation type 1.5
MIS: Minimally invasive surgery
mMG: modified McCormick grade
MRI: Magnetic resonance imaging

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Supplementary digital content available online.

Citation: World Neurosurg. (2018) 115:407-413.
https://doi.org/10.1016/j.wneu.2018.04.100

*Journal homepage: **www.WORLDNEUROSURGERY.org***

*Available online: **www.sciencedirect.com***

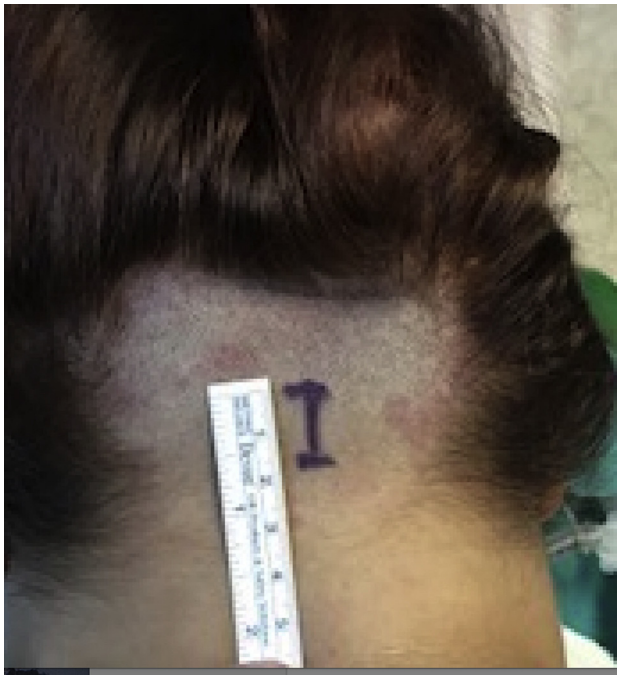

Figure 1. Minimal skin incision.

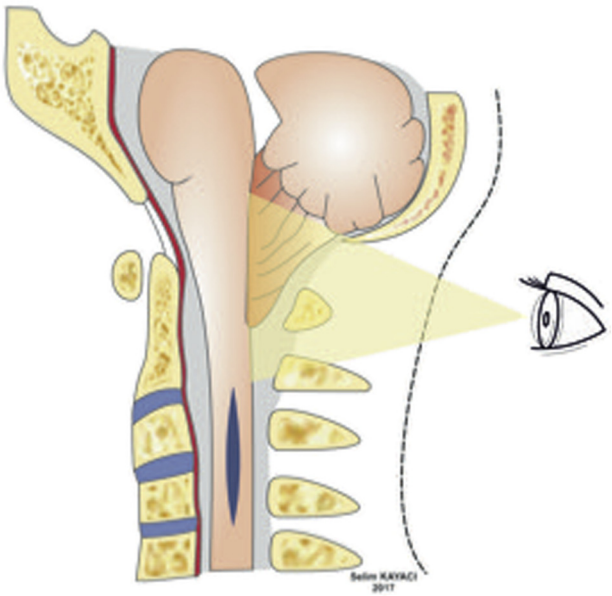

Figure 2. Angle of vision field from skin incision.

In the absence of severe syringomyelia in patients with CM1 and CM1.5, the first treatment option is suboccipital craniectomy, posterior atlanto-occipital membrane resection, and removal of the posterior ring of the atlas.[2,5] This surgical procedure is routinely performed as an open microscopic surgery; however, some complications may occur. Therefore, research on MIS approaches, such as foramen magnum and C1 decompression and dural widening, has been conducted. Here we present the long-term clinical outcomes of patients with CM1 and CM1.5 treated using MIS.

METHODS

A total of 61 patients admitted to our clinics between February 2009 and December 2016 with a diagnosis of CM1 or CM1.5 who underwent MIS were investigated retrospectively. All patients had suboccipital headache and lower central neck pain that was particularly provoked by coughing. The headache and neck pain could not be reduced with such treatment modalities as antineuropathic drugs or local injections. The neurologic and radiologic findings were recorded on the first postoperative day and at 6 months, 2 years, and 5 years thereafter. The general neurologic status of each patient was evaluated according to the modified McCormick grade (mMG).[6] The surgical benefits were assessed according to the Odom scale.[7] In addition, the volume and regression of the syrinx cavities, dimensions of the tonsillar herniations, and preoperative and postoperative dimensions of the cisterna magna were recorded.

All patients were followed up clinically and radiologically for a mean of 5.5 years (range, 44–88 months). Patients with complex Chiari malformations, such as an unstable craniovertebral junction and retroflex dense basilar invagination, were excluded from this study.

Radiologic Findings

All patients were evaluated preoperatively and postoperatively with magnetic resonance imaging (MRI) and preoperatively with computed tomography. Measurements were made by the same radiologist with Osirix software. The inclusion criterion for CM1 was the herniation of the cerebellar tonsils >5 mm below the foramen magnum. Syringomyelia is usually present, but not associated with brainstem or fourth ventricular herniation or hydrocephalus. CM1.5 is specifically describes patients with CM1 malformations but with the addition of an elongated brainstem and fourth ventricle. The degree of tonsillar ectopia was classified as follows: grade 1, tonsils extend to the posterior neural arcus of the atlas[8]; grade 2, tonsils exceed the neural arcus of the atlas; grade 3, tonsils completely exceed the neural arcus of the atlas.

Each syrinx was classified by its shape as A, distended; B, moniliform; C, circumscribed; or D, slender.[9]

Syringomyelia was graded as follows: grade 0, tonsillar herniation present, but syrinx cavity not developed; grade 1, syrinx in the form of central channel widening that was <3 mm and shorter than the third vertebral segment; grade 2, syrinx >3 mm and longer than the third to fifth vertebral levels; grade 3, syrinx >5 mm and longer than the fifth vertebral segment; grade 4, the holocord causing the massive spinal cord