

# Pearls & Oysters: Cough headache secondary to Chiari malformation type I

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## PEARLS

- Cough headache is a rare condition, but its presence should prompt thorough evaluation for intracranial pathology, given a high prevalence of secondary causes of headache in this population.
- Chiari malformation type 1 is a common diagnosis in which many patients are asymptomatic, though headache is the most common neurologic complaint when symptoms are seen.

## OY-STER

- Headaches are exceptionally common; the coexistence of headache and Chiari malformation type 1 does not alone implicate the malformation as the cause of the headache. Headache symptoms that are positional or associated with transient increases in intracranial pressure (e.g., cough, Valsalva) may indicate association of the headache with the malformation.

**CASE REPORT** A 17-year-old girl with migraine headaches and Noonan syndrome presented for neurologic consultation regarding worsened headaches in the setting of a hospitalization for pneumonia. The patient reported a 3-year history of paroxysmal, debilitating, midline headaches typically less than 1 minute in duration, which were increased in frequency and severity at the time of consultation. Pain began at the base of the occiput and then spread anteriorly to the brow line. These brief headaches were associated with coughing, laughing, and neck extension. They were different in nature from her migraine headaches, which were prolonged and associated with photophobia and vomiting. In addition, the patient reported more recent problems with choking while eating. Physical examination was unremarkable with the exception of difficulty with tandem gait. Brain MRI revealed 20 mm of tonsillar herniation below the foramen magnum, with crowding of the foramen magnum, consistent with Chiari malformation type 1 (CM-I) (figure). A diagnosis of cough headache secondary to CM-I was made. Indomethacin was started at a dose of 25 mg twice

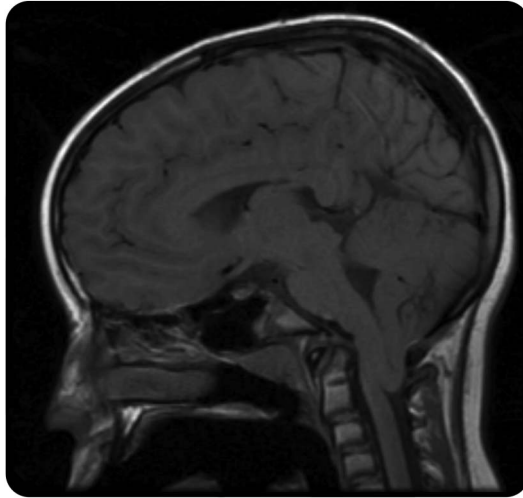
daily as needed. Upon follow-up 1 month later, headaches had significantly decreased in frequency.

**DISCUSSION** Headaches directly associated with cough may be primary or secondary. According to International Classification of Headache Disorders (ICHD-2) diagnostic criteria, primary cough headaches are paroxysmal headaches brought on by coughing, straining, or Valsalva, lasting 1 second to 30 minutes, and not attributable to another disorder.<sup>1</sup> Primary cough headache most frequently presents as sudden-onset, bilateral, posterior, explosive pain without associated nausea, vomiting, photophobia, or phonophobia.<sup>2,3</sup> Cough headache with similar clinical characteristics, but caused by another disorder or structural lesion, is referred to as secondary cough headache. Of those with cough headache, the majority are adults with primary cough headache.

Reported secondary causes of cough headache include CM-I, subdural hematoma, acute sphenoid sinusitis, and other posterior fossa structural lesions including primary intracranial malignancy, metastatic malignancy, arachnoid cyst, and os odontoideum.<sup>2,3</sup> Of 7,100 consecutive adult patients in a Taiwanese headache clinic evaluated over a period of 8 years, only 83 (1.2%) were diagnosed with cough headache. Intracranial pathology was found in only 9 of these patients (10.8% of those with cough headache), including 2 with symptoms attributable to CM-I.<sup>2</sup> Similar cough headache prevalence of 1% was found in 6,412 consecutive adult patients from a headache clinic in Spain, though in this cohort, 58.8% of patients with cough headache demonstrated intracranial pathology on MRI. Of these, 80% had CM-I.<sup>3</sup> Similar data regarding the prevalence of cough headache are not available specifically for pediatric populations nor are data available for the incidence of cough headache in patients with CM-I. Overall, epidemiologic studies point to a very low prevalence of cough headache, even within the context of specialty headache clinics.

Differentiation between primary and secondary cough headache (of which headache attributed to CM-I is a subtype) is important as secondary causes may require intervention beyond pain management.

**Figure** Parasagittal T1-weighted MRI of the head demonstrates 20-mm tonsillar herniation, crowding of the foramen magnum, and compression of the cervicomedullary junction



In the ICHD-2 criteria for CM-I-related headache, one criterion includes improvement of the headache within 3 months following surgery, which presents a challenge for definitive presurgical diagnosis.<sup>1</sup> It can be difficult to differentiate primary from secondary cough headache based on clinical symptoms alone. In the Taiwanese cohort, there were no differences in sex, age at headache onset, pain location, pain characteristics, or associated features between patients with primary and secondary cough headache.<sup>2</sup> In the Spanish cohort, the duration of headache symptoms prior to presentation was greater in those with secondary cough headache (5 years vs 11 months).<sup>3</sup> Common features among patients with cough headache in both cohorts include sudden, severe, posterior headaches lasting less than a minute, most commonly arising in the fifth or sixth decade of life.

Current research suggests that both primary and secondary cough headache are related to paroxysmal increases in intracranial pressure. In the case of secondary cough headache, structural abnormalities may lead to alteration in CSF flow dynamics during cough or Valsalva, resulting in the development of the signs and symptoms of cough headache. The underlying pathophysiology behind primary cough headache is less clear. Lane and Davies<sup>4</sup> reported a modified Valsalva maneuver that was successful in differentiation of primary from secondary cough headache. Sixteen patients with cough headache were asked to exhale into the connecting tube of an aneroid sphygmomanometer to a pressure of 60 mm Hg. This elicited headache in 11 patients, 10 of whom had identifiable intracranial pathology on MRI, 8 of those having CM-I. Further validation of this technique could be

helpful in differentiating primary from secondary cough headache without expensive neuroimaging studies.

CM-I itself is an uncommon finding; in a retrospective study of 5,248 pediatric patients receiving MRI of the head or cervical spine, 51 were found to have tonsillar herniation greater than 5 mm below the foramen magnum, consistent with CM-I, or approximately 9.7 per 1,000 children imaged. Of those, 37% were asymptomatic. Headache, found in 55% of patients, was the most common presenting symptom. Distinction between secondary cough headache and other headache types was not reported.<sup>5</sup>

CM-I is typically a stable malformation. A retrospective study of 147 children with CM-I followed for an average of 4.6 years found no change in mean tonsillar herniation or number of symptomatic patients between initial consultation and final follow-up; however, 7 children developed a syrinx during that time.<sup>6</sup> In a patient with cough headache and any other neurologic symptom consistent with CM-I or focal neurologic deficit, suspicion should be elevated for an underlying etiology. Of particular interest to the case presented here, CM-I may be a rare complication of Noonan syndrome.<sup>7</sup>

The optimal treatment of cough headache, whether primary or secondary, is poorly understood. Most patients with primary cough headache have remission of symptoms after 4 years without treatment. Individual episodes, however, can be debilitating despite the brief nature of the headaches, making symptomatic treatment useful. Patients with intracranial pathology may require surgical management for resolution of symptoms. First-line pharmacological therapy is typically indomethacin in a total daily dose of 50–150 mg. The mechanism of action for indomethacin in cough headache is unknown, though its effect may be mediated through decreasing intracranial pressure.<sup>8</sup> Both aforementioned large case series of patients with cough headache have shown that patients with primary cough headache respond more frequently to indomethacin (78%–100%) than do those with a secondary cause (0%–38%).<sup>2,3</sup> Topiramate, methysergide, propranolol, naproxen, and IV metoclopramide have also been shown to be useful for symptomatic relief of cough headache.<sup>8</sup>

Surgical management of CM-I typically involves posterior cranial fossa decompression with an enlargement of the foramen magnum. In a series of 177 adult patients undergoing primary decompression for Chiari malformation, strain-related headache was the second most common presenting symptom (54.2%). Three months postintervention, 98.9% of those with headache had improvement of symptoms, although at 1 year benefit was sustained in only 62.5%. Surgical intervention showed a 0.6% acute (within 7 days of surgery)

complication rate and a 10.1% delayed complication rate. Pseudomeningocele was the most common complication in both the acute and delayed setting.<sup>9</sup> Of note, in a pediatric population of 130 patients surgically treated for CM-I, 83% had postoperative relief of symptoms.<sup>10</sup> The limited evidence available suggests that surgery is the most efficacious treatment for CM-I-associated headache, though due to the nature of the intervention, this should be considered carefully. While the data are mixed regarding the efficacy of indomethacin in patients with CM-I-associated cough headache, we continue to recommend a trial of indomethacin as first-line therapy due to its mild side effect profile in comparison to surgical intervention and due to challenges in distinguishing secondary from primary cough headache, which may demonstrate spontaneous resolution over time without surgical intervention.

#### AUTHOR CONTRIBUTIONS

J.E. Bates: drafting/ revising the manuscript, study concept or design, analysis or interpretation of data, acquisition of data. Dr. Augustine: drafting/ revising of manuscript, study concept or design, study supervision or coordination.

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#### DISCLOSURE

The authors report no disclosures relevant to the manuscript. Go to [Neurology.org](http://Neurology.org) for full disclosures.

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