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Review

# The Role of Neurophysiology in Managing Patients with Chiari Malformations

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**Abstract:** Chiari malformation type 1 (CM1) includes a series of congenital anomalies that share ectopia of the cerebellar tonsils below the foramen magnum, in some cases associated with syringomyelia or hydrocephalus. CM1 can cause dysfunction of the brainstem, spinal cord, and cranial nerves. This functional alteration of the nervous system can be detected by various modalities of neurophysiological tests, such as brainstem auditory evoked potentials, somatosensory evoked potentials, motor evoked potentials, electromyography and nerve conduction studies of the cranial nerves and spinal roots, as well as brainstem reflexes. The main goal of this study is to review the findings of multimodal neurophysiological examinations in published studies of patients with CM1 and their indication in the diagnosis, treatment, and follow-up of these patients, as well as their utility in intraoperative monitoring.

**Keywords:** blink reflex; brainstem auditory evoked potentials; brainstem reflexes; Chiari type 1 malformation; electromyography; evoked potentials; intraoperative monitoring; motor evoked potentials; somatosensory evoked potentials; syringomyelia

## Introduction

Chiari malformations (CMs) comprise a series of neurodevelopmental disorders characterized by a descent of the cerebellar tonsils through the foramen magnum (FM) [1–5]. CM type 1 (CM1) was first described by the Austrian pathologist Hans Chiari in two papers published in 1891 and 1895 [2–4]. Nevertheless, the term “Arnold-Chiari malformation” must still be introduced to search in PubMed or other databases for literature on this abnormality. However, due to Arnold’s minor role in the original description, “Chiari malformation” is the most widely accepted eponym [6]. In Chiari’s original description, four types of malformations were differentiated (CM1, CM2, CM3, and CM4). CM1 is traditionally defined as a tonsillar descent of 3–5 mm below the FM, quantified in a mid-sagittal section of magnetic resonance imaging (MRI). The choice of cut-off point for tonsillar descent (3 or 5 mm) is somewhat arbitrary and varies depending on the criteria of the different authors [7–13]. CM type 2 (CM2) is characterized by a descent of the structures of the brainstem or vermis below the FM and is always associated with spinal dysraphisms (spina bifida) in addition to tonsillar ectopia. Patients with CM2 also present a series of brain anomalies associated with spina bifida that are not observed in any other type of CM (gray matter heterotopia, polygyria, and descended tentorium, among others). CM type 3 (cranio-cervical encephalocele) and CM type 4 (cerebellar hypoplasia) [14] are severe malformations with a low incidence that most authors consider unrelated

to CM1 and CM2. The recent International Consensus Conference recommended considering CM3 and CM4 as separate entities [15].

Other variants of CM1 have been described as minor or major forms of CM1. In 1998, Iskandar et al. coined the term “Chiari 0” (CM0) to describe five pediatric patients with syringomyelia, no tonsillar herniation, and a “tight” posterior fossa (PF), who improved after PF decompression. Subsequent studies by this and other groups have confirmed this entity and observed a significant volumetric reduction in the PF and alterations in cerebrospinal fluid (CSF) dynamics around the FM as the common etiopathogenic factor in CM0. Some authors suggested the term “tight cisterna magna” to define the same entity [16–19]. In 2004, Tubbs et al. coined the term “Chiari 1.5” to define those patients without spina bifida that presented, in addition to the tonsillar ectopia, a variable descent of the brainstem and displacement of the obex below the FM [13]. We found that once this variant was defined, a considerable percentage of patients in our series initially classified as CM1 actually corresponded to CM1.5 [19–22].

An additional type, often not included in canonical classifications, is comprised of patients in which any type of CM is associated with different osseous malformations of the craniovertebral junction (CVJ). In a previous paper, we proposed the term ‘complex CVJ abnormalities’ when patients present tonsillar herniation and at least two of the following abnormalities: a significant retroflexed odontoid, a basilar impression (BI), platybasia, severe bone abnormalities in the C0–C2 complex, uni- or bilateral occipital condyle hypoplasia, atlantooccipital assimilation, or other abnormalities that condition an anterior compression of the cervical-medullary junction [19,23] (Figure 1). These patients need different clinical management and often require multiple surgical procedures (such as anterior approaches and occipito-cervical fusions) [24].

**Figure 1.** Patient with a complex craniovertebral junction (CVJ) malformation. This 17-year-old male with a VACTERL association (<https://rarediseases.info.nih.gov/diseases/5443/vacterl-association>)—a non-random association of congenital disabilities that affects multiple parts of the body—was referred to our institution due to a Chiari malformation type 1 (CM1) detected as the patient presented recurrent episodes of severe occipital headaches. The neurological examination was normal. The patient had CM1 with an asymmetrical tonsillar descent 11 mm below the McRae line. (A): T1\*weighted sagittal midline image; (B): T2\*weighted sagittal, and (C): T2\*weighted coronal slices; (D) and (E): Three-dimensional CT reconstruction of the CVJ. White asterisks in (B) and (C) indicate the right cerebellar tonsil. The patient presented complete assimilation of the anterior and posterior C1 arches with an associated C2-C3 fusion and partial agenesis of the right C2 lamina. C0: posterior part of the foramen magnum. C1 labels the right lateral mass of the atlas. The red arrow in (D) shows the assimilated posterior C1 arch. Arrow in (F) indicates the valve (Polaris® adjustable valve, Sophysa, Orsay, France) of a ventriculoperitoneal shunt implanted one month before because of hydrocephalus. This patient was treated with halo-vest stabilization for three weeks, followed by a one-step surgical procedure in which a posterior fossa reconstruction was conducted and instrumented occipito-C4 posterior fusion with bone-bank allograft (F).

A common feature of CMs is that the cerebellar tonsillar descent causes compression of the neural structures and hinders CSF passage at the cervico-medullary junction, consequently altering brainstem and upper spinal cord function. The compressive effect on neural structures at this level may produce alterations in brainstem pathways, cranial nerve nuclei, or their exit from the brainstem, sleep-regulating regions, and cardiorespiratory centers [25]. The frequent association of syringomyelia with any form of CM can induce additional damage to the spinal cord and the spinal roots that emerge from the gray matter. These functional alterations of the nervous system can be detected by multimodal neurophysiological tests: brainstem auditory evoked potentials (BAEPs), somatosensory evoked potentials (SEPs), motor evoked potentials (MEPs), electromyography (EMG) of the cranial nerves/ spinal roots, and brainstem reflexes.

Our paper aims to review the literature and summarize the findings reported in multimodal neurophysiological examinations in patients with CM0, CM1, and CM1.5 and their role in managing these patients, establishing indications for surgical treatment and follow-up. In addition, we will update their role in intraoperative neurophysiological monitoring (IONM). The role of

neurophysiology in the diagnosis of sleep-related disorders, a frequent finding in CM1 [22,25,26], has deliberately been excluded from this review. We also excluded articles referring to cognitive evoked potentials.

## Methods

This study is an updated narrative review of neurophysiological studies (except for polysomnography and sleep disorders) published up until 30/08/2022. We searched the following databases: PubMed, EMBASE, and WoS. The search strategy used a combination of the keywords: "Arnold Chiari malformation," "Chiari type 1 malformation\*," "Chiari type 1.5 malformation\*," "Type I Arnold Chiari malformation\*," "Chiari malformation type I with Syringomyelia," "CM1 with Syringomyelia," "Syringomyelia" and "evoked potentials," "blink reflex\*," "brainstem auditory evoked potential\*," "brainstem reflex\*," "electromyography," "motor evoked potential\*," "intraoperative neuromonitor\*." The flowchart used is shown in Figure 2.

**Figure 2.** Flowchart. Diagram of the process used to select the articles for this review. CM1: Chiari malformation type 1. IONM: Intraoperative neurophysiological monitoring.

## Exploring Brainstem and Spinal Cord Functionality

### *Brainstem Auditory Evoked Potentials (BAEPs)*

BAEPs were first described in humans in 1970 [19]. They allow the recording of a series of five to seven positive electrical signals generated in response to sounds, each of which has a well-defined electrical generator [20,21] (Figure 3); because waves VI and VII are not constant in the healthy population, they are not routinely evaluated. BAEPs explore the functional integrity of a limited portion of the brainstem, both in the rostrocaudal direction—from the VIII cranial nerve entry at the pontomedullary junction to the upper part of the pons-midbrain— as well as in the transversal plane. The ventral part of the brainstem is not explored with BAEPs. It is widely accepted that alterations in waves I to III reflect the involvement of neural structures ipsilateral to the auditive stimulation. In contrast, abnormalities in the amplitude or latency of waves IV and V indicate contralateral structural impairment [22]. BAEPs can be abnormal in several processes: 1) peripheral auditory pathology (conductive or cochlear hearing loss); 2) primary anatomical modifications of the brainstem due to congenital pathologies (CM, Joubert syndrome, Dandy-Walker syndrome, etc.) or secondary (vascular, tumor, demyelinating, or degenerative); 3) anoxia and ischemia; and 4) intracranial hypertension with transtentorial herniation [23]. Due to the close relationship between the cranial nerves and the brainstem, BAEPs are routinely used in IONM and are important in surgical procedures in which the brainstem and cranial nerves are at risk [27].

**Figure 3.** Example of normal brainstem auditory evoked potentials. The image summarizes the waves recorded after applying the acoustic stimulus with their respective neural generators. Wave I: organ of Corti of the cochlea (the distal portion of the auditory nerve). Wave II: cochlear nucleus (and the proximal intracranial portion of the auditory nerve). Wave III: superior olivary complex, trapezoid body. Wave IV: ventral nucleus of the lateral lemniscus. Wave V: inferior colliculus. Wave VI: medial geniculate nucleus of the thalamus. Wave VII: thalamocortical radiations and primary auditory cortex (Heschl's) or Brodmann's Area 41. Ai: ipsilateral earlobe. (Modified from Moncho et al., with permission [23]).

### *Somatosensory Evoked Potentials (SEPs)*

SEPs explore the conduction of the electrical impulse through the lemniscal or posterior cordonal system (Figure 4), collecting the integrated responses obtained at different anatomical levels after applying a repetitive electrical stimulus on a peripheral sensory nerve or the sensory portion of a mixed nerve—usually the median nerve (MN) for the upper limbs and the posterior tibial nerve (PTN) for the lower limbs. In addition, other nerves, such as the cubital, trigeminal, saphenous, or pudendal, can be explored [28–31]. SEPs explore the somesthetic pathway from the peripheral nerve,

preganglionic or postganglionic plexus, roots, spinal cord, brainstem, thalamus, and supratthalamic structures [23]. In anoxic or traumatic coma, they are useful for evaluating cortical and subcortical function. SEPs are now routinely used in the IONM of surgical procedures with a risk of damaging structures that generate and transmit the electrical signal [32]. In addition, 'dorsal column mapping' has also been introduced to guide the surgical team in procedures with a risk of spinal cord damage [33,34].

**Figure 4.** Example of normal somatosensory evoked potentials. The image shows the responses recorded at different levels after stimulation of the median nerve in the wrist and its main generators. N9: brachial plexus. N11: entry of roots in dorsal horns and posterior columns. N13: high cervical and medullary structures (nuclei of the posterior columns). P14: most caudal part of the medial lemniscus. N18: medial lemniscus at the mesencephalic level. N20: primary somatosensory cortex (Brodmann areas 3, 1, 2). Cc': contralateral parietal cortex. Hc: Contralateral shoulder. Cv7: the spinous process of the seventh cervical vertebra. (Modified from Moncho et al., with permission [23]).

Cutaneous heat stimulations with a laser beam can selectively activate thermoalgesic A-delta and C nociceptors, leading to the generation of laser-evoked potentials (LEPs) in the cortex [35,36]. LEPs have been recognized as reliable neurophysiological tools for investigating neuropathic pain [37]. Pure spinothalamic lesions, such as syringomyelia, brainstem syndromes, or small fiber neuropathy, are characterized by normal SEPs but abnormal LEPs [36,38]. However, spinal injuries involving the cervical or lumbar dorsal horn can also alter or abolish components of classical SEPs, such as N13 or N22 [31,39]. Despite their potential utility, LEPs have not been widely adopted due to technical difficulties, the need for skilled personnel, and associated risks like skin burns and hyperpigmentation. Contact heat-evoked potentials (CHEPs) have emerged as a safer alternative to LEPs and are easier to use but require patient cooperation and are unsuitable for IONM [40]. In recent research, Leandri et al. introduced a new noninvasive electrode that selectively stimulates nociceptive nerve-free endings, enabling the evaluation of disorders affecting the nociceptive pathway by enlarging the stimulated surface area [41].

### *Motor Evoked Potentials*

The term "motor evoked potentials" refers to potentials recorded from muscle or nerve after stimulation of the primary motor areas in the CNS. MEPs are the responses elicited distally along the spinal cord, peripheral nerve, or muscle with stimulation of the central motor pathways (spinal cord or cerebral hemispheres) using either transcranial magnetic stimulation (TMS) or electrical stimulation. Electrical stimulation is utilized routinely for IONM, whereas magnetic stimulation is generally used for diagnostic studies in awake patients because of its better tolerance.

TMS is performed with coils of different sizes and shapes. A magnetic field applied through the skull induces a stimulus in the underlying brain tissue with minimal current affecting the skin and subcutaneous tissue. For lower extremity MEPs, the coil is centered in the Cz region; for the upper limb, it is just lateral to this reference. With peripheral recordings, three MEP characteristics are used: latency, amplitude, and threshold. MEP amplitude is highly variable but is generally considered abnormal if it is less than 20% of the compound muscle action potential (CMAP) amplitude obtained on peripheral neurography [42]. Central motor conduction time (CMCT) is the most commonly used parameter to identify CNS disorders [43] and is calculated by subtracting the latency of spinal MEPs from the latency of cortical MEPs [44]. TMS MEPs are used to study central demyelinating disease, motoneuron disease, epilepsy, movement disorders, ataxia, myelopathies, neuropathies, and radiculopathies [45].

In order to elicit an MEP by transcranial electric stimulation (TcMEP) for IONM, subcutaneous electrodes are placed on the scalp in C1-C2 or C3-C4 (10-20 EEG International System). In general, a 50% reduction in amplitude from the average baseline value during surgery is considered significant and a warning sign of damage to the corticospinal tract. Similarly, corticobulbar motor evoked potentials (CoMEPs) elicit cranial nerve responses [46]. In addition to the two aforementioned

techniques, which assess the integrity of the corticospinal tract, several mapping methods have been described for identifying the corticospinal tract by directly stimulating the exposed neural structures with a bipolar or monopolar electrode [47,48].

#### *Electromyography and Nerve Conduction Studies*

Virtually all primary neuromuscular diseases cause changes in the electrical activity recorded in muscle fibers. These changes can best be explored using needle electrodes inserted into the muscle to record spontaneous and voluntary electromyography (EMG). EMG is one of the major diagnostic tools for identifying and characterizing motor unit disorders, including anterior horn cells, peripheral nerves, neuromuscular junctions, and muscles. EMG complements nerve conduction studies (NCS) in the localization of neuromuscular disorders [44,45]. Normal muscle fibers at rest show no spontaneous electrical activity outside the end plate region. All voluntary muscle activity is mediated by the lower motor neurons and their corresponding innervated muscle fibers that form the 'motor unit potentials' (MUPs). Voluntary MUPs are the sum of the potentials of individual muscle fibers innervated by a single anterior horn cell that occurs in the region of the needle electrode. MUPs are characterized by their firing pattern, rates of fire (recruitment), and configuration and appearance; MUPs fire in a semi-rhythmic pattern. Neuromuscular diseases may show abnormal spontaneous discharges, abnormal voluntary MUPs, or both. Abnormal spontaneous activity includes fibrillation potentials, fasciculation potentials, myotonic discharges, complex repetitive discharges, myochemical discharges, cramps, and neuromyotonic discharges. MUPs may have abnormal duration and amplitude, an abnormal number of phases, or morphology. The extent and distribution of these abnormalities allow us to define the type and severity of the disease, its stage, and the anatomical location of the lesion [44].

EMG is also used in IONM via continuous free-run and stimulated EMG. Free-run EMG consists of recording spontaneous muscle activity in real time. In intraoperative neurophysiology, we use this technique as a monitoring tool to detect surgically induced mechanical irritation of the peripheral nervous system and cranial nerves before irreversible damage to these structures may occur. Stimulated EMG consists of applying an electrical stimulus directly to the peripheral motor nerves or roots to be registered in the corresponding muscles and can be used as a mapping tool to detect the location of peripheral or cranial nerves that are difficult to distinguish from tumors, fibers, or fatty tissues. Stimulated EMG is also used to check the function of injured (or at risk of injury) nerves, roots, or trunks by evaluating electrical transmission through these structures and comparing it to the baseline [49].

#### *Brainstem Reflexes*

**Blink reflex.** The blink reflex (BR) is the neural response obtained in the orbicularis oculi muscle after stimulating the trigeminal nerve with electrical, mechanical, or other stimuli. The BR is the most widely used brainstem reflex in clinical practice. The trigeminal BR is mediated by the first division of the trigeminal nerve (afferent branch) and facial nerve (efferent branch). In normal individuals, two responses (R1 and R2) are obtained when recording from the ipsilateral muscles to the stimulated nerve, and a single (R2) response is obtained from the contralateral muscles [50]. R1 is constituted by the oligosynaptic reflex arc, which includes trigeminal afferents, brainstem connections between the sensory part of the trigeminal nucleus, the motor nucleus of the facial nerve, the facial nerve proper, and the orbicularis oculi muscle (Figure 5). The R2 component has polysynaptic connections within the brainstem but has the same afferent/efferent pathways as R1. BR is useful in the study of trigeminal and facial nerve lesions, peripheral neuropathy, posterior fossa lesions, multiple sclerosis, and extrapyramidal diseases [51]. Under general anesthesia in the operating room, only the R1 response can be recorded [52–55].

**Figure 5.** Blink reflex and laryngeal adductor reflex. Panel (A) shows exemplary traces of the R1 response of the blink reflex (BR) (A1) and the R1 and R2 components of the laryngeal adductor reflex (LAR) (A2) intraoperatively recorded under general anesthesia. Panel (B) illustration

represents the setup used to stimulate and record the BR (top scheme, green lines) and the LAR (bottom scheme, purple lines), including a schematic representation of the arc reflex pathways (please, note that discontinued lines are used for afferent fibers and continued lines for efferent pathways), and the brainstem structures involved. V1 is the first branch of the cranial nerve V; CN: cranial nerve; NA: ambiguous nucleus; NST: nucleus of the solitary tract; RLN: recurrent laryngeal nerve; SLN: superior laryngeal nerve; VF: vocal fold.

**Trigeminal-hypoglossal reflex.** Brainstem trigeminal-hypoglossal reflexes (THR), also known as jaw-tongue reflexes, coordinate the tongue's position in the mouth during chewing, swallowing, vocalization, and breathing. Recently, a novel methodology for obtaining jaw-opening THR of the brainstem under general anesthesia has been described. This technique could be helpful for the intraoperative monitoring of surgeries involving the trigeminal, hypoglossal nerves, and lower brainstem lesions, however, further studies are still required to validate it [56–60].

**H reflex in the masseter muscle.** The H reflex in the masseter muscle is a monosynaptic trigeminal-trigeminal reflex transmitted through the mesencephalic nucleus of the trigeminal nerve. It reflects conduction through the midbrain and the median pons [61–63]. It can be helpful in studying patients with brainstem or trigeminal lesions, and can be obtained under general anesthesia, thus representing a new method for intraoperative monitoring, especially for lesions involving the midbrain and pons. Together with the BR, the H reflex is a straightforward method for intraoperative monitoring of brainstem reflex circuits and the functional integrity of the trigeminal and facial nerves. Nevertheless, yet more studies are necessary to demonstrate their usefulness [64].

**Laryngeal adductor reflex.** Recording of the laryngeal adductor reflex (LAR), a brainstem reflex entirely governed by the vagus nerve, has been introduced to monitor the vagus and laryngeal nerves in thyroid surgery [65,66]. Preservation of the reflex prevents potentially harmful substances from entering the tracheobronchial tree by activating the adductor muscles of the larynx to close both vocal folds [67,68]. Recently, a study was published using this reflex in surgeries around the brainstem to monitor cranial nerve X and related intrinsic brainstem pathways (**Figure 5**), showing that IONM with continuous LAR monitoring, together with vocal-CoMEPs, may be useful to reduce swallowing and laryngeal complications, such as aspiration and pneumonia, enhancing safety in resecting complex PF lesions [69,70].

### Neurophysiological Limitations in the Developing Central Nervous System

Neurophysiological test references change during the maturational process of the CNS and, therefore, exhibit some peculiarities depending on the patient's age. In term newborns, the most reproducible waves in BAEPs are I, III, and V [71]. However, unlike adults, it is normal for wave I to have a greater amplitude than complex IV/V. The myelin maturation process shortens the latency of the successive waves and the central conduction time (CCT or I-V interval). It has also been demonstrated that the auditory pathway responsible for generating the first and last components of BAEP responses matures at a different rate [72]. In a normal subject, wave I latency reaches the adult value around six months of life, while wave V does so by one year of life [71].

A similar process occurs in SEPs, the maturation of peripheral segments of the sensory pathway progresses more rapidly than that of central segments. N9 latency—brachial plexus potential—shortens during the first year of life and subsequently increases as the patient's size increases. The N9-N11 interval—conduction between the brachial plexus and the dorsal column—decreases with age. In contrast, the N13-N20 (CCT) presents a rapid shortening during the first year of life, reaching adult characteristics at four years of age [73].

Independent of the changes described, the motor stimulation threshold is higher in children than in adults under 18 years of age [74]. CMCT is a dependent value of CNS maturation but has different times. For example, with relaxation (resting motor threshold), children reach adult values approximately at the age of ten, however, with facilitation (active motor threshold), they reach normal values at two years of age [74,75]. Also, MEPs could be obtained at a resting motor threshold

in the upper limb after the first year of life and in the lower limb after four years. Maturation also affects the duration, morphology, and amplitude of MEPs.

When evaluating peripheral nerves in children, the myelination process also affects parameters commonly used for assessing NCS [76]. For instance, conduction velocity increases in proportion to nerve fiber diameters and the distance between the nodes of Ranvier, achieving faster motor and sensitive velocities with growth; distal latencies and amplitudes also change. For example, the CMAP increases threefold in the upper limbs and twofold in the lower limbs [76].

The LAR in children is vital for airway protection. In humans, the larynx descends from the neck at 4-6 months. This descent necessitates a change in the airway protection mechanism from a pattern in which an upright epiglottis in the newborn is grasped by an intact palatopharyngeal sphincter to one in which the epiglottis folds down over the aditus to the larynx and the closing vocal folds. This also coincides with the transition from obligate nasal breathing in early infancy to the adult pattern of facultative nasal or oral breathing. It has been suggested that difficulties in adjusting to this transition may be a factor in sudden infant death syndrome, which peaks in incidence at approximately the same age that laryngeal descent is complete (4–6 months) [77,78]. In theory, as of six months of life, we should record the LAR (either in the clinic or the operating theatre), although it may be necessary to do so with hook wire electrodes if there is no age-appropriate endotracheal tube [79,80].

A discussion of the changes from children to adults is beyond the scope of this paper. However, it is essential to bear in mind that neurophysiological tests have some peculiarities according to age, basically due to myelin maturation, which must be considered when evaluating children.

### Follow-Up Neurophysiological Studies in Patients with CM1

There are many publications on the follow-up of CM1 patients, however, practically half of them refer to case reports or series with fewer than ten patients (**Table 1**). Here, we summarize the most relevant information.

#### *Brainstem Auditory Evoked Potentials*

To our knowledge, the first paper was published in 1983 by Stone et al. [81], describing BAEP findings in a 16-year-old boy with CM2 and symptoms suggestive of brainstem involvement and neuroimaging revealing brainstem compression. The authors diagnosed CM2, however, as the reported patient did not have any visible spinal lesion, we believe that CM1.5 was the most likely diagnosis [13]. BAEPs were abnormally bilaterally, with I-V interpeak latency (IPL) prolongations and no visible wave III identified upon stimulation of the right ear. The patient underwent bilateral posterior fossa decompression (PFD) and cervical laminectomy. Six months later, he presented neurological improvement and bilateral normalization of his BAEPs. A few authors have also described case reports with retrocochlear findings in the BAEPs of CM1 patients, in both adults [82,83] and children [84,85] (Table 1).

In 1999, Hort-Legrand and Emery [86] reported 79 patients with syringomyelia, of which 48 cases were associated with CM1. BAEPs were abnormal in 13 of the 59 patients studied, and the most frequent finding was prolongation of the I-V IPL, more often unilaterally. Another study on syringomyelia and CM1 [87] found that BAEP abnormalities had a better correlation with clinical and radiological findings than SEPs.

In 2006, Henriques Filho and Pratesi used BAEPs to study 75 CM1 and CM2 patients [88]. Abnormalities were detected in most cases (71%); however, more were observed in CM2 patients. According to these authors, these findings reinforce the concept that pontine alterations are a consequence of brainstem dysgenesis in CM2. They also observed that patients with CM1 preferentially had peripheral alterations—cochlea or cochlear nerve close to the cochlea—with less in the area located between the cochlear nerve and the pontomedullary junction. This study concludes that the assessment of the V/I amplitude ratio, together with other abnormalities, allows the identification of a more significant number of BAEP alterations in patients with CM1 and is a valuable tool for the diagnosis. Vidmer et al. [89] in 2011, described BAEP findings in a single

pediatric patient with CM1 (a nine-year-old girl), in whom the abnormalities were located at the peripheral or cochlear level, supporting the results of Henriques Filho and Pratesi [88].

In studies carried out by our group—the latest including 200 patients—we found that BAEPs were altered in 38.5% of patients with CM1 [19,20]. The most common finding was increased I–V interpeak latency and increased wave V latency (31%). The logistic regression model used to predict the probability of an abnormal BAEP value showed that age, the degree of tonsillar ectopia, and the clinical detection of lower cranial nerve dysfunction had a statistically significant influence on predicting abnormal BAEPs. This finding could be explained by the fact that a more severe distortion of brainstem structures induces more BAEP abnormalities. Therefore, patients with a greater degree of tonsillar ectopia or lower cranial nerve dysfunction had a higher frequency of abnormal BAEPs.

Di Stefano et al. studied BAEPs and MRI in three groups of patients: with intracranial hypotension (n = 18), CM1 without intracranial hypotension (n = 18), and sensorineural hearing loss (n = 20) vs. controls (n = 52). In the CM1 group, they found abnormal BAEPs in 33% of patients, the main finding being a prolonged latency of wave V and I-III and III-V intervals [90], comparable results to those obtained by our group [19,20].

### *Somatosensory Evoked Potentials*

In 1986, Anderson et al. [91] described the SEP alterations in nine patients with syringomyelia, eight of whom had an associated CM1. The most frequent findings were unilateral or bilateral reduction or absence of the cervical potential. In all but one patient with CM1 and syringomyelia, a prolonged or asymmetric CCT was found; in 1988, Forcadas et al. reported similar results [92]. In 1990, Jabbari et al. published a study with SEPs from the MN and PTN in 22 patients with syringomyelia, four with CM. Three of the four patients with CM1 and syringomyelia had normal SEPs [93]. Restuccia and Mauguère [39] studied the MN SEPs of 24 patients with syringomyelia, 16 associated with CM1. They observed several anomalies, the most frequent being an alteration or absence of the N13 wave in 83% of patients with cervical syringomyelia. When syringomyelia was associated with CM, these authors, in contrast to Jabbari et al. [93], observed an increase in the P14-N20 interval; Morioka et al. reported similar results [94]. Several case reports and small series with SEP investigations in CM1 patients show abnormal SEPs at a central level, with an absence of cortical responses and an increased latency of cortical potentials or CCT [89,95–97].

Moncho et al. found that SEPs were altered in 43.5% of CM1 patients. The most frequent alteration in MN SEPs was an increase in N13-N20 IPL, whereas the most frequent alteration in PTN SEPs was an increase in N22-P37 IPL, sometimes associated with an alteration of the cervical potential. In a logistic regression model, only age and degree of tonsillar herniation were statistically significant in predicting abnormal SEPs. These findings indicated a greater probability of finding abnormal SEPs in older patients with greater tonsillar descent [19,20]; recently, Guvenc et al. described similar findings [98].

Some studies specify the relation between idiopathic scoliosis and CM1, in which abnormal SEPs can be due to tonsillar ectopia or syringomyelia rather than spinal deformity [95,99]. Cheng et al. found a relationship between tonsillar ectopia and SEP alterations when studying 164 patients with idiopathic scoliosis, 12 of whom had associated CM1 [99]. Utzig et al. reported the case of a 15-year-old girl complaining of intense headache, recurrent left-sided paresthesia, and progressing scoliosis, who had CM1 with cervical syringomyelia. Ulnar and PTN SEPs showed an absence of cortical responses that improved after surgical treatment with PFD, C1 laminectomy, and partial removal of the cerebellar tonsils with the reappearance of cortical responses of acceptable latency, however with reduced amplitude for the left extremities [95].

The somatosensory tract in CM1 can also be evaluated by trigeminal SEPs from V3, which can present prolonged latency. Hort-Legrand and Emery found that trigeminal SEPs were much more sensitive than BAEPs, being altered when patients had bulbar symptoms [86].

Berciano et al. reported a CM1 patient who presented with sudden lancinating episodes of left cervicobrachial pain provoked by coughing fits, correlated with cervico-dorsal syringomyelia extending into the left posterolateral quadrant in axial sections passing through vertebral levels C7

and D1. Pre- and postoperative upper limb dermatomal SEPs (dSEPs) elicited by stimulation at levels C6, C7, and C8 showed N20 attenuation only after stimulation of the left fifth finger (left C8 level). Given the preservation of peripheral Sensory nerve action potentials (SNAPs) in both upper limbs, such a selective dSEP attenuation is probably caused by extension of the paracentral posterior horn cavity into the C8 posterior entry zone. Under such circumstances, dSEP is a useful method of evaluating this radicular semiology [100]. In 2015, Awai and Curt reported seven patients with syringomyelia showing differences in dSEP and CHEP affectations. Dermatome CHEPs were abnormal in all patients in at least one dermatome [101].

#### *Motor Evoked Potentials*

Few authors have described TMS MEP alterations in patients with CM1, most of them with syringomyelia. The most frequent findings were increased CMTCs [86,102–104].

Referring to pre- and post-surgical MEP evaluations, Cristante et al. [105] described the MEP findings in eight CM1 patients. Preoperative TMS MEPs showed that functionally impaired muscles exhibited neurophysiologic abnormalities, and even one patient without any motor deficit had abnormal MEPs. Interestingly, the postoperative functional motor recovery of five of the eight patients—mostly partial—was not reflected by a similar improvement in MEPs.

#### *NCS, EMG, and other Peripheral Nerve Studies*

In 1992, Gerard et al. described the case of a five-year-old girl—with CM1 and BI—admitted due to insufficiency of the soft palate [106]. EMG activity was recorded bilaterally in the levator palatini and anterior faucial pillars, showing ample biphasic or polyphasic action potentials at rest (probably spontaneous activity). When the child cried, the frequency of these potentials increased poorly, and recruitment was impaired. Clinical examination and EMG findings led to a suspicion of denervation of the IX, X, and XI cranial nerves.

CM1 should be a differential diagnosis in patients with adult dysphagia onset requiring MRI examination, even when presenting with “typical” lower motor neuron signs in bulbar muscles. SEPs, NCS, and EMG can help diagnose these cases [107].

In patients with CM1 and associated syringomyelia, NCS and EMG are useful in detecting anterior horn affection caused by the syringomyelia. This syndrome characteristically presents with spontaneous activity while performing needle EMG if acute or denervating activity occurs, with large, polyphasic (neurogenic) MUPs, reduced voluntary recruitment, and normal sensory nerve conduction [108–113]. Sometimes, this can be misdiagnosed as focal mononeuropathy if only motor NCSs are performed since a common finding is a small amplitude CMAP [108–110,112,113], although just one group reported unaffected CMAPs [111]. Another study found a confounding ulnar neuropathy with a tricky focal slowing at the elbow in a patient with cervical syringomyelia at C7-D1. A few studies reported a clinical foot drop syndrome in children, with neurogenic findings in most muscles that were dependent on the common peroneal nerve but also the PTN, and with normal sensory evaluation. The MRI confirmed the presence of holocord syrinx [110–114]. Therefore, it is essential to keep in mind that foot drop is a common problem that can present, and even though the most frequent cause is peroneal neuropathy, other causes include anterior horn cell disease, lumbar plexopathies, L5 radiculopathy, partial sciatic neuropathy, and, rarely, parasagittal lesion.

Painful cutaneous nerve stimulation can suppress EMG activity in voluntarily contracting muscle [115–119]. This phenomenon is designated the “cutaneous silent period” (CSP). In 1996, Kaneko et al. [120] studied five male patients (mean age 18 years) with cervical syringomyelia using CSPs, CMAPs, F waves, MEPs, and SEPs of upper limbs. MEP latency and amplitude were within normal limits in all patients. Three of the five patients presented absent or reduced cervical N13 potentials from SEPs of the upper limbs with preserved cortical responses. In the symptomatic hand, all patients exhibited reduced CSPs, constituting the only abnormal finding in two subjects. The laterality of the abnormal CSPs and the reduced N13 potentials of the SEPs were consistent with the site of the syrinx on MRI. Previous reports demonstrated that absent or reduced cervical N13 potentials with preserved scalp responses of upper limb SEPs are characteristic of early cervical

syringomyelia [39,91,92]. Kanebo et al. further demonstrated that the absence of CSPs with normal conduction in large, myelinated fibers that form motor and sensory pathways is characteristic of patients with cervical syringomyelia. In addition, the CSP decrease was more sensitive to syringomyelic changes than abnormal cervical N13 potentials were.

#### *Brainstem Reflexes*

We only detected two reports regarding CM1 follow-up with brainstem reflexes, specifically, the blink reflex. Amoiridis et al. [121] reported a case of a 25-year-old man with CM1 and holocord syrinx who experienced dysesthesia and weakness with urinary retention during shooting practice. The PTN SEPs showed no lumbar potential (N24; L1/iliac crest), whereas a high cervical potential (N33; C2/Fz) and a cortical potential (P40; Cz'/Fz) with normal latencies were registered. The MN SEP was normal bilaterally. The amplitude of the H reflex in the soleus muscle was low bilaterally, and H reflex latency was prolonged on the right. Motor and sensory NCS were normal. F waves in legs and arms were abolished. They performed the BR that showed R1 absent bilaterally and R2 latency with left-side stimulation prolonged on both sides. However, neurological examination of the cranial nerves was unremarkable. Jacome, in 2001, described four CM1 patients presenting with blepharoclonus, one with altered R1, two with altered R2, and one with normal BR. Also, facial EMG showed complex repetitive discharges of the right mentalis muscle in one patient [122].

#### **Intraoperative Neurophysiological Monitoring in CM1**

Most articles on this topic refer to neuromonitoring during PF surgeries. A few refer to neuromonitoring during the insertion of shunts for the treatment of syringomyelia. A third category comprises CM1 and scoliosis surgery, as well as studies reporting new techniques (**Table 2**).

##### **Posterior fossa surgery**

The role of intraoperative neuromonitoring (IONM) during PF surgery for CM1 is controversial, and there is still no consensus on the usefulness of the technique for the prevention of new neurological deficits nor on the most effective modality to use. Anderson et al. studied changes in intraoperative BAEPs and SEPs in 11 pediatric patients undergoing suboccipital decompression for CM1. BAEP conduction times were compared at four different surgical stages: before positioning, after positioning, after craniotomy, and after durotomy. Their data indicated that significant improvement in conduction times occurs after bony decompression and division of the atlantooccipital membrane rather than after dura opening. They also highlighted the risk of altering SEPs and BAEPs during positioning [123]. Zamel et al. reviewed the records of 80 pediatric patients, with or without syringomyelia, who underwent suboccipital decompression to treat CM1. They divided patients into two groups according to whether or not the dura mater was opened. Their data suggest that PFD with bone removal alone significantly improves BAEP conduction times in most pediatric patients with CM1 and that using duraplasty led to a further, albeit slight, improvement in conduction times in only 20% of patients [124]. Kim et al. [125] performed an alternate strategy for treating 11 symptomatic children presenting with basilar invagination associated with CM1. Their technique involved suboccipital decompression, C1 laminectomy, and duraplasty to treat CM1 and subsequent intraoperative manual reduction using accentuated cervical distraction and extension. Their results showed that immediately after reduction and fusion, SEPs improved significantly in 10 patients, while remaining unchanged from baseline in one.

In 2012, Chen et al. analyzed BAEP and SEP parameters in 13 consecutive pediatric patients who underwent suboccipital decompression to treat symptomatic CM1. They recorded the MN SEP, PTN SEP, and BAEP latencies at four stages: preoperatively, following craniotomy, following durotomy, and following closure [126]. In contrast to that reported by Anderson et al. [123] and Zamel et al. [124], they demonstrated clinically notable improvements in overall latency times in both MN SEP N20 and BAEP wave V, not only after bony decompression but also after duraplasty. Since their results showed more significant improvements in MN SEPs, they consider them the most sensitive for evaluating sufficient decompression. Incorporating an additional replacement of the bone flap

retained the improvements in MN SEP and BAEPs, suggesting that this technique does not compromise the decompressive effect of the surgery.

The largest pediatric population series—with 156 patients—was reviewed in 2015 by Kennedy et al., in children with PFD but not dural opening [127]. IONM with SEPs and BAEPs was performed before and after prone positioning and during surgery. They found that 78% of patients exhibited significant BAEP conduction time improvement after bony decompression. SEPs were used to assess the lack of problems with patient positioning during surgery.

The first study evaluating the efficacy of multimodality IONM using both TcMEPs and SEPs during PFD in 21 pediatric patients was presented in 2016 by Barzilai et al. [128]. They found significant signal changes in three patients, all concomitant with patient positioning on the surgical table: in two cases, the signals recovered at closure, whereas in one case, SEPs remained attenuated even after repositioning. No new neurological deficits were observed post-operatively. They concluded that IONM could be useful during patient positioning in PFD surgery.

In 2019, Rasul et al. also analyzed IONM data with SEPs and BAEPs at two time points—baseline before skin incision and final at the time of skin closure—in 37 symptomatic patients who underwent PFD for CM1. They found significant SEP latency reduction in all patients. As for BAEPs, 13 patients had a reduction in their conduction time [129]. However, they could not establish a definite link between clinical outcomes and IONM changes, unlike other authors [130].

In 2020, a case report published by Krishnakumar et al. [131] evaluated the use of IONM to identify neurological safety during critical surgical steps. They reported a 13-year-old patient diagnosed with atlantooccipital dislocation with BI, hypoplastic C1 arch with CM1, and thoracic and lumbar scoliosis. Loss of TcMEPs was recorded during positioning and screw fixation, recognizing the need for a new surgical strategy. Changes in TcMEPs were reverted, and even an increase in amplitude was recorded at the end of surgery compared with baseline. During the three-month follow-up, the patient showed no new neurological deficit.

Contrary to most studies presented in the pediatric population—in which a beneficial role of IONM is suggested—the literature regarding the adult population is more skeptical as to the benefits of IONM during PFD for CM1 treatment. The results of these studies are more heterogeneous than those in pediatric patients; however, most are single case reports and, therefore, represent anecdotal evidence.

Grossauer et al. [132] published the case of a 32-year-old woman who underwent surgery for CM1 associated with extensive cervicothoracic syringomyelia and whose SEPs did not notably improve following craniotomy or durotomy. Instead, they only improved after opening the fourth ventricle and restoring CSF flow at the level of the craniocervical junction.

Roser et al. [133] presented IONM data from a small series of 39 consecutive adult patients (84.6% presenting with syringomyelia) who underwent suboccipital decompression and duraplasty for the treatment of symptomatic CM1. SEPs and TcMEPs were monitored in all patients, reporting significant changes in both SEPs and TcMEPs in only two cases related to patient positioning. They found no significant change in SEP and TcMEP amplitudes or latencies between baseline and final measurements.

Heffez et al. examined the relationship between the extent of tonsillar ectopia and intraoperative changes in BAEP conduction times in an extensive series of adult CM1 cases [134]. They divided the patients into four groups depending on the position of the cerebellar tonsil. They noted a reduction of > 0.1 ms in conduction times in 35 - 49% of patients with no statistical difference between groups or clinical outcomes. This suggests that, even with minimal tonsillar herniation, a disruption in the function of at least one pathway within the brainstem may occur and that this disturbance is improved by brainstem decompression.

In 2022, Schaefer et al. [135] suggested that CM1 suboccipital decompression surgery may be performed safely without IONM after evaluating SEPs, TcMEPs, and/or BAEPs in 93 adult patients, as they found only one instance of a transient decrease in TcMEPs, which resolved without intervention.

Some articles describe MEPs or SEPs during IONM in CM1 patients treated with other surgical techniques, like atlantoaxial stabilization surgery or endoscopic suboccipital decompression, with inconclusive results [136,137] (Table 2).

#### **Surgery for the direct treatment of syringomyelia in MC1**

The first articles about IONM for syringomyelia in CM1 patients were published by Milhorat et al., with 32 syringomyelia patients (21 with CM1). IONM with MN SEPs demonstrated a significant decrease in N20 latency and a nonsignificant increase in N20 amplitude 30 minutes after syrinx decompression. They concluded that the improvement in N20 latency was indirect evidence of preexisting long tract compression. However, these conclusions are questionable in patients with CM1, in whom SEP improvements may have been caused, in part, by previous PF surgery [138,139].

Pencovich et al., in 2013, were the first to address the benefits of multimodality IONM during syringomyelia surgery. They monitored SEPs and MEPs in 13 patients with syringomyelia, six of whom also had CM1. One of the CM1 patients presented severe attenuation of left leg MEP data noted upon the midline approach to the syrinx. The catheter was subsequently removed and PFD was completed without syrinx drainage. IONM might be a valuable adjunct to spinal cord surgeries for the treatment of syringomyelia, supporting the routine use of multimodal IONM in this kind of surgery [140,141].

#### **CM1 in scoliosis surgery**

The association between Chiari malformation syrinx and scoliosis has been widely documented during the past few decades [99,142–144]. The concomitant rate of scoliosis ranges from 15 to 50% in patients with Chiari malformation [145–147]. There is a potential risk from scoliosis surgery in patients with syringomyelia-associated scoliosis, which might be higher with longer and wider syrinxes. We found two articles with different conclusions concerning IONM during scoliosis surgery in patients with CM1. Tan et al. [148] compared radiographic features, clinical outcomes, and surgical complications after posterior spinal fusion between 21 patients with CM1 and syringomyelia-associated scoliosis and 21 with idiopathic syringomyelia-associated scoliosis. As for the IONM data, TcMEPs were implemented in all patients. They noted no differences in the successful recording of baseline TcMEPs or evident TcMEP deterioration between the two groups. Patients with CM-associated scoliosis had longer syrinxes than the syringomyelia-associated scoliosis group; however, their preoperative neurological status and IONM signals during the operation were similar. In addition, no progression of the original neurological deficits or neurological complications were detected in either group. Shi et al. [149] studied 60 neurologically asymptomatic CM-associated scoliosis (CMS) patients (48 presented with syringomyelia) and 210 idiopathic scoliosis (IS) patients. CMS patients showed similar absolute values of IONM, including SEPs latency and amplitude and MEPs amplitude compared to IS patients, however, the syringomyelia in CMS patients indicated a more severe curvature and lower SEP amplitude, even though these patients had previously undergone PFD.

#### **Exploratory Research on the Subject**

Two articles stand out in this section. In the first, Brînzeu and Sindou carried out a research study on the functional anatomy of the accessory nerve (XI CN) via IONM (mapping) [150]. Forty-nine patients undergoing surgery with intradural exposure at the craniocervical junction—22 CM1 patients—were enrolled in the study. The authors concluded that the function of each of the XI CN rootlets appears to be specific. Thus, the cranial root contributes, independently of the spinal root, to the innervation of the vocal folds, which makes it a specific entity. The spinal root innervates the sternocleidomastoid and trapezius muscles with a craniocaudal motor organization of its cervical rootlets. Giampiccolo et al. conducted an exploratory and preliminary study on cerebello-cortical stimulation in 10 patients undergoing PF surgery, one of them with CM1. A third of children undergoing cerebellar resections can present cerebellar mutism. According to recent evidence, it is suggested that this may arise from damage to cerebellar efferents, either uni- or bilaterally, affecting the cortex along the cerebello-dento-thalamo-cortical pathway. There is currently no neurophysiological procedure available to intraoperatively monitor this pathway. This study has

promising implications for pediatric PF surgery to preserve cerebello-cortical paths and thus prevent cerebellar mutism [151].

## Conclusions

Many published articles deal with the techniques performed in the neurophysiology laboratory in CM1 patients, however, most are of limited usefulness as they focus on heterogeneous, non-systematized series or case reports, where only the most affected patients (most with syringomyelia) are investigated. Most studies are about SEPs, probably due to the frequent association of CM1 with syringomyelia. The most common SEP alteration (both MN and PTN) was increased CCT, occasionally associated with altering the cervical potential. We did not find any publication regarding LEPs and only one about CHEPs and CM1 with syringomyelia, although, as we mentioned previously, the sensibility of these tests is superior to classic SEPs in detecting pure spinothalamic lesions. BAEPs are the second most frequently studied modality in the literature, with the most common finding being an increased I–V interval, suggesting central or retrocochlear involvement. MEPs are the third most frequently studied parameter, and the most commonly observed finding is an increased CMCT. Regarding EMG, most results indicate major or minor denervation, confirming the presence of an anterior horn cell lesion when patients have syringomyelia.

Our view is that the utility of neurophysiological studies in patients with CM1 depends on the clinical context. Thus, when there is apparent symptomatology, our findings suggest that neurophysiological explorations do not add clinically relevant information, nor are they useful to establish which patients should undergo surgical treatment. Hence, it is not necessary to include them in the routine study. Nevertheless, an important finding in our series was that half of the patients in whom MC1 was discovered incidentally had abnormal evoked potentials, establishing objective evidence of subclinical dysfunction. Therefore, this is useful in the follow-up of patients whose symptoms have apparently stabilized and who require follow-up to define further the subclinical progression.

Concerning PF surgery IONM, the published studies we found were also heterogeneous series, not only as regards patient characteristics (such as different age ranges, with or without syringomyelia, and CVJ malformations) but also the type of neurophysiologic modality used for monitoring. Most authors used SEPs, with BAEPs or MEPs having been used less frequently. Surprisingly, we did not find any publication on the control of cranial nerves by reflexes (such as BR, LAR), Co-MEP, or by evaluating free-EMG activity. Since the lower cranial nerves can be affected by this pathology and may be at risk in certain patients in whom tonsillar coagulation or resection are required or in patients at high risk from surgery, such as patients with CM1.5, with the currently available equipment, when intraoperative monitoring is decided, the goal is multimodal IONM including monitoring of the lower cranial nerves. Some authors have proposed IONM during surgery in patients with CM1 in the following three scenarios: 1) during patient placement before surgery, 2) to determine when adequate decompression has been achieved and, therefore, to design surgery on an individualized basis, and 3) to detect intraoperative evoked potential worsening when intradural manipulation is planned. For surgical positioning in patients with CM1 (scenario 1), the rationale for IONM is that it can reduce the risk of neurological injury while it can still be reversed, particularly in patients with CM1.5 in whom retrocurved odontoids and partial anterior compression of the cervico-medullary junction are present. In scenario 2, the most controversial aspect is, in our opinion, the use of IONM to limit the degree of PFD and guide the decision of whether or not to open the dura mater. Analyzing some series that compare MRI studies of CM1 pediatric patients treated with an extradural procedure and others undergoing duraplasty, the normalization rate of tonsillar descent is seen to be clearly higher in the group of patients treated with duraplasty [124,152,153]. Given these results, we cannot consider that optimal treatment can be decided based on IONM findings. Regarding the use of IONM for monitoring neurological worsening during surgery (scenario 3), none of the reports reviewed concerning CM1—without associated complex CVJ malformations—showed any significant worsening during surgery that would have prompted the surgical team to modify their surgical strategy. These findings confirm that the incidence of neurological complications in the PFD

surgery of CM1 patients without complex CVJ malformations is very low once the patient has been positioned.

In conclusion, the evidence to date shows that the only potential benefit of IONM in CM1 is during patient positioning and only in certain patients (CM1.5), making the routine use of IONM difficult to justify, at least with the IONM modalities studied so far.

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## References

1. Cleland, null Contribution to the Study of Spina Bifida, Encephalocele, and Anencephalus. *J Anat Physiol* **1883**, *17*, 257–292.
2. Chiari H Über Veränderungen Des Kleinhirns Infolge von Hydrocephalie Des Grosshirns. *Dtsch Med Wochenschr* **1891**, *17*, 1172–1175.
3. Chiari H Über veränderungen des Kleinhirns, des Pons un der Medulla Oblongata in folge von congenitaler Hydrocephalie des Grosshirns. *Denkschr Akad Wiss Wien* **1896**, *63*, 71–116.
4. Chiari, H. Concerning Alterations in the Cerebellum Resulting from Cerebral Hydrocephalus. 1891. *Pediatr Neurosci* **1987**, *13*, 3–8.
5. Cruveilhier J *L'anatomie Pathologique Du Corps Humain; Descriptions Avec Figures Lithographiées et Coloriées; Diverses Altérations Morbides Dont Le Corps Humain et Susceptible*; Bailliere: Paris, 1829;
6. Mortazavi, M.M.; Tubbs, R.S.; Brockerhoff, M.A.; Loukas, M.; Oakes, W.J. The First Description of Chiari I Malformation with Intuitive Correlation between Tonsillar Ectopia and Syringomyelia. *J Neurosurg Pediatr* **2011**, *7*, 257–260, doi:10.3171/2010.12.PEDS10579.
7. Barkovich, A.J.; Wippold, F.J.; Sherman, J.L.; Citrin, C.M. Significance of Cerebellar Tonsillar Position on MR. *AJNR Am J Neuroradiol* **1986**, *7*, 795–799.
8. Bogdanov, E.I.; Heiss, J.D.; Mendelevich, E.G.; Mikhaylov, I.M.; Haass, A. Clinical and Neuroimaging Features of "Idiopathic" Syringomyelia. *Neurology* **2004**, *62*, 791–794.
9. Botelho, R.V.; Bittencourt, L.R.A.; Rotta, J.M.; Tufik, S. A Prospective Controlled Study of Sleep Respiratory Events in Patients with Craniovertebral Junction Malformation. *J. Neurosurg.* **2003**, *99*, 1004–1009, doi:10.3171/jns.2003.99.6.1004.
10. Meadows, J.; Kraut, M.; Guarnieri, M.; Haroun, R.I.; Carson, B.S. Asymptomatic Chiari Type I Malformations Identified on Magnetic Resonance Imaging. *J. Neurosurg.* **2000**, *92*, 920–926, doi:10.3171/jns.2000.92.6.0920.

11. Milhorat, T.H.; Nishikawa, M.; Kula, R.W.; Dlugacz, Y.D. Mechanisms of Cerebellar Tonsil Herniation in Patients with Chiari Malformations as Guide to Clinical Management. *Acta Neurochir (Wien)* **2010**, *152*, 1117–1127, doi:10.1007/s00701-010-0636-3.
12. Sekula, R.F.; Jannetta, P.J.; Casey, K.F.; Marchan, E.M.; Sekula, L.K.; McCrady, C.S. Dimensions of the Posterior Fossa in Patients Symptomatic for Chiari I Malformation but without Cerebellar Tonsillar Descent. *Cerebrospinal Fluid Res* **2005**, *2*, 11, doi:10.1186/1743-8454-2-11.
13. Tubbs, R.S.; Iskandar, B.J.; Bartolucci, A.A.; Oakes, W.J. A Critical Analysis of the Chiari 1.5 Malformation. *J. Neurosurg.* **2004**, *101*, 179–183, doi:10.3171/ped.2004.101.2.0179.
14. Tubbs, R.S.; Demerdash, A.; Vahedi, P.; Griessenauer, C.J.; Oakes, W.J. Chiari IV Malformation: Correcting an over One Century Long Historical Error. *Childs Nerv Syst* **2015**, doi:10.1007/s00381-015-2765-8.
15. Ciaramitaro, P.; Massimi, L.; Bertuccio, A.; Solari, A.; Farinotti, M.; Peretta, P.; Saletti, V.; Chiapparini, L.; Barbanera, A.; Garbossa, D.; et al. Diagnosis and Treatment of Chiari Malformation and Syringomyelia in Adults: International Consensus Document. *Neurol Sci* **2022**, *43*, 1327–1342, doi:10.1007/s10072-021-05347-3.
16. Isik, N.; Elmaci, I.; Kaksi, M.; Gokben, B.; Isik, N.; Celik, M. A New Entity: Chiari Zero Malformation and Its Surgical Method. *Turk Neurosurg* **2011**, *21*, 264–268, doi:10.5137/1019-5149.JTN.2705-09.1.
17. Kyoshima, K.; Kuroyanagi, T.; Oya, F.; Kamijo, Y.; El-Noamany, H.; Kobayashi, S. Syringomyelia without Hindbrain Herniation: Tight Cisterna Magna. Report of Four Cases and a Review of the Literature. *J. Neurosurg.* **2002**, *96*, 239–249.
18. Tubbs, R.S.; Elton, S.; Grabb, P.; Dockery, S.E.; Bartolucci, A.A.; Oakes, W.J. Analysis of the Posterior Fossa in Children with the Chiari 0 Malformation. *Neurosurgery* **2001**, *48*, 1050–1054; discussion 1054–1055.
19. Moncho, D.; Poca, M.A.; Minoves, T.; Ferré, A.; Cañas, V.; Sahuquillo, J. Are Evoked Potentials Clinically Useful in the Study of Patients with Chiari Malformation Type 1? *J. Neurosurg.* **2016**, 1–14, doi:10.3171/2015.11.JNS151764.
20. Moncho, D.; Poca, M.A.; Minoves, T.; Ferré, A.; Rahnama, K.; Sahuquillo, J. Brainstem Auditory and Somatosensory Evoked Potentials in Relation to Clinical and Neuroimaging Findings in Chiari Type 1 Malformation. *J Clin Neurophysiol* **2015**, *32*, 130–138, doi:10.1097/WNP.0000000000000141.
21. Ferré, Á.; Poca, M.A.; de la Calzada, M.D.; Moncho, D.; Romero, O.; Sampol, G.; Sahuquillo, J. Sleep-Related Breathing Disorders in Chiari Malformation Type 1: A Prospective Study of 90 Patients. *Sleep* **2017**, *40*, doi:10.1093/sleep/zsx069.
22. Ferré, Á.; Poca, M.A.; de la Calzada, M.D.; Moncho, D.; Urbizu, A.; Romero, O.; Sampol, G.; Sahuquillo, J. A Conditional Inference Tree Model for Predicting Sleep-Related Breathing Disorders in Patients With Chiari Malformation Type 1: Description and External Validation. *J Clin Sleep Med* **2019**, *15*, 89–99, doi:10.5664/jcsm.7578.
23. Moncho, D.; Poca, M.A.; Minoves, T.; Ferré, A.; Rahnama, K.; Sahuquillo, J. [Brainstem auditory evoked potentials and somatosensory evoked potentials in Chiari malformation]. *Rev Neurol* **2013**, *56*, 623–634.
24. Sahuquillo, J.; Moncho, D.; Ferré, A.; López-Bermeo, D.; Sahuquillo-Muxi, A.; Poca, M.A. A Critical Update of the Classification of Chiari and Chiari-like Malformations. *Journal of Clinical Medicine* **2023**, *12*, 4626, doi:10.3390/jcm12144626.
25. Ferré Masó, A.; Poca, M.A.; de la Calzada, M.D.; Solana, E.; Romero Tomás, O.; Sahuquillo, J. Sleep Disturbance: A Forgotten Syndrome in Patients with Chiari I Malformation. *Neurologia* **2011**, doi:10.1016/j.nrl.2011.01.008.
26. Ferré, Á.; Poca, M.A.; de la Calzada, M.D.; Moncho, D.; Romero, O.; Sampol, G.; Sahuquillo, J. Sleep-Related Breathing Disorders in Chiari Malformation Type 1. A Prospective Study of 90 Patients. *Sleep* **2017**, doi:10.1093/sleep/zsx069.
27. Møller, A.R. Monitoring Auditory Evoked Potentials. In *Intraoperative Neurophysiological Monitoring*; Humana Press: Totowa, New Jersey, 2006; pp. 85–124 ISBN 978-1-59745-018-8.
28. Guérit, Jean-Michel *Les potentiels évoqués*; 2e éd.; Masson (Paris), 1993; Vol. 1 vol; ISBN 2-225-84107-1.
29. Desmedt, J.E. [Physiology and Physiopathology of Somatic Sensations Studied in Man by the Method of Evoked Potentials]. *J. Physiol. (Paris)* **1987**, *82*, 64–136.
30. Cracco, R.Q.; Cracco, J.B. Somatosensory Evoked Potential in Man: Far Field Potentials. *Electroencephalogr Clin Neurophysiol* **1976**, *41*, 460–466.
31. Cruccu, G.; Aminoff, M.J.; Curio, G.; Guerit, J.M.; Kakigi, R.; Mauguiere, F.; Rossini, P.M.; Treede, R.-D.; Garcia-Larrea, L. Recommendations for the Clinical Use of Somatosensory-Evoked Potentials. *Clin Neurophysiol* **2008**, *119*, 1705–1719, doi:10.1016/j.clinph.2008.03.016.
32. Cruccu, G.; Aminoff, M.J.; Curio, G.; Guerit, J.M.; Kakigi, R.; Mauguiere, F.; Rossini, P.M.; Treede, R.-D.; Garcia-Larrea, L. Recommendations for the Clinical Use of Somatosensory-Evoked Potentials. *Clin Neurophysiol* **2008**, *119*, 1705–1719, doi:10.1016/j.clinph.2008.03.016.

33. Nair, D.; Kumaraswamy, V.M.; Braver, D.; Kilbride, R.D.; Borges, L.F.; Simon, M.V. Dorsal Column Mapping via Phase Reversal Method: The Refined Technique and Clinical Applications. *Neurosurgery* **2014**, *74*, 437–446; discussion 446, doi:10.1227/NEU.0000000000000287.
34. Simon, M.V.; Chiappa, K.H.; Borges, L.F. Phase Reversal of Somatosensory Evoked Potentials Triggered by Gracilis Tract Stimulation: Case Report of a New Technique for Neurophysiologic Dorsal Column Mapping. *Neurosurgery* **2012**, *70*, E783–788, doi:10.1227/NEU.0b013e31822e0a76.
35. Cruccu, G.; García-Larrea, L. Clinical Utility of Pain--Laser Evoked Potentials. *Suppl Clin Neurophysiol* **2004**, *57*, 101–110, doi:10.1016/s1567-424x(09)70348-4.
36. Garcia-Larrea L Pain Evoked Potentials. In *Handbook of neurology*; Elsevier: Amsterdam, 2006; Vol. 81, pp. 439–462.
37. Cruccu, G.; Anand, P.; Attal, N.; Garcia-Larrea, L.; Haanpää, M.; Jørum, E.; Serra, J.; Jensen, T.S. EFNS Guidelines on Neuropathic Pain Assessment. *Eur J Neurol* **2004**, *11*, 153–162, doi:10.1111/j.1468-1331.2004.00791.x.
38. Kakigi, R.; Inui, K.; Tamura, Y. Electrophysiological Studies on Human Pain Perception. *Clin Neurophysiol* **2005**, *116*, 743–763, doi:10.1016/j.clinph.2004.11.016.
39. Restuccia, D.; Mauguière, F. The Contribution of Median Nerve SEPs in the Functional Assessment of the Cervical Spinal Cord in Syringomyelia. A Study of 24 Patients. *Brain* **1991**, *114* ( Pt 1B), 361–379.
40. Dotson, R.M.; Sandroni, P. CONTACT HEAT EVOKED POTENTIALS. In *Clinical Neurophysiology*; Contemporary Neurology Series; OUP USA, 2009; pp. 688–692 ISBN 978-0-19-538511-3.
41. Leandri, M.; Marinelli, L.; Siri, A.; Pellegrino, L. Micropatterned Surface Electrode for Massive Selective Stimulation of Intraepidermal Nociceptive Fibres. *J Neurosci Methods* **2018**, *293*, 17–26, doi:10.1016/j.jneumeth.2017.08.032.
42. *Magnetic Stimulation in Clinical Neurophysiology*; Hallett, M., Chokroverty, S., Eds.; 2nd ed.; Elsevier Butterworth-Heinemann: Philadelphia, Pa, 2005; ISBN 978-0-7506-7373-0.
43. Claus, D. Central Motor Conduction: Method and Normal Results. *Muscle Nerve* **1990**, *13*, 1125–1132, doi:10.1002/mus.880131207.
44. Strommen, J.A. Motor Evoked Potentials. In *Clinical Neurophysiology*; Contemporary Neurology Series; OUP USA, 2009; pp. 385–397 ISBN 978-0-19-538511-3.
45. Kimura, J. Motor Evoked Potentials. In *Electrodiagnosis in Diseases of Nerve and Muscle: Principles and Practice*; Oxford University Press, 2013; pp. 526–551 ISBN 978-0-19-935316-3.
46. Deletis, V.; Fernández-Conejero, I. Intraoperative Monitoring and Mapping of the Functional Integrity of the Brainstem. *J Clin Neurol* **2016**, *12*, 262–273, doi:10.3988/jcn.2016.12.3.262.
47. Deletis, V. Intraoperative Neurophysiology of the Corticospinal Tract of the Spinal Cord. *Suppl Clin Neurophysiol* **2006**, *59*, 107–112, doi:10.1016/s1567-424x(09)70019-4.
48. Deletis, V.; Seidel, K.; Sala, F.; Raabe, A.; Chudy, D.; Beck, J.; Kothbauer, K.F. Intraoperative Identification of the Corticospinal Tract and Dorsal Column of the Spinal Cord by Electrical Stimulation. *J. Neurol. Neurosurg. Psychiatry* **2018**, doi:10.1136/jnnp-2017-317172.
49. Simon, M.V. *Intraoperative Clinical Neurophysiology: A Comprehensive Guide to Monitoring and Mapping*; Demos Medical: New York, 2010; ISBN 978-1-933864-46-4.
50. Kugelberg, E. [Facial reflexes]. *Brain* **1952**, *75*, 385–396, doi:10.1093/brain/75.3.385.
51. Smith, B.E. Cranial Reflexes and Related Techniques. In *Clinical Neurophysiology*; OUP USA, 2009; pp. 529–542 ISBN 978-0-19-538511-3.
52. Deletis, V.; Urriza, J.; Ulkatan, S.; Fernandez-Conejero, I.; Lesser, J.; Misita, D. The Feasibility of Recording Blink Reflexes under General Anesthesia. *Muscle Nerve* **2009**, *39*, 642–646, doi:10.1002/mus.21257.
53. Aydinlar, E.I.; Kocak, M.; Soykam, H.O.; Mat, B.; Dikmen, P.Y.; Sezerman, O.U.; Elmaci, İ.; Pamir, M.N. Intraoperative Neuromonitoring of Blink Reflex During Posterior Fossa Surgeries and Its Correlation With Clinical Outcome. *J Clin Neurophysiol* **2022**, *39*, 299–306, doi:10.1097/WNP.0000000000000777.
54. Fernández-Conejero, I.; Ulkatan, S.; Deletis, V. Chapter 10 - Monitoring Cerebellopontine Angle and Skull Base Surgeries. In *Handbook of Clinical Neurology*; Nuwer, M.R., MacDonald, D.B., Eds.; Intraoperative Neuromonitoring; Elsevier, 2022; Vol. 186, pp. 163–176.
55. Karakis, I.; Simon, M.V. Neurophysiologic Mapping and Monitoring of Cranial Nerves and Brainstem. In *Intraoperative Neurophysiology*; Simon, M.V., Ed.; Springer Publishing Company: New York, 2018; pp. 345–388 ISBN 978-1-62070-117-1.
56. Mirallave Pescador, A.; Téllez, M.J.; Sánchez Roldán, M. de L.Á.; Samusyte, G.; Lawson, E.C.; Coelho, P.; Lejarde, A.; Rathore, A.; Le, D.; Ulkatan, S. Methodology for Eliciting the Brainstem Trigeminal-Hypoglossal Reflex in Humans under General Anesthesia. *Clin Neurophysiol* **2022**, *137*, 1–10, doi:10.1016/j.clinph.2022.02.004.

57. Ishiwata, Y.; Ono, T.; Kuroda, T.; Nakamura, Y. Jaw-Tongue Reflex: Afferents, Central Pathways, and Synaptic Potentials in Hypoglossal Motoneurons in the Cat. *J Dent Res* **2000**, *79*, 1626–1634, doi:10.1177/00220345000790081701.
58. Lowe, A.A. The Neural Regulation of Tongue Movements. *Prog Neurobiol* **1980**, *15*, 295–344, doi:10.1016/0301-0082(80)90008-8.
59. Morimoto, T.; Kawamura, Y. Properties of Tongue and Jaw Movements Elicited by Stimulation of the Orbital Gyrus in the Cat. *Arch Oral Biol* **1973**, *18*, 361–372, doi:10.1016/0003-9969(73)90160-x.
60. Luo, P.; Zhang, J.; Yang, R.; Pendlebury, W. Neuronal Circuitry and Synaptic Organization of Trigeminal Proprioceptive Afferents Mediating Tongue Movement and Jaw-Tongue Coordination via Hypoglossal Premotor Neurons. *Eur J Neurosci* **2006**, *23*, 3269–3283, doi:10.1111/j.1460-9568.2006.04858.x.
61. Godaux, E.; Desmedt, J.E. Human Masseter Muscle: H- and Tendon Reflexes. Their Paradoxical Potentiation by Muscle Vibration. *Arch Neurol* **1975**, *32*, 229–234, doi:10.1001/archneur.1975.00490460045005.
62. Kimura, J. H, T, and Masseter Reflexes and the Silent Period. In *Electrodiagnosis in Diseases of Nerve and Muscle: Principles and Practice*; Oxford University Press, 2013; pp. 216–218 ISBN 978-0-19-935316-3.
63. Szentagothai, J. Anatomical Considerations on Monosynaptic Reflex Arcs. *J Neurophysiol* **1948**, *11*, 445–454, doi:10.1152/jn.1948.11.5.445.
64. Ulkatan, S.; Jaramillo, A.M.; Téllez, M.J.; Goodman, R.R.; Deletis, V. Feasibility of Eliciting the H Reflex in the Masseter Muscle in Patients under General Anesthesia. *Clin Neurophysiol* **2017**, *128*, 123–127, doi:10.1016/j.clinph.2016.10.092.
65. Sinclair, C.F.; Téllez, M.J.; Tapia, O.R.; Ulkatan, S.; Deletis, V. A Novel Methodology for Assessing Laryngeal and Vagus Nerve Integrity in Patients under General Anesthesia. *Clin Neurophysiol* **2017**, *128*, 1399–1405, doi:10.1016/j.clinph.2017.03.002.
66. Sinclair, C.F.; Téllez, M.J.; Ulkatan, S. Noninvasive, Tube-Based, Continuous Vagal Nerve Monitoring Using the Laryngeal Adductor Reflex: Feasibility Study of 134 Nerves at Risk. *Head Neck* **2018**, *40*, 2498–2506, doi:10.1002/hed.25377.
67. Ludlow, C.L.; Van Pelt, F.; Koda, J. Characteristics of Late Responses to Superior Laryngeal Nerve Stimulation in Humans. *Ann Otol Rhinol Laryngol* **1992**, *101*, 127–134, doi:10.1177/000348949210100204.
68. Sasaki, C.T.; Suzuki, M. Laryngeal Reflexes in Cat, Dog, and Man. *Arch Otolaryngol* **1976**, *102*, 400–402, doi:10.1001/archotol.1976.00780120048004.
69. Téllez, M.J.; Mirallave-Pescador, A.; Seidel, K.; Urriza, J.; Shoakazemi, A.; Raabe, A.; Ghatan, S.; Deletis, V.; Ulkatan, S. Neurophysiological Monitoring of the Laryngeal Adductor Reflex during Cerebellar-Pontine Angle and Brainstem Surgery. *Clin Neurophysiol* **2021**, *132*, 622–631, doi:10.1016/j.clinph.2020.10.021.
70. Kaneoka, A.; Pisegna, J.M.; Inokuchi, H.; Ueha, R.; Goto, T.; Nito, T.; Stepp, C.E.; LaValley, M.P.; Haga, N.; Langmore, S.E. Relationship Between Laryngeal Sensory Deficits, Aspiration, and Pneumonia in Patients with Dysphagia. *Dysphagia* **2018**, *33*, 192–199, doi:10.1007/s00455-017-9845-8.
71. Tarantino, V.; Stura, M.; Vallarino, R. [Development of Auditory Evoked Potentials of the Brainstem in Relation to Age]. *Pediatr Med Chir* **1988**, *10*, 73–76.
72. Reroñ, E.; Sekuła, J. Maturation of the Acoustic Path-Way in Brain Stem Responses (ABR) in Neonates. *Otolaryngol Pol* **1994**, *48*, 363–369.
73. Boor, R.; Goebel, B. Maturation of Near-Field and Far-Field Somatosensory Evoked Potentials after Median Nerve Stimulation in Children under 4 Years of Age. *Clin Neurophysiol* **2000**, *111*, 1070–1081.
74. Hameed, M.Q.; Dhamne, S.C.; Gersner, R.; Kaye, H.L.; Oberman, L.M.; Pascual-Leone, A.; Rotenberg, A. Transcranial Magnetic and Direct Current Stimulation in Children. *Curr Neurol Neurosci Rep* **2017**, *17*, 11, doi:10.1007/s11910-017-0719-0.
75. Garvey, M.A. Transcranial Magnetic Stimulation in Children. In *Magnetic stimulation in clinical neurophysiology*; Hallett, M., Chokroverty, S., Eds.; Elsevier Butterworth-Heinemann: Philadelphia, Pa, 2005; pp. 429–433 ISBN 978-0-7506-7373-0.
76. Kuntz, N.L. Chapter 6 - Clinical Neurophysiology of the Motor Unit in Infants and Children. In *Clinical Neurophysiology of Infancy, Childhood, and Adolescence*; Holmes, G.L., Jones, H.R., Moshé, S.L., Eds.; Butterworth-Heinemann: Philadelphia, 2006; pp. 130–145 ISBN 978-0-7506-7251-1.
77. Sasaki, C.T.; Levine, P.A.; Laitman, J.T.; Crelin, E.S. Postnatal Descent of the Epiglottis in Man. A Preliminary Report. *Arch Otolaryngol* **1977**, *103*, 169–171, doi:10.1001/archotol.1977.00780200095011.
78. Wang, X.; Guo, R.; Zhao, W.; Pilowsky, P.M. Medullary Mediation of the Laryngeal Adductor Reflex: A Possible Role in Sudden Infant Death Syndrome. *Respir Physiol Neurobiol* **2016**, *226*, 121–127, doi:10.1016/j.resp.2016.01.002.
79. Costa, P.; Gaglini, P.P.; Tavormina, P.; Ricci, F.; Peretta, P. A Method for Intraoperative Recording of the Laryngeal Adductor Reflex during Lower Brainstem Surgery in Children. *Clin Neurophysiol* **2018**, *129*, 2497–2498, doi:10.1016/j.clinph.2018.08.028.

80. Ulkatan, S.; Téllez, M.J.; Sinclair, C. Laryngeal Adductor Reflex and Future Projections for Brainstem Monitoring. Reply to "A Method for Intraoperative Recording of the Laryngeal Adductor Reflex during Lower Brainstem Surgery in Children." *Clin Neurophysiol* **2018**, *129*, 2499–2500, doi:10.1016/j.clinph.2018.09.017.
81. Stone, J.L.; Bouffard, A.; Morris, R.; Hovsepian, W.; Meyers, H.L. Clinical and Electrophysiologic Recovery in Arnold-Chiari Malformation. *Surg Neurol* **1983**, *20*, 313–317, doi:10.1016/0090-3019(83)90087-3.
82. Brookler, K.H. Vestibular Findings in a 62-Year-Old Woman with Dizziness and a Type I Chiari Malformation. *Ear Nose Throat J* **2005**, *84*, 630–631.
83. Hendrix, R.A.; Bacon, C.K.; Sclafani, A.P. Chiari-I Malformation Associated with Asymmetric Sensorineural Hearing Loss. *J Otolaryngol* **1992**, *21*, 102–107.
84. Johnson, G.D.; Harbaugh, R.E.; Lenz, S.B. Surgical Decompression of Chiari I Malformation for Isolated Progressive Sensorineural Hearing Loss. *Am J Otol* **1994**, *15*, 634–638.
85. Ahmed, A.U.; Mackenzie, I.; Das, V.K.; Chatterjee, S.; Lye, R.H. Audio-Vestibular Manifestations of Chiari Malformation and Outcome of Surgical Decompression: A Case Report. *J. LARYNGOL. OTOL.* **1996**, *110*, 1060–1064, doi:10.1017/S0022215100135753.
86. Hort-Legrand, C.; Emery, E. Motor and sensory evoked potentials in syringomyelia. *Neurochirurgie* **1999**, *45*, 95–104.
87. Isik, N.; Elmaci, I.; Isik, N.; Cerci, S.; Basaran, R.; Gura, M.; Kalelioglu, M. Long-Term Results and Complications of the Syringopleural Shunting for Treatment of Syringomyelia: A Clinical Study. *BRITISH JOURNAL OF NEUROSURGERY* **2013**, *27*, 91–99, doi:10.3109/02688697.2012.703350.
88. Henriques Filho, P.S.A.; Pratesi, R. Abnormalities in Auditory Evoked Potentials of 75 Patients with Arnold-Chiari Malformations Types I and II. *Arq Neuropsiquiatr* **2006**, *64*, 619–623.
89. Vidmer, S.; Sergio, C.; Veronica, S.; Flavia, T.; Silvia, E.; Sara, B.; Valentini, L.G.; Daria, R.; Solero, C.L. The Neurophysiological Balance in Chiari Type 1 Malformation (CM1), Tethered Cord and Related Syndromes. *Neurol. Sci.* **2011**, *32 Suppl 3*, S311–316, doi:10.1007/s10072-011-0692-3.
90. Di Stefano, V.; Ferrante, C.; Telese, R.; Caulo, M.; Bonanni, L.; Onofri, M.; Franciotti, R. Brainstem Evoked Potentials and Magnetic Resonance Imaging Abnormalities in Differential Diagnosis of Intracranial Hypotension. *Neurophysiol Clin* **2019**, *49*, 217–226, doi:10.1016/j.neucli.2019.04.003.
91. Anderson, N.E.; Frith, R.W.; Synek, V.M. Somatosensory Evoked Potentials in Syringomyelia. *J. Neurol. Neurosurg. Psychiatr.* **1986**, *49*, 1407–1410.
92. Forcadas, I.; Hurtado, P.; Madoz, P.; Zarranz, J.J. [Somatosensory Evoked Potentials in Syringomyelia and the Arnold-Chiari Anomaly. Clinical and Imaging Correlations]. *Neurologia* **1988**, *3*, 172–175.
93. Jabbari, B.; Geyer, C.; Gunderson, C.; Chu, A.; Brophy, J.; McBurney, J.W.; Jonas, B. Somatosensory Evoked Potentials and Magnetic Resonance Imaging in Syringomyelia. *Electroencephalogr Clin Neurophysiol* **1990**, *77*, 277–285.
94. Morioka, T.; Kurita-Tashima, S.; Fujii, K.; Nakagaki, H.; Kato, M.; Fukui, M. Somatosensory and Spinal Evoked Potentials in Patients with Cervical Syringomyelia. *Neurosurgery* **1992**, *30*, 218–222, doi:10.1227/00006123-199202000-00011.
95. Utzig, N.; Burtzlaff, C.; Wiersbitzky, H.; Lauffer, H. [Evoked Potentials in Chiari-Malformation Type I with Syringomyelia--a Case History]. *Klin Padiatr* **2003**, *215*, 241–243, doi:10.1055/s-2003-41402.
96. Akakin, A.; Yilmaz, B.; Ekşi, M.Ş.; Kılıç, T. Treatment of Syringomyelia Due to Chiari Type I Malformation with Syringo-Subarachnoid-Peritoneal Shunt. *J. Korean Neurosurg. Soc.* **2015**, *57*, 311–313, doi:10.3340/jkns.2015.57.4.311.
97. Strowitzki, M.; Schwerdtfeger, K.; Donauer, E. The Value of Somato-Sensory Evoked Potentials in the Diagnosis of Syringomyelia. *Acta Neurochir (Wien)* **1993**, *123*, 184–187.
98. Guvenc, G.; Kızmazoglu, C.; Aydın, H.E.; Coskun, E. Somatosensory Evoked Potentials and Cerebrospinal Fluid Flow in Chiari Malformation. *Annals of Clinical and Analytical Medicine* **2019**, *10*, 348–353, doi:10.4328/JCAM.5913.
99. Cheng, J.C.; Guo, X.; Sher, A.H.; Chan, Y.L.; Metreweli, C. Correlation between Curve Severity, Somatosensory Evoked Potentials, and Magnetic Resonance Imaging in Adolescent Idiopathic Scoliosis. *Spine* **1999**, *24*, 1679–1684.
100. Berciano, J.; Poca, M.-A.; García, A.; Sahuquillo, J. Paroxysmal Cervicobrachial Cough-Induced Pain in a Patient with Syringomyelia Extending into Spinal Cord Posterior Gray Horns. *J. Neurol.* **2007**, *254*, 678–681, doi:10.1007/s00415-006-0412-8.
101. Awai, L.; Curt, A. Preserved Sensory-Motor Function despite Large-Scale Morphological Alterations in a Series of Patients with Holocord Syringomyelia. *J. Neurotrauma* **2015**, *32*, 403–410, doi:10.1089/neu.2014.3536.

102. Stancanelli, C.; Mazzeo, A.; Gentile, L.; Vita, G. Unilateral Hyperhidrosis as Persistently Isolated Feature of Syringomyelia and Arnold Chiari Type 1. *Neurol. Sci.* **2018**, *39*, 1607–1608, doi:10.1007/s10072-018-3403-5.
103. Nogués, M.A.; Pardal, A.M.; Merello, M.; Miguel, M.A. SEPs and CNS Magnetic Stimulation in Syringomyelia. *Muscle Nerve* **1992**, *15*, 993–1001, doi:10.1002/mus.880150904.
104. Cocito, D.; Peci, E.; Garbossa, D.; Ciaramitaro, P. Neurophysiological Correlates in Patients with Syringomyelia and Chiari Malformation: The Cortico-Diaphragmatic Involvement. *J Clin Med* **2022**, *11*, 5080, doi:10.3390/jcm11175080.
105. Cristante, L.; Westphal, M.; Herrmann, H.D. Cranio-Cervical Decompression for Chiari I Malformation. A Retrospective Evaluation of Functional Outcome with Particular Attention to the Motor Deficits. *Acta Neurochir (Wien)* **1994**, *130*, 94–100.
106. Gerard, C.L.; Dugas, M.; Narcy, P.; Hertz-Pannier, J. Chiari Malformation Type I in a Child with Velopharyngeal Insufficiency. *Dev Med Child Neurol* **1992**, *34*, 174–176, doi:10.1111/j.1469-8749.1992.tb14985.x.
107. Paulig, M.; Prosiegel, M. Misdiagnosis of Amyotrophic Lateral Sclerosis in a Patient with Dysphagia Due to Chiari I Malformation. *JOURNAL OF NEUROLOGY NEUROSURGERY AND PSYCHIATRY* **2002**, *72*, 270–270, doi:10.1136/jnnp.72.2.270.
108. Bagnato, S.; Rizzo, V.; Quartarone, A.; Majorana, G.; Vita, G.; Girlanda, P. Segmental Myoclonus in a Patient Affected by Syringomyelia. *Neurol Sci* **2001**, *22*, 27–29, doi:10.1007/s100720170033.
109. Scelsa, S. Syringomyelia Presenting as Ulnar Neuropathy at the Elbow. *CLINICAL NEUROPHYSIOLOGY* **2000**, *111*, 1527–1530, doi:10.1016/S1388-2457(00)00361-8.
110. Muhn, N.; Baker, S.K.; Hollenberg, R.D.; Meaney, B.F.; Tarnopolsky, M.A. Syringomyelia Presenting as Rapidly Progressive Foot Drop. *J Clin Neuromuscul Dis* **2002**, *3*, 133–134, doi:10.1097/00131402-200203000-00006.
111. Jayamanne, C.; Fernando, L.; Mettananda, S. Chiari Malformation Type 1 Presenting as Unilateral Progressive Foot Drop: A Case Report and Review of Literature. *BMC Pediatr* **2018**, *18*, 34, doi:10.1186/s12887-018-1028-8.
112. Saifudheen, K.; Jose, J.; Gafoor, V.A. Holocord Syringomyelia Presenting as Rapidly Progressive Foot Drop. *J Neurosci Rural Pract* **2011**, *2*, 195–196, doi:10.4103/0976-3147.83594.
113. McMillan, H.J.; Sell, E.; Nzau, M.; Ventureyra, E.C.G. Chiari 1 Malformation and Holocord Syringomyelia Presenting as Abrupt Onset Foot Drop. *Child's Nerv. Syst.* **2011**, *27*, 183–186, doi:10.1007/s00381-010-1275-y.
114. Panda, A.K.; Kaur, M. Rapidly Progressive Foot Drop: An Uncommon and Underappreciated Cause of Chiari I Malformation and Holocord Syrinx. *BMJ Case Rep* **2013**, *2013*, doi:10.1136/bcr-2013-009644.
115. Leis, A.A.; Kofler, M.; Ross, M.A. The Silent Period in Pure Sensory Neuronopathy. *Muscle Nerve* **1992**, *15*, 1345–1348, doi:10.1002/mus.880151209.
116. Leis, A.A.; Štětárová, I.; Berić, A.; Stokić, D.S. Spinal Motor Neuron Excitability during the Cutaneous Silent Period. *Muscle Nerve* **1995**, *18*, 1464–1470, doi:10.1002/mus.880181218.
117. McLellan, D.L. The Electromyographic Silent Period Produced by Supramaximal Electrical Stimulation in Normal Man. *J Neurol Neurosurg Psychiatry* **1973**, *36*, 334–341, doi:10.1136/jnnp.36.3.334.
118. Shefner, J.M.; Logigian, E.L. Relationship between Stimulus Strength and the Cutaneous Silent Period. *Muscle Nerve* **1993**, *16*, 278–282, doi:10.1002/mus.880160306.
119. Uncini, A.; Kujirai, T.; Gluck, B.; Pullman, S. Silent Period Induced by Cutaneous Stimulation. *Electroencephalogr Clin Neurophysiol* **1991**, *81*, 344–352, doi:10.1016/0168-5597(91)90023-q.
120. Kaneko, K.; Kawai, S.; Fuchigami, Y.; Morita, H.; Ofuji, A. Cutaneous Silent Period in Syringomyelia. *MUSCLE NERVE* **1997**, *20*, 884–886, doi:10.1002/(SICI)1097-4598(199707)20:7<884::AID-MUS14>3.0.CO;2-S.
121. Amoiridis, G.; Meves, S.; Schols, L.; Przuntek, H. Reversible Urinary Retention as the Main Symptom in the First Manifestation of a Syringomyelia. *JOURNAL OF NEUROLOGY NEUROSURGERY AND PSYCHIATRY* **1996**, *61*, 407–408, doi:10.1136/jnnp.61.4.407.
122. Jacome, D.E. Blepharoclonus and Arnold-Chiari Malformation. *Acta Neurol Scand* **2001**, *104*, 113–117, doi:10.1034/j.1600-0404.2001.104002113.x.
123. Anderson, R.C.; Dowling, K.C.; Feldstein, N.A.; Emerson, R.G. Chiari I Malformation: Potential Role for Intraoperative Electrophysiologic Monitoring. *J Clin Neurophysiol* **2003**, *20*, 65–72.
124. Zamel, K.; Galloway, G.; Kosnik, E.J.; Raslan, M.; Adeli, A. Intraoperative Neurophysiologic Monitoring in 80 Patients with Chiari I Malformation: Role of Duraplasty. *J Clin Neurophysiol* **2009**, *26*, 70–75, doi:10.1097/WNP.0b013e31819f9058.
125. Kim, I.-K.; Wang, K.-C.; Kim, I.-O.; Cho, B.-K. Chiari 1.5 Malformation: An Advanced Form of Chiari I Malformation. *J Korean Neurosurg Soc* **2010**, *48*, 375–379, doi:10.3340/jkns.2010.48.4.375.

126. Chen, J.A.; Coutin-Churchman, P.E.; Nuwer, M.R.; Lazareff, J.A. Suboccipital Craniotomy for Chiari I Results in Evoked Potential Conduction Changes. *Surg Neurol Int* **2012**, *3*, 165, doi:10.4103/2152-7806.105277.
127. Kennedy, B.; Kelly, K.; Phan, M.; Bruce, S.; McDowell, M.; Anderson, R.; Feldstein, N. Outcomes after Suboccipital Decompression without Dural Opening in Children with Chiari Malformation Type I. *JOURNAL OF NEUROSURGERY-PEDIATRICS* **2015**, *16*, 150–158, doi:10.3171/2014.12.PEDS14487.
128. Barzilai, O.; Roth, J.; Korn, A.; Constantini, S. The Value of Multimodality Intraoperative Neurophysiological Monitoring in Treating Pediatric Chiari Malformation Type I. *Acta Neurochir (Wien)* **2015**, doi:10.1007/s00701-015-2664-5.
129. Rasul, F.T.; Matloob, S.A.; Haliasos, N.; Jankovic, I.; Boyd, S.; Thompson, D.N.P. Intraoperative Neurophysiological Monitoring in Paediatric Chiari Surgery—Help or Hindrance? *Childs Nerv Syst* **2019**, *35*, 1769–1776, doi:10.1007/s00381-019-04312-y.
130. Kawasaki, Y.; Uchida, S.; Onishi, K.; Toyokuni, M.; Okanari, K.; Fujiki, M. Intraoperative Neurophysiologic Monitoring for Prediction of Postoperative Neurological Improvement in a Child With Chiari Type I Malformation. *J Craniofac Surg* **2017**, *28*, 1837–1841, doi:10.1097/SCS.00000000000003926.
131. Krishnakumar, M.; Ramesh, V.; Goyal, A.; Srinivas, D. Utility of Motor Evoked Potential in Identification and Treatment of Suboptimal Positioning in Pediatric Craniovertebral Junction Abnormalities: A Case Report. *A&A Practice* **2020**, *14*, e01323, doi:10.1213/XAA.0000000000001323.
132. Grossauer, S.; Koeck, K.; Vince, G.H. Intraoperative Somatosensory Evoked Potential Recovery Following Opening of the Fourth Ventricle during Posterior Fossa Decompression in Chiari Malformation: Case Report. *JNS* **2015**, *122*, 692–696, doi:10.3171/2014.10.JNS14401.
133. Roser, F.; Ebner, F.H.; Liebsch, M.; Tatagiba, M.S.; Naros, G. The Role of Intraoperative Neuromonitoring in Adults with Chiari I Malformation. *Clinical Neurology and Neurosurgery* **2016**, *150*, 27–32, doi:10.1016/j.clineuro.2016.08.019.
134. Heffez, D.S.; Golchini, R.; Ghorai, J.; Cohen, B. Operative Findings and Surgical Outcomes in Patients Undergoing Chiari I Malformation Decompression: Relationship to the Extent of Tonsillar Ectopia. *Acta Neurochir (Wien)* **2020**, *162*, 1539–1547, doi:10.1007/s00701-019-04172-0.
135. Schaefer, J.; Atallah, E.; Tecce, E.; Thalheimer, S.; Harrop, J.; Heller, J.E. Utility of Intraoperative Neuromonitoring for Decompression of Chiari Type I Malformation in 93 Adult Patients. *Journal of Neurosurgery* **2022**, 1–6, doi:10.3171/2022.3.JNS22127.
136. Di, X. Endoscopic Suboccipital Decompression on Pediatric Chiari Type I. *Minim Invasive Neurosurg* **2009**, *52*, 119–125, doi:10.1055/s-0029-1224170.
137. Shah, A.; Patil, A.; Vutha, R.; Thakar, K.; Goel, A. Recovery of Transcranial Motor Evoked Potentials After Atlantoaxial Stabilization for Chiari Formation: Report of 20 Cases. *World Neurosurg* **2019**, *127*, e644–e648, doi:10.1016/j.wneu.2019.03.236.
138. Milhorat, T.H.; Kotzen, R.M.; Capocelli, A.L.; Bolognese, P.; Bendo, A.A.; Cottrell, J.E. Intraoperative Improvement of Somatosensory Evoked Potentials and Local Spinal Cord Blood Flow in Patients with Syringomyelia. *J Neurosurg Anesthesiol* **1996**, *8*, 208–215, doi:10.1097/00008506-199607000-00004.
139. Milhorat, T.H.; Capocelli, A.L.; Kotzen, R.M.; Bolognese, P.; Heger, I.M.; Cottrell, J.E. Intramedullary Pressure in Syringomyelia: Clinical and Pathophysiological Correlates of Syrinx Distension. *Neurosurgery* **1997**, *41*, 1102–1110, doi:10.1097/00006123-199711000-00014.
140. Pencovich, N.; Korn, A.; Constantini, S. Intraoperative Neurophysiologic Monitoring during Syringomyelia Surgery: Lessons from a Series of 13 Patients. *Acta Neurochir (Wien)* **2013**, *155*, 785–791; discussion 791, doi:10.1007/s00701-013-1648-6.
141. Sánchez-Roldán, M.A.; Moncho, D.; Rahnama, K.; Santa-Cruz, D.; Lainez, E.; Baiget, D.; Chocron, I.; Gandara, D.; Bescos, A.; Sahuquillo, J.; et al. Intraoperative Neurophysiological Monitoring in Syringomyelia Surgery: A Multimodal Approach 2023, doi: 10.20944/preprints202307.0052.v1.
142. Zhu, Z.; Qiu, Y.; Wang, B.; Yu, Y.; Qian, B.; Zhu, F. Abnormal Spreading and Subunit Expression of Junctional Acetylcholine Receptors of Paraspinal Muscles in Scoliosis Associated with Syringomyelia. *Spine (Phila Pa 1976)* **2007**, *32*, 2449–2454, doi:10.1097/BRS.0b013e3181573d01.
143. Eule, J.M.; Erickson, M.A.; O'Brien, M.F.; Handler, M. Chiari I Malformation Associated with Syringomyelia and Scoliosis: A Twenty-Year Review of Surgical and Nonsurgical Treatment in a Pediatric Population. *Spine (Phila Pa 1976)* **2002**, *27*, 1451–1455, doi:10.1097/00007632-200207010-00015.
144. Hausmann, O.N.; Böni, T.; Pfirrmann, C.W.A.; Curt, A.; Min, K. Preoperative Radiological and Electrophysiological Evaluation in 100 Adolescent Idiopathic Scoliosis Patients. *Eur Spine J* **2003**, *12*, 501–506, doi:10.1007/s00586-003-0568-1.

145. Zhu, Z.; Yan, H.; Han, X.; Jin, M.; Xie, D.; Sha, S.; Liu, Z.; Qian, B.; Zhu, F.; Qiu, Y. Radiological Features of Scoliosis in Chiari I Malformation Without Syringomyelia. *Spine (Phila Pa 1976)* **2016**, *41*, E276-281, doi:10.1097/BRS.0000000000001406.
146. Tubbs, R.S.; Beckman, J.; Naftel, R.P.; Chern, J.J.; Wellons, J.C.; Rozzelle, C.J.; Blount, J.P.; Oakes, W.J. Institutional Experience with 500 Cases of Surgically Treated Pediatric Chiari Malformation Type I. *J Neurosurg Pediatr* **2011**, *7*, 248–256, doi:10.3171/2010.12.PEDS10379.
147. Sha, S.; Zhu, Z.; Lam, T.P.; Sun, X.; Qian, B.; Jiang, J.; Cheng, J.C.Y.; Qiu, Y. Brace Treatment versus Observation Alone for Scoliosis Associated with Chiari I Malformation Following Posterior Fossa Decompression: A Cohort Study of 54 Patients. *Eur Spine J* **2014**, *23*, 1224–1231, doi:10.1007/s00586-014-3265-3.
148. Tan, H.; Lin, Y.; Rong, T.; Shen, J.; Zhang, J.; Feng, E.; Jiao, Y.; Liang, J.; Li, Z. Surgical Scoliosis Correction in Chiari-I Malformation with Syringomyelia Versus Idiopathic Syringomyelia. *J Bone Joint Surg Am* **2020**, *102*, 1405–1415, doi:10.2106/JBJS.20.00058.
149. Shi, B.; Qiu, J.; Xu, L.; Li, Y.; Jiang, D.; Xia, S.; Liu, Z.; Sun, X.; Shi, B.; Zhu, Z.; et al. Somatosensory and Motor Evoked Potentials during Correction Surgery of Scoliosis in Neurologically Asymptomatic Chiari Malformation-Associated Scoliosis: A Comparison with Idiopathic Scoliosis. *Clin Neurol Neurosurg* **2020**, *191*, 105689, doi:10.1016/j.clineuro.2020.105689.
150. Brinzeu, A.; Sindou, M. Functional Anatomy of the Accessory Nerve Studied through Intraoperative Electrophysiological Mapping. *JOURNAL OF NEUROSURGERY* **2017**, *126*, 913–921, doi:10.3171/2015.11.JNS15817.
151. Giampiccolo, D.; Basaldella, F.; Badari, A.; Squintani, G.M.; Cattaneo, L.; Sala, F. Feasibility of Cerebello-Cortical Stimulation for Intraoperative Neurophysiological Monitoring of Cerebellar Mutism. *Child's Nerv. Syst.* **2021**, *37*, 1505–1514, doi:10.1007/s00381-021-05126-7.
152. Caldarelli, M.; Novegno, F.; Vassimi, L.; Romani, R.; Tamburrini, G.; Di Rocco, C. The Role of Limited Posterior Fossa Craniectomy in the Surgical Treatment of Chiari Malformation Type I: Experience with a Pediatric Series. *J. Neurosurg.* **2007**, *106*, 187–195, doi:10.3171/ped.2007.106.3.187.
153. Barzilai, O.; Roth, J.; Korn, A.; Constantini, S. Letter to the Editor: Evoked Potentials and Chiari Malformation Type 1. *J. Neurosurg.* **2016**, 1–4, doi:10.3171/2016.4.JNS161061.

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