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Neuroradiologic Review of Arnold Chiari-Malformations: The Importance of Neurophysiologic Diagnosis

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Abstract

Arnold Chiari, also known as Chiari Malformation (CM), is the name given to a group of deformities of the hindbrain (Cerebellum, Pons, And Medulla Oblongata) that range from herniation of the posterior fossa contents outside of the cranial cavity to absence of the cerebellum with or without other associated intracranial or extra cranial defects such as hydrocephalus, syrinx or spinal. In this malformation group CM in all of its forms may produce neuronal dysfunctions of the brainstem, cerebellum, cranial nerves, medulla, and upper spinal cord. Evoked Potentials (EPs) can be useful for informing the clinician and the patient about the existence of objective neurophysiological abnormalities. In this review, we aim to review CM neuro-radiologically at the diagnosis stage and to emphasize the importance of neurophysiological recognition of CM in the early stage, especially in asymptomatic patients.

Keywords: Arnold-Chiari malformation type; Visual evoked potential; Somatosensory evoked potential; Brain auditory evoked potentials; Cranial MRI

Introduction

CM comprises various pathologies that have in common anatomical deformities of the brainstem and cerebellum. Hans Chiari, an Austrian pathologist, was the first to describe the conditions that now bear his name. His initial manuscript published in 1891, described Chiari malformations 1, 2, and 3. In 1896 publication, Chiari postulated the pathogenesis of CM and described Chiari 4 malformation [1-3]. These anomalies are characterized by downward elongation or displacement of the cerebellar tonsils or the vermis into the cervical spinal canal. Abnormalities can be associated with CM, such as hydrocephalus, syringomyelia, spina bifida, hydromyelia, kyphosis, scoliosis, and tethered cord syndrome. Moreover, CM may be associated with hereditary syndromes and other disorders that affect growth and bone formation, such as craniosynostosis, Ehlers-Danlos syndromes, and Klippel-Feil syndrome. Classically, the CMs are classified into 4 types: Chiari types 1, 2, 3, and 4. Chiari Malformation type 1 (CM1) is by far the most common type of Chiari Malformation. CM1 is characterized by extension of the cerebellar tonsils by at least 5 mm below the foramen magnum. Chiari Malformation type 2 (CM2) is found in patients with myelomeningocele and involves a greater degree of hindbrain displacement, which may include the cerebellar vermis, brainstem, and fourth ventricle. CM3 is an encephalocele of the posterior fossa with herniation of portions of the cerebellum and brainstem into the encephalocele sac. CM4 is aplasia or hypoplasia of the cerebellum [4]. By Jerry Oakes et al., 2 additional types of Chiari Malformation have been described; Chiari Malformation 0 (CM0) which have syringomyelia without displacement of the cerebellar tonsils, caudal displacement of the cervicomedullary junction and intradural obstruction of Cerebrospinal Fluid (CSF) flow and Chiari Malformation. 1.5 is a severe variant of CM1 in which there is a caudal displacement of the brainstem in addition to the cerebellar tonsils below the foramen magnum [5-7] (Table 1). At the diagnosis stage, demonstrating the tip of the cerebellar tonsils 5 mm below the foramen magnum, associated possible syrinx, and regional malformations on Magnetic Resonance Imaging (MRI) is crucial [8].

Other useful tests in the management of patients with CM include myelography which has value as an alternative in patients in whom an MRI cannot be obtained, CT or X-rays of the neck and head which may reveal common associated bony defects, particularly of the craniocervical junction relevant for surgical planning and Cine MRI that useful in the evaluation of cerebrospinal fluid dynamics also may demonstrate blockage of flow at the foramen magnum.

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EPs are noninvasive studies that measure the electrophysiological response of the nervous system to different sensory stimuli, including Brainstem Auditory Evoked Potentials (BAEP), Visual Evoked Potentials (VEP), and Somatosensory Evoked Potentials (SEP). EPs have been used especially in early-stage CM patients as CM-1 as initial diagnostic support and are especially useful for determining the extent of neurologic involvement in asymptomatic or oligosymptomatic cases. They are also useful for identifying changes that may indicate surgical intervention in the follow-up in whom no symptoms and detected incidentally, as well as oligosymptomatic patients, is only a few studies, describe the findings of BAEP and SSEP testing in CM-1 [9-12] and unfortunately none for VEP. In this review, a clinico-radiological view of patients with CM will be presented and the importance of using EPs in patients diagnosed with early-stage CM and their place in the prognosis will be mentioned.

Neurological Symptoms and Physical Examination

CM-1is the mildest of the hindbrain malformations and a number of subgroups have been defined. In the first group, intrauterine hydrocephalus causes tonsillar herniation. Once myelinated, the tonsils retain this pointed configuration and ectopic position. Patients tend to be present in childhood with hydrocephalus and usually with syringomyelia. A second group involves those with associated craniocervical dysgenesis. They usually present later than children or young adults with occipital headaches, especially when straining (cough-laugh headache), cranial nerve palsies, or dissociated sensory abnormalities secondary to syringomyelia. The third group relates to acquired deformities of the foramen magnum, such as basilar invagination. These are usually adults who develop syringomyelia and have headaches and cranial neuropathies. Although the spectrum of Chiari I malformations is not usually associated with other cerebral abnormalities, syringomyelia is found in 20% to 70% of patients, depending on the degree and extent of disruption of normal Cerebrospinal Fluid (CSF) flow between the spine and cranium [13].

CM-2 is a more extensive and complex abnormality than CM-1, with infratentorial and supratentorial abnormalities. The cerebellar tonsils, inferior vermis, IV ventricle, and brainstem are herniated from a shallow posterior fossa through a wide foramen magnum with obstruction to CSF flow at the exits of the IV ventricle. There is virtually always a meningocele or meningomyelocele present with some associated hydrocephalus. [14]. Partial or complete agenesis of the corpus callosum is found in most patients, while falx hypoplasia, fused enlarged massa intermedia, colpocephaly, abnormal gyral patterns, and interdigitation of the paramedial gyri across the midline are all associated features. Infratentorial, there is breaking of the tectum, petrous bone scalloping, a low torcula, and cervicomedullary kinking. A degree of spinal dysraphism is usually present with a tethered cord and filum lipoma. Signs of brainstem compression with swallowing difficulties, stridor, apneic spells, a weak cry, or arm weakness can be found. If presenting as an adult bilateral limb weakness and wasting followed by sensory disturbance are most common, dysphasia and ataxia are less common.

CM-3 is very rare and herniation of the posterior fossa contents into an associated occipital or high cervical cephalocele with the other features of a Chiari II malformation [14]. Patients have severe neurological defects and a poor prognosis [15].

CM-4 is controversial and extremely rare [14,16]. Many authors consider the features of primary cerebellar agenesis as a Chiari IV malformation. In primary cerebellar agenesis, there are remnants of residual cerebellum-for example, anterior quadrangular lobule-a normal brainstem and a normal-sized posterior fossa, and a normal brain and spine.

Diagnosis and Treatment

Diagnosis of anomalies of the craniovertebral junction includes a number of research methods:

1. Clinical neurological examination and clinical and genealogical analysis

2. MRI of the cranio-vertebral junction, brain and spinal cord in sagittal and axial projections in T1 and T2 modes: MR-angiography is added as needed

3. Computed tomography of the cranio-vertebral junction if it is impossible to perform MRI, if it is necessary to assess the bone structures of this area

4. X-ray examination of the skull and cervical spine if CT or MRI of the cranio- vertebral region is impossible

5. Examination of the fundus

6. Otoneurologic examination

7. Prenatal echography is performed according to indications

8. Tran cranial Doppler sonography, evoked stem potentials are performed according to indications [17-20].

Clinico-Radiological Evaluation

MRI of the Head and Cervical Spine is the most informative and widely used method for the diagnosis of CM [19-23]. Conventional studies are performed with 1.5- or 3-T MRI units and with the patient supine on the examining table. In certain special situations, as in the case of patients with suspected cranio-cervical instability or recent trauma, there may be a benefit of obtaining an upright MRI study with or without flexion and extension of the neck. With MRI, in addition to detecting the characteristic dislocation of the tonsils, we can get useful information about the presence or absence of pathology (syringomyelia, hydrocephalus). There has been much emphasis on the measurement of the extent of tonsillar descent below the foramen magnum, based on the original studies of Barkovich, who stated that patients with 5 mm or more of tonsillar descent were more likely to have a Chiari anomaly [24]. More recently, it has become clear that the measurement of tonsillar descent does not necessarily correlate with the clinical picture. The pointed shape of the cerebellar tonsil's indicative of compression, the presence or absence of CSF spaces surrounding the tonsils, and evidence of brainstem compression on axial images at the level of the foramen magnum may be as significant as a measurement of tonsillar descent. Cine MRI is useful in the evaluation of cerebrospinal fluid dynamics. May demonstrate blockage of flow at the foramen magnum and distinguish symptomatic ACA types 0 and 1 from asymptomatic cerebellar ectopia, as well as clarify the indications for surgical decompression and predict the outcome of surgical treatment [25,26].

Computed Tomography (CT) with myelography/cisternography can be performed for patients who cannot undergo MRI (if there are contraindications to this diagnostic method). However, in this case, the use of high-speed (for e.g., 64-slice) spiral CT (MSCT) will

Table 1: Summary of types of Chiari malformations.

Description	Chiari Malformation Type
Syrinx without caudal descent of cerebellar tonsils. Syrinx resolves with posterior fossa decompression	0
Caudal displacement of the cerebellar tonsils at least 5 mm below the foramen magnum	1
Caudal displacement of the brainstem and cerebellar tonsils below the foramen magnum	1.5
Caudal displacement of hindbrain structures below the foramen magnum in a patient with myelomeningocele	2
Posterior fossa encephalocele containing brainstem and cerebellar tissue	3
Aplasia or hypoplasia of the cerebellum	4

be optimal, which makes it possible to diagnose ACA without the use of a contrast agent, while non-contrast Multi-Slice Computed Tomography (MSCT) with sagittal reconstruction eliminates the need for myelography. It may reveal common associated bony defects, particularly of the cranio-cervical junction relevant for surgical planning.

Neurophysiological Evaluation

Evoked potentials are noninvasive studies that measure the electrophysiological response of the nervous system to different sensory stimuli. EPs have been used in CM-0, CM-1, and CM-1.5 patients as initial diagnostic support and are especially useful for determining the extent of neurologic involvement in asymptomatic or oligo-symptomatic cases. They are also useful for identifying changes that may indicate surgical intervention in the follow-up of patients, especially in children in whom no symptoms were present and CM-1 was detected incidentally, as well as oligo-symptomatic patients. At the time of writing, however, only a few studies, all with very limited and heterogeneous series of patients, describe the findings of BAEP and SSEP testing in CM-1, and most of them refer to their use during intraoperative neurophysiological monitoring [9-12]. As a result, at present, there are still many unknowns regarding the role of EPs in the diagnosis and follow-up of CM-0, CM-1, and CM-1.5; the prognostic value of EPs and their relationship with clinical findings and the severity of malformation has not yet been well established.

Most of the studies have been retrospective studies with a limited number of patients and diverse patient populations and focused on especially SSEPs in patients with syringomyelia [27-31]. Restuccia and Maguire [30] observed findings consistent with those reported by Anderson et al. Restuccia and Mauguière described the most frequent disturbance as an alteration or absence of the cervical potential and an increased N13 to N20. In another study, a high percentage of patients (60%) with CM-1 exhibited EP alterations regardless of their clinical or radiological findings and they concluded that BAEP and SSEP studies play an important role in incidentally detected patients with CM (especially in CM-0, CM-1, and CM-1.5) [32]. They concluded that EPs may help to establish objective evidence of subclinical dysfunctions and neurophysiological studies may help to define subgroups of patients who require further testing and follow-up to personalize strategies for the management of incidental and oligosymptomatic patients.

SSEP

SSEPs, elicited from the upper and lower limbs within 30 ms and 60 ms, respectively, of percutaneous electrical stimulation, are considered resulting from action potentials and synaptic potentials from successive anatomic neural generators within the dorsal-lemniscus's thalamocortical sensory system [33]. SSEPs are typically named by their negative or positive polarity at the peak latency, and the time of the peak latency (e.g., N20 is a negative deflection in the EEG waveform, usually peaking at 20 ms post stimulus) as typically observed in the normal population. The actual latency value for a SEP may be different from that implied by the component's name [34]. SEPs evaluate the dorsal column- medial lemniscus pathway, including the spinal cord and brainstem levels. The lemniscal pathways are typically affected in patients with CM, such as displacement of the cerebellar tonsils, hydrocephalus and syringomyelia. In EPs studies, it is found that most of the patients with a thermalgesic disturbance-the most frequent neurological finding in patients with syringomyelia also exhibited altered SSEPs [31,34,35]. Also, in one study, it is found that the degree of tonsillar herniation was statistically significant in predicting abnormal SSEPs [32]. SSEPs were also abnormal in 30.4% of patients in whom CM had been found incidentally.

VEP

VEP is used to assess the visual conduction pathways through the optic nerves and brain. To measure VEP, visual fields are stimulated, usually with a checkerboard visual stimulus, and the evoked response is recorded using surface recording electrodes over the occipital lobe [36]. The visual perception stimulation method was used to manage cortical visual impairment. Many causes that cause changes in cerebrospinal fluid, such as hydrocephalus, can cause changes in VEP [37]. Hydrocephalus is commonly associated with CMs and it can produce marked changes in the EPs. Several groups of researchers have recorded EPs in hydrocephalic patients in attempts to find a sensitive measure of increased intracranial pressure, of associated pathological changes, or as an indication of the need for neurosurgical intervention [38-41]. Although some of the above studies have found consistent correlations between Visual Evoked Potential (VEP) abnormalities and raised intracranial pressure [42], others have found VEPs to be more useful in only monitoring hydrocephalic patients. Ehle and Sklar [38] found that the 15 infants they studied all had abnormally delayed VEPs, which improved quickly post-shunting. Guthkelch [40] found that increased latencies were present only when the hydrocephalus was accompanied by increased head size (above the 98th percentile); hydrocephalic neonates who were normocephalic usually had normal VEPs. In a later study, they also found that hydrocephalic children had slower maturation of the VEP [39]. They repeated the VEP testing post-shunting in infants <4 months and found only small decreases in latency. Thus, several studies have found VEPs to change with shunting, and therefore it is important in myelomeningocele patients who develop hydrocephalus to record EPs following shunting. In one study, VEPs were recorded in 47 infants with myelomeningocele to determine if the evoked potentials reflected the early neurological status and if they had prognostic value as to the children's neurological outcome. VEPs were abnormal in only 55% of symptomatic infants. Of the infants who did

not have symptomatic AC malformation, 69% had normal VEPs. Of the patients with normal VEPs, 63% were normal on follow-up; of the patients with abnormal VEPs, 71% were abnormal on follow-up. It was concluded that the VEPs studied early in the neonatal course do not appear to be sufficiently sensitive to be valuable prognostically in these infants [41]. As we know, there are no studies on the use of VEP in adult patients with CM.

BAEP

The Brainstem Auditory Evoked Response (BAER) or Potential (BAEP) reflects the electrophysiological activity of many neurons in the brainstem auditory pathway following acoustic stimulation. The BAER is the far-field reflection of sequentially activated neurons at successively higher levels of this pathway and can be used to assess peripheral auditory function, also the functional integrity and development of the brain in general in conditions that affect the brainstem auditory pathway. There is evidence to suggest that BAER waves I and II are generated in the extra cranial and intracranial portions of the VIIIth nerve, respectively [42]. Subsequent waves IIIeVII are generated in auditory centers at gradually higher levels of the pathway, with partially overlapping contributions to individual waves. Wave III is derived from the cochlear nucleus; wave IV is generated in the superior olivary complex and wave V e together with the negative potential that follows is generated in the region of the lateral lemniscus and possibly inferior colliculus. Waves VI and VII are likely to originate from the inferior colliculus, although the exact origins remain to be determined. This close relationship between waveforms and anatomical structures makes it possible to localize accurate conduction defects in the brainstem [42]. BAEPs are extremely useful in the diagnosis and localization of a number of brainstem lesions [43]. Quite frequently, they reveal abnormalities even when CT evidence of disease is lacking or inconclusive. In one study, abnormal responses were obtained in 75% of cases. This figure agrees with the 50% to 86% given in other reports in the literature [44,45]. Analysis of symptomatic and asymptomatic cases shows that both groups had prolonged III-V Inter-Peak Latency (IPL). Furthermore, in the symptomatic and 64% of asymptomatic patients, the I-III IPL was normal. In another study, the frequency and degree of severity of abnormalities in the auditory pathways in patients with Chiari malformations type I and II was evaluated in 75 patients (48 children and 27 adults) by means of auditory evoked potential evaluation. Among the 75 patients studied, 27 (36%) disclosed Arnold-Chiari malformations type I and 48 (64%) showed Arnold-Chiari malformations type II. 53 (71%) of these patients showed some degree of auditory evoked potential abnormalities. Tests were normal in the remaining 22 (29%) patients. They concluded as auditory evoked potential testing can be considered a valuable instrument for the diagnosis and evaluation of brain stem functional abnormalities in patients with Arnold-Chiari malformations type I and II. The determination of the presence and degree of severity of these abnormalities can contributory to the prevention of further handicaps in these patients either through physical therapy or by means of precocious corrective surgical intervention [46]. In this study, it is reported that the most frequent abnormality found in BAEPs in classic CM-1 was on a cochlear or auditory peripheral level [47]. Also, in another study, it is found that the degree of tonsillar herniation, and lower cranial nerve dysfunction, had a statistically significant influence in predicting abnormal BAEPs [29]. BAEPs were abnormal in 39.1% of asymptomatic patients on a retro cochlear level. They concluded that the more severe distortion of the brainstem structures induced more BAEP abnormalities and, therefore, as expected, patients with lower cranial nerve dysfunction presented with a higher frequency of abnormal BAEPs.

Conclusion

EPs play an important diagnostic role in asymptomatic/oligosymptomatic patients such as CM-0, CM-1, and CM-1.5 and are also useful in determining prognosis before and after surgery in other types of CMs. MRI is widespread available and crucial for the first diagnosis. In patient follow-up and treatment response, only clinical-radiological follow-up will be insufficient in terms of prognosis and guiding treatment. At this stage, it is important to add neurophysiological evaluation to radiological examinations and to determine the necessity of using each one separately. We believe that the importance of EPs will be better understood with large-scale studies in this area.

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