



ORIGINAL ARTICLE

Audio-Vestibular Signs and Symptoms in Chiari Malformation Type I. Case Series and Literature Review[☆]



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KEYWORDS

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Abstract

Introduction: Chiari malformation is an alteration of the base of the skull with herniation through the foramen magnum of the brain stem and cerebellum. Although the most common presentation is occipital headache, the association of audio-vestibular symptoms is not rare. The aim of our study was to describe audio-vestibular signs and symptoms in Chiari malformation type I (CM-I).

Materials and methods: We performed a retrospective observational study of patients referred to our unit during the last 5 years. We also carried out a literature review of audio-vestibular signs and symptoms in this disease.

Results: There were 9 patients (2 males and 7 females), with an average age of 42.8 years. Five patients presented a Ménière-like syndrome; 2 cases, a recurrent vertigo with peripheral features; 1 patient showed a sudden hearing loss; and 1 case suffered a sensorineural hearing loss with early childhood onset. The most common audio-vestibular symptom indicated in the literature in patients with CM-I is unsteadiness (49%), followed by dizziness (18%), nystagmus (15%) and hearing loss (15%). Nystagmus is frequently horizontal (74%) or down-beating (18%). Other audio-vestibular signs and symptoms are tinnitus (11%), aural fullness (10%) and hyperacusis (1%). Occipital headache that increases with Valsalva manoeuvres and hand paresthesias are very suggestive symptoms.

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Conclusions: The appearance of audio-vestibular manifestations in CM-I makes it common to refer these patients to neurotologists. Unsteadiness, vertiginous syndromes and sensorineural hearing loss are frequent. Nystagmus, especially horizontal and down-beating, is not rare. It is important for neurotologists to familiarise themselves with CM-I symptoms to be able to consider it in differential diagnosis.

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PALABRAS CLAVE

Vértigo;
Mareo;
Hipoacusia;
Inestabilidad;
Acúfeno;
Plenitud ótica;
Malformación
de Chiari tipo I

Manifestaciones audiovestibulares en la malformación de Chiari tipo I. Serie de casos y revisión bibliográfica

Resumen

Introducción: La malformación de Chiari es una alteración de la base del cráneo, en la que se produce herniación del cerebelo y del tronco cerebral a través del foramen magnum. Aunque su forma de presentación más frecuente es la cefalea occipitounal, no es rara la asociación de síntomas audiovestibulares. El objetivo de nuestro estudio fue conocer las manifestaciones audiovestibulares en la malformación de Chiari tipo I (MCH-I).

Material y métodos: Se realizó un estudio retrospectivo observacional de los pacientes remitidos a nuestra unidad en los últimos 5 años, así como una revisión bibliográfica de las manifestaciones audiovestibulares de esta enfermedad.

Resultados: Se presentan 9 pacientes (2 varones y 7 mujeres) de 42,8 años de edad media. Cinco de los pacientes consultaron con un síndrome menieriforme, 2 casos como vértigo recurrente de características periféricas, otro como hipoacusia súbita y el último caso como hipoacusia neurosensorial de inicio en la infancia. La manifestación audiovestibular más frecuentemente descrita en la literatura en pacientes es la inestabilidad (49%), seguida de vértigo (18%), nistagmo (15%) e hipoacusia (15%). Dentro del nistagmo, el más frecuente es el nistagmo horizontal (74%) seguido del vertical hacia abajo (18%). Otras manifestaciones audiovestibulares son acúfenos (11%), plenitud ótica (10%) e hiperacusia (1%). La cefalea occipitounal que aumenta con las maniobras de Valsalva y las parestesias en las manos son muy sugestivos de esta enfermedad.

Conclusiones: La aparición de síntomas audiovestibulares en la MCH-I hace que sea relativamente frecuente su derivación al otoneurólogo. Estos pacientes presentan inestabilidad, síndrome vertiginoso e hipoacusia neurosensorial. No es rara la presencia de nistagmo, sobre todo horizontal y vertical hacia abajo. Es importante la familiarización de los otoneurólogos con la sintomatología de esta enfermedad de cara a su diagnóstico diferencial.

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Introduction

Chiari malformation is a congenital disease, consisting of an anatomical alteration in the skull base, in which there is a herniation of the cerebellum or the brainstem through the foramen magnum up to the cervical canal.¹

For several decades, the eponyms Arnold and Chiari have been used as synonyms to define cases with ectopy of the cerebellum tonsils below the level indicated by the posterior border of the foramen magnum. Chiari malformation was described for the first time by Cleland² in 1883. In 1891 Chiari,³ in his study *About the cerebellar alterations resulting from cerebral hydrocephalus*, defined this entity as *an elongation in the shape of a wedge of the cerebellum tonsils and of the medial part of the inferior cerebellum lobules, which run throughout the medulla within the cervical canal*.

Later on (1907), Schwalbe and Gredig, under Julius Arnold at the University of Heidelberg,⁴ described 4 new cases, putting the name of their mentor before that of Chiari in the designation of the malformation, today recognised for type II.

Classically, Chiari malformation has been considered typical of young adults and adolescents. Its frequency is unknown; however, thanks to the introduction of nuclear magnetic resonance (NMR) from 1985 on, its detection has increased considerably, with some studies estimating a prevalence of between 0.1% and 0.5%.⁵

According to the 2009 consensus document,⁶ the classification of Chiari malformation covers 5 subtypes, from 0 to IV (Table 1), among which type I is undoubtedly the most frequent, except in early childhood, a period in which type II is more common.⁷

Table 1 Chiari Malformation Classification According to the Consensus Document on Malformations of the Cranial-Cervical Junctionure.

Type 0	Alteration of hydrodynamics (CSF)	Syringomyelia with minimum data on tonsillar herniation or without
Type I	Caudal cerebellar tonsillar herniation >5 mm	Characteristically associated with hydro-syringomyelia
Type II	Caudal herniation through the foramen magnum of the cerebellar vermis, brainstem and 4th ventricle	Myelomeningocele and hydrocephalus, and less frequently, with hydro-syringomyelia. Other types of intracranial alterations can be observed (tentorium hypoplasia, craniolacunia, anomalies of the Sylvian aqueduct)
Type III	Occipital encephalocele with part of the intracranial anomalies associated with Chiari II	
Type IV	Aplasia or hypoplasia of the cerebellum associated with aplasia of the tentorium cerebelli	

Source: Amado Vázquez et al.⁶

This malformation can be asymptomatic and be discovered incidentally in an imaging study. However, its most common form of presentation is occipital and neck pain exacerbated by physical activity or Valsalva manoeuvres. Loss of superficial abdominal reflexes, radicular pain and hand dysesthesias are frequent.⁸

As a consequence of the development of cranial neuropathies or brainstem compression, it is possible that symptoms of the otorhinolaryngological area can appear, such as hoarseness, vocal paralysis, dysarthria, palatine weakness, lingual atrophy, aspirations, etc.⁶ Specifically, in the otoneurological area, there are descriptions of cases of presentation as nystagmus (characteristically vertical downwards), neurosensory hypoacusis and even clinical pictures suggestive of peripheral vestibular disease.^{6,9,10} It is therefore important for the otoneurologist to be familiar with the symptoms of this disease for its differential diagnosis and multidisciplinary management.

Recent studies have been unable to correlate the degree of herniation of the cerebellar tonsils and the seriousness of the clinical symptoms.⁸ These findings, and the appearance of symptoms in patients with herniation <5 mm, make it plausible that the underlying physiopathological mechanism has to do with decoupling between the volumetric cranial-spinal compensation circuit and the spinal fluid blend circuit, an event that happens as a result of the distortion of cerebrospinal fluid (CSF) circulation between the cisterna magna and the cervical subarachnoid space.

The objective of our study was to review the audiovestibular signs and symptoms of Chiari type I malformation.

Materials and Methods

Retrospective Observational Study

We reviewed the clinical histories and neuroradiological studies of the patients referred to the Otoneurological Area of the Hospital Universitario Marqués de Valdecilla with the diagnosis of Chiari type I malformation (CHM-I) between 1 January 2008 and 31 December 2012. Only patients who

fulfilled the diagnostic criteria for this disease established by Avellaneda-Fernández et al.⁶ were chosen.

Demographic and clinical variables were analysed, including family and personal antecedents, age at diagnosis, first symptom and treatment received. Information about the otoneurological examination performed was gathered from all the patients, seeking spontaneous nystagmus with or without visual fixation, evoked by the gaze, triggered by cephalic agitation or positional manoeuvres (supine, hyper-extended, right and left lateral decubitus and Dix-Hallpike). The assessment included a caloric test and a tonal audiometry tonal. In one of the cases (a paediatric patient) a behavioural audiometry was also performed, as well as an objective study using automatic brainstem auditory evoked potentials (BAEP). The patients included in the study received a joint follow-up from the Neurology and Neurosurgery services at the same hospital. In all the cases a neuro-radiological imaging study using NMR was available and, in 3 of the cases, a functional study of the CSF flow through cine-MR imaging.

The data were stored in an Apache OpenOffice database (Apache Software Foundation, Forest Hill, MD, USA) and later processed using the tools available in that computer software package.

Literature Review

To characterise the otoneurological signs and symptoms of CHM-I we reviewed the literature in PubMed, which included all the articles in which audiovestibular signs and symptoms of this disease were classified. We used the search chain ("arnold-chiari malformation" [MeSH Terms] OR ("arnold-chiari" [All Fields] AND "malformation" [All Fields]) OR "arnold-chiari malformation" [All Fields] OR ("chiari" [All Fields] AND "malformation" [All Fields]) OR "chiari malformation" [All Fields]) AND Type [All Fields] AND 1 [All Fields] AND ("vertigo" [MeSH Terms] OR "vertigo" [All Fields] OR "dizziness" [All Fields] OR "dizziness" [MeSH Terms] OR "tinnitus" [MeSH Terms] OR "tinnitus" [All Fields] OR "hearing loss" [MeSH Terms] OR "hearing" [All Fields] AND "loss" [All Fields] OR "hearing loss" [All Fields] OR ("hearing" [All Fields] AND "impairment" [All Fields]) OR "hearing impairment"

Case	Age (years) /sex	Ev.	AV	PF+	CT+	Others	NMR*
1	41/F	1 year	LNSHL fluct Tinnitus Fullness PV Instability				Occipital-cervical head-ache-Parasthesias 12 mm
2	51/F	1 year	Tinnitus B RV Instability	Left B-W		Temporal dysesthesias and right concha	8 mm SRM-D4-
3	57/F	10 Years	RNSHL fluct Tinnitus B Fullness Instability				7 mm
4	48/M	2 Years	RNSHL fluct Tinnitus izq. Instability	Left B-W THI 62%		Obstacles	5 mm Flow: < posterior
5	47/F	5 Months	Acúfeno VR Instability				5-15 mm
6	53/F	24 horas (1 month)	HNSizqda. progr. HNS drcha. tinnitus				12.7 mm
7	55/M	8 years	RNSHL Progr. RNS/L fluct. Right tinnitus PV Instability				9.9 mm Flow: < posterior
8	68/F	10 years	BNSHL-aud- PV/RV Instability				17-22 mm Flow: < post
9	3/M	3 years	LNSHL		· ABAEP LE NSHL-70 dB · Behavioural LTA: moderate LNSHL	Central apnoeas, dysphagia	18.5 mm

Figure 1 Series of Chiari type I malformation cases with audiovestibular manifestations referred to the Otoneurology Unit at the Hospital Universitario Marqués de Valdecilla. *Measurement in mm of the degree of cerebellar herniation through the foramen magnum; LTA: liminal tonal audiometry; AV: audiovestibular symptoms; B-W: Babinski-Weil; B: bilateral; PE+: pathological findings in the physical examination; Ev.: time of evolution; Fluct.: fluctuating; Hypofx: hypofunction; RNSHL: right neurosensory hearing loss; LNSHL: left neurosensory hearing loss; SNSHL: sudden neurosensory hearing loss; mm: millimetres; UL: upper limbs; RE: right ear; CT+: pathological findings in the complementary tests; ABAEP: automatic brainstem auditory evoked potentials; Progr: progressive; NMR: nuclear magnetic resonance; SRM: syringomyelia; THI: Tinnitus Handicap Inventory; PV: vertigo with positional characteristics; RV: recurrent vertigo.

[All Fields] OR (“aural” [All Fields] AND “fullness” [All Fields]) OR “aural fullness” [All Fields]. Afterwards, we selected the originals written in English or in Spanish.

Results

Case Series

Nine patients (2 males and 7 females) diagnosed with CHM-I that presented audiovestibular signs and symptoms were

identified. Mean age at diagnosis was 42.8 years (range, 3–57 years) and the mean cerebellar tonsil descent was 12.2 mm (range, 5–22 mm). The clinical-exploratory characteristics of our series are shown in Fig. 1. Fig. 2 shows the radiological image (NMR) of 1 of the patients.

Clinical Presentation

Five of the patients came to consultation for a Ménière-like syndrome. In 2 patients the key otoneurological symptom was vertigo; in 1 case it was recurrent with peripheral

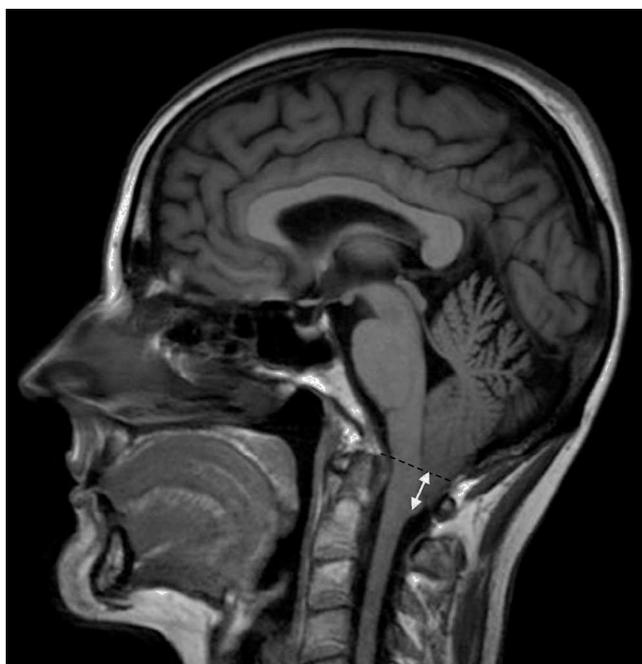


Figure 2 Nuclear magnetic resonance image, sagittal slice, of Case 8 in our series. The McRae line is represented by the dashed line. The arrow marks the degree of cerebellar tonsillar herniation, of 22 mm in this case.

characteristics, and in the other it was of positional characteristics. These 2 patients suffered hearing loss, 1 in the form of sudden hypoacusis and the other as childhood-onset neurosensory hypoacusis.

In 8 of the 9 cases, the patients associated occipital-neck headaches of years of evolution and paresthasias in the hands. In 1 case, these worsened together upon exacerbation of the audiovestibular symptoms.

The 3-year-old male was diagnosed in the context of a complex clinical picture of central apnoeas, episodes of loss of consciousness non-seizure in appearance, psychomotor delay and dysphagia. A moderate unilateral neurosensory hypoacusis was found in the auditory exploration.

Audiovestibular Signs and Symptoms

The most prevalent audiovestibular signs and symptoms in our series were hearing loss and tinnitus, which appeared with the same frequency ($n=7$; 77.8%). The hearing loss was unilateral in 4 cases and bilateral in 3. In 5 patients it fluctuated, with tinnitus and instability, while in 2 patients (22.2%) it was also associated with sensation of aural fullness. In 1 case (a 53-year-old women with a history of left progressive neurosensory hypoacusis of long evolution), the symptom that motivated the consultation was a sudden-onset contralateral hearing loss with a sudden 24-h onset; she recovered to her original condition in 30 days.

Five patients reported vertigo, understood to be the illusion of movement of objects: in 2 patients it was positional, in another 2 it could be characterised as a recurrent type, while another case mixed both types of vertigo.

The otoneurological functional and clinical examination was normal in all cases. However, the auditory assessment

Table 2 Audiovestibular Signs and Symptoms in Chiari Type I Malformation, by Order of Appearance ($n=627$).

Audiovestibular signs	Frequency (%)
<i>Instability</i>	49
<i>Vertigo</i>	18
<i>Nystagmus</i>	15
Horizontal	74
Vertical downwards	18
Vertical upwards	5
Not specified	3
<i>Hearing loss</i>	15
Bilateral	44
Unilateral	30
Not specified	26
<i>Tinnitus</i>	11
<i>Aural fullness</i>	10
<i>Hyperacusis</i>	1

Source: Literature review.⁹⁻¹⁸

showed alterations of greater or lesser seriousness in 8 of the 9 cases. Fig. 1 details the pathological findings in the physical examination (PE+) and the complementary tests (CT+).

Literature Review

The search chain identified 444 articles related to Chiari type I malformation, of which 19 reported data on neurotoxic symptomatology or semiology. Of these, 10 fulfilled the criteria needed to be included in our literature review.⁹⁻¹⁸

The different forms of otoneurological presentation of the CHM-I described in the literature are shown summarised by order of frequency of appearance in Table 2.⁹⁻¹⁸ The most common audiovestibular symptom was instability (49%), followed by vertigo (18%), nystagmus (15%) and hearing loss (15%). Within nystagmus, the most common seems to be horizontal nystagmus horizontal (74%), followed by vertical downwards (18%). The type of nystagmus (horizontal, vertical downwards or vertical upwards) found in the patients was not characterised in some studies. Likewise, not all the articles reported the laterality of the hearing loss, although it appears that bilateral hearing loss is more prevalent than unilateral (44% vs 30%).

Other audiovestibular signs are tinnitus (11%), aural fullness (10%) and hyperacusis (1%).

Discussion

According to the 2009 consensus document,⁶ the Chiari malformation classification includes 5 subtypes (0-IV), of which type I is undoubtedly the one diagnosed with greatest frequency in adults. Type II is necessarily associated with spinal dysraphism and, consequently, it is almost invariably diagnosed in childhood.⁸

The most frequent symptom in patients with CHM-I is suboccipital headache, which typically increases with Valsalva manoeuvres (60%-70%).⁸ In our series this symptom was found in 8 of the 9 patients (88.9%). Cerebellar

syndrome is also a common finding, described in almost 3/4 of the patients. It usually runs with instability and dysmetria and seems to be due to both the conflict of space in the posterior fossa and to the anomalous transmission of energy that happens during cardiac systole as a consequence of the decoupling of the spinal fluid volumetric buffer circuit.

Symptoms due to dysfunction of motor and sensory pathways or cranial nerves are not unknown. In fact, paresthesias in the upper limbs are the second symptom that patients usually report with greater frequency, appearing in 61% of the cases.⁶ In this series it was detected with the same prevalence as occipital-neck headache.

The affectionation of brainstem nerve nuclei, as well as of the nerve fascicles that run through them to reach the medulla or diencephalon, can provoke a variegated clinical picture with differing symptoms, which include sudden falls without loss of consciousness (drop attacks), apnoeas, dysphonia, dysphagia, dysarthria, heart arrhythmias or alteration in sphincter control.⁸ One of the patients in our series suffered one of these complex symptom sets, consisting of central apnoeas, episodes of loss of consciousness, psychomotor delay and dysphagia.

Cranial nerves are involved in 15%–25% of the patients with Chiari malformation in general¹ and in 20% of the CHM-I.¹¹ It can present as, among others, neuralgia of the trigeminal or glossopharyngeal nerves, vocal fold paralysis, dysarthria, hoarseness, sleep apnoea, cricopharyngeal achalasia, soft palate weakness, lingual atrophy, facial hypoesthesia, frequent aspirations and absence of gag reflex.^{11,19–21}

The association of syringomyelia is relatively frequent, although its prevalence and exact physiopathological mechanism are not completely known.²² Although syringomyelia can be asymptomatic in some cases, the usual clinical presentation is a progressive intramedullary syndrome. In our series, only 1 case presented syringomyelia, at level D4.

Audiovestibular Symptoms

Audiovestibular symptoms are not rare in patients with CHM-I. They usually begin in the 2nd or 3rd decade of life, insidiously and progressively. The mean age of the patients in our series coincides with these literature data, although it must be remembered that all of them presented other, non-audiovestibular symptoms of years of evolution. Besides, it is complex to differentiate the origin of these audiovestibular symptoms in these patients, given that it often is impossible to rule out the presence of concomitant peripheral disease.

In the majority of the studies reviewed, the characterisation of the audiovestibular symptoms was far from being optimal, probably as a result of a pragmatic approach from the neurological or neurosurgical points of view. Perhaps the clinical sign that was best systemised was nystagmus, which is considered typically vertical inferior or vertical downwards in CHM-I.⁸ However, systematic review of the literature seems to demonstrate that horizontal nystagmus is much more frequent: it appears in 74% of the patients and quadruples the presence of vertical inferior nystagmus (18%). Curiously, we were unable to detect nystagmus in any of the patients in our series, probably because of the intermittence that is typical of this exploratory sign.

The laterality of the hearing loss was described in 3/4 of the articles analysed. There was a dominance of bilateral auditory deficit over unilateral, in contrast to what was found in the present series, in which unilateral hearing loss was dominant. The onset of this auditory signs and symptoms (sudden, fluctuating, progressive, etc.) was mentioned in practically none of the studies.

In the literature, there was not normally a syndromatic classification of the vertigo that affects these patients. However, it is accepted that the vertigo characteristic in CHM-I is positional or triggered by head movements.¹ Our findings are congruent with this, given that positional vertigo affected a third of the patients in our series and accounted for 40% of the dizziness syndromes in these patients.

The systematisation of tinnitus or the sensation of aural fullness is even more deficient; neither the intensity nor the laterality of these symptoms is normally reported, which emphasises the importance of a multidisciplinary approach for this type of patients, allowing better characterisation and monitoring of the symptoms.

Neuro-Radiological Study

For the characterisation of the herniation of the cerebellar tonsils, McRae's line was used as a reference.²³ This is an imaginary line that is obtained joining the basion (anterior border of the foramen magnum over the middle line) with the opisthion (posterior border of the foramen magnum over the middle line). By consensus, a descent of the cerebellar tonsils >5mm below this line is considered pathological.¹ McRae's line is represented by the dashed line in Fig. 2.

The popularisation in the middle of the 1990s of NMR sequences sensitive to CSF flow, using phase contrast techniques with electrocardiographic triggers and dynamic images in closed cycle cine format, has made it possible to characterise the circulatory behaviour of CSF in the cranial-cervical junction in both normal subjects and patients affected by Chiari malformation. Under normal conditions, a significant percentage of CSF displacement that happens during a cardiac cycle (and that makes up one of the main mechanisms of encephalic volumetric buffering) occurs between the cisterna magna and cervical subarachnoid space, in the cranial-caudal direction in systole and caudal-cranial in diastole.^{24,25} In Chiari malformation various alterations have been found in the spinal fluid circulatory pattern, which has made it possible to go deeper into the physiopathology of this disease. In addition, the restriction of space that exists in the posterior fossa of these patients and the circulatory decoupling that takes place between the cisterna magna and the cervical subarachnoid space condition an increase in the peak of diastolic and systolic speed of the CSF in the foramen magnum.²⁶ This also conditions a reduction in the length of the systole and an increase in the duration of the diastole at the level of the cervical subarachnoid space. The presence of arachnoid adhesions in the cisterna magna provokes the appearance of CSF jets of anomalous direction, which collide against the cerebellar tonsils themselves and against the bulbo-medullary union. In addition, the caudal displacement that is produced in the cerebellar tonsils in these patients during systole is, as a mean, 13% higher than in healthy subjects. This is probably

a result of the incapability to balance the increase in volume produced during cardiac systole with CSF displacements in the cranial-cervical junction.

Several studies seem to demonstrate that the development of clinical signs and their intensity are related more to the presence of an anomalous spinal fluid circulation pattern than with the absolute degree of cerebellar tonsil descent.^{8,27-30} This relationship can be seen in our series: Patient 4, with a tonsillar descent just at the limit of normality (5 mm) presented an alteration in CSF flow dynamics and a poorly tolerated Ménière-like clinical picture; however, Patient 8, with a cerebellar tonsillar herniation of 22 mm (Fig. 2) showed a less severe alteration of CSF circulation and a clinical picture of hearing loss, vertigo and instability, subjectively well tolerated with the use of bilateral auditory prostheses. In this sense, studying CSF flow through cine-MR seems to be appearing as a diagnostic tool of great value in clinical practice, given that it provides information potentially relevant for both the selection of surgical candidates (above all in oligosymptomatic and asymptomatic patients⁸ with associated syringomyelia²²) and also for the determination of the most appropriate surgical technique,²⁷ especially in complex malformations of the cranial-cervical junction. Likewise, it permits monitoring the evolution of the patients managed conservatively and ensures the appropriate correction of spinal fluid circulation following surgical treatment.^{26,30} A specific situation in which this technique could be highly useful is the subjects with syringomyelia or clinical manifestations compatible with Chiari malformation, but without significant cerebellar tonsillar herniation (Chiari type 0). In these patients the cine-MR would be capable of detecting alterations in CSF circulation in the cranial-cervical union, backing the indication of a surgical treatment⁸ that would otherwise be controversial.

There are no studies relating audiovestibular symptoms with the findings of cine-MR in CHM-I or the evolution of otoneurological manifestations with the intensity of the problem present in CSF circulation. At any rate, controlled and randomised trials are needed to study the role of CSF flow in the diagnosis, prognosis and treatment of patients with Chiari malformation.

Treatment

Treatment of Chiari malformation attempts to improve the relation between the container and the content of the foramen magnum, and to normalise CSF flow in the cranial-cervical junction, so as to lessen the symptoms that these patients exhibit or, at least, to prevent their progression. There is a certain consensus in considering the possibility of surgical treatment for every patient with CHM-I and clearly correlated clinical signs and symptoms. Unfortunately, the evolution of the audiovestibular manifestations following surgery is not appropriately characterised in the literature. Furthermore, indicating surgical treatment for patients in which these symptoms are dominant is subject to controversy. The scenario for asymptomatic patients with syringomyelia is similar, as there is no unanimity of criteria for advising surgery, with the rates of indication ranging between 9% and 75%.⁸

In the absence of a clear compromise in the pre-stem circulation of the CSF, the procedure of choice is osseous decompression of the foramen magnum associated with an expander duraplasty with arachnoid preservation.³¹ Opening the arachnoids is generally unnecessary and should only be performed when there is evidence of an extensive arachnoid reaction that would condition the existence of asynchronous and multidirectional jets in the spinal fluid circulation of the cranial-cervical junction.

Conclusions

The fact that Chiari type I malformation presents with audiovestibular symptoms means that referral to the otoneurologist is relatively frequent for these patients. Consequently, becoming familiar with the symptoms of this disease is interesting, considering its differential diagnosis. Studying CSF flow through cine-MR could provide complementary information relevant for selecting surgical candidates and for monitoring surgical results in these patients.

Conflict of Interests

The authors have no conflicts of interest to declare.

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References

1. Fernández AA, Guerrero AI, Martínez MI, Vázquez ME, Fernández JB, Chesa i Octavio E, et al. Malformations of the craniocervical junction (Chiari type I and syringomyelia: classification, diagnosis and treatment). *BMC Musculoskelet Disord*. 2009;10 Suppl. 1:S1.
2. Cleland J. Contribution to the study of spina bifida, encephalocele and anencephalus. *J Anat Physiol*. 1883;17:257.
3. Chiari H. Über Veränderungen des Kleinhirns infolge von Hydrocephalie des Grosshirns. *Dtsch Med Wochenschr*. 1891;17:1172-5.
4. Arnold J. Myelocyste, Transposition von Gewebskeimen und Sympodie. *Beitr Path Anat Allgem Path*. 1894;16:1-28.
5. Speer MC, Enterline DS, Mehlretter L. Chiari type I malformation with or without syringomyelia: prevalence and genetics. *J Genet Couns*. 2003;12:297.
6. Amado Vázquez ME, Avellaneda Fernández A, Barrón Fernández J, Chesa i Octavio E, de la Cruz Labrado J, Escribano Silva M, et al. Malformaciones de la unión craneo-cervical (Chiari tipo I y siringomielia). Documento de consenso. Madrid: AWWE; 2009. p. 5.
7. Sarnat HB. Disorders of segmentation of the neural tube: Chiari malformations. *Handb Clin Neurol*. 2008;87:89.
8. Khoury C, Patterson MC, Dashe JF. Chiari malformations; 2013. UpToDate [Internet based]. Available from: <http://www.uptodate.com> [accessed 11.03.13].
9. Naya Gálvez MJ, Fraile Rodrigo JJ, Liesa RF, Vicente González EA, Garrido CM, Sampérez LC, et al. Otorhinolaryngologic manifestations in Chiari malformation. *Am J Otolaryngol*. 2002;23:99-104.
10. Kumar A, Patni AH, Charbel F. The Chiari I malformation and the neurologist. *Otol Neurotol*. 2002;23:727-35.

11. Steinbok P. Clinical features of Chiari I malformations. *Childs Nerv Syst.* 2004;20:329.
12. Chang CZ, Howng SL. Surgical outcome of Chiari I malformations, an experience sharing and literature review. *Kaohsiung J Med Sci.* 1999;15:659–64.
13. Hendrix RA, Bacon CK, Sclafani AP. Chiari-I malformation associated with asymmetric sensorineural hearing loss. *J Otolaryngol.* 1992;21:102–7.
14. Milhorat TH, Chou MW, Trinidad EM, Kula RW, Mandell M, Wolpert C, et al. Chiari I malformation redefined: clinical and radiographic findings for 364 symptomatic patients. *Neurosurgery.* 1999;44:1005–17.
15. Weber PC, Cass SP. Neurotologic manifestations of Chiari 1 malformation. *Otolaryngol Head Neck Surg.* 1993;109:853–60.
16. Bertrand RA, Martinez SN, Robert F. Vestibular manifestations of cerebellar ectopia (sub-group of Chiari I). *Adv Otorhinolaryngol.* 1973;19:355–66.
17. Chait GE, Barber HO. Arnold–Chiari malformation, some otoneurological features. *J Otolaryngol.* 1979;8:65–70.
18. Spooner JW, Baloh RW. Arnold–Chiari malformation: improvement in eye movements after surgical treatment. *Brain.* 1981;104:51–60.
19. Combarros O, Albarez de Arcaya A, Berciano J. Isolated unilateral hypoglossal nerve palsy: nine cases. *J Neurol.* 1998;245:98–100.
20. Dyste GN, Menezes AH, VanGilder JC. Syntomatic Chiari malformations. An analysis of presentation, management and long term outcome. *J Neurosurg.* 1989;71:159–68.
21. Erdogan E, Cansever T, Secer HI, Temiz C, Sirin S, Kabatas S, et al. The evaluation of surgical treatment options in the Chiari malformation type I. *Turk Neurosurg.* 2010;20:303–13.
22. Clarke EC, Stoodley MA, Bilston LE. Changes in temporal flow characteristics of CSF in Chiari malformation type I with and without syringomyelia: implications for theory of syrinx development. *J Neurosurg.* 2013;118:1135–40.
23. Rydell RE, Pulec JL. Arnold–Chiari malformation. Neurotologic symptoms. *Arch Otolaryngol.* 1971;94:8–12.
24. Urbizu A, Toma C, Poca MA, Sahuquillo J, Cuenca-León E, Cormand B, et al. Chiari malformation type I: a case–control association study of 58 developmental genes. *PLOS ONE.* 2013;8:e57241.
25. Cousins J, Haughton V. Motion of the cerebellar tonsils in the foramen magnum during the cardiac cycle. *Am J Neuroradiol.* 2009;30:1587–8.
26. Haughton VM, Korosec FR, Medow JE, Dolar MT, Iskandar BJ. Peak systolic and diastolic CSF velocity in the foramen magnum in adult patients with Chiari malformations and in normal controls participants. *Am J Neuroradiol.* 2003;24:169–76.
27. Panigrahi M, Reddy BP, Reddy AK, Reddy JJ. CSF flow study in Chiari I malformation. *Childs Nerv Syst.* 2004;20:336.
28. Ventureyra EC, Aziz HA, Vassilyadi M. The role of cine flow MRI in children with Chiari I malformation. *Childs Nerv Syst.* 2003;19:109.
29. Bhadelia RA, Frederick E, Patz S, Dubey P, Erbay SH, Do-Dai D, et al. Cough-associated headache in patients with Chiari I malformation: CSF flow analysis by means of cine phase-contrast MR imaging. *Am J Neuroradiol.* 2011;32:739–42.
30. Martin BA, Kalata W, Shaffer N, Fischer P, Luciano M, Loth F. Hydrodynamic and longitudinal impedance analysis of cerebrospinal fluid dynamics at the craniovertebral junction in type I Chiari malformation. *PLOS ONE.* 2013;8:e75335.
31. Sindou M, Gimbert E. Decompression for Chiari type I-malformation (with or without syringomyelia) by extreme lateral foramen magnum opening and expansile duraplasty with arachnoid preservation: comparison with other technical modalities (literature review). *Adv Tech Stand Neurosurg.* 2009;34:85–110.