



## Headache Patterns in Chiari Malformations - Epidemiology, Pathophysiology, and Treatment Insights: a literature Review

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**Categoria:** Cefaleias Secundárias

### Introduction

Chiari malformations comprise a group of congenital conditions characterized by anatomic abnormalities of the cerebellum, brainstem and craniocervical junction. The specific structural defects identified allow further classification in type I to type III. Clinical manifestations can range from asymptomatic to severe neurologic impairment, and headache is one of the most common presentations.

### Objective

To describe the patterns of headache associated with Chiari malformations, detailing the epidemiology, underlying pathophysiology, radiological findings and treatment options based on a literature review. Methodology This is a literature review which analyzed articles in English published in the last 10 years, between August and September 2023. The descriptors used were ("Arnold" AND "chiari" AND "malformation" AND "headache") in the databases PubMed and Embase. A total of 15 articles were found to be relevant to the topic.

### Results

Out of the 15 articles analyzed, two estimated the prevalence of headache among patients diagnosed with Chiari Malformation I (CM), as defined by the cerebellar tonsils located at least 5 mm below the level of the foramen magnum in imaging exams, to vary between 32% and 92%. However, determining the prevalence and incidence of Chiari Malformation (CM) itself, which may or may not be associated with headache, is a challenge, as many of the prevalence estimates are based on imaging tests. Regarding the pathophysiology of CM, it can be summarized as a divergence between the contents of the brain and those of the supratentorial cranial vault and posterior fossa. As a rule, the pathological mechanisms that can lead to CM1 fall into 4 groups: 1) structural anomalies of the skull base, 2) abnormal segmentation of the vertebral and cervical bodies, 3) overcrowding caused by a cranial vault and/or posterior fossa and 4) excess tissue in the posterior fossa or throughout the cranial vault. These alterations compromise the circulation of cerebrospinal fluid (CSF), leading to a poorly regulated CSF pressure: this is the mechanism thought to explain the so-called "classic" Chiari headache, characterized by intense occipital pain triggered by Valsalva-like maneuvers. A study has identified 1,913 patients with Chiari Malformation (CM), of whom 1430 complained of headaches. While 912 (63,77%) had typical headaches, the most prevalent being headaches associated with coughing, another 453 patients (31,6%) had atypical headaches, such as migraine, tension headaches, cluster headaches, trigeminal neuralgia, etc. Regarding the treatment of typical headaches associated with CM, the study compared conservative or surgical treatment, comprising 3 techniques: 1) extradural decompression (DED), 2) intradural decompression (DID) and 3) decompression of the foramen magnum; the rate of improvement in headaches was much higher with surgical treatment. In addition, one article found a correlation between radiological findings in magnetic resonance imaging (MRI) and headache improvement, showing that the greater the preoperative tonsillar descent, the greater the improvement in the minimum intensity of headache after decompression of the foramen magnum.

### Conclusion

CM is closely associated with symptomatic cases of headache. However, determining the overall prevalence and incidence of CM, whether related to headaches or not, remains a challenge due to the dependence on imaging tests for diagnosis. The pathophysiology of CM is linked to various structural anomalies and disrupts the circulation of cerebrospinal fluid, leading to a range of symptoms. These results highlight the complexity of CM and the need for adapted diagnostic and treatment strategies to address both the malformation and the associated symptoms, particularly headaches.

**Keywords:** Chiari Malformation; Valsalva Headache; Literature Review; Chiari Malformation treatment.