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"Management: opinions from different centers"—the Sankt Augustin experience

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Abstract

Objective In this "how we do it" survey, we review our management regimen of symptomatic CM1 and provide an analysis of our institutional case series of "bony only" decompression of the craniocervical junction without dural opening.

Methods In regard to the latter clinical symptomatology, neurological status, electrophysiology data, and pre- and post-surgical MRI were analyzed. Surgery was performed in standard fashion under IOM, evaluated by intraoperative ultrasound.

Results We reviewed 22 patients (mean age at surgery 13 ± 7 years; 11 female, 11 male). Neck pain, occipital headaches, sensory symptoms, and dizziness were the predominating symptoms; 9% had central apnea, 5 patients had scoliosis, and 2 patients had a history of premature synostosis. On MRI, preoperative mean tonsillar herniation was 16.55 ± 6.19, compared to 14.25 ± 6.75 after surgery. About half of patients with syringomyelia ($n = 11$) experienced imagining improvement after surgery. Patients with neck pain, occipital headaches, dizziness, and sensory abnormalities benefited most from surgery. Of the 6 cases that presented with pathological SSEPs, 4 exhibited improved measurements after surgery. There were no postoperative complications.

Conclusion To conclude bony decompression for CM1 resulted in clinical and imaging wise improvement and can be viewed as a safe first-lane option for symptomatic CM1.

Keywords Chiari Malformation Type I · Bony-only · Outcome · Clinical symptoms · Electrophysiology · MRI

What symptoms are considered typical

It is well accepted that especially in mild or incidental forms of Chiari Malformation Type I (CM1), one has to carefully evaluate whether there is a causal link between imaging findings and general clinical symptoms like headaches or even more unspecific complaints. In general, symptoms regarded typical for CM1 like occipital headaches, neck pain, shoulder pain, sensory symptoms (especially dysesthesia like numbness), and dizziness were also commonly observed in our patient cohort as outlined in the following (and Table 1). Further clinical symptoms and findings, which were associated with CM1 in our series, were torticollis, frontal headaches, holocephalgia, dysphagia, ataxia, and pain in the lower extremities. In our view, pathological electrophysiological and polysomnographic findings like central apnea are strong indicators for symptomatic CM1. As such, besides high-resolution MRI imaging, these technical investigations are of high importance in the diagnostic work-up of CM1 in our department.

Criteria for surgical indications

Clinical symptoms as listed above that can be reasonably associated with problems in the craniocervical junction and related to CM1 typical imaging findings are indicative for surgery. In addition, pathological findings in polysomnography and electrophysiological examinations reinforced the decision for surgery. In asymptomatic, i.e., incidental CM1 cases, further imaging findings like syringomyelia or medullary edema are considered an indication for bony decompression of the craniocervical junction.

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Table 1 Clinical findings and their post-surgical course in our institutional case series

Symptoms	Pre-op	Post-op		
		Resolved	Improved	Unchanged
Occipital headaches	7	3	4	–
Neck pain	9	1	7	1
Torticollis	1	1	–	–
Frontal headaches/atypical headaches	3	–	2	1
Dysphagia	1	1	–	–
Behavioral symptoms/developmental delay	1	–	–	1
Sensory symptoms	6	2	3	1
Ataxia	2	–	2	–
Leg pain	2	1	–	1
Back pain	1	1	–	–
Dizziness/loss of consciousness	5	3	2	–
Nausea	2	–	1	1
Central apnea (PSG)	4	–	2	2
Fatigue	3	–	2	1

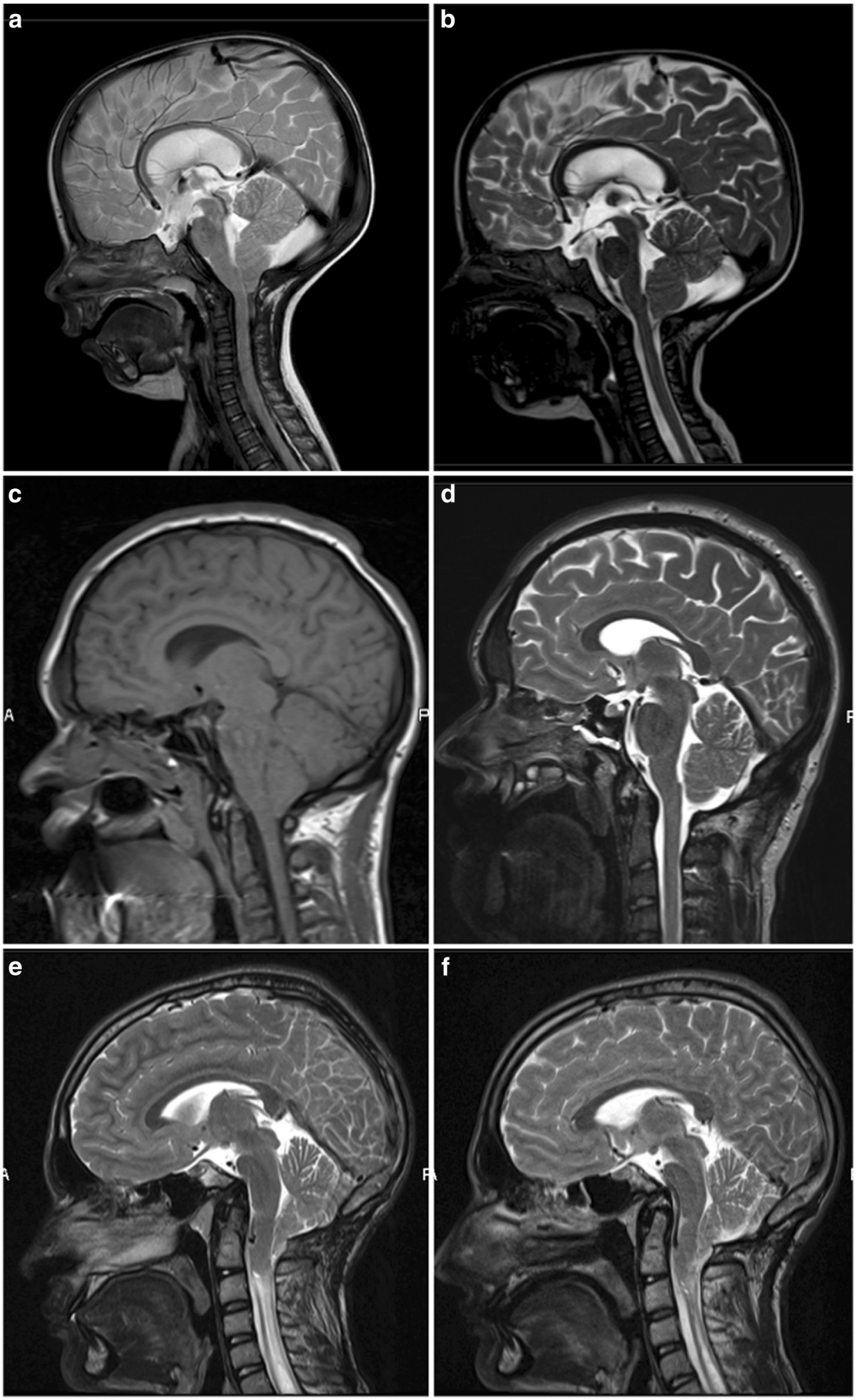
Table 2 Cases with preoperative pathological SSEPs and their post-surgical course

Case	Symptoms	SSEP (*N. medianus* and *N. tibialis*)	
		Pre-op	Post-op
Case 6	Progressive dysesthesia of the left body ➔ Improved after surgery	*N. medianus* (left) reduced amplitudes *N. tibialis* (left) pathol.	Normal (improved)
Case 9	Neck pain, numbness of the right side/leg ➔ Completely recovered	*N. tibialis* (right) pathol.	Normal (improved)
Case 14	Torticollis, tingling extremities ➔ Completely resolved	*N. tibialis* (left) pathol.	Unchanged
Case 16	Occipital headaches, neck pain, back pain, sleep apnea, numbness (non-specific) ➔ Pain resolved ➔ Apnea improved	*N. tibialis* (left) pathol.	Improved
Case 17	Neck pain, intermitted pain, and numbness in the arms and legs ➔ Pain and sensory deficits improved	*N. tibialis* pathol.	*N. tibialis* (left) improved

Type of surgery commonly used

In our institution, bony decompression of the craniocervical junction without dural opening is the preferred first-line option for symptomatic CM1. The primary aim is the sufficient enlargement of the foramen magnum with lateral decompression and removal of the posterior arch of C1. Thereby, foramen magnum decompression is performed via suboccipital craniectomy (usually measuring about 3–4 cm wide and 3–4 cm above the foramen magnum) plus laminectomy of the atlas. The dural band at the foramen magnum is meticulously

Fig. 1 Illustrative cases of MRI imaging courses after bony decompression for symptomatic Chiari 1. Sagittal T2-MRI of the preoperative (**a**) status of a 2 years old patient with typical Chiari 1 findings in comparison with 8-month postoperative imaging (**b**), demonstrating sufficient release of the craniocervical junction after bony decompression. In comparison, **c** sagittal T1 MRI and **d** sagittal T2 MRI depict pre- and postoperative MRI imaging in an older patient (15 years of age at surgery) with resolved herniation of tonsils after surgery in the longer follow-up (4 years). **e** A sagittal MRI-Imaging (T2) of a 16-year-old patient who received bony decompression for Chiari 1 already but had the progression of a pre-existing syringomyelia in the 3-month follow-up imaging. After the redo-surgery with the widening of the occipital bony decompression, the syrinx appeared not as extended as before the second operation (**f**) ▶


a
b
c
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e
f

released, and the superficial layer of the dura is split longitudinally. In this procedure, intraoperative ultrasound is viewed as mandatory to verify sufficient release (judged from the grade of pulsatility and configuration of tonsils). Further, all cases operated in our institution in this way were and are monitored during surgery by intraoperative monitoring (IOM; SSEP and MEP).

When the result is considered good

The surgical outcome is considered effective if preoperative symptoms resolve or improve after the procedure. Pure impact on imaging findings like the resolution of the syrinx or upper movement of the cerebellar tonsils is considered less relevant.

Short presentation of the series (results and complications)

Due to the fact that bony decompression (as outlined above) is the favored first-line option for symptomatic CM1 in our institution, we screened the hospital database for patients who were surgically treated for CM1 with decompressive surgery without dural opening from 2015 to 2018. Patients' records were analyzed in regard to the pre- and postsurgical clinical history and clinical courses, including neurological findings, polysomnography investigations, and electrophysiological studies. Pre- and post-surgical MRI studies (3 months after surgery and later) were reviewed for the degree of cerebellar tonsil herniation and the existence of syringomyelia or medullary edema. Only patients who received high-resolution standard T1 and T2 sagittal and axial imaging of the whole neuraxis pre-surgically and high-resolution post-surgical imaging were considered in this survey.

Further, only patients with a minimum of a 3-month clinical and imaging follow-up after surgery were considered. As such, the clinical and imaging courses of 22 patients, who received bony decompression as outlined above, were analyzed in respect of clinical outcome and complications.

Mean patients' age at surgery was 13 ± 7 years. Female to male ratio 11:11. Five patients presented with scoliosis, 2 patients had a history of premature synostosis, 11 patients presented with syringomyelia on initial MRI, 1 presented with edema in the cervical spinal cord, and 1 patient had received bony decompression before ("redo"). Considering clinical findings, neck pain (19%), occipital headaches (15%), sensory symptoms (13%), and dizziness (11%) were the predominating symptoms. In PSG, 9% confirmed central apnea; 6% of the patients complained about frontal headaches. All clinical symptoms before surgery are summarized in Table 1.

In general, the surgical outcome was good as no intra- or postoperative complications occurred. Patients recovered fast after surgery (hospital stay ≤ 5 days). None exhibited worsening of IOM recordings during surgery or during further follow-up. One patient needed second surgery with duroplasty due to the persistence of cervical headaches.

In most of the investigated cases, there was an improvement of clinical complaints as summarized in Table 1: Patients with symptoms like neck pain, occipital headaches, dizziness, and sensory abnormalities benefited most from bony decompression of the craniocervical junction. In regard to electrophysiological findings, 6 cases revealed pathological SSEPs before surgery, from which 4 exhibited improved measurements after surgery (details are provided in Table 2).

On MRI, the mean tonsillar position in relation to the craniocervical junction before surgery was 16.55 ± 6.19 (range 5–29 mm). After surgery, the mean position changed to 14.25 ± 6.75 (range 0–24 mm). Thereby in 7 out of 22 cases tonsils ascended more than 2 mm upwards after surgery (Fig. 1 a to d depicts exemplarily 2 cases, in which tonsils appeared in an improved or normal position during the postoperative course). Eleven patients in our cohort presented with syringomyelia before surgery. In 5 of them, the syrinx became smaller after surgery (especially in the longitudinal direction). Figure 1e and f exemplarily depict 1 patient, who received a bony re-decompression for Chiari 1 due to persistent syringomyelia before scoliosis surgery. After the second decompression, the cervical syrinx became obviously smaller.

To summarize, bony decompression for clinically relevant CM1 resulted in improvement of clinical symptoms in most of the investigated cases. Also, in some of the patients, tonsils ascended after surgery, and syringomyelia, if present pre-surgically, improved in about half of theses of the cases.

Thus, bony decompression can be viewed as a safe first-line option for patients who exhibit clinically relevant CM1. However, this should of course be proven by prospective, ideally multicenter studies.

Compliance with ethical standards

Conflict of interest The authors declare that they have no conflict of interest.

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