Original Research

Various manifestation of Chiari I malformation in children and improvement after surgery

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Abstract

Introduction: Chiari I malformation is defined as caudal displacement of the cerebellar tonsils into the foramen magnum. The most commonly associated finding is cervical syringomyelia. The most common presenting symptom is occipital and upper cervical pain. The incidence of syringomyelia is between 30% and 70%. Surgery is recommended for symptomatic patients. The main purpose of the study is to present the data of children with Chiari type I malformation that were treated surgically in low-volume center and to describe some atypical presentations in children. **Materials and methods**: At University Medical Centre Maribor, Slovenia, we performed a retrospective study of children with Chiari type I malformation that were treated surgically in the period from 2012 until 2021. The indication for surgery was symptomatic Chiari type I malformation. Suboccipital decompression with laminectomy of at least C1 (in one case also C2) with splitting of dura or duraplasty was performed. In a few cases coagulation of both tonsillar tips was necessary to achieve sufficient decompression and restoration of adequate cerebrospinal fluid (CSF) flow. **Results**: Ten children (under 18 years of age) underwent surgery for Chiari I malformation. Four patients had atypical presentation. In nine patients there was improvement after surgery. Complication rate was zero with no revision surgery. In one case suboccipital headache persisted. In all cases with preoperative syringomyelia it improved after surgery. Scoliosis improved in two out of three cases. There was no mortality after the procedure. **Discussion**: When patients with Chiari I malformation become symptomatic, it is recommended to perform surgery as soon as possible. Comparing the techniques; dura-splitting technique has advantages of lesser operation duration, lesser intraoperative bleeding and lower complication rates than duraplasty. **Conclusions**: The majority of patients with Chiari I malformation improve after surgery. Surgical procedure is safe with very low morbidity and mortality. Surgical technique must be an individualized patient tailored choice.

Keywords: Chiari I malformation; surgical techniques; posterior fossa decompression; duraplasty; autism; torticollis; attack of jerks

1. Introduction

Chiari I malformation consists of various degrees of caudal displacement of the cerebellar tonsils into the foramen magnum [1–3]. The most commonly associated finding is cervical syringomyelia. The foramen magnum potentially compresses the herniated cerebellar tissue and restricts normal CSF flow across the craniocervical junction [2]. Hydrocephalus occurs in 7–9% of cases [4]. The most common presenting symptom is pain (60–70%), which is usually located occipitally and in the upper cervical region. It is often induced by Valsalva manoeuvre and resolves shortly after the manoeuvre is completed. Other common symptoms include weakness or numbness, spasticity, loss of temperature sensation and unsteadiness. Spinal cord dysfunction is the result of direct cord compression or syringomyelia. The incidence of syringomyelia is between 30% and 70%. Syrinx location is most often cervical, followed by cervicothoracic. Permanent spinal cord damage can occur if left untreated. Some patients have various ophthalmologic and otologic disturbances, downbeat nystagmus, lower cranial nerve dysfunction, vocal cord paralysis with stridor or hoarseness or sleep apnea, which may be a

cause of sudden death. Scoliosis due to underlying syrinx is also a common presentation [2].

The imaging method of choice for diagnosis of Chiari I malformation is MRI. It is important to rule out other possible causes of tonsillar displacement due to secondary raised intracranial pressure (intracranial mass lesion, Dandy-Walker malformation, hydrocephalus) [5]. The exact incidence of Chiari I malformation is not known, although Meadows and colleagues found that out of 22,591 patients who underwent MRI of the head, 175 (0.775%) were found to have tonsillar herniation extending more than 5 mm below the foramen magnum [6]. The average age at the time of presentation was 41 years with female to male ratio being 1.3:1 [3].

Surgery is recommended for symptomatic patients and they respond best when operated on within 2 years of the onset of symptoms. Asymptomatic patients may be followed and operated upon if and when they become symptomatic. Patients who have been symptomatic and stable for years may be considered for observation, with surgery indicated when signs of deterioration occur. The most frequently performed operation is posterior fossa decompression (suboc-

cipital craniectomy), combined with or without other procedures: laminectomy of C1, possible to C2 or even C3, dural splitting, dural patch grafting (pericranium, artificial dura or fascia lata) [3].

The main purpose of the study is to present the data of children with Chiari type I malformation that were treated surgically in low-volume centre.

2. Materials and methods

At the University Medical Centre Maribor, Slovenia, we performed a retrospective study of our patients with Chiari type I malformation that were treated surgically in the period from 2012 until 2021. Our indication for surgery was symptomatic Chiari type I malformation. Before the operation all patients had confirmed Chiari malformation on MRI. Ten children underwent surgery. Six of them also had preoperative syringomyelia of the cervical or/and thoracic part of spinal cord. Two of the authors performed surgery on all of the analysed patients, JR with 20 years of experience and TŠ with 9 years of experience.

In an almost ten-year long period, we have all together operated 28 patients with Chiari I malformation at our institution. Median age of all patients was 34 years (from 2 to 68 years). With a cut-off of 18 years of age, we then split patients in paediatric (10 cases) and adult (18 cases) groups. For the purpose of this study, the paediatric group was further analysed.

Surgical procedure

Patients were positioned in prone position with head secured in the Mayfield head rest and flexed. We performed medial skin incision from inion to at least the level C2. In some cases, we prepared and harvested cervical fascia for duraplasty. Dissection of dorsal cervical muscles was performed through the linea alba down to spinal processes and occipital muscles were dissected from occipital bone. In all cases we performed suboccipital craniectomy with three centimetres in diameter around the foramen magnum. This was followed by C1 laminectomy and in more severe cases also C2 laminectomy, depending on the degree of tonsillar herniation (Fig. 1). We split the two dural layers in the exposed region and the movement of the cerebellar tonsils was inspected; if sufficient movement of cerebellar tonsils was present the decompression was stopped at that point, and we began with wound closure (Fig. 2). In other cases, we continued the procedure with duroplasty. Incision of the dura was made in Y shape. We released arachnoid membranes and mobilised cerebellar tonsils and established communication to the fourth ventricle. In larger herniations we coagulated or dissected herniated part of cerebellar tonsils. Usually, we did not release the syrinx. In only one severe adult case of syringomyelia we performed fenestration of the syrinx and created a syringo-subarahnoid shunt. After release of the tonsils, we started with duroplasty. It was done either with artificial dural grafts or autologous grafts (fascia,

periosteum) (Fig. 3). To prevent possible CSF leakage, we put a fibrin patch and fibrin glue over the suture line in dura. Wound closure was done meticulously to prevent haemorrhage and CSF leak through skin.
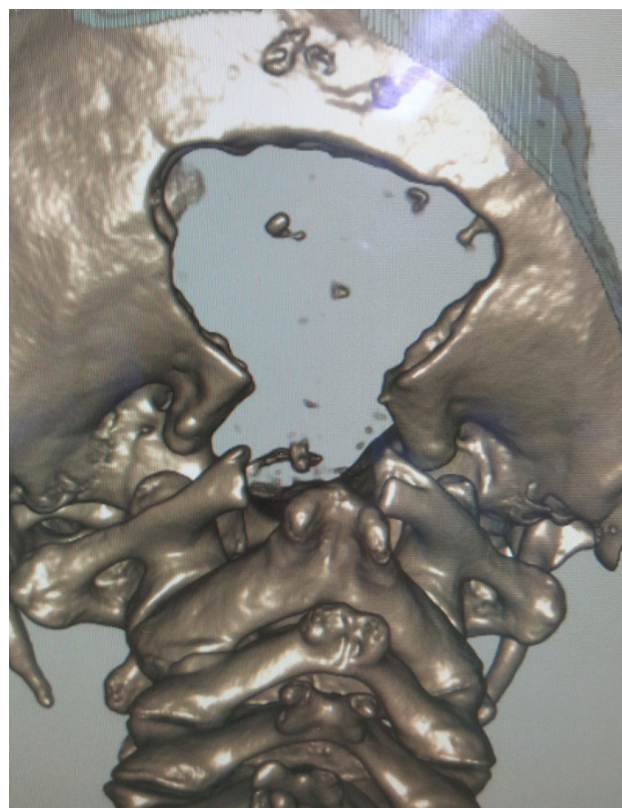

Fig. 1. Postoperative CT reconstruction showing degree of suboccipital decompression and laminectomy of C1.
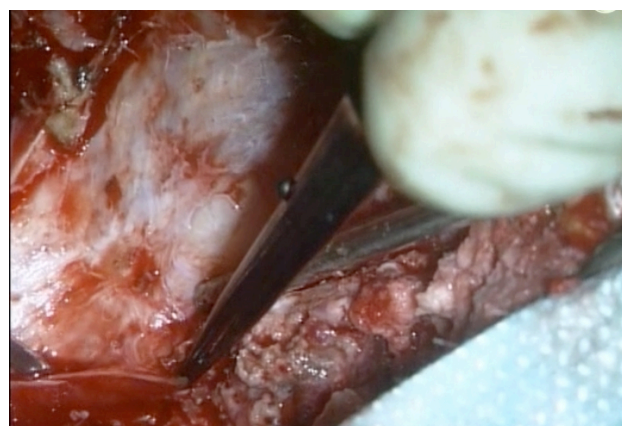
Fig. 2. Separation of dural leaves.

After surgery patients were hospitalised for at least seven days, during which they underwent early postoperative rehabilitation. They had their first follow up three months after surgery, re-occurring at six- and twelve-month
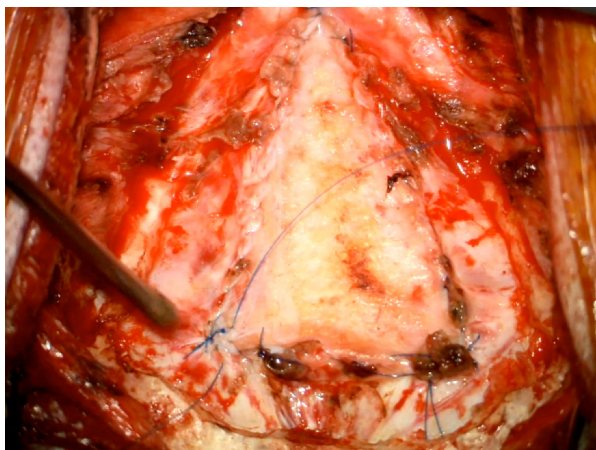
Fig. 3. Duroplasty with muscle fascia in atypical case shown in Fig. 5.

Fig. 4. Extend of syrinx before (A) and after (B) surgery.

intervals. Further follow-ups depended on patient's condition.

We analysed patients' data, duration, and type of symptoms before operation, type of operation, presence of syringomyelia, whether duroplasty was performed and the type of duroplasty, symptoms after surgery, follow-up period and final outcome (Table 1). We performed descriptive statistic of data. We retrospectively gathered the mentioned data from our hospital information system called Medis.

3. Results

The median age in the paediatric group was 12 years (from 2 to 16 years). Only one child was younger than 10 years. The majority of operated children were girls (8 out of 10). Duration of symptoms was on average 12 months (from 2 months to 24 months). Most common symptoms were headache (8 patients) and scoliosis (3 patients). Other symptoms were gait disturbance, limb pain, paraesthesia, hand tremor, auto-aggression, and torticollis. Syrinx was present in 6 patients, among them were three patients with scoliosis and one patient with paraparesis.

In all cases we have performed suboccipital decompression and C1 laminectomy. Additional C2 laminectomy was performed in one case. In six patients the operation procedure stopped after separation of the two dural layers. Four patients had additional duraplasty. In one patient the coagulation of the tonsils was performed additionally. Duraplasty was done with bovine pericardium in two cases and with suboccipital muscle fascia in two cases.

In the majority of patients there was improvement after surgery. Patients usually experienced transient headaches and nausea after surgery which subsided in a short period of time (within one month). In one patient the suboccipital headaches persisted. In three patients with scoliosis, the regression of scoliosis was achieved in two of them; in the third case, after initial arrest of scoliosis progression after Chiari I surgery, the surgery for scoliosis

was still needed 20 months after initial Chiari I surgery due to the late progression of the scoliosis. In all cases with syringomyelia we achieved regression of syringomyelia (Fig. 4).
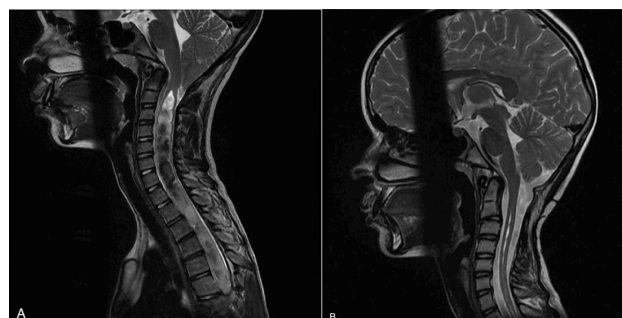
The median period of follow up was 13 months. Success rate of surgery was 90% (one patient had persistent suboccipital headaches). There were no postoperative complications with no revision surgeries and no mortality. The details of our patients' data analysis are presented in Table 1.

Atypical Chiari I malformation cases

Chiari I malformation can manifest with atypical clinical picture [7,8]. Some of our patients had atypical clinical picture too. The details of those patients are presented here.

CASE 1

We had a 12-year old autistic girl that had more severe auto-aggression. Because of change in her clinical picture and progression of aggression despite medication in six-month period, her neurologist decided to perform head MRI. On her MRI, it was seen Chiari type I malformation with syrinx. There were no other neurological deficits. Because of progression of her aggression we decided to operate her. We performed suboccipital decompression with laminectomy of C1 and duroplasty. After surgery her behaviour was less aggressive, and syrinx was smaller on head and neck MRI taken 4 months after surgery. Her clinical picture is after more than one year of follow up still stable and improved.

CASE 2

The second atypical case was a two years old girl with attacks of torticollis, gait disturbance and vomiting. Her symptoms started eight months after birth. Severity and frequency of attacks increased in following years. At the end attacks were almost every week with torticollis, headache, vomiting, gait disturbance. After that she was hole day tired. EEG was normal. On her MRI, there was Chiari I malformation without syrinx. Because of progression of symptoms without other causes of torticollis, we decided to perform suboccipital decompression, laminectomy C1 and duroplasty. After surgery she improved, her torticollis subsided.

Table 1. Analysed data of children with Chiari I malformation that were treated surgically at UMC Maribor.

Patient	Age (years)	Sex (F-female, M-male)	Surgery	Fenestration of syrinx	Duroplasy	Syringomielia preop.	Duration of clinical problems preop (months)	Clinical signs/symptoms preop.	Revision surgery	Complications	Improvement postop.	Follow-up (months)
1	10	F	subocc. craniectomy, C1 laminectomy, duroplasty	no	yes	yes	12	headache, scoliosis	no	no	Yes (less syrinx and scoliosis)	46
2	11	F	subocc. craniectomy, C1 and C2 laminectomy	no	no	yes	24	headache, scoliosis	no	no	yes (less syrinx)	20
3	13	M	subocc. craniectomy, C1 laminectomy	no	no	no	24	headache, hand tremor	no	no	yes (less headache)	7
4	15	F	subocc. craniectomy, C1 laminectomy	no	no	no	9	headache	no	no	yes (less headache)	13
5	16	F	subocc. craniectomy, C1 laminectomy	no	no	yes	21	headache	no	no	yes (less headache)	7
6	10	F	subocc. craniectomy, C1 laminectomy	no	no	yes	12	headache, scoliosis	no	no	yes (less headache)	24
7	15	F	subocc. craniectomy, C1 laminectomy	no	no	no	2	headache	no	no	no	14
8	12	F	subocc. craniectomy, C1 laminectomy, duroplasty	no	yes	yes	6	autism, aggression,	no	no	yes (less aggression)	13
9	12	M	subocc. craniectomy, C1 laminectomy, duroplasty	no	yes	yes	5	paraparesis, attacks of jerks in lower limbs	no	no	yes (no more jerks, the strength in his legs is improving)	5
10	2	F	subocc. craniectomy, C1 laminectomy, duroplasty	no	yes	no	16	cervicalgia, torticollis, gait disturbance and vomiting	no	bacteremia, not related to surgery	yes (less torticollis)	4

CASE 3

A 12-years old boy suffered from attacks of jerks in lower limbs, pain in legs and a progressive unsteady walk. His clinical picture gradually progress in 5 months with more frequent jerks in legs and unsteady, shaking walking. He performed detailed diagnostic in other clinical centre and despite small Chiari type I malformation and small syrinx on level C6 and C7, all other results were normal. Psychosomatic cause was also in role. When patients came to us, because of future progression of symptoms, we decided for surgery and performed suboccipital decompression, laminectomy C1 and duroplasty. After surgery his clinical picture and MRI image improved, and there were no more jerks, the strength in his legs is now improving (Fig. 5).

Fig. 5. Case of a 12-year old boy with a small Chiari type I and syrinx before (A) and after (B) surgery.

4. Discussion

At the Department of neurosurgery at the University Medical Centre Maribor, Slovenia, we retrospectively analysed 10 paediatric patients with posterior fossa decompression for the treatment of Chiari I malformation, operated during the time period between 2012 to 2021. That represent 36% of all surgicaly treated patients with Chiari I malformation in ten years' period. Median age of all cases was 34 years and majority (78.6%) were females. This is comparable with known epidemiology of Chiari I malformation, where average age of presentation is 41 (in a range from 12 to 73), with female preponderance (female:male = 1.3:1) [9]. In all cases suboccipital decompression with laminectomy of at least C1 was performed, with additional C2 laminectomy in one case. The main reason for that was that with C1 laminectomy alone we were not able to fully visualize the caudal tip of the cerebellar tonsil and therefore the decompression would not be sufficient with just C1 laminectomy. Separation of dural leaves was performed in six cases. In the other four cases we performed duraplasty with grafting. The main reasons for that decision were the insufficient movement of cerebellar tonsils, cases with greater descensus of tonsils, or more severe syringomyelia. It has been shown that dura-splitting technique has clinical and radiological outcomes comparable to conventional dura opening techniques with duraplasty. This technique has advantages of lesser operation duration, lesser intraoperative bleeding, and lower complication rates compared to the aforementioned technique [10]. In very few cases, we encountered severe tonsillar ectopia and in those cases we needed to coagulate both tonsillar tips to achieve sufficient decompression and restoration of adequate CSF flow. Prior surgery decision on type of surgery can be made also according to the mechanism of ptosis of the hindbrain, based on a preoperative MRI morphometric study of the posterior cranial fossa and craniovertebral junction, that classified Chiari I malformation in type A (normal volume of posterior fossa, volume of foramen magnum and occipital bone size), type B (normal volume of posterior fossa, small volume of foramen magnum and occipital bone size) and type C (small volume of posterior fossa, foramen magnum and small occipital bone size). For the first two types foramen magnum decompression is recommended and expansive suboccipital cranioplasty for type C [11]. During surgery Colour Doppler ultrasonography imaging could help neurosurgeon to tailor the surgery according to patient-specific variables as are adequate decompression of cervicomedullary junction, creation of a sufficient volume of retrocerebellar space and establishment of optimal cerebrospinal fluid flow between the cranial and spinal compartments [12]. Others have shown that in the absence of hydrocephalus and craniocervical region instability, posterior fossa decompression with duraplasty provides better clinical outcomes but with higher risk of complications [13]. Thereby, it can be an optimal surgical strategy because of its higher clinical improvement and lower recurrence rate in the patients with syringomyelia. In patients without syringomyelia, posterior fossa decompression alone can be a preferred choice because of its similar clinical improvement and lower costs. It is also associated with lower risk of complications [14]. Under the circumstance of failed posterior fossa decompression, additional surgery with duraplasty tends to be worth considering [15].

Syringomyelia was present in six out of ten cases. Among them three patients had scoliosis. The aetiology of Chiari related syringomyelia is still not fully understood, but it was greatly enhanced by the advent of MRI [16]. Alikely explanation is that by Oldfield and colleagues, who proposed that the pulsatile movement of the cerebellar tonsils acting on an enclosed spinal subarachnoid space drives fluid into the cord parenchyma [17]. Stoodley's research [18,19] pointed to the role of arterial pulsations in the spinal subarachnoid space as contributing to the trans-

parenchymal propagation of CSF.

In three patients with scoliosis, we achieved arrest of scoliosis progression initially. In two cases regression of scoliosis in the follow up time period of 24 and 46 months was observed. In the third case the progression of scoliosis was observed 20 months after the Chiari I surgery and additional surgery for scoliosis was needed. Regression of scoliosis is reported in 55–64.8% of Chiari type I patient after posterior fossa and foramen magnum decompression with or without syrinx drainage. Factors for progression of scoliosis after decompressive surgery are age more than 10 years, Cobb angel more than 30–44.5° and double major curve pattern.

In the clinical picture the majority of patients had headache (8 out of 10), followed by scoliosis, paraesthesia, limb pain, gait disturbance, hand tremor, breathing disorders, auto-aggression and torticollis. Chiari I malformation can manifest with atypical clinical picture and we had three cases with more pronounced aggression in autistic girl, attacks of torticollis and attacks of jerks in lower limbs with pain in legs. There are reports of other atypical clinical presentations of Chiari I malformations, for instance with recurrent syncopes and seizures. Regarding syncopal episodes, various pathophysiological mechanisms have been proposed; compression of the midbrain ascending reticular activating system, vascular compromise from vertebrobasilar artery compression, hypotension secondary to cardiorespiratory centre involvement are examples of these [7]. Another rare manifestation of Chiari I malformation is opisthotonos. It is usually seen in infants, but it is possible in older children too. The mechanism by which opisthotonos is related to Chiari malformation is thought to be associated with hindbrain sensitivity to fluctuations in intracranial pressure [8].

The mean time from the presentation of symptoms to surgery was 12 months. When we decided to operate on a patient, we tried to do it as soon as possible as it has been shown that the neurological outcomes are better when the patients are operated on within 2 years of the onset of symptoms [3]. The results after surgery were good with no evident worsening of neurological symptoms in all of our patients. In the paediatric group the success rate of surgery was 90% (only one patient had persistent headaches after surgery) and there were no postoperative complications. We have not observed any postoperative craniocervical instability or cervical kyphosis.

5. Conclusions

We conclude that our experience with pediatric Chiari I malformation patients and our results of surgical treatment are similar to other studies. Surgical results in the presented paediatric group of patients after Chiari I decompression are good. In majority of cases an improvement of symptoms and signs was achieved. The surgical procedure is safe with very low morbidity and mortality. We should be also aware of broad presentation of Chiari I malformation and be prepared to act also in atypical cases. If patients are left untreated, malformation can progress to irreversible deficits. Because of that early treatment is important and the chosen surgical technique must be an individualized patient tailored choice.

Author contributions

JR—designed the research study and performed the research, analysed the data and wrote the manuscript, contributed to editorial changes in the manuscript. TŠ—designed the research study and performed the research, analysed the data and wrote the manuscript, contributed to editorial changes in the manuscript. BH—designed the research study and performed the research, analysed the data and wrote the manuscript, contributed to editorial changes in the manuscript. All authors read and approved the final manuscript.

Ethics approval and consent to participate

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Conflict of interest

The authors declare no conflict of interest.

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