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SPECIAL ANNUAL ISSUE

Clinical diagnosis—part I: what is really caused by Chiari I

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Abstract

Purpose Chiari malformation is a group of congenital malformations involving the brainstem, cerebellum, and upper spinal cord, frequently identified in both young adults and in children. Chiari I malformation (CM1), classically defined as a caudal displacement of the cerebellar tonsils through the foramen magnum into the spinal cord, is the most common clinical type. A syringomyelia can be associated at the time of the diagnosis or appear secondarily and manifest with medullary symptoms. The aim of this paper is to update the knowledge on clinical manifestations specifically related to Chiari I malformation with or without syringomyelia in the pediatric population.

Methods Current literature with focus on relevant clinical pediatric issues is reviewed and discussed, comparing with those related to adults; we include the results of a 10-year single-center experience on 600 CM1 patients.

Results and conclusions Herniation of the cerebellar tonsils may lead to significant clinical symptoms, including neck and cervical pain, short-lasting occipital "cough" headache, dizziness, and gait impairment; in children younger than 3 years, oropharyngeal symptoms are prevalent (sleep apnea, feeding problems) whereas in those older than 3 years, a higher incidence of cough headache and scoliosis is reported. CM1 clinical features, both in children and in adults, have in common the presence of anatomical deformities of the brainstem and cerebellum. Clinical myelopathy (sensory/autonomic disorders, motor weakness) can result from direct compression of the cervical spinal cord by the herniated cerebellar tonsils or can be due to the presence of a syrinx, reported in association with Chiari I between 35 and 75% of pediatric patients. Similarly, in our series (440 females, 160 males, 98% > 18 years), syringomyelia associated with Chiari I was ranging from 40 to 60% (respectively in asymptomatic and symptomatic groups); headache was reported in 65%. Sensory disturbances (48%), cranial nerve deficits (45%), motor weakness (32%), and autonomic disorders (35%) were the most frequent neurological signs in our cohort. In Chiari I malformation, cervical pain and occipital cough headache are the most characteristic presenting symptoms, both in old children and in adults; however, headache is often multifactorial, and CM1 patients can report a wide variety of non-specific symptoms and signs. Clinical diagnostic CM1 criteria, shared at the national and international level, are recommended with the aim to avoid consequent controversies on diagnosis and on surgical decision making.

Keywords Chiari I malformation · Hindbrain hernia · Clinical diagnosis · Chiari syndrome · Syringomyelia

Introduction

Chiari malformations constitute a group of entities with very different etiologies, pathophysiology, and clinical features that have in common the presence of anatomical deformities of the brainstem and cerebellum. Chiari I malformation (CM1) is classically defined as caudal displacement of the cerebellar tonsils at least 3 mm into the upper cervical spinal canal; it may be congenital or acquired. Although Chiari I more often presents symptomatically in early adult life, the widespread use of magnetic resonance imaging (MRI) has led to increased recognition of this disorder in children [1]. At the same time, given this

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increasing frequency of radiological diagnosis, care must be taken to differentiate between patients with recognizable symptoms and those with symptoms that are unrelated to the findings of tonsillar herniation on imaging. In the MRI era, Chiari I malformation in a child is frequently an incidental diagnosis without associated symptoms; in two studies of 49 and 53 children with Chiari I, more than 50% appeared asymptomatic and the rate of very young children was higher in asymptomatic groups [2, 3].

The aim of this paper is to review the current knowledge on clinical diagnosis more specifically related to pediatric presentation and the differences with that of adults. In this comparison, we include the results of a personal 10-year experience from data prospectively collected at the Centro Regionale Esperto Siringomielia-Sindrome di Chiari (CRESSC) in Torino, Italy.

Clinical manifestations

Patients with Chiari I may present with a variety of symptoms and signs, ranging from headache and neck pain to signs of brainstem compression, cerebellar syndrome, and myelopathy [4–6]. Clinical myelopathy can result from direct compression of the upper cord, by the herniated cerebellar tonsils, or can be due to the presence of a syrinx, reported in association with Chiari I between 35 and 75% of pediatric patients [7]. In a series of 500 CM1 children surgically treated, 57% were found to have syringomyelia, most commonly located in the cervicothoracic cord, while 39% had holocord syringes [8]. The major difference in pediatric clinical presentation between age groups is the more common occurrence of sleep apnea and feeding problems in younger patients: children younger than 3 years of age had significant oropharyngeal symptoms, whereas those older than 3 years had a higher incidence of cough headache and scoliosis [9]. Otherwise, the clinical manifestations of children and adults appear similar.

Pain It is the most common presenting symptom (60–70%), usually occipital and upper cervical, often induced or exacerbated by Valsalva maneuvers such as laughing, sneezing, and coughing. Other common symptoms include non-radicular pain in the shoulder, back, and limbs [10]. Older children tend to present with neurological symptoms attributable to the development of syringomyelia and the onset of cord symptoms [11].

Headache It is the most common complaint occurring in 80% of patients, less frequently infants and young children. In infants and in children who are unable to communicate verbally, headache may be manifested by crying and irritability [10]. In Chiari I malformation, headache is typically posterior, short lasting, exacerbated by cough or Valsalva maneuvers, or induced by cervical motion; it can be isolated or associated with vertigo, ataxia, limb weakness, or numbness.

Diagnostic criteria for headache attributed to Chiari I malformation were established by the International Classification of Headache Disorders, third edition (ICHD-3). These criteria are the evidence of causation demonstrated by at least two of the following three clinical criteria:

1. Headache has developed in temporal relation to the CM1 or led to its discovery (a), headache has resolved within 3 months after successful treatment of the CM1 (b), either or both.
2. Headache has one or more of the following three characteristics: precipitated by cough or Valsalva-like maneuver (a), occipital or suboccipital location (b), lasting < 5 min (c).
3. Headache is associated with other symptoms and/or clinical signs of the brainstem, cerebellar, lower cranial nerve, and/or cervical spinal cord dysfunction.

Radiological CM1 criteria by MRI requires 5 mm caudal descent of the cerebellar tonsils or 3 mm caudal descent of the cerebellar tonsils plus crowding of the subarachnoid space at the craniocervical junction as evidenced by compression of the cerebrospinal fluid (CSF) spaces posterior and lateral to the cerebellum, reduced height of the supraocciput, increased slope of the tentorium, or kinking of the medulla oblongata [12].

Syncope may occur with cough; Pascual has posited that headache in Chiari I is caused by the descent of the cerebellar tonsils during cough that compresses the upper cervical nerve roots [13]. However, headache is often polymorphous and poorly characterized, and distinguishing headache actually related to Chiari I from other forms that coexist independently with Chiari I (i.e., primary headache, such as migraine or tension headache) is mandatory [14].

Neurological signs may be divided into three different presentations: brainstem, spinal cord, and cerebellar syndrome, reported in 22, 65, and 11% respectively in an adult series [15]. Presenting signs include motor and sensory loss (30–92%), hyporeflexia (38%), hyperreflexia (40–52%), clonus (18%), Babinski response (28%), ataxia (20–40%), respiratory irregularities (10%), and nystagmus (23–70%) [10]. Focal motor deficits, such as claw hand, are very unusual as presenting signs, reported in few Chiari I children associated with syrinx [16, 17]. Sensory-neural hearing loss as a presenting symptom is also a relatively rare (37%) and well-characterized phenomenon, with a 28% post-operative improvement rate from the data available, although the majority not in journals commonly read by the neurological community [18]. Acute neurological presentation in previously asymptomatic Chiari I patients is also rarely reported: a short series of three pediatric cases, two of three following a mild head injury [19], and one

Table 1 Demographic data in patients with Chiari I malformation (CRESSC)

	All, $n = 600$ (100%)	Symptomatic, $n = 240$ (40%)	Asymptomatic, $n = 360$ (60%)
Age (%)			
Pediatric (< 18 years)	16 (3)	5 (30)	11 (70)
Adult (18–60 years)	429 (71)	172 (40)	257 (60)
Over 60 years	155 (26)	63 (40)	92 (60)
Gender (%)			
Male	160 (27)	76 (47)	84 (53)
Female	440 (73)	164 (37)	276 (63)
MRI morphology (%)			
Tonsillar herniation ≥5 mm	558 (93)	235 (42)	323 (58)
Tonsillar herniation 3–4 mm	32 (5)	5 (17)	27 (83)
Tonsillar herniation ≤ 3 mm	10 (2)	0	10 (100)
Types (%)			
CMI A—CMI + Syr	262 (44)	156 (60)	106 (40)
CMI B—isolated	338 (56)	84 (25)	254 (75)

Abbreviations: *CMI* Chiari I malformation, *Syr* syrinx (syringomyelia or hydromyelia)

case after upper respiratory viral infection have been described [20].

Brainstem syndrome Ocular and otological signs and symptoms are reported in approximatively 70% of patients, such as oscillopsia, nystagmus, diplopia, isolated abducens nerve palsy, field cuts and tinnitus, fluctuating hearing loss, vertigo, and nausea [10]. In particular, nystagmus, both vertical and downbeating, occur in 23–70% [21]. Less commonly reported manifestations are hypertension and sinus bradycardia. Lower cranial nerve dysfunction is reported in 15–26% of patients: oropharyngeal dysfunction is common (palatal weakness, tongue atrophy, cricopharyngeal achalasia); in contrast, glossopharyngeal and trigeminal are rare. Children under 3 years old typically present with lower cranial nerve dysfunction, manifesting with poor feeding, failure to thrive, recurrent aspiration pneumonia, dysphagia, or stridor; vocal cord paralysis with stridor may be present in 8% [10]. Drop attacks and central sleep apnea in young children with sudden death have been reported; in a large series of 500 patients, 5% presented with central apnea confirmed by a sleep study [8]. Dysphagia occurred in 4/11 adults (36%), all four with fluids, when prospectively studied before and after surgical decompression for Chiari I in a recent report [22], with a frequency similar to the ranges reported in previous children [8] and adult studies [22].

Spinal cord syndrome Spinal cord syndrome is the result of direct cord compression or a syrinx. Loss of pain and

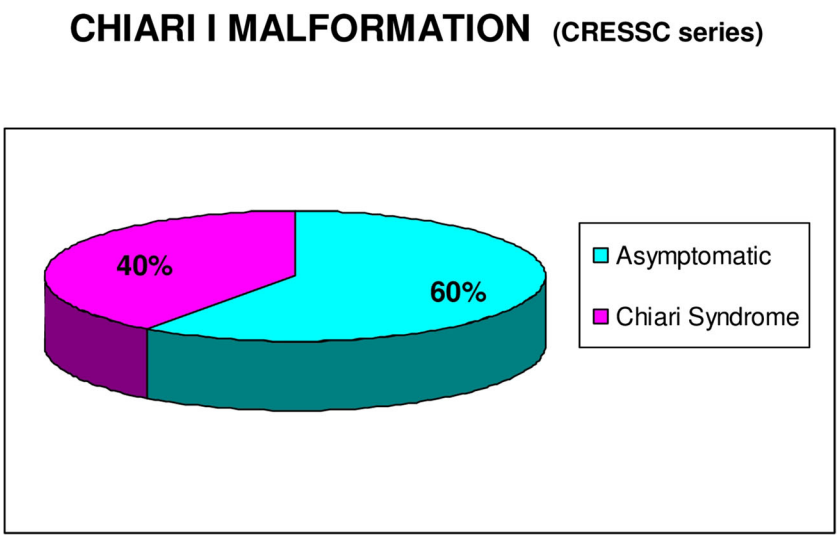

Fig. 1 Percentage of symptomatic and asymptomatic patients in CRESSC series ($n = 600$)

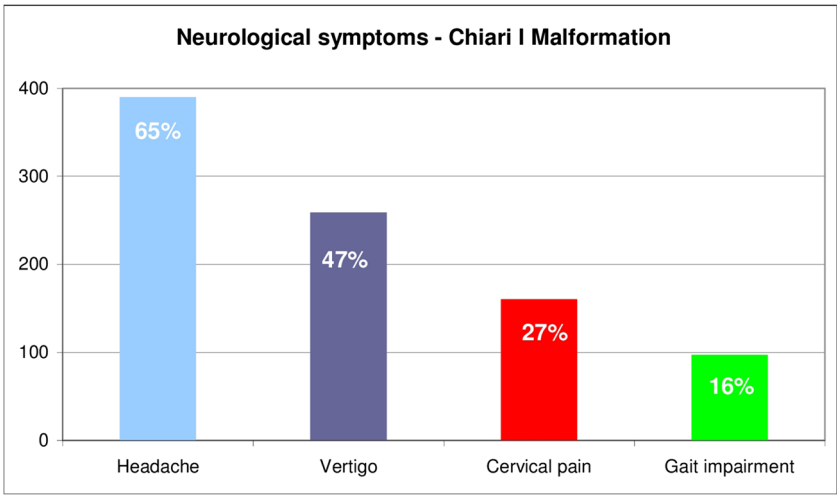

Fig. 2 Major neurological symptoms in Chiari I malformation (CRESSC series)

temperature sensation in the upper extremities, with preserved light touch and proprioception, in Chiari I should alert to the presence of associated syringomyelia; scoliosis in a Chiari I patient may also signal the presence of a syrinx, particularly in children. In fact, scoliosis was identified in four out of five syringomyelia patients under 20 years compared with one in six older patients and was the presenting feature of syringomyelia in about two thirds of children [23, 24]. A left convex thoracic curve in a male, abnormal or absent abdominal reflexes, or non-dermatomal pain should also raise the suspicion of an underlying syrinx. There is a causal relationship between syringomyelia and scoliosis, supported by many studies, but it is not always clear whether a syrinx is the cause of or a consequence of scoliosis; furthermore, which is the interaction and the primary anomaly between hindbrain hernia, syringomyelia and scoliosis is also unknown [25]. Tubbs et al. reported scoliosis in 18% of pediatric Chiari I associated with syrinx [8]; in 2011, Brockmeyer described only 2 of 22 patients with scoliosis and Chiari I malformation without syrinx at MRI [26]. In other series, syrinx and associated scoliosis is much more commonly reported in children less than 15 years of age (81%) than in adults (45%), but while only 6% of children complain of associated pain, 39% of adults did; moreover, older age and increased curve size predict poor response to Chiari decompression [27]. Finally, older children (13–18 years) presented with a higher percentage of cord symptoms or syrinx-related neurological deficits (50%) compared with 20% in the 0 to 12 year age group [11].

Cerebellar syndrome Cerebellar compression causes ataxia, limb clumsiness, and nystagmus [6]. Ataxia and cerebellar signs are described in 40% of a pediatric series of 20 patients operated on for Chiari I [11], but only in 6% of 49 adults followed before and after Chiari I surgery [14].

Personal experience

A review of the experience at our Department during the last 10 years has been performed. Between March 2009 and March 2019, 600 patients (440 females, 160 males, 98% > 18 years) with Chiari I malformation have been screened at the Centro Regionale Esperto Siringomielia-Sindrome di Chiari (CRESSC), Neuroscience Department, Turin, Italy (Protocol n. 7837, 01/02/2010; protocol n. 52554, 20/05/2015, Città della Salute e della Scienza di Torino Hospital, Turin). All patients underwent a multidisciplinary evaluation by the CRESSC team (neurologist, neurophysiologist, neurosurgeon, neuroradiologist, physiatrist, neuro-urologist, speech pathologist) to support the diagnosis and the following therapy (surgery/drugs/rehabilitation), according to the shared Recommendations identified by the Interregional Chiari and Syringomyelia Consortium [28].

Five hundred and ninety patients (98%) met ICHD-3 radiological criteria for Chiari I malformation definition with tonsillar herniation ≥ 3–5 mm [12]. Demographics, MRI parameters (morphology/level), and clinical phenotypes are reported in symptomatic (40%) and asymptomatic (60%) cases (Table 1; Fig. 1). All patients were prospectively evaluated with clinical scales (disability, pain, quality of life) and neuroimaging (brain and whole spinal cord cine-MRI), before and after surgery (posterior fossa decompression and duraplasty).

A syrinx (syringomyelia or hydromyelia) associated with Chiari I was reported in 44% of all cases, 60% symptomatic; isolated Chiari was 56%, only 25% symptomatic. Major neurological symptoms were headache (65%), dizziness (47%), cervical pain (27%), and loss of balance (16%) (Fig. 2). Scoliosis was reported in 29% of all CM1 patients, 43 patients presented with scoliosis, CM1, and syringomyelia (25%). Two hundred and forty patients (40%) had pathological objective neurological

Table 2 Neurological signs in patients with symptomatic Chiari I, Chiari Syndrome (CRESSC series)

	All *n*=240	Surgery (PFDD) *n*=142	No Surgery *n*= 98
Sensory disorders (%) [a]	**115 (48)**	**80 (56)**	**35 (36)**
Sensory Levels	52 (22)	27 (19)	25 (26)
Hypoesthesia UL	55 (23)	30 (21)	25 (26)
Hypoesthesia LL	34 (14)	30 (21)	4 (4)
Paraesthesia/Dysesthesia	19 (8)	10 (7)	9 (9)
Radicolar/Truncal deficit	16 (6)	3 (2)	13 (13)
Dissociated Thermoalgesic loss	10 (4)	10 (7)	0
Deep sensitive disorders	2 (1)	1 (2)	1 (1)
Motor weakness (%) [a]	**78 (32.5)**	**52 (36)**	**26 (26)**
Hyposthenia UL	22 (9)	17 (12)	5 (5)
Upper MTN signs	36 (15)	31 (22)	5 (5)
Lower MTN signs	46 (19)	30 (21)	16 (16)
Hyposthenia LL	15 (6)	15 (11)	0
Spasticity	18 (7.5)	15 (11)	3 (3)
Muscle atrophy	12 (5)	10 (7)	2 (2)
Paraparesis	12 (5)	12 (8)	0
Tetraparesis	10 (3)	10 (7)	0
Cranial Nerves disorders (%) [a]	**109 (45)**	**77 (54)**	**32 (33)**
Dysphagia	62 (26)	40 (28)	22 (22)
Nystagmus	50 (21)	43 (30)	7 (7)
Dyspnea	25 (10)	25 (18)	0
Visual disorders	15 (6)	10 (7)	5 (5)
Hearing loss	9 (4)	5 (3.5)	4 (4)
Sleep Disorders (%)	**36 (15)**	**31 (22)**	**5 (5)**
OSAS	24 (10)	20 (14)	4 (4)
CSAS	12 (5)	12 (8)	0
Autonomic Disorders (%) [a]	**74 (31)**	**70 (49)**	**4 (4)**
Incontinence	22 (9)	20 (14)	2 (2)
Urge-incontinence	52 (22)	50 (35)	2 (2)
Neurogenic bladder	10 (4)	10 (7)	0
Gait Ataxia/cerebellar signs	**18 (7.5)**	**13 (9)**	**5 (5)**

[a] Patients who have at least one of neurological signs.

Abbreviations: UL-Upper Limbs; LL-Lower Limbs; MTN-Motoneurons; PFDD-Posterior Fossa Decompression and Duraplasty.

examination, defining the "symptomatic Chiari" or Chiari syndrome. The most frequent neurological signs were sensory disturbances (48%), cranial nerve deficits (45%), motor weakness (32%), and autonomic disorders (35%). Paraparesis and tetraparesis, prevalent in the surgery group, was 11% and 7% respectively (Table 2).

There is a general consensus in the literature that the clinical diagnosis in Chiari I malformation cannot be done without specific neurological symptoms and signs that define the Chiari syndrome. Symptomatic cases are more common in adults than in children, particularly in Chiari I with syringomyelia. In our study cohort, 60% of Chiari I with syringomyelia patients were symptomatic, with the same frequency reported in Taylor et al. [29]. We noted hypoesthesia at upper limbs in 21–23%, lower motoneuron signs (such as muscle atrophy, areflexia, and weakness at upper limbs) in 19–21%, and upper motoneuron signs (such as spasticity, iperreflexia, Babinski reflex, and clonus at lower limbs) in 15–22%; autonomic disorders (in prevalence bladder urge incontinence) were much more common in syringomyelia patients, with a defined neurogenic bladder in 4–7% (Table 2, Fig. 3). In the early stage, the spinal signs were rather often unilateral and syringomyelia presented with apparent involvement of a single segmental level and focal motor deficits; isolated lower motoneuron signs at upper limbs, mimicking

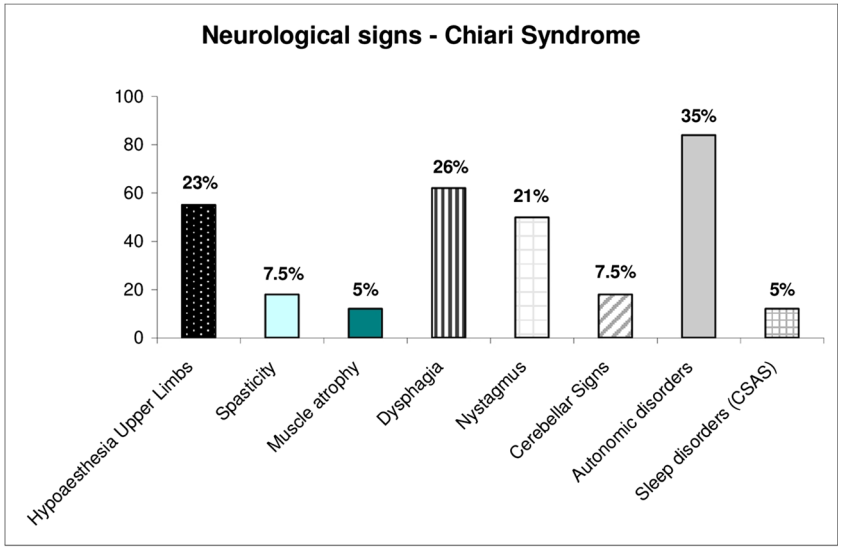

Fig. 3 Percentage of neurological signs in Chiari syndrome (CRESSC series)

entrapment mononeuropathies (such as ulnar neuropathy at the elbow), were reported both in our cohort and in literature [17, 30]. In the chronic stage, upper motoneuron signs at the lower limbs could be also present, mimicking typical clinical manifestations of amyotrophic lateral sclerosis, another multilevel neuromuscular disease. Brainstem syndromes were frequently reported in Chiari syndrome and in siringobulbia, with nystagmus in 21–30% and dysphagia in 26–28%, similarly shown in previous series [21, 22]; central sleep apnea syndromes (CSAS) were evidenced in 5–8% by polysomnography, with the same frequency reported in a large pediatric study [8].

Conclusions

Chiari I malformation can present with a wide variety of symptoms, also non-specific, with consequent controversies on diagnosis. The potential anatomy changes during maturation also influence the decision to treat a younger versus older patient with Chiari I, particularly in isolated forms, not associated with syringomyelia, and in asymptomatic or minimally symptomatic cases.

The analysis of the literature and our results from a prospective CM1 series of 600 patients indicate that cervical pain and headache are the most characteristic presentation symptoms, both in older children and in adults; however, headache is often multifactorial and the Chiari-distinctive, posterior "cough" headache is about rare in the study cohorts.

We recommend shared clinical diagnostic CM1 criteria, standardized at National and International level (i.e., ICHD-3), and multidisciplinary assessments with the aim to support the diagnosis and the surgical decision-making.

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Compliance with ethical standards

Conflict of interest There was no financial support nor industry affiliations involved in this work. None of the authors has any personal or institutional financial interest in drugs, materials, or devices.

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