Acta Neurol Scand 2009: 120: 295–299 DOI: 10.1111/j.1600-0404.2009.01183.x

ACTA NEUROLOGICA
SCANDINAVICA

Long-term outcome after surgery for Chiari I malformation

Tisell M, Wallskog J, Linde M. Long-term outcome after surgery for Chiari I malformation.
Acta Neurol Scand 2009: 120: 295–299.

Objective – To examine the long-term effects of suboccipito-cervical decompression on an intention-to-treat basis in patients with Chiari I malformation (CMI). *Patients and methods* – Twenty-four consecutive patients, 14 females and 10 males with a median age of 26 years, underwent decompressive surgery for CMI during 1998–2006. All patients were contacted by an independent examiner and asked to complete a questionnaire regarding headache, other neurological symptoms and negative impact of the disease on the daily life before and after surgery. The median follow-up time after surgery was 3.2 years (range 1.7–9.2 years). Twenty-three patients (96%) completed the questionnaire. *Results* – On an intention-to-treat basis there was an improvement in headache in 75%, decreased associated neurological symptoms in 88% and less negative impact on daily life in 75% of the 24-operated patients. *Conclusions* – More than three-quarters of the patients still considered their situation improved at long-term follow-up after surgery. These results support surgical intervention in symptomatic Chiari I patients.

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Key words: Chiari I malformation; complications; headache; long-term outcome; treatment

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Accepted for publication February 18, 2009

Introduction

Chiari I malformation (CMI), herniation of the cerebellar tonsils through the foramen magnum (1) was originally described after autopsy by the Austrian pathologist Hans Chiari in 1891 in a patient who had succumbed to hydrocephalus. Currently, the diagnosis is most often established by MRI (1), showing obliteration of retrocerebellar CSF spaces, tonsillar herniation of at least 5 mm, and different degrees of dysplasia of the cranial base (1, 2).

The aetiology is unknown (3). It has previously been considered that the posterior fossa is underdeveloped with reduced supraoccipital distance, increased deviation of the tentorium, hypoplasia of clivus and malformation of bone structures in the craniovertebral region, but no primary malformation of the nervous tissue (2–4). However, Sgouros et al. recently reported that children with CMI without syringomyelia have normal posterior fossa volumes (5). Up to 65% of the patients have syringomyelia (1, 6, 7), explained by an obstructed subarachnoidal CSF flow at the level of the foramen magnum, leading to an increased flow of CSF through perivascular and interstitial tissue of the central canal of the spinal cord. The CSF obstruction could eventually cause hydrocephalus (1). Syringomyelia might be associated with scoliosis (8), and a relation has been observed between the scoliosis and the extent of neurological symptoms (6).

Epidemiological data are scarce. Most patients become symptomatic during the second or third decade of life. Milhorat et al. reported a mean age of symptom debut of 24.9 ± 15.8 years (1), referring to the term 'adult type' of Chiari malformation. There is female predominance in most studies (1, 4). A subgroup of CMI is hereditary (1, 9, 10). Acquired CMI after lumbo-peritoneal shunting has been reported (11).

The dominant symptom in CMI is headache (1, 12, 13), in the typical case localized occipitally and radiating to vertex and retroorbitally, or to the neck

and shoulders. The headache is frequently triggered by physical activity, valsalva manoeuvre, coughing, laughter or changes of body position. The duration varies from minutes to chronic daily headache (1, 14, 15). However, there are also asymptomatic patients with CMI (1). Common differential diagnoses are benign cough headache and migraine (16, 17). Furthermore, some patients have a combination of headaches, including primary headache syndromes and to select patients to treatment and predict the outcome of surgery, a detailed analysis of the headache is mandatory (12).

Other symptoms found in CMI include ocular disturbances (retroorbital pain, obscurations, light flashes, blurred vision, photophobia and loss of visual field), and otoneurological disturbances (vertigo, balance disturbances, pressure in the ears, tinnitus, reduced hearing, hyperacusia and oscillopsia). Other neurological deficits include dysphagia, dysartria, reduced coordination, paresis, spasticity, hyperreflexia, sensory dysfunction and hyperhidrosis (1, 14, 18).

The first-line surgical treatment for CMI is suboccipito-cervical decompression with or without duraplasty, to increase the retro-cerebellar space and facilitate CSF flow between the cranial and spinal CSF spaces (19). Improvement after surgery has been observed in larger series in more than 80% of the patients (13, 20–22) with a low rate of complications (8, 13, 22). However, there are no controlled studies. Data on long-term outcome is scarce. Recently, Kumar et al. reported that 79 of 87-operated patients experienced an improvement or stabilization of their disease process after a mean follow-up of 34.21 $\pm$ 21.53 months (22).

The aim of this study of 24-consecutive patients was to examine the long-term effects of suboccipito-cervical decompression on an intention-to-treat basis in patients with CMI.

Patients and methods

Patients

All patients in western Sweden diagnosed with CMI are referred to Sahlgrenska University Hospital for evaluation and treatment. During 1998–2006, 24 patients (14 females and 10 males) were operated on with suboccipito-cervical decompression and duraplasty with a duragraft (16 Gore-tex®, W. L. Gore and Associates, Inc., Flagstaff, AZ, USA, seven Durepair®, Medtronic Neurosurgery, Goleta, CA, USA and one muscle-fascia-graft). In 23 patients, total or partial laminectomy of atlas was also performed. In five of these patients, the laminectomy was extended to C2, and in one patient to C3. The median age was 26 years (range 4–54 years) and the median duration of symptoms prior to surgery was 38 months (range 0–300 months). Twenty-two of the patients fulfiled the diagnostic criteria for headache-related CMI according to the International Classification of Headache Disorders (ICHD, 2nd edition) (12), while two patients had no headache but symptomatic syringomyelia.

All patients underwent MRI preoperatively. The median herniation of the tonsils (measured from the caudal limit on foramen magnum on sagittal images) was 15.5 mm (range 4–35 mm). Twelve patients (50%) had syringomyelia. Three patients had scoliosis. None was aware of having a relative with CMI. None of the patients had secondary causes of CMI.

Six patients (25%) were re-operated; two extended decompressions due to relapsing symptoms after 10 months and 7 years respectively, one ventriculo-peritoneal shunt due to concomitant intracranial hypertension, one syringo-subarachnoidal shunt due to progressive myelopathia from a cervical syrinx, one superficial revision of a CSF fistula and one revision of an infected duragraft. In addition, there was one case of suspected post-operative meningitis, treated successfully with antibiotics.

Short-term follow-up

Minimum 3 months after the latest (in case of re-operation) surgery the patients underwent short-term follow-up with post-operative MRI and clinical examination. Twenty-two patients (91%) reported that they had improved (13 were asymptomatic), one was unchanged and one was lost to clinical follow-up since he had moved from the region.

Long-term follow-up

In 2007–2008, all 24 patients were contacted by an independent examiner, an experienced research nurse with no involvement in the surgical treatment nor in the pre- or postoperative care of the patients. The patients were asked to complete a questionnaire assessing their previous headache and other neurological symptoms and present condition in comparison with their condition before surgery. In addition, the patients were asked to rate the negative impact from the disease on daily life before and after surgery on a visual analogue scale (0–100). There were separate scales for work/studies, family life and leisure activities.

Twenty-three (96%) of the patients completed the questionnaire. The median follow-up time was 3.2 years (range 1.7–9.2 years).

The study was approved by the local ethical committee.

Statistical analysis

Descriptive statistics were performed. Pre and post-operative data were compared using the McNemar test for categorical variables and the Wilcoxon Signed Ranks test for numerical variables. $P < 0.05$ was considered significant. Analyses were performed using the Statistical Package for Social Sciences (SPSS™) software program (version 14.0 for Windows; SPSS, Inc., Chicago, IL, USA).

Results

Twelve patients (50%) had no remaining symptoms. The headache had decreased post-operatively in eighteen patients (75%) and only three claimed significant remaining headache (Table 2).

Table 1 Clinical characteristics of the preoperative headache in 21 patients with Chiari I malformation

Description of preoperative headache (n = 21)	Number of patients
Characteristics of headache (one or more)	
Localized	19
Enhanced by physical activity	15
Pulsating	12
Nausea	6
Neurological symptoms	5
Lateralized	5
Vomitting	4
Photophobia	1
Sound sensitive	2
Localization of headache (one or more)	
Occiput	16
Neck	13
Temporal	7
Parietal	6
Forehead	4
Factors triggering headache (one or more)	
Movement of neck	15
Cough/sneeze	15
Body position	14
Valsalva	11
Laughter	6
Trauma	7
Psychological stress	2
Duration of headache (one or more)	
<1 h	9
1–3 h	9
4–12 h	7
12–24 h	4
>24 h	4
Recovery after headache attacks (one or more)	
Complete recovery	8
Some recovery	9
No recovery	4

Table 2 Symptoms and signs in 23 patients with Chiari I malformation before surgery and 3.2 years (median) post-operatively

	Before surgery (number of patients)	Long-term follow-up (number of patients)	P (McNemar)
Headache	21	3	<0.001
Ocular symptoms	13	2	<0.01
Retroorbital pressure	7	1	<0.05
Diplopia	4	1	n.s
Visual field defects	3	1	n.s
Blurred vision	2	0	n.s
Flashes	2	0	n.s
Otoneurological symptoms	17	6	<0.01
Vertigo	13	3	<0.01
Balance disturbance	10	2	<0.01
Ocillopsia	7	2	n.s
Pressure in ears	3	0	n.s
Tinnitus	2	1	n.s
Reduced hearing	2	0	n.s
Other neurological deficits	17	5	<0.001
Paresis	11	1	<0.01
Coordination disturbance	10	1	<0.01
Sensoric dysfunction	9	2	<0.05
Dysphagia	7	0	<0.05
Dysartri	2	0	n.s
Hyperhidrosis	2	1	n.s

The improvement in headache had been experienced after a median of 3.5 weeks (range: 0–52 weeks). Before surgery, the patients had headache on median 30 days a month. At long-term follow-up after surgery, the median amount of headache was 1 day a month ($P < 0.001$, Wilcoxon signed ranks test). The characteristics of the preoperative headache are listed in Table 1.

Twenty-one (88%) patients reported having fewer other neurological symptoms. There was a decrease of all registered symptoms as listed in Table 2.

The patients visual-analogue estimation of negative impact on daily life of their disease in three indices is shown in Table 3. Overall 18 patients (75%) reported improvement in at least two indices, three were unchanged and two experienced deterioration.

Table 3 The negative impact of the disease on daily life in 23 patients with Chiari I malformation before and after surgery estimated using visual analogue scale (0–100)

	Before surgery (median VAS)	After surgery (median VAS)	Individual changes
Impact on work/studies	62	20	19 I/3 U/1 W
Impact on family life	68	14	18 I/4 U/1 W
Impact on leisure activities	74	27	18 I/3 U/2 W

I, improved; U, unchanged; W, worse.

One patient did not return the questionnaire. He had moved before the 3-month check-up.

Thus, on an intention-to-treat basis there was an improvement in headache in 75%, decreased associated neurological symptoms in 88%, and less negative impact on daily life in 75% of the 24-operated patients.

Discussion

The symptom debut of CMI has a peak incidence among young adults. The cardinal symptom is headache with a distinctive profile. Despite a wide variety of associated neurological symptoms, the clinical features are frequently misdiagnosed and sometimes considered psychogenic (1). In a large cohort of 270 patients (87% female) previously diagnosed with fibromyalgia, MRI demonstrated that 20% of the patients had CMI with an average herniation of the tonsils on 7.1 ± 1.8 mm (23). In the present study, the median time from symptom debut to treatment was 38 months, implying delayed diagnosis.

The dominant manifestation of CMI is headache, which is experienced intermittently and not demonstrable in a clinical examination. Thus, both the indications before and the evaluations after surgery are largely based on the patients' history. One challenge is to differentiate headache caused by CMI from other intermittent headache, such as migraine. In this case series, for example, five patients (20%) had recurrent episodes of severe throbbing headaches associated with nausea and/or increased sensitivity to sound and light, but with freedom from most symptoms between the attacks, which *per se* resembles migraine according to the ICHD (12). In a future revision of the diagnostic criteria for headache due to CMI, we recommend that headaches with short duration (less than 1 h) should also be included in order to enhance the positive predictive value of the diagnostic criteria. We base this on the finding that nine (41% of those with headache) of our patients described headaches sometimes or always being shorter than 1 h, which, in adults, rules out migraine.

Both headache and the other reported symptoms are likely to be influenced on a short term-basis by a surgical procedure. However, such placebo and nocebo effects are known from previous studies of surgery to decrease over time, and be less influential on long-term results (24–28). In this study, the median follow-up time was 3.2 years and the minimum follow-up time was 1.7 years.

We found at long-term follow-up of the 24 patients that 75% of the patients had a significant decrease in headache, 88% of the patients had fewer other neurological symptoms and 75% of the patients had less negative impact of the disease on daily life. These results are comparable to other reports on long-term follow-up (22) and support surgical treatment of symptomatic patients with CMI. But there are significant remaining symptoms, and it is an important task to better identify which symptoms are likely to improve after surgery.

There was a relatively high rate of complications. The most common problems were leakage and infections attributable to the duraplasty, and a lesser amount of complications can probably be achieved by improving the techniques of the dura closure. Only one patient received a fascia graft, and during the study-period the choice of dura substitute was changed from Gore-tex® to Durepair®. In addition, five different surgeons operated the relatively small number of patients, and it is possible that a higher numbers of operations per surgeon would lead to fewer complications. Both remaining symptomatic syringomyelia and benign intracranial hypertension requiring further surgery have been reported by others (29–31). The need for long-term follow-up is exemplified by the two patients who improved initially, had relapses of symptoms owing to fibrous scarring, and were re-operated 10 months and 7 years after the first operation, respectively. Longer follow-up times might increase the possibility to identify late complications, such as relapse due to fibrous scarring.

In this follow-up the patients completed a questionnaire after contact from an independent examiner. Questionnaires are inherently subjective and less reliable than clinical evaluations and objective measures. However, as previously mentioned, the symptomathology in CMI is predominantly subjective, and we, therefore, believe self-administered instruments are the least invasive and most time and cost-effective way to obtain data. The proportion of missing responses was small (4%).

The patients were included consecutively from a well-defined geographical region and we consider them representative of individuals with the disease. The patients' age, sex distribution and symptomathology as well as the frequency of syringomyelia are in accordance with previous reports (1). The detailed profile of the headache as described in Table 1, together with clinical and MRI findings, confirms that the patients suffered from CMI headache as defined in the ICHD (12). One major limitation of this study is the lack of data on patients not operated on, and so an epidemiolog-

ical survey is an important subject for future studies.

Conclusion

The long-term intention-to-treat effects of surgery in CMI were positive in 75% regarding headache, 88% regarding other neurological symptoms and 75% regarding negative impact on daily life. These results support surgical intervention in symptomatic Chiari I patients.

Acknowledgement

This study was supported by Göteborg Medical Society, Edit Jacobsson Foundation and John and Brit Wennerström Foundation.
We thank Gudrun Barrows for her invaluable help and patience.

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