



# Chiari Malformation Type 1: Clinical Features, Diagnosis and Treatment- An Italian Case Series

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## Abstract

Chiari type 1 Malformation (CM 1) is characterized by caudal displacement of the cerebellar tonsils through the foramen magnum with obstruction of cerebrospinal fluid (CSF) outflow. The lack of guidelines treatment and surgical indication of the patients affected by CM 1 can cause controversies in management. Moreover, CM1 can occur in asymptomatic individuals. We multidisciplinary evaluated 341 adult patients suffering from CM 1 in a collaborative project of a Chiari Special Outpatient Service started between neurologists, neurosurgeons and neuroradiologists. Clinical presentation, diagnosis, and treatment of these cases are discussed. A multidisciplinary approach should be the first step for a correct evaluation of patients with CM 1.

**Keywords:** Chiari malformation type 1; Cerebrospinal fluid; MIDAS

## Introduction

Chiari type 1 Malformation (CM 1) is characterized by caudal displacement of the cerebellar tonsils through the foramen magnum associated with crowding of the craniocervical junction with obstruction of cerebrospinal fluid (CSF) outflow. This variation of hind-brain development can sometimes occur in asymptomatic individuals [1]. Conventional treatment for symptomatic CM1 is surgical decompression but in several cases the natural history of the condition is relatively benign and non-progressive [1]. Most patients with CM 1 have not surgical indication and they need to be treated by clinicians, especially neurologists [2]. CM 1 can cause a variety of symptoms most likely cough headache which is caused by the sagging of the cerebellar tonsils below the foramen magnum [3,4]. CM 1 can be associated with syringomyelia and spinal deformities, including scoliosis [5].

## Aim of the Study

In 2015 started a collaborative project in which our group of neurologists, neurosurgeons and neuroradiologists contribute to create a Chiari Special Outpatient Service [2]. This was set up to provide a multidisciplinary evaluation, treatment and follow-up of patient suffering from CM 1. Here we reported our experience, observations and outcome of the patients.

## Patients and Methods

Patients with diagnosis of CM 1 documented by a previous MRI imaging scan were evaluated at our Institution Chiari Special Outpatient Service. Clinical history, charts and neuroimaging of the patients were reviewed. The visit, neurological examination and review of the charts was performed by a neurologist who decided during the evaluation to eventually brief discuss the case with a neurosurgeon at the same time. By the end of the visit, the two specialists suggested the treatment. Patients without indication for surgical treatment were managed by the neurologist whereas patients with surgery indication were discussed in a new following multidisciplinary meeting and the surgical treatment was defined. Visual analogue scale (VAS) [6] was applied in all patients to measure patient's pain intensity and migraine disability assessment test (MIDAS) was performed in patient with chronic migraine [7].

## Results

From September 2015 to May 2017, 341 adult patients (99 males, 242 females) mean age 44.7 years (range 16-65), suffering from CM 1 were multidisciplinary evaluated. Average duration of the symptoms prior to evaluation at our Outpatient Service was 7.6 years (range 11 months-12 years)

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61 (17.8%) patients out of 341 evaluated had had a previous surgical treatment and of the remaining 280 patients 87 had had surgical indications during previous evaluations performed elsewhere 337 patients (99%) reported headache with different quality (pulsating, dull, throbbing, even binding or lancinating) and duration (from few minutes to several days) Dizziness was present in 296 (87%), nausea in 139 (41%), tinnitus 92 (27%), gait impairment in 57 (17%), neck pain 303 (89%), numbness 139 (41%), dysphagia 51 (15%).

A suboccipital-occipital headache of variable quality and duration aggravated by Valsalva's maneuver, effort, cough, or postural changes was described in 286 (84%). Extremity paresthesia was referred by 269 (79%) Pain in the arms was present in 228 (67%) and restriction of neck movements in 265 (78%).

Previous history of 55 patients (16%) had foramen magnum decompression (FMD) performed elsewhere with improvement of the symptoms in 50 cases. The six patients underwent elsewhere a filum terminale sectioning all without improvement.

Due to chronic pain 33 patients (9.6%) had medication overuse (MO) with 17 of them involving drugs of dependence.

At the first visit VAS resulted in moderate pain in 143 patients (42%), severe in 93 (27%) and mild in 105 (31%). Among the 131 patients without surgical indication re-evaluated after a three months period of pharmacological treatment the VAS resulted in mild (66 patients, 50%), moderate (44 patients, 33%), severe (21 patients, 17%). MIDAS questionnaire was filled in by the 41 patients suffering from chronic migraine. In 39 patients the score was 21+ (grade IV, severe disability) while in 2 it was between 6 and 10 (grade II, mild disability).

As regards neurological findings, in 13 (4%) out of 341 patients who had previously received a diagnosis of CM 1 the degree of tonsillar herniation on magnetic resonance imaging was 3 up to 5 mm below McRae's line which can be defined as "low-lying cerebellar tonsils" Chiari Malformation 0 classification instead of CM 1. Syringomyelia associated with 235 patients (69%), 31 patients with dissociated sensory loss, 14 with weakness of the hands and 11 with cord atrophy. With cord syndrome 2 (0.5%) had tethered and 3 (0.9%) had hydrocephalus 27 (8%) patients on 33 patients with MO underwent after evaluation an inpatient detoxification program at our Institute with pharmacological treatment after withdrawal from MO. The MIDAS questionnaire filled in by these 27 patients after a three months period of prophylactic treatment resulted in improvement with 49% in 21 patients.

After our evaluation 17 patients (5%) had surgical indication and 8 of them underwent a posterior fossa decompression at our Institute [8]. The surgical treatment was performed in all patients by the same two senior neurosurgeons with the same procedure: after occipital bone craniectomy, the posterior arch of the atlas was routinely removed, but C2 and related muscle attachments were preserved. The dura was opened with a midline incision, the arachnoid plane was preserved; a tonsil resection or coagulation was performed when the tonsils extended behind the posterior arch of the axis or in case of arachnoiditis. A dural patch was therefore tightly sutured just at the craniovertebral passage. Fibrin glue and different sealant products were applied on the suture to prevent cerebrospinal fluid leak [8].

After our prescription of further examinations 135 patients were re-evaluated: MRI with peak velocity of CSF flow in both the anterior

and posterior subarachnoid space through foramen magnum, motor and somatosensory cortical evoked potentials (MEP, SSEP); 102 of them had a significant reduction of CSF flow in posterior fossa while 24 had loss in SSEP wave form and 9 had central conduction abnormalities at MEP.

We decide to perform an investigation for central apneas with poly-sonography in over weighted/obese 15 patients resulting in OSAS (obstructive sleep apnea syndrome) in 7, while in 8 patients was normal.

For adjustment of pharmacological treatment 189 patients are still in follow-up. As regards conservative management of the patients, we administrated antidepressant (venlafaxine, amitriptyline, escitalopram), antiepileptic drugs (gabapentin, pregabalin, topiramate), atypical neuroleptic (quetiapine) at low dosage for treatment of chronic pain and pain related mood disorders.

## Discussion

Managing of patients with CM 1 can be difficult, first because some may have no symptoms. Other patients show poor or mild symptoms that often are not Chiari Malformation related. Moreover many patients receive surgical indication while they don't have any. The main problem is the lack of guidelines in the surgical indication of CM 1. We established in our study group few guidelines for surgical indication: patients with CM1 symptoms (cough/Valsalva manoeuvre precipitated headache, cerebellar dysfunction, dysphagia, incoordination, motor or sensory disturbances) associated with evolving syringomyelia or with rapid clinical deterioration associated with significant reduction of CSF flow in posterior fossa and clinical/neurological signs of intracranial hypertension were suitable for craniovertebral decompression. Despite many debates about indication for surgery in CM1, we believe that treatment is indicated just in patients with symptomatic CM1 and progressive syringomyelia while conservative management should be the most frequent choice in all the other patients. Along the 61 patients who had a previous surgical treatment before our evaluation, we found out that only 7 of them fitted our indication guidelines for surgery. According to literature and in agreement with neurosurgeons, surgical treatment is indicated just in patients with symptomatic CM1 and progressive syringomyelia [8]. In our case serie only few cases needed a surgical treatment and conservative management was generally proposed as the best choice. Clinical presentations of CM 1 can include a wide variety of manifestations with headache most common [2,9,10]. Headache is the most common and leading symptom in CM1 [2]. The typical CM1-related headache is usually occipital-suboccipital severe and induced by cough, laughing, straining, Valsalva manoeuvre. Other CM2 related symptoms are dysphagia, visual disturbance, sleep apnea; dizziness, gait impairment, pain in the arms and extremity paresthesias are also observed but many of these features can be reported by patients affected by other disease such as cervicopathy, multiple sclerosis and mielopathy; so it is difficult, except for typical CM1 headache, to operate a specific distinction and differential diagnosis can be hard. The high frequency of not CM1 related symptoms such as migraine (41 patients), nausea (139 patients), neck pain (303 patients), restriction of neck movements (265 patients) in our case serie can be explained remembering that patients with CM1 have prevalence of common primary headache disorder and cervicopathy similar to that of the general population [2,9,10]. A multidisciplinary approach should be the first step for a correct evaluation of patients with CM 1 [8].

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