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Nummular headache: a narrative review

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ABSTRACT

Background: Nummular headache (NH) is a primary headache disorder, although secondary cases have also been reported. It was first described in 2002 and included in the International Classification of Headache Disorders, 3rd Edition (ICHD-3), 2018.

Methods: This narrative review examines epidemiological and clinical features, secondary cases, pathogenesis, and treatment of NH, drawing on updated literature. We conducted a systematic review searching the PubMed database and carefully reviewing the reference lists of all identified articles.

Results: Although NH is considered a rare condition, its incidence and prevalence are likely underreported. More than 700 cases have been described so far, and in a tertiary headache clinic, the diagnosis of NH is not uncommon. In other settings, some cases may be missed, and many patients with NH do not seek medical attention due to mild pain intensity. The clinical features include continuous or intermittent pain, generally of mild to moderate intensity, sometimes with exacerbations. The pain is localized in a round or elliptic area of the scalp, typically measuring 1-6 cm, mainly situated in the parietal, temporal, or occipital regions. Although most cases are unilateral, bilateral or midline localizations have also been reported.

Conclusions: The pathogenesis of NH is poorly understood; however, some clinical data suggest a peripheral origin of pain, such as a dysfunction of C-fibers in the epicranial cutaneous nerves. Diagnosis, primarily clinical, requires exclusion of other causes through comprehensive patient history, clinical examination, neuroimaging, and blood tests. Secondary cases due to underlying lesions, including systemic diseases or previous surgical treatments, must be ruled out. The most commonly used and effective prophylactic treatments are onabotulinumtoxinA and gabapentin, although various other drugs and non-pharmacological treatments have been proposed over the years.

Key words: nummular headache, epicrania, secondary cases, onabotulinumtoxinA, gabapentin.

Introduction

Nummular headache (NH) is a rare and distinctive type of headache, characterized by mild but persistent pain typically located in a small, well-defined, coin-shaped area of the scalp. The anatomic features of pain led to the choice of the term "nummular", derived from the Latin "*nummus*", meaning "coin", reflecting the characteristic presentation of the pain's location. This headache type was first described by Pareja *et al.* in 2002 (1) and, in 2004, was included in the Appendix of The International Classification of Headache Disorders (ICHD-2) at point A13.7.1. (2)

Following the publication of several case reports and case series, NH was recognized as a separate clinical entity and included in the main body of the International Classification of Headache Disorders, 3rd Edition (ICHD-3) beta (2013), (3) and ICHD-3 (2018), (4) within "Other Primary Headaches", at point 4.8 (**Table 1**).

The aim of this article is to review the epidemiological and clinical features of NH, including cases in children and adolescents, as well as secondary cases. It also discusses potential pathogenic mechanisms of this entity and provides an updated narrative review of proposed treatments. A systematic review was performed using the PubMed database, with search terms including "nummular headache", "epicrania", "epicranial headache", "coin-shaped headache", "secondary nummular headache", "gabapentin", and "onabotulinumtoxinA". The reference lists of the identified articles were also carefully examined. Only articles written in English were included, except for one article published in Spanish. (5)

Epidemiology

Due to the relatively recent description and to the frequent association with other primary headaches, epidemiological data on NH are limited, and no population-based studies are available. Most of the information derives from case reports and small or large clinical series, mainly collected by Spanish authors. One of the first large case series reported an incidence of NH of 4.6% out of 1,560 headache patients over three years. (6) Another large series involving 5,515 patients evaluated in an outpatient clinic over ten years found that 225 (4.1%) were diagnosed with NH. (7) In a subsequent series, an annual incidence of 6.64 cases per 100,000 population and a prevalence of 0.03% within the urban area served by the hospital were reported. (5) Notably, all these series were conducted by Spanish clinicians with specific expertise in diagnosing NH. Cases may be missed, and the diagnosis underestimated, especially when seen by headache specialists who are not experts in this condition. Furthermore, many patients with NH may not seek medical attention due to the mild intensity of their pain.

According to one review on this clinical entity, (8) more than 540 cases of NH had been described up to that point. The female-to-male (F/M) ratio showed a slight female prevalence of 1.6:1, which is comparable to previous studies where the range varied from 0.7:1 (9) to 1.8:1. (10) An exception is the second case series by Pareja *et al.*, (11) which reported a much higher F/M ratio of 3.67:1. The mean age at onset was approximately 48 years, with a wide range from 4 to 86 years. A relatively uniform distribution across different age groups has also been reported. (5)

A prospective, observational study conducted in Southern India found no significant differences in epidemiological or clinical features between the study population and previously reported data. (12) This observation supports the concept of NH as a primary headache with no distinctive features across different populations. An Italian study (9) involving 19 patients diagnosed with NH (11 males and 8 females) reported an average headache onset at approximately 39 years, with no significant gender differences in clinical presentation. However, the time from headache onset to diagnosis was significantly longer in men than in women (13.5 *vs.* 0.9 years).

Treatment issues of NH have been discussed in a recent case series published in 2023 (13) and in a new case report from 2024, (14) bringing the total number of published cases to over 720. However, some cases might have been included in more than one publication. These data suggest that NH is an uncommon, but not very rare, headache that is not associated with a specific period of life.

A frequent association with previous or concurrent diagnoses of other primary or secondary headaches – including migraine, tension-type headache (TTH), medication-overuse headache (MOH), or primary stabbing headache – has been reported in approximately half of NH patients at diagnosis (ranging from 46.7% to 54.1%). (15-16) Conversely, trigeminal neuralgia (9-11) or occipital neuralgia (17) are rarely associated with NH. An earlier update (16) and a case series (7) reported that in about 12.8% of NH cases, a history of head trauma was identified as a potential precipitating factor. No data are available regarding the latency period between trauma and the onset of headache.

Clinical features and diagnostic criteria

In NH, pain is localized to a limited area of the scalp, typically with a round or, more rarely, elliptical shape and well-defined borders. The diameter of the affected area generally ranges from 1 to 6 cm. This area is most often located in the parietal region, but other locations are also common, including the occipital or temporal regions, and more rarely, the frontal area. Regarding laterality, unilateral pain is most frequently reported; however, cases of bifocal, (7,18-22) multifocal, (23-25) or crossing the midline (6,7,9,11,17,26,27) locations have also been documented. Notably, the pain is usually situated in regions innervated by multiple cranial nerves. The involved area tends to remain stable over time, even over long periods.

The pain intensity is usually described as gradually increasing from the onset. (28) Although the pain can be continuous or intermittent during the day, it remains chronic in approximately 75% of cases, sometimes with sporadic, temporary remissions. (6) The temporal course of the disease may be continuous and unremitting, episodic, or follow a remitting pattern that eventually evolves into a continuous form. The pain intensity is typically mild to moderate and is described by most patients as pressing or throbbing; however, some patients may experience severe pain, characterized as stabbing or burning, sometimes accompanied by brief exacerbations or superimposed paroxysms. (6-9,12,15,20,26) There is no information suggesting a circadian pattern. A peculiar case of menstrual-related NH has been reported. (29)

Sensory symptoms within the painful area, such as hypoesthesia, hyperesthesia, dysesthesia, tenderness, and allodynia, may also be experienced. (6,28) Typically, NH is not associated with migraine-like autonomic features such as nausea, vomiting, photophobia, or phonophobia, and very rarely with lacrimation or rhinorrhea. (26) Early reports on NH primarily described its clinical features, detailing the symptoms reported by small patient groups. One of the first case series was published by Pareja *et al.*, (11) followed shortly by other case reports. Some reports described peculiar cases, such as patients with trophic changes of the skin or hair, like skin atrophy, alopecia, or focal hair heterochromia (30-32) or local skin temperature changes within the scalp region involved in pain sensation. (33) Other reports described symptoms different from those initially observed, including high-intensity pain with severe exacerbations. (20) Variations in pain intensity were sometimes spontaneous or triggered by local stimuli or other factors such as head movements, physical effort, Valsalva maneuver, (9,34) coughing, or sexual activity. (35) Some cases also exhibited migraine-like features. (36) A pressure algometer used to assess the pressure pain threshold in the affected area, compared to other cranial regions in NH patients, showed a lower pain threshold in the painful area. (37)

Since its first observations, NH has been classified among primary headaches. Establishing the diagnosis requires exclusion of systemic diseases, endovascular interventions, (14,38-40) or underlying structural lesions through a comprehensive clinical history, general and neurological examination (including assessment of touch and pain sensitivity within the symptomatic area), blood tests (evaluating inflammatory markers and immunology screening), and neuroimaging. Differential diagnosis must particularly rule out other conditions labeled as epicranias, such as epicrania fugax, (8) supraorbital neuralgia, or occipital neuralgia. If the pain area is circumscribed, temporal arteritis can be distinguished from NH using inflammatory markers. Although NH and primary stabbing headache may share some symptoms – particularly episodic stabbing pain – the temporal pattern of NH is typically chronic and continuous.

In **Table 2**, the demographic and clinical features of NH patients from major case series (including at least 10 patients) are summarized.

Table 1. Diagnostic criteria of the International Classification of Headache Disorders, 3rd Edition, 2018 (retrieved from: https://ichd-3.org/other-primary-headache-disorders/4-8-nummular-headache/).

Nummular headache
Description:
Pain of highly variable duration, but often chronic, in a small circumscribed area of the scalp and in the absence of any underlying structural lesion.
Diagnostic criteria:
A. Continuous or intermittent head pain fulfilling criterion B
B. Felt exclusively in an area of the scalp, with all of the following four characteristics:
1. sharply-contoured
2. fixed in size and shape
3. round or elliptical
4. 1-6 cm in diameter
C. Not better accounted for by another ICHD-3 diagnosis.

ICHD-3, International Classification of Headache Disorders, 3rd Edition, 2018.

Table 2. Demographic and clinical features of NH patients reported in the main case series.

Ref.	n	F/M	Age at onset, yrs (M±SD or range)	Association with other primary headache (%)	Associated precipitating factors (%)	Site of pain (%)	Laterali-zation of pain (%)	Shape (%)	Size, cm (M±SD or range)	Quality of pain (%)	Intensity of pain, VAS (M±SD or range)	Disease course (%)	Associated sensitive symtoms (%)
1	13	1.6:1	52 (26-70)	54	Head trauma (31)	Pa (62), Oc (23), Te (15)	Mono (100)	Ci (70), El (30)	3.3 (2-6)	Pr (69), S (31)	3.0 (1-7)	CU (62), Ep (38)	Dysesthesia (69)
5	83	1.02:1	46 (20-86)	Tension-type headache (2)	n.r.	Pa (47), Te (24), Fr (12), Oc (6), Te-Pa (6), Pa-Oc (2.5), Fr-Te (2.5)	Mono (95), Midline (4)	Ci (82), El (13), Other (4)	2.4±0.7	Pr (59), S (20), Th (14), B (6), Others (4)	5.3 (2-9)	CU (51), Ep (26), R (18)	Hypoesthesia (60), Hyperesthesia (40)
7	225	1.8:1	49.6±18.3	n.r.	Trauma (13)	Pa (31), Oc (24), Fr (20), Te (10), V (3)	Mono (90.3), Bil (0), Midline (9.3)	Ci (81), El (19)	4.4±1.3	Pr (59), B (18), S (17), T (5)	5.2±1.6	CU (43), Ep (23), Undef. (34)	Allodynia (36), Hyperesthesia (31)
9	19	0.7:1	40±13.4	Migraine (42), Tension-type headache (11), Trigeminal neuralgia (11)	n.r.	Oc (32), Pa (26), V (16), Te (11), Fr (5)	Mono (74), Midline (26)	n.r.	n.r.	S (32), Co (26), Th (21), B (21)	6.3 (2-10)	CU (47), R (53)	Hypoestesia (5)
11	14	3.7:1	38 (13-72)	Migraine (14), Trigeminal neuralgia (7)	Benign head injury (7)	Pa (50), Te (36), Fr (7), Oc (7)	Mono (93), Midline (7)	Ci (92), El (8)	1-6	Undef.	Mild (64), Moderate (29), Severe (14)	CU (50), Ep (50)	Allodynia (21), Hypoesthesia (21), Hyperalgesia (14)
12	29	1.9:1	47.6±11.9	Migraine (24), Tension-type headache (7), Trigeminal neuralgia (3)	Previous head injury (3)	Pa (52), Oc (24), Other (24)	Mono (100)	Ci (76), El (24)	3.2±0.3	B (41), Pr (35), S (36)	5.0±0.8	CU (72)	Dysesthesia or Allodynia (31)
13	183	1.5:1	49.5±16.8	Migraine (15), Epicrania fugax (11), Tension-type headache (4), Other (3)	n.r.	Pa (42), Fr (24), Oc (22), Te (15), V (4)	Mono (89), Midline (15)	Ci (85), El (14)	4 (3-5)	Pr (51), B (21), S (20), Th (8)	5 (4-6)	CU/Ep (89), R (11)	Allodynia (36), Dysesthesia (32)
15	24	1.2:1	50.3±15.2	Migraine (21), Tension-type headache (13), Trigeminal neuralgia (4), Cluster headache (4), Other (13)	Previous head injury (16)	Pa (62), Te (17), Oc (13), Fr (8)	Mono (100)	Ci (60), El (40)	3.2 (1-6)	B (21), Pr (21), Th (8), Other (50)	Mild (60), Moderate (29), Severe (4), Variable (7)	CU (60), Ep (40)	Dysesthesia or Allodynia (31), Hypoesthesia (8)
27	16	1.6:1	50 (19-79)	Migraine (56), Medication overuse headache (25)	Previous head injury (88)	Pa (38), Te (25), V (19), Fr (12), Oc (6)	Mono (100)	Ci (56), El (6), n.r. (38)	3.9 (2-10)	n.r.	n.r.	CU (75), Ep (25)	Dysesthesia (94)

M, mean; SD, standard deviation; VAS, Visual Analog Scale; n.r., not reported; F, frontal; Pa, parietal; Te, temporal; Oc, occipital; V, vertex; Ci, circular; El, elliptical; Pr, pressing/oppressive; S, stabbing; B, burning; Th, throbbing; Co, constricting; CU, continuous/unremitting; Ep, episodic; R, remitting.

Nummular headache in children and adolescents

NH is rarely reported in children and adolescents. Jeong *et al.*, (41) in a headache clinic in Korea, retrospectively observed a frequency of 0.3% for NH diagnosis among 2,466 patients aged 3 to 18 years (mean age 10.9), with 60% females and 40% males. A recent paper (42) described a case series of seven patients under 18 years diagnosed with NH at a tertiary pediatric headache center. The authors also conducted a systematic review of the literature, analyzing previously reported cases of NH in children and adolescents. In total, 9 pediatric cases with onset during childhood had been previously reported, with an additional case from India, (43) raising the total number of published cases to 17. The mean age at onset ranged between 9.7 and 11.9 years, with a female-to-male ratio of approximately 2:1.

Headache features in these pediatric cases were generally similar to those described in adults, with more patients reporting pain on the midline or at the vertex and frequently experiencing allodynia. (42) Comorbid headaches included migraine without aura (either episodic or chronic), MOH in three cases, and TTH in one. Some cases had associated intra- or extracranial lesions, usually considered incidental findings, such as a bone cholesterol cyst in the painful area, (44) focal hair heterochromia, (32) a pineal cyst, (42) and one case of mega-cisterna magna. (42) One case was presumed to be secondary to Langerhans cell histiocytosis (eosinophilic granuloma), as pain subsided following complete resection of the intracranial lesion. (45)

Most pediatric cases reported in the literature were refractory to treatment, except for the case previously described, (43) which responded fully to a combination of carbamazepine and gabapentin. In some cases with migraine comorbidity, (42) NH responded well to migraine-specific therapy. Additionally, spontaneous remission was observed in one case. (44)

Secondary cases

Since 2007, (46) numerous cases have been reported in which typical pain was associated with or secondary to underlying pathologies affecting the skin, subcutaneous tissue, bones, or intracranial structures. Following these case reports and series, patients presenting with NH-like symptoms have been found to have superficial aneurysms, (47) cranial or cutaneous malformations, (48,49) calcific hematomas of the scalp, (50) varicella-zoster shingles, (51) and eosinophilic granuloma. (45) Among intracranial lesions, arachnoid cysts, (52) pituitary adenomas, (53) and meningiomas (46) have also been reported. A recent review (44) described 8 new cases – including bone hemangiomas, superficial inflammatory or cholesterol cysts, osteomas, cavernomas, and arachnoid granulations – and discussed previous observations of secondary NH.

Secondary NH associated with systemic diseases has also been reported, primarily in the context of autoimmune disorders such as rheumatoid arthritis, Sjögren syndrome, sicca syndrome, and antiphospholipid antibody syndrome. (54) No specific features of pain were identified in these patients. An interesting review is dedicated to post-traumatic cases of NH. (7) These cases are not strictly classified as secondary, as head injury is generally considered a precipitating event rather than the direct cause of pain. (15,44) Following these observations, patients with NH preceded by head trauma tend to have a higher mean age and are more often affected by cutaneous allodynia. (7)

Among precipitating factors, surgical manipulations (55-57) and insect bites (30) have also been described. A recent article by Ighodaro *et al.* reported a young woman with a history of migraine who developed a new-onset, persistent, circumscribed parietal headache following intracranial carotid aneurysm pipeline stenting. The pain features were typical of NH, and other secondary causes – as well as procedure-related complications or local vascular abnormalities – were excluded. (40) Another case report by Liu *et al.* described a 40-year-old man who developed a new-onset pinprick parietal headache with characteristics of NH shortly after undergoing percutaneous transluminal angioplasty and stenting for middle cerebral artery atherosclerotic stenosis. Other potential causes of secondary headache were ruled out, and hypotheses regarding the pathogenetic mechanisms were proposed. (14)

Pathophysiology

The pathophysiology of NH is not fully understood. However, research has identified some characteristic features of the pain that suggest underlying mechanisms. One hypothesis, supported by Pareja *et al.* in Spain, describes it as an "epicrania" resulting from dysfunction of C-fibers in the terminal branches of cutaneous nerves within the epicranial tissues, (58) similar to other painful entities such as supra-orbital neuralgia, occipital neuralgia, trochleitis, epicrania fugax, and primary stabbing headache, which are consistent with neuropathic pain. (8,30,59) Supporting this hypothesis are observations indicating that symptoms and signs are restricted to a sharply delimited scalp area, without diffuse hypersensitivity of the pericranial structures, unlike migraine and tension-type headache. (60) Additionally, algometric measurements show that the pain threshold is lowered only in the symptomatic area. (37,61) The presence of trophic skin changes or hair loss – closely related in time and space to the pain – further supports a peripheral mechanism, given the importance of innervation for maintaining normal skin and hair structure. (30,32) These data strongly suggest a peripheral origin, rather than a central one, such as activation of the trigeminovascular system.

This hypothesis appears inconsistent with observations of some cases where the painful area is located in the midline, (6,11,26) and with the limited, partial, or temporary effectiveness of anesthetic infiltration of the affected area. (1,5,20,26,44,55,62-65) However, nerve fibers extending across cranial bones and crossing the midline have been identified. Furthermore, it has been observed (66) that some nerve branches responsible for pain run within the inner periosteum and through transdiploic or intradiploic layers, penetrating the skull and making them inaccessible to the action of local anesthetics. In a recent case series discussing the pathogenesis, (5) the origin of NH from epicranial structures was confirmed, but the concomitant presence of pain, sensory dysfunction, and trophic changes led to the hypothesis that NH may be part of a complex regional pain syndrome.

Structural abnormalities detected in secondary forms of NH may also shed light on the pathogenesis of primary NH. In fact, most symptomatic NH cases show lesions located within or near the scalp or skull bones, in proximity to the painful area. These lesions could damage peripheral fibers of cutaneous, pericranial, or epicranial nerves. In diagnosed primary forms, such lesions might be unremarkable upon macroscopic examination. There are also post-traumatic forms of NH, where trauma may precipitate pain by injuring epicranial tissues and nerve branches. (7) In patients with associated autoimmune disorders, a distal autoimmune-associated sensory axonopathy has been hypothesized. (54)

In NH secondary to endovascular stent placement, local manipulation of the vessel wall, causing endothelial injury and subsequent activation of sensory afferents, has been considered. This process could possibly lead to persistent inflammation of the vessel wall. (14) A dedicated study (67) found no differences in mood state between NH patients and controls, nor any correlation between clinical parameters of NH and levels of anxiety or depression.

Treatment and prognosis

Currently, no controlled clinical trials on the treatment of NH are available; most therapeutic approaches are based on case reports or case series. The first cases described by Pareja's group were either untreated or managed only with analgesics (paracetamol) or non-steroidal anti-inflammatory drugs (NSAIDs), which generally proved to be ineffective or only minimally effective. (1,11) Notably, in many cases, patients did not require active treatment due to the low intensity of their pain. (9,11,18,44) Paracetamol and NSAIDs may still be useful as acute treatments for pain exacerbations. (7-9,11,14,18,68) In one case with migraine features, triptans used as an acute therapy and topiramate as a prophylactic agent were reported to be effective. (36)

Various pharmacological prophylactic therapies have been proposed for NH, including: gabapentin, (5,7,9,13,15,19,20,23,28,33,43-45,51,55,56,68-71) onabotulinumtoxinA, (15,20,31,52,61,63,72,73) tricyclic antidepressants (TCA) (5,7,9,26,53,55,65,74,75), other antidepressants, (9,15,20) pregabalin, (7,9,15,49,52) topiramate, (20,36) indomethacin, (9,18,34,76) and other NSAIDs, (9,50) with mixed or inconclusive results. Non-pharmacological treatments have also been proposed, including acupuncture, (76) transcutaneous electrical nerve stimulation, (63) and percutaneous electrical nerve stimulation. (77) As previously reported, infiltration with local anesthetics generally provides partial or no benefit. (1,5,20,26,44,55,62-65,75,77)

Other treatments include palmitoylethanolamide, both as monotherapy (78) and in combination with topiramate, (79) neurotropin – a Japanese extract from animal skin used as an analgesic, (80,81) carbamazepine, (15,43,52,76,82) oxcarbazepine, (9,83) lamotrigine, (7) and metoprolol. (84) Recently, a case was reported of a NH patient with coexisting high-frequency episodic migraine who did not respond to gabapentin and BoNT-A but showed a beneficial response to galcanezumab, an anti-CGRP monoclonal antibody, which also improved NH symptoms. (85)

In two cases of primary NH, (86) the pharmacological response appeared to vary according to the temporal course of the disease: when pain was episodic with spontaneous remissions, the pain bouts responded to indomethacin; when NH became continuous and unremitting, it responded to gabapentin. This led to the hypothesis – later not confirmed – that NH pain might have different origins: central when episodic and peripheral when continuous.

Some cases have shown positive responses to surgical excision of underlying lesions. (45,47) A series of 49 primary NH patients with associated vascular Doppler signals within the painful area was successfully treated with minimally invasive arterectomy under local anesthesia, resulting in significant benefit. (38) A systematic review of both primary and secondary NH cases treated surgically has also been published. (39) When surgical removal of the underlying lesion is not feasible, medical therapy generally remains similar to that used in primary cases. (44)

A review article analyzed treatments reported in 110 published cases of NH from 2002 to 2019, (71) proposing that patients with complete resolution after treatment were younger, more likely to be female, and diagnosed earlier. These treatments were evaluated based on their frequency of use and efficacy; gabapentin, NSAIDs, BoNT-A, and TCA were the most frequently used and effective therapies, while carbamazepine did not demonstrate comparable efficacy.

A recent case series focusing on the treatment of primary unifocal NH (the NUMITOR Study) was published. (13) This real-world, observational study aimed to assess the responder rate of preventive drugs in 183 NH patients retrospectively screened from the registry of Valladolid University Hospital, covering the period from 2002 to 2022. Treatment response was estimated by the reduction in headache days per month at 30%, 50%, and 75%, between weeks 8 and 12, compared to baseline. Tolerability and rates of treatment discontinuation were also evaluated. The most used oral drugs for prevention were gabapentin, amitriptyline, and lamotrigine. Other medications included pregabalin, beta-blockers, flunarizine, topiramate, BoNT-A, and anesthetic nerve blocks. The most effective and best-tolerated treatment was BoNT-A, with responder rates of 62.5% (50%) and 47.5% (75%), respectively. Regarding oral therapies, gabapentin also showed efficacy, with responder rates of 43.7% (50%) and 35.2% (75%). These results, consistent with previous literature, support the use of BoNT-A as a first-line treatment for NH. Based on the presented cases and their clinical outcomes, BoNT-A and gabapentin at medium dosages (600-1200 mg) appear to be the most frequently used and effective treatments to date. In refractory cases, TCA may also be considered. A specific protocol for BoNT-A dosing and injection localization has been proposed, consisting of five injections of 5 IU within the painful area – one centrally and four in the peripheral regions. (73)

In **Table 3**, we provide a summary of response rates to the most commonly used preventive pharmacological treatments in primary adult NH, based on the most representative case series.

The clinical outcome and prognosis of NH have been poorly described. In most cases, NH is considered a benign condition, with either temporary or long-lasting remissions that can occur spontaneously (5-7,44) or following treatment. (69,75) However, in some cases, NH may be refractory to treatment and persist for years. A recent case series demonstrated that patients with bifocal NH exhibited higher rates of spontaneous remission in the first pain area compared to those with unifocal NH. (87) Secondary cases caused by treatable underlying lesions may respond well to surgical intervention, whereas the few cases of secondary NH related to endovascular procedures described so far seem to be resistant to pharmacological therapy.

Table 3. Treatment response rates to the most used preventive medical treatments in primary adult NH, from the most representative case series.

Drug	No response (<30%)	Partial response (31–50%)	Good response (>51%)
OnabotulinumtoxinA	13	33	54
Gabapentin	18	35	47
CBZ/OXC	39	15	46
TCA/other antidepressants	48	17	35
Anaesthetic blockades	23	45	32
Pregabalin	39	39	22
Topiramate	63	25	12

CBZ, carbamazepine; OXC, oxcarbazepine; TCA, tricyclic antidepressants.

Conclusions

NH can be defined as an organic, primary headache syndrome characterized by a distinct symptomatology: the pain is localized to a rounded or elliptical area of 1-6 cm in diameter, typically with a stable unilateral localization (although some bilateral and multifocal cases were reported). The pain is usually of mild to moderate intensity, but severe pain and local exacerbations can also occur. Cases with onset in pediatric age, as well as some secondary NH cases or associated with other clinical conditions or interventions, have been documented. The typical topographic distribution and associated symptoms suggest a peripheral pathogenesis, such as dysfunction of terminal C-fibers innervating the scalp or skull bones. Recently, a hypothesis proposing NH as a complex regional pain syndrome has also been suggested. (5) The diagnosis is primarily clinical; however, it is essential to exclude secondary forms through comprehensive clinical examination, neuroimaging, and blood tests, including inflammatory markers and immunology screening. Currently, treatment is supported by limited and sporadic evidence due to the absence of controlled clinical trials. The most commonly used and effective medical therapies for primary NH are BoNT-A and gabapentin, although surgical treatments have also been proposed for secondary cases and, in some instances, for selected patients with primary NH.

Currently, two clinical studies (registered on ClinicalTrials.gov) (88) are ongoing, carried out by Valladolid Group: the *Observational Prospective Study on the Presence of Typical Migraine Features in Nummular Headache Patients: The Nunamig Study* and the *NUMITOR study: Headache Iberian Study on the Treatments and Outcomes in Real-World Setting*.

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