Surgical Decompression for Chiari Malformation Type I: An Age-Based Outcomes Study Based on the Chicago Chiari Outcome Scale

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■ BACKGROUND: **There is currently inadequate evidence on the efficacy of surgical decompression for Chiari malformation type I (CM1) in different age groups of patients. In this study, we compared postoperative outcomes across 3 different age groups using the Chicago Chiari Outcome Scale (CCOS).**

■ METHODS: **A total of 144 patients who underwent Chiari decompression at our institution between 2008 and 2014 were divided into 3 groups: group A, children age 0–18 years; group B, younger adults age 19–40 years; and group C, older adults, age 41+ years. Patient outcomes were assigned a numerical value based on the CCOS and subjected to statistical analysis. Direct comparisons were made across the 3 age groups.**

■ RESULTS: **The mean overall score was 14.0 over a mean follow-up of 27.2 months. All 3 groups demonstrated clinical improvement following Chiari decompression; however, group A demonstrated significantly better postoperative improvements than groups B and C in total CCOS scores (7.8% and 12.2%, respectively; $P < 0.001$) and all the component scores except complications. Group B was not significantly different from group C in total score or any of the component scores. There was a logarithmic relationship between age and outcome ($R^2 = 0.64$), in which the outcome scores experienced an initial decline with increasing age but leveled off by early adulthood.**

■ CONCLUSIONS: **A direct comparison among the age groups revealed a negative age effect on surgical decompression outcomes in CM1 patients. Children performed significantly better than younger and older adults. This finding supports early surgical intervention for symptomatic pediatric patients to achieve long-term surgical benefit.**

INTRODUCTION

Chiari malformation type I (CM1) is characterized by congenital hypoplasia of the posterior fossa and caudal displacement of the cerebellar tonsils below the foramen magnum.[1,2] The etiology of CM1 remains unknown, although numerous clinical presentations have been described. Occipital headaches are the most common symptom, reported in 73%–98% of patients.[3-5] These headaches tend to be exacerbated by Valsalva maneuvers, such as coughing, laughing, or bending. Other common symptoms include diplopia, dizziness, ringing in the ears, tinnitus, ear pain, pain in the neck and shoulders, upper extremity weakness, paresthesias, clumsiness, dysphagia, drooling, sleep apnea, poor fine motor movements, and episodic nausea and vomiting. Common physical signs include nystagmus, decreased palate elevation, decreased or absent gag reflex, dysmetria, hyperreflexia, ataxia, and weakness, particularly in the hands. Syringomyelia is found in 24%–75% of CM1 cases and may lead to the classic cape-like loss of upper extremity nociception as well as hand intrinsic muscle atrophy.[3,4,6,7] Scoliosis is present in approximately 25% of patients with CM1.[3,4,8] Although this is a congenital abnormality, patients may become symptomatic at any age, and symptoms may wax and wane, or change, over time. Close questioning of older patients who "suddenly" become symptomatic usually reveals that Chiari symptoms have actually been present since birth or childhood, and have simply worsened.

Key words
- Chiari decompression
- Chiari malformation type I
- Chicago Chiari Outcome Scale
- Outcome study

Abbreviations and Acronyms
CCOS: Chicago Chiari Outcome Scale
CMI: Chiari malformation type I
MRI: magnetic resonance imaging

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Magnetic resonance imaging (MRI) is the gold standard for diagnosing CM1. CM1 may be characterized by tonsillar herniation as small as 3 mm or as large as 25 mm or larger. No correlation has been demonstrated between the extent of tonsillar herniation and the severity of clinical symptoms, and in fact patients with long herniations are frequently asymptomatic. Conversely, some of the most symptomatic patients have relatively small tonsillar herniations.[2,4] Nonetheless, MRI in conjunction with history and physical examination remains the most effective tool available for identifying candidates for surgical decompression. Patients who are consistently symptomatic with tonsillar descent below the foramen magnum detected on MRI are traditionally offered surgical decompression.

Surgical decompression is the most commonly pursued approach for CM1.[9] This is achieved with or without duroplasty.[10,11] Although the current literature provides ample evidence of the efficacy of Chiari decompression in the pediatric and adult populations separately, few direct comparisons between age groups have been reported.[7,12] This is due in part to the fact that surgical outcomes have been difficult to assess in the absence of a standardized outcome measurement system. Various attempts have been made using a variety of reporting methods, including gestalt impression of overall postoperative performance ("good", "fair", etc.), description of signs and symptoms, and standardized scales.[13] Of these, only the Chicago Chiari Outcome Scale (CCOS) has been validated in both adults and children.[14,15] Based on this quantitative measurement system, in the present study we were able to compare Chiari decompression outcomes across different age groups.

METHODS

Patient Selection

This retrospective, cross-sectional study was carried out under the auspices of the Human Investigation Committee at Oakland University William Beaumont School of Medicine. Informed consent was obtained from each patient before surgery. A total of 144 patients treated with surgical decompression for CM1 between 2008 and 2014 were included in the study. This cohort comprised 113 females (78.5%) and 31 males (21.5%), with a mean age at surgery of 25.1 ± 14.3 years (range, 1–61 years).

The patients were divided into 3 age groups: group A, children age 0–18 years; group B, younger adults age 19–40 years; and group C, older adults age 41+ years. There were 63 subjects in group A (42 females, 21 males), 57 in group B (51 females, 6 males), and 24 in group C (20 females, 4 males). The vast majority (93.1%) of the patients presented with the typical occipital headache exacerbated by Valsalva maneuvers. All patients presented with 1 or more of the following, in decreasing order: occipital headaches accentuated by Valsalva maneuvers, neck pain, ataxia, paresthesias, sleep apnea or snoring, dysphagia, weakness, dizziness, visual disturbances, nausea/vomiting, and tinnitus (Table 1). MRI was used to identify patients exhibiting posterior fossa crowding and tonsillar ectopia (Figure 1A). Patients with previous surgical decompression and/or subsequent spinal cord untethering performed elsewhere were excluded from the study.

Table 1. Symptoms at Initial Presentation

Symptom	Number (%)
Valsalva headache	134 (93.1)
Neck pain	69 (47.9)
Ataxia/clumsiness	62 (43.1)
Paresthesias	57 (39.6)
Sleep apnea or snoring	56 (38.9)
Dysphagia	55 (38.2)
Weakness	40 (27.8)
Dizziness	39 (27.1)
Visual disturbances	36 (25.0)
Nausea/vomiting	26 (18.1)
Tinnitus	22 (15.3)

Surgery

All surgeries were performed by the senior author at William Beaumont Hospital (Royal Oak, Michigan). Suboccipital craniectomy, bovine pericardium duraplasty, reduction of the cerebellar tonsils, and C1 laminectomy were performed routinely using microsurgical technique. The cerebellar tonsil tips were resected with a subpial technique, reduced by coagulation, or a combination of the 2 techniques based on the extent required to accomplish patency and complete decompression of the fourth ventricle. The fourth ventricle was carefully inspected for arachnoid adhesions or further obstructing tissue. Free flow of fluid from the fourth ventricle laterally and into the subarachnoid space was ensured. The dural suture line was coated with DuraSeal (Medtronic, Minneapolis, Minnesota) (Figure 1B).

Outcome Measurement and Analysis

A retrospective chart review was conducted for all patients. Written records from the surgeon were converted into numerical values from 1 to 4 in four separate components: pain symptoms, non-pain symptoms, functionality, and complications. Assignment of numerical scores was based on the same criteria set forth in the original CCOS description. These criteria are briefly outlined below.

A score of 4 indicated, in its respective category, complete resolution of pain or non-pain symptoms, return to full activity level before disease, or complete absence of complications. A score of 3 indicated some improvement of preoperative pain or non-pain symptoms, postoperative pain or non-pain symptoms controlled with nonsurgical interventions as needed; return to >50% of activity level before disease; or occurrence of transient complications. A score of 2 indicated no change in pain, no change in non-pain symptoms, moderate impairment of physical function (<50% activity level before disease), or prolonged complication that was improved with nonsurgical treatment or therapy. A score of 1 indicated worsening of pain, worsening of non-pain symptoms, complete absence of functional improvement, or permanent complications unresponsive to subsequent non-surgical treatment or therapy.