

# Nummular Headache and Its Surgical Treatment

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**Background:** Nummular headache (NH) is an uncommon primary headache characterized by pain limited to a precise small area of the scalp. There is no global consensus on its pathogenesis, but its extracranial origin is the most accepted theory. Moreover, peripheral mechanism is supported by the overlapping symptomatology of secondary forms of NH and is well described in the literature. However, a standard effective treatment is still lacking.

**Methods:** A literature search according to Preferred Reporting Items for Systematic Reviews and Meta-Analysis guidelines was conducted to evaluate surgical strategies for NH. Inclusion criteria were English language, diagnosis of primary NH according to International Classification of Headache Disorders, 3rd Edition, or of secondary NH, and follow-up at a minimum of 3 months. The treatment had to consist of peripheral surgery.

**Results:** One hundred eighty-seven records were identified after duplicates were removed, 15 full-text articles were assessed for eligibility, and 4 records were selected for inclusion. A total of 53 patients were included in this review, 50 of whom were diagnosed with primary NH. The general positive response after surgery (>50% reduction in occipital migraine headaches) was about 70.0% for primary NH, while secondary NH always showed complete pain relief. However, many variations in patient selection and type of surgery were described.

**Conclusions:** Neurovascular relationship in the extracranial tissues seems to be involved in the onset of NH. However, only limited data from meager literature and from few patients are currently available. Shared multicentric research protocols are badly required. (*Plast Reconstr Surg Glob Open* 2020;8:e2989; doi: 10.1097/GOX.0000000000002989; Published online 28 July 2020.)

## INTRODUCTION

Since first described in 2002 by Pareja et al,<sup>1</sup> nummular headache (NH) has been the center of many debates. Its uncommon presentation creates difficulty in providing reliable answers. To date, about 400 cases have been published, of which only a few studies had an acceptable number of patients, allowing for statistically significant results.<sup>2,3</sup> Most of the studies in the literature, in fact, are retrospective case reports or case series with <20 patients.<sup>4-6</sup>

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According to ICHD-3,<sup>7</sup> NH is a well-circumscribed headache and is also called “coin-shaped headache.” It is part of epicranial headaches,<sup>8</sup> and is characterized by a chronic pain that lasts for a highly variable time and by being limited to a precise, small round (or elliptical) area of the scalp (diameter: 1–6 cm) that cannot be better accounted for by any another ICHD-3 diagnosis. All underlying structural lesions must be absent.

The most important debates concerning NH arise from the analysis of the official definition. The particular topography and sensory abnormalities, such as variable combinations of hypoesthesia, dysesthesia, paresthesia, allodynia, and/or tenderness, suggest an origin from epicranial tissues. However, for many authors, the central mechanism does not seem to be completely excludable. In support of this theory, the ineffectiveness of local anesthetic injections and the extension of the area of pain beyond the midline as opposed to multiple coexistent (albeit rare) locations<sup>9</sup> have been noted.

Another topic of discussion regarding NH concerns the absolute overlap of its symptoms with some cases secondary to underlying extracranial and intracranial structural lesions, including Langerhans cell histiocytosis,<sup>10</sup> arachnoid cyst,<sup>11</sup> subtentorial meningioma,<sup>12</sup> localized

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calcific hematoma of the scalp,<sup>13</sup> craniosynostosis,<sup>14</sup> and aneurysmatic dilatations of arteries supplying blood to scalp.<sup>15</sup> Many of these secondary forms certainly support the peripheral mechanism for NH, considering especially that, in animal models, pain fibers crossing through the skull or running longitudinally within the endosteum are described.<sup>16,17</sup>

To date, NH pathogenesis remains unclear, and there is a lack of a standard treatment protocol.<sup>18</sup> Antiepileptic medications such as Gabapentin<sup>19</sup> at a high dose and onabotulinum toxin type A<sup>3</sup> seem to be the only therapies able to make at least a partial remission. However, targeted surgical therapy has often proved to be decisive in secondary cases. This review aims to explore studies in which surgical therapy of peripheral tissues has been used to treat primary and secondary NH.

## METHODS

### Search Criteria

A thorough literature search was conducted in March 2020 across four databases (PubMed, MEDLINE, Scopus, and Cochrane Library), without date limits. The search terms used to identify all citations reporting outcomes of NH were “nummular headache” and “coin-shaped headache”. Results were independently analyzed by 2 of the authors. Double references were excluded. After reading titles and abstracts of citations, a list of articles was generated for review. Additional articles reviewing the reference list of relevant abstracts were included. This study was conducted according to Preferred Reporting Items for Systematic Reviews and Meta-Analysis (PRISMA) guidelines for systematic reviews.

### Selection Criteria

Before conducting literature search, inclusion and exclusion criteria were defined to avoid selection bias.

#### Inclusion Criteria

- English language;
- Diagnosis of NH according to International Classification of Headache Disease, 3rd edition;
- Diagnosis of secondary NH;
- Preoperative and outcome data with a follow-up of at least 6 months;
- Studies on peripheral surgery; and
- Primary data from case reports, prospective/retrospective observational studies, and randomized control trials.

#### Exclusion Criteria

- Studies on radiosurgery, cryosurgery, and botulinum toxin injection without surgery.

## RESULTS

A total of 128 citations from PubMed, 153 from Scopus, and 3 from Cochrane Library were initially identified. After a title and abstract review was analyzed by 2 different reviewers and a further search on the bibliography of

full-text articles was done, 15 records were considered relevant. Full-text analysis excluded further 11 articles. Only 4 articles of the initial research, published from 2013 to 2018, fulfilled our inclusion criteria and were included in the systematic review (Fig. 1).

Of the 4 selected studies (Table 1), 1 was a case series<sup>20</sup> and the other 3 were case reports.<sup>2,10,15,21</sup> A total of 53 patients were included in the review on peripheral surgical management of NH. The sample size ranged from 1 to 49 patients. Demographic data examined were sex, with a total prevalence of women (42 women and 11 men), and age, reported as mean and as range by Guyuron et al (mean age, 45 years; range, 21–65 years). The youngest patient was 12 years old and the oldest was 67. Patients included were those with an ICHD-3 diagnosis of NH or fulfilling the clinical criteria of NH but with an underlying structural peripheral lesion diagnosed after suspected of having NH. However, most of the patients not only suffered from NH but also reported a variable headache history (92.8% in Guyuron et al).<sup>20</sup> Specific preoperative investigations were described in 3 of 4 studies: careful clinical and neurological examination associated to a Doppler signal within the painful area<sup>20</sup>; skeletal studies, immunoglobulin level and paraprotein search, computed tomography scan, and magnetic resonance imaging (MRI)<sup>10</sup>; and blood tests, including erythrocyte sedimentation rate, C-reactive protein, MRI, and ultrasound examination<sup>15</sup>. Characteristics such as dimensions, type of pain, and specific alterations of the affected area were described in only 3 case reports, while Guyuron et al did not report any information. Surgical strategies were also different with respect to the type of diagnosis: complete surgical removal of 2 eosinophilic granulomas (focal histiocytosis) and 1 fusiform aneurysm of the superficial temporal artery was described for secondary cases of NH<sup>10,15</sup>; Dai et al removed the surface of the scalp from the symptomatic area to the tendinous part of the epicranial muscle and grafted it with healthy skin. Guyuron et al made an arterectomy/artery cauterization and a neurolysis/neurectomy through a 0.6- to 1.2-cm scalp incision in correspondence to the Doppler signal in all their patients.

Surgical excision of the identified lesions led to the complete disappearance of the symptoms in 3 of the 3 secondary NH cases.<sup>10,15</sup> Dai et al reported pain recurrence several days after excision of the clinically identified affected area. Finally, Guyuron et al<sup>20</sup> reported a significant reduction in headache frequency and severity, with a surgical success rate of about 70%. However, 92.8% of patients had concomitantly undergone unilateral or bilateral auriculotemporal decompression or had undergone a previously not well-defined surgery for migraine. Follow-up ranged from 6 months to 5 years. Complications were not described in any of the studies except for the study by Lopez-Ruiz et al,<sup>15</sup> which reported a gradually receding mild discomfort over the surgical area after aneurysm excision.

## DISCUSSION

NH is described as an uncommon, even rare, type of primary headache. It affects a small area and patients often do not require treatment. However, cases characterized by a moderate to severe pain have been reported,<sup>2,22</sup> and

when a concurrent or unrelated headache diagnosis coexists, NH tends to manifest independently.<sup>23</sup> Refractoriness to multiple therapeutic approaches is significant,<sup>24,25</sup> and, as for other types of headache, standard effective treatment is still lacking.

There are suggestions that NH might be a localized neuralgia of peripheral sensory nerves,<sup>26,27</sup> but the exact pathogenesis is still unclear. Currently, the identification of an underlying anatomical cause leads to a clear distinction between primary and secondary NH. However, a wide range of diagnostic investigations have not been described for the majority of the patients, and, even when neuroimaging procedures (such as MRI) are performed, a diagnosis delay can occur for years due to the nonvisibility of some lesions in common radiological neuroimaging. In fact, imaging targeted to the extracranial vessels is not usually performed. However, our review highlights a predominant vascular origin of focal characteristic pain, both in primary and secondary NH. Therefore, it can be assumed that, in many studies, similar lesions have never been diagnosed. The series of patients described by Guyuron et al, characterized by 49

patients with a positive Doppler signal in the exact point or significantly close to the indicated painful area, may even raise the doubt that a close neurovascular relationship is the cause of all, if not most, primary NH identified. Furthermore, arterectomy for the elimination of irritation on peripheral nerves that are responsible for migraine headache or trigeminal neuralgia has recently been widely reported.<sup>28-34</sup> However, in disagreement with the literature data<sup>18,23</sup> (range, 15.1%–46.7%), the series of Guyuron’s patients presented a very high percentage of NH coexistent with other headaches: 92.8% of all patients who had undergone concomitant (36.6% auriculotemporal decompression) or had undergone a previously not well-defined surgery for migraine. The outcome may therefore have been conditioned.

This review has limitations: a limited number of selectable research articles, and a limited number of patients and their unbalanced distribution between studies (3 case reports and 1 case series of 49 patients). Differences in the accuracy of preoperative diagnostic assessments are not negligible, given the need to carefully evaluate the presence of secondary causes of localized epicranial pain.

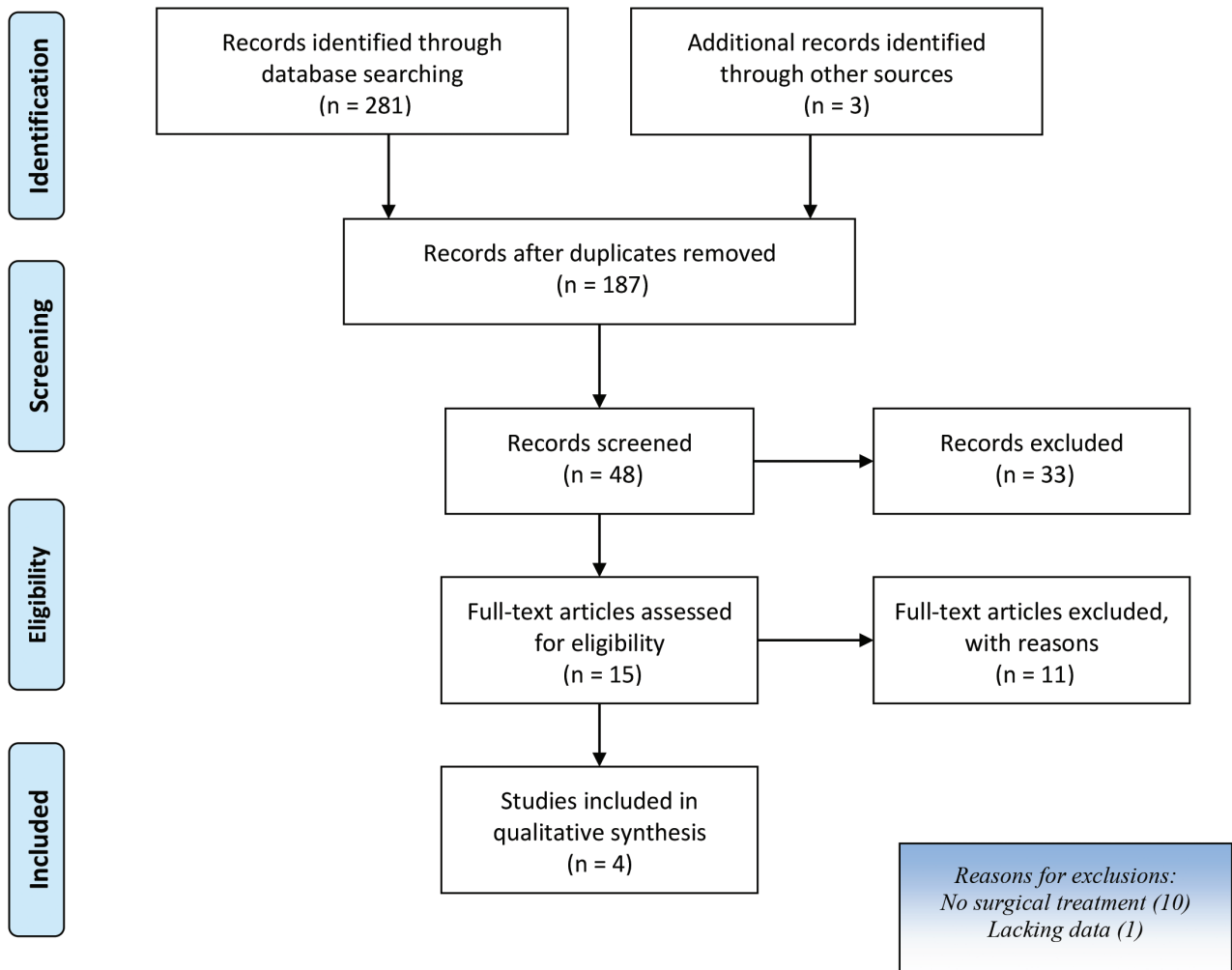


Fig. 1. PRISMA flow diagram.

**Table 1. Studies Included in Qualitative Synthesis**

Study/ Ref No.	Year	Type	Sample (patients)	Diagnostic Strategy	Surgical Strategy	Outcomes Measurements	Follow-up (mo)	Results	Complications
López-Ruiz et al. <sup>15</sup>	2014	Case report	67-year-old man with a 25-year history of depressive pain and local allodynia in a small round 4-cm area at the right temple. No or at least mild relief with medications.	Inspection and palpation revealed a throbbing bulge at the center of this area that was easily compressible and tender. Physical and neurological examinations were otherwise normal. Blood tests, including erythrocyte sedimentation rate and C-reactive protein, did not show any abnormalities. MRI of the brain was also normal. An ultrasound examination of the right STA revealed a fusiform aneurysm of 2.5 cm in diameter located at the origin of the frontal branch, just where the bulge could be palpated.	Complete surgical removal of the aneurysm.	VAS preoperative and postoperative diameter painful area duration, frequency, intensity	8-mo follow up	Frequency: 2–3 episodes per week Duration: 3 hours, if left untreated. Occasionally, for days. Intensity: severe (8/10). Histopathologic examination of the excised specimen showed fibrous atherosclerotic plaques but no signs of giant cell arteritis. After surgery, the patient remained asymptomatic.	
Guyuron et al. <sup>20</sup>	2018	Care series	49 patients (42 women and 7 men) ICHD diagnosis; with an average age of 45 y (range 21–65 y)	Doppler exam. Nerve block.	All patients had an associated Doppler signal within the identified area of pain and subsequently, under local anesthesia, underwent surgical arterectomy or cauterization plus neurolysis, independently from nerve block success.	Data on site-specific preoperative NH frequency, severity, duration, and headache-free days were collected. Data of postoperative headache frequency, severity, duration, and headache-free days were obtained. Headache index was calculated by multiplying frequency by severity and length of headache.	The average follow-up was 16 months with a range of 8–33 mo	Side of headache: Right for 55% Left for 44% 7.2% of patients underwent NH surgical treatment alone; 92.8% of patients underwent concomitant or previous migraine surgeries. NH frequency significantly decreased by an average of 10.7 days, from 19.7 to 9 days (54.3% reduction; $P < 0.001$ ). Significant increase in the number of headache-free days per month (10 vs. 21; $P < 0.001$ ). NH severity was significantly improved with an average reduction of 3.5 from 8.2/10 to 4.7/10 based on the visual analog scale ( $P < 0.001$ ). NH duration decreased by 0.3 hours ( $P = 0.4$ ). Headache index decreased by 39.6%, from 378.6 to 228.4 ( $P < 0.05$ ). n = 15 patients (30.6%) nonresponders; n = 22 patients (44.9%) reduction in headache index score by 50%; n = 12 patients (24.5%) free from headache	There were no complications identified during the follow-up period.

(Continued)

Table 1. (Continued)

Study/ Ref No.	Year	Type	Sample (patients)	Diagnostic Strategy	Surgical Strategy	Outcomes Measurements	Follow-up (mo)	Results	Complications
Silva Rosas et al. <sup>10</sup>	2018	Case report	2 male patients Patient 1: A 35-year-old man with a 4-month history of pain (continuous sensation of ice) in an area of 5 cm diameter in the right parietal region. No headache history. Patient 2: A 12-year-old boy with a 3-month history of pain in an oval-shaped area of 3 cm in the left lateral frontal region. No headache history.	Patient 1: Skeletal studies, immunoglobulin level, and paraprotein results were normal. A CT scan and MRI showed a lesion underlying the symptomatic area. Patient 2: Examination of the area was normal, with no swelling and no skin lesions, apart from allodynia in the affected area. Skull radiography, CT scan, and MRI showed a circumscribed lesion underlying the symptomatic area. Skeletal survey, immunoglobulins, and protein immunoelectrophoresis were normal.	Patient 1: Cranietomy with a margin of healthy bone and a cranioplasty with acrylic. Patient 2: A craniectomy was performed, leaving a healthy bone margin	VAS	Patient 1: 5 years Patient 2: 6 months' follow-up.	Patient 1: Preoperative Intensity: 5/10 (He had a partial response to gabapentin at a dose of 900 mg per day) Frequency: continuous Histopathologic examination showed eosinophilic granuloma (histiocytosis) with complete resection. Postoperatively, the patient was immediately free of pain and was fully recuperated 2 days after surgery. Patient 2: Preoperative Intensity: 3/10–9/10 in case of contact. Histopathologic examination showed eosinophilic granuloma (focal histiocytosis) with complete resection. Pain relief was immediate. After surgery, the headache disappeared, but several days later, the pain reappeared in a circular area (diameter, 5 cm) in the left temporal region. The new symptomatic area overlapped with the former one. All the other characteristics were the same as before.	None
Dai et al. <sup>21</sup>	2013	Case report	1 patient: 63-year-old man suffering from pain in a focal head area of 10-cm diameter in the vertex after taking a cold shower in 1994. Hypertension for 10 years. No or at least mild relief with nerve blocks, acupuncture, and a variety of medications.	None.	In 2004, surgical removal from the scalp to the tendinous part of the epicranial muscle of the symptomatic area and skin grafting.	VAS preoperative and postoperative diameter painful area	NA		None

CT, computed tomography; STA, superficial temporal artery; VAS, visual analogue scale.

Since certainties about NHs are lacking, a more careful assessment of patients presenting with this type of headache is mandatory, although the pathology is often not as disabling as other forms tend to be. Specific peripheral vascular examination (ultrasonographic or angiographic studies) could easily become part of the diagnostic investigation before undertaking any therapeutic strategy. The knowledge derived from studying other forms of headache can support researchers in identifying the most effective specific treatment. Medications are often expensive and associated with non-negligible side effects. Arterectomy is a minimally invasive procedure that can be performed under local anesthesia without any noteworthy complications. It seems to be effective in some forms of NH, especially when a pharmacological approach has failed.

### CONCLUSIONS

Research on NH is still very scarce. Greater attention should be paid to the analysis of patients who present the characteristic symptomatology. Shared multicentric research protocols could overcome the difficulties related to the limited number of patients suffering from this uncommon pathologic condition.

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