The Neck and Headaches

Nikolai Bogduk, MD, PhD, DSc

KEYWORDS

• Headache • Cervical • Cervicogenic • Diagnosis • Treatment

KEY POINTS

- The mechanism of cervicogenic headache is convergence between cervical afferents and trigeminal or cervical afferents in the dorsal horn of the C1-3 segments of the spinal cord.
- No clinical features have been validated for the diagnosis of cervicogenic headache.
- Definitive diagnosis requires evidence of a cervical source of pain, usually by the application of controlled, diagnostic blocks.
- The C2-3 zygapophysial joint is the most common source of cervicogenic headache to have been identified.
- Of conservative therapies, the best evidence supports exercise, with or without manual therapy.
- Headache stemming from the C2-3 or C3-4 zygapophysial joint can be successfully treated with thermal radiofrequency neurotomy.

CASE STUDY

Patient 001 was a 23-year-old female nurse, who attributed the onset of her headaches to prolonged periods of wearing heavy lead aprons in a radiology suite. Her headache was constant and centered on the right occipital region, spreading to the forehead and right orbit. The headaches had persisted for 3 years and were not relieved by physical therapy or analgesics. The patient could not work and was involved in a worker's compensation claim. Examination revealed tenderness maximal over the C2-3 region of the cervical spine; headache was aggravated by rotation of the head. The headache was completely relieved by anesthetizing the right third occipital nerve. Repeat blocks, on 3 occasions, consistently relieved the headache completely, in accordance with the duration of action of the agents used: lignocaine or bupivacaine. Intra-articular injection of steroids temporarily relieved her headache for a few weeks, whenever they were used as a palliative measure. Thermal radiofrequency third occipital neurotomy completely relieved her headaches, for 9 months in the first instance. On recurrence of the headache, repeat neurotomy relieved the headache for 12 months after the first repeat, and then 14 months after the second repeat.

Disclosure: The author has nothing to declare.

Newcastle Bone and Joint Institute, Royal Newcastle Centre, University of Newcastle, PO

Box 664J, Newcastle, New South Wales 2300, Australia *E-mail address:* vicki.caesar@hnehealth.nsw.gov.au

Neurol Clin 32 (2014) 471–487 http://dx.doi.org/10.1016/j.ncl.2013.11.005

neurologic.theclinics.com

Having been relieved of her headache, the patient completed a university entrance examination, and then a university degree, before returning to full-time employment. Yearly repeat neurotomy has kept her free of headache.

DEFINITION

Pain arising from the upper cervical spine may be referred into regions of the head. The patient may be unaware of a cervical problem, and headache becomes the presenting feature. Technically, such headaches should be classified as referred pain from the cervical spine, but the term of reference that has largely been adopted in the literature, and in clinical practice, is cervicogenic headache. 1–4

MECHANISM

The anatomic basis of cervicogenic headache is convergence, onto second-order neurons in the C1-C3 segments of the spinal cord, between nociceptive afferents of the first division of the trigeminal nerve and nociceptive afferents of the C1, C2, and C3 spinal nerves.⁵ Convergence between trigeminal and cervical afferents explains referral of pain from cervical sources to the forehead, orbit, and temporal regions of the head. Convergence between other cervical afferents and those of C2 explains referral of pain to the occiput and parietal regions.

Physiologic convergence has been shown in laboratory animals, between trigeminal afferents from the dura mater of the skull and cervical afferents in the greater occipital nerve. $^{6-9}$ The convergence largely involves A δ and C fibers, onto neurons in laminae I, II, V, and VI of the dorsal horn at C2. Stimulation of trigeminal afferents sensitizes the response to cervical input, and stimulation of cervical afferents sensitizes trigeminal input.

In human volunteers, pain in the head has been evoked experimentally by electrical stimulation of the dorsal rootlets of C1¹⁰ and by noxious stimulation of the greater occipital nerve¹¹ or the suboccipital muscles of the neck.^{12–15} Noxious stimulation of the C2-3 intervertebral disk, but not lower disks, produces pain in the occipital region.^{16,17} Distending the C2-3 zygapophysial joint with injections of contrast medium produces pain in the occipital region,¹⁸ as does distending the lateral atlantoaxial joint or the atlanto-occipital joint.¹⁹ All segments from the occiput to C4-5 are capable of producing referred pain to the occiput, but referral to the forehead and orbital regions more commonly occurs from segments C1 and C2.¹³

In patients with suspected cervicogenic headache, headache can be relieved by anesthetizing the C2-3 zygapophysial joint^{20–22} or the lateral atlantoaxial joint.^{23–28} The C2-3 zygapophysial joint is the most common source,^{22,27–29} followed by the lateral atlantoaxial joint,^{27,28} and occasionally, the C3-4 zygapophysial joint.^{27,29,30}

From a given joint, pain can be perceived in various regions of the head, but certain trends are evident (Fig. 1).²⁷ Pain from C2-3 tends to be perceived across the lateral occipital region and into the forehead and orbital region. Pain from C1-2 also tends to gravitate to the orbital region but otherwise more often occurs in the vertex or around the ear. Pain from C3-4 tends to focus in the suboccipital region and upper cervical spine; when it does spread to the head, it is largely restricted to the posterior regions, sparing the forehead and orbit.

CLINICAL FEATURES

The essential clinical feature of cervicogenic headache is dull, aching pain perceived in some combination of the occipital, temporal, parietal, frontal, or orbital regions of

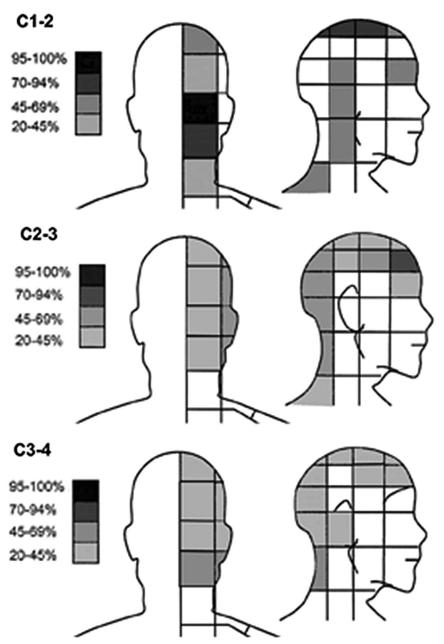


Fig. 1. Maps of the frequency with which pain from the synovial joints indicated is distributed to various regions of the head. (*From* Cooper G, Bailey B, Bogduk N. Cervical zygapophysial joint pain maps. Pain Med 2007;8:344–53; with permission.)

the head. Although a cervical source of pain is necessary for the diagnosis, its features may be cryptic or elusive. Certain, putatively distinctive, clinical features have been listed, 31–33 but none has been validated, 1–4 and none has been accepted by the International Headache Society in its taxonomy. 34 As a result, cervicogenic

headache is essentially a headache for which a cervical source of pain needs to be shown.

DISTINCTIONS

Although technically a form of cervicogenic headache, because they involve a cervical nerve, 2 conditions are distinguished from common, or idiopathic, cervicogenic headache, by their unique, clinical features and pathology. Both involve the C2 spinal nerve.

Headache is a feature of neck-tongue syndrome, but its cardinal and distinctive feature is numbness of the tongue on rotating the head.³⁵ The numbness is caused by stretching of cervical afferents from the hypoglossal nerve by a subluxating lateral atlantoaxial joint; the temporary headache is probably caused by strain of the joint.³⁶

C2 neuralgia is distinguished by intermittent, lancinating pain into the occiput, which may be accompanied by lacrimation and ciliary injection.³⁷ The condition can be caused by various disorders that affect the C2 spinal nerve where it runs behind the lateral atlantoaxial joint. Inflammatory disorders of the joint may result in the nerve becoming incorporated in the fibrotic changes of chronic inflammation.^{38–40} Otherwise, the C2 spinal nerve can be affected by meningioma,⁴¹ neurinoma,⁴² anomalous vertebral arteries,³⁹ and venous abnormalities, ranging from single to densely interwoven, dilated veins surrounding the C2 spinal nerve and its roots⁴³ to U-shaped arterial loops or angiomas compressing the C2 dorsal root ganglion.^{38,39,43}

So-called occipital neuralgia is a variously and poorly defined entity that was a popular, anecdotal diagnosis of occipital headache in the past. However, no explanation has been provided for why a disorder of a cutaneous nerve should cause deep, dull, aching pain; no pathology has been proved; and no controlled studies of diagnosis or treatment have been published.^{2–4} Some cases in the past may have been instances of C2 neuralgia before that entity was defined; other cases may have been examples of referred pain from upper cervical skeletal structures.

SOURCES

The neuroanatomy of cervicogenic headache dictates that any of the structures innervated by the C1-C3 spinal nerves could be a source of headache. ²⁻⁴ These structures include the posterior neck muscles, the C2-3 and C3-4 zygapophysial joints, the atlantoaxial joints, the C2-3 and C3-4 intervertebral disks, the dura mater of the upper cervical spine, and the vertebral artery. Importantly, the dura mater and vessels of the posterior cranial fossa are innervated by cervical nerves, and disorders of these structures are critical in the differential diagnosis of cervicogenic headache. Similarly, for present purposes, the common carotid artery can also be considered a cervical structure.

From time to time, structures below C3 have been implicated as a source of cervicogenic headache, but the evidence is indirect. The structures for which there is the most abundant and most rigorous evidence lie within the catchment of the C1-C3 spinal nerves.

CAUSES

In conventional pathology terms, many causes of cervicogenic headache have been reported, but each in small numbers. Pathologically specific causes underlie only a few cases.

Accepted as possible causes of cervicogenic headache are tumors and infections of the upper cervical spine,³⁴ but these are rare. Also accepted is rheumatoid arthritis

of the upper cervical joints, but this diagnosis arises in patients with manifest arthritis elsewhere in the limbs. Theoretically, rheumatoid arthritis or gout might present initially in the neck, but such cases have not been reported in the headache literature. A case report warns that metastasis to a cervical lymph node can cause cervicogenic headache. 44

Although, in the past, congenital anomalies have been listed in the differential diagnosis of cervicogenic headache, there is no evidence to implicate them so.^{2–4} Likewise, trigger points in the upper cervical muscles have been advanced as a cause of cervicogenic headache, but no controlled studies have vindicated this contention.

The C2-3 intervertebral disk has been implicated in several studies, ^{16,17,45} as a source of cervicogenic headache, but the pathology that renders a cervical disk painful has not been established. Several studies have implicated the lateral atlantoaxial joint as a source.^{23–26} In posttraumatic cases, the responsible lesions might include capsular rupture, intra-articular hemorrhage, and bruising of intra-articular meniscoids, or small fractures through the superior articular process of the axis.⁴⁶

The most extensively, and most rigorously, studied form of cervicogenic headache is pain from the C2-3 zygapophysial joint, mediated by the third occipital nerve, and therefore known as third occipital headache. Studies in experimental animals have shown that zygapophysial joints can become a source of persistent, nociceptive pain when subject to submaximal strain injuries, such as whiplash. The lesion affects the capsule of the joint, but is not manifest radiographically as arthopathy. Studies using controlled diagnostic blocks have shown that in 53% of patients with headache after whiplash, their pain can be traced to a C2-3 zygapophysial joint. This figure renders the C2-3 zygapophysial joint the most common source of cervicogenic headache, with microscopic capsular injury being the pathology.

DIAGNOSIS

The essential requirement for the diagnosis of cervicogenic headache is a cervical source or cervical cause for the pain. The International Headache Society explains that this diagnosis is typically not possible using a conventional approach to the diagnosis of headache, relying on history, physical examination, or medical imaging (Box 1).³⁴ Controlled diagnostic blocks are required to establish a cervical source of pain.³⁴

Certain clinical features can be used to suspect cervicogenic headache with different certainties of diagnosis (Box 2).⁴⁹ A diagnosis of possible cervicogenic headache can be entertained if patients have unilateral headache and pain starting in the neck. Satisfying any 3 additional criteria promotes the diagnosis to probable cervicogenic headache. The clinical features that most strongly indicate cervicogenic headache are pain radiating to the shoulder and arm; varying duration or fluctuating continuous pain; moderate, nonthrobbing pain; and history of neck trauma.⁴⁹

A definitive diagnosis can be established using controlled, diagnostic blocks, protocols for which have been defined. 50–52 Such blocks include intra-articular blocks of the lateral atlantoaxial joint (Fig. 2), blocks of the third occipital nerve to anesthetize the C2-3 zygapophysial joint (Fig. 3), and blocks of the medial branches of the C3 and C4 dorsal rami, which innervate the C3-4 zygapophysial joint. Suitable controls include using local anesthetic agents with different durations of action, or injections of normal saline, or anesthetizing an adjacent structure that is not the source of pain. 50–52 To be convincing, diagnostic blocks should completely relieve the headache whenever the target structure is anesthetized with an active agent, with relief lasting for the duration of action of the agent used, and no relief if normal saline is used, or if an alternate structure is anesthetized.

Box 1

Diagnostic criteria for cervicogenic headache, as proposed by the International Headache Society

Diagnostic Criteria

- A. Pain referred from a source in the neck and perceived in 1 or more regions of the head and/ or face, fulfilling criteria C and D
- B. Clinical, laboratory, and/or imaging evidence of a disorder or lesion within the cervical spine or soft tissues of the neck known to be, or generally accepted as, a valid cause of headache¹
- C. Evidence that the pain can be attributed to the neck disorder or lesion based on at least 1 of the following:
 - 1. Demonstration of clinical signs that implicate a source of pain in the neck²
 - 2. Abolition of headache following diagnostic blockade of a cervical structure or its nerve supply using placebo- or other adequate controls³
- D. Pain resolves within 3 months after successful treatment of the causative disorder or lesion

Notes

- 1. Tumors, fractures, infections, and rheumatoid arthritis of the upper cervical spine have not been validated formally as causes of headache, but are nevertheless accepted as valid causes when demonstrated to be so in individual cases. Cervical spondylosis and osteochondritis are NOT accepted as valid causes fulfilling criterion B. When myofascial tender spots are, the headache should be coded under 2. *Tension-type headache*.
- 2. Clinical signs acceptable for criterion C1 must have demonstrated reliability and validity. The future task is the identification of such reliable and valid operational tests. Clinical features such as neck pain, focal neck tenderness, history of neck trauma, mechanical exacerbation of pain, unilaterality, coexisting shoulder pain, reduced range of motion in the neck, nuchal onset, nausea, vomiting, photophobia, and so forth are not unique to cervicogenic headache. These may be features of cervicogenic headache, but they do not define relationship between the disorder and the source of the headache.
- Abolition of headache means complete relief of headache, indicated by a score of zero on a visual analogue scale (VAS). Nevertheless, acceptable as fulfilling criterion C2 is ≥90% reduction in pain to a level of <5 on a 100-point VAS.

From Headache Classification Committee of the International Headache Society (IHS). The International Classification of Headache Disorders, 3rd edition (beta version). Cephalgia 2013;33(9):629–808; with permission.

Target structures can be selected from epidemiologic data. Contemporary data implicate the C2-3 zygapophysial joint as the most common, and therefore most likely, source of cervicogenic headache. ^{22,27–30} Next most likely are the lateral atlantoaxial joint and the C3-4 zygapophysial joint. These pretest probabilities may change in the future as diagnostic blocks of the lateral atlantoaxial joints become more widely used. If synovial joints prove not to be the source of pain, the C2-3 intervertebral disk can be tested using diskography. ^{16,17,45}

A significant feature of patients in whom third occipital nerve blocks have been positive is that all had a history of trauma. This finding reinforces history of trauma as a cardinal clinical feature for probable cervicogenic headache (see **Box 2**). No studies have shown that third occipital headache occurs without a history of trauma.

Differential Diagnosis

Certain serious conditions need to be considered in the differential diagnosis of cervicogenic headache. These conditions include space-occupying lesions of the posterior

Box 2
Criteria for identifying possible and probable cervicogenic headache, as proposed by Antonaci and colleagues

- 1. Unilateral headache without side-shift
- 2. Symptoms and signs of neck involvement:

Pain triggered by neck movement or sustained awkward posture and/or external pressure of the posterior neck or occipital region

Ipsilateral neck, shoulder, and arm pain

Reduced range of motion

- 3. Pain episodes of varying duration or fluctuating continuous pain
- 4. Moderate, nonexcruciating pain, usually of a nonthrobbing nature
- 5. Pain starting in the neck, spreading to oculofrontotemporal areas
- 6. Anesthetic blockades abolish the pain transiently provided complete anesthesia is obtained or sustained neck trauma a relatively short time prior to the onset
- 7. Various attack-related phenomena: autonomic symptoms and signs, nausea, vomiting, ispilateral edema, and flushing in the periocular area, dizziness, photophobia, phonophobia, blurred vision in the ipsilateral eye

From Antonaci F, Ghirmai S, Bono S, et al. Cervicogenic headache: evaluation of the original diagnostic criteria. Cephalalgia 2001;21:573–83; with permission.

cranial fossa and aneurysms of the vertebral artery or internal carotid artery. Technically, these conditions constitute a form of cervicogenic headache, in that the pain is mediated by cervical nerves, and early in their evolution, these conditions can cause headache that is indistinguishable clinically from other forms of cervicogenic headache. However, they are distinguished from conventional cervicogenic headache by having detectable lesions that can have serious neurologic sequelae.

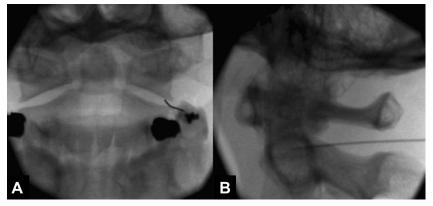


Fig. 2. Radiographs of a needle in place for performing an intra-articular block of the right lateral atlantoaxial joint. (*A*) Anteroposterior view. (*B*) Lateral view. (*From* International Spine Intervention Society. Lateral atlantoaxial joint blocks. In: Bogduk N, editor. Practice guidelines for spinal diagnostic and treatment procedures. 2nd edition. San Francisco (CA): International Spinal Intervention Society; 2013. p. 43; with permission.)



Fig. 3. A lateral radiograph of a needle in place for performing a third occipital nerve block. (*From* International Spine Intervention Society. Third occipital nerve blocks. In: Bogduk N, editor. Practice guidelines for spinal diagnostic and treatment procedures. 2nd edition. San Francisco (CA): International Spinal Intervention Society; 2013. p. 155; with permission.)

Space-occupying lesions of the posterior cranial fossa are distinguished from cervicogenic headache by the neurologic signs and increased intracranial pressure that they cause. However, early in the presentation, these signs might still be developing. Therefore, practitioners need to be alert to the subsequent onset of neurologic signs that declare these conditions.

Sixty percent of patients with aneurysms of the vertebral artery or the internal carotid artery present with headache as the sole feature. 53–56 Within a matter of a few days, aneurysms typically declare themselves by the onset of neurovascular features. However, during this period, the headache may be misdiagnosed as common cervicogenic headache, unless the practitioner is alert to the possibility of aneurysm. This dilemma is particularly relevant for practitioners who perform manipulative therapy for the neck. In the event of a subsequent stroke, it is impossible to determine if the aneurysm and stroke were caused by the manipulation or if it was a spontaneous aneurysm that would have caused stroke irrespective of the aneurysm.

Imaging

There is no evidence that medical imaging is diagnostic of any cause of cervicogenic headache. Imaging is indicated only in patients who show neurologic signs. However, in that context, the indication for imaging is the neurologic signs, not the pain.

In patients with cardiovascular risk factors or a history of neck distortion or cervical manipulation, aneurysm needs to be considered. For this entity, magnetic resonance angiography is the appropriate investigation.

Manual Examination

Manual therapists contend that they can diagnose symptomatic joints by examining the cervical spine. Previously, this belief was based on 1 small study, which ostensibly validated manual examination of the cervical spine.⁵⁷ However, that study has now been refuted by a larger study,⁵⁸ using more rigorous diagnostic criteria and more

rigorous statistical analysis. Manual examination, therefore, lacks a proven foundation as a diagnostic test for cervicogenic headache.

TREATMENT

Two approaches have been used for the treatment of cervicogenic headache. Conservative therapies have been used in patients in whom a clinical diagnosis of cervicogenic headache was made, based on diagnostic criteria that have not been validated. Consequently, not all patients who were treated may have had a cervical source of headache. Furthermore, these therapies were applied without a specified source of pain being treated. Targeted treatments have been used in patients in whom the putative source of pain was identified with various degrees of rigor, ranging from clinical suspicion to single diagnostic blocks or controlled diagnostic blocks.

CONSERVATIVE THERAPY

No drugs have been proved to be effective for cervicogenic headache. Infliximab has been tested but not in a controlled study.⁵⁹ Eighty percent of patients treated with transcutaneous electrical nerve stimulation reported at least 60% reduction in their headache index, at 1 month,⁶⁰ but no longer-term data are available, and none from controlled studies. Injections of onabotulinum toxin are no more effective than placebo.⁶¹

Manual therapy has been advocated for headache believed to be of cervical origin, but most of the literature consists of case reports or case series. ⁶² The few randomized controlled studies have provided follow-up of only 1 or 3 weeks, and systematic reviews ^{62–64} have variously found the evidence for manual therapy to be limited, or of varying quality, with conflicting results, rendering the efficacy of manual therapy uncertain.

The largest and strongest study of conservative therapy for cervicogenic headache showed that treatment with manual therapy, specific exercises, or manual therapy plus exercises were each significantly more effective at reducing headache frequency and intensity than was no specific care by a general practitioner. However, manual therapy alone was not more effective than exercises alone, and combining the 2 interventions did not achieve better outcomes. Seventy-six percent of patients achieved greater than 50% reduction in headache frequency at the 7-week follow-up, and 35% achieved complete relief. At 12 months, 72% had greater than 50% reduction in headache frequency, but the proportion who had complete relief was not reported. Corresponding figures for reduction in pain intensity were not reported.

Targeted Treatment

Various interventions have targeted the greater occipital nerve in the treatment of cervicogenic headache. These interventions include injections and various surgical procedures.

Greater occipital nerve

Ninety percent of patients treated with an injection of 160 mg of depot methylprednisolone onto the greater occipital nerve obtained relief, but only for 10 to 77 days. ⁶⁶ Surgical liberation of the nerve initially relieved headache in 80% of cases but the relief had a median duration of only 3 to 6 months. ⁶⁷ Excision of the greater occipital nerve provided relief in some 70% of patients, but for a median duration of only 244 days. ⁶⁸

In 1 study, patients were selected for surgery if they satisfied the clinical criteria for cervicogenic headache and obtained relief of headache from diagnostic blockade of

the C2 spinal nerve. ⁶⁹ They underwent decompression and microsurgical neurolysis of the C2 spinal nerve, with excision of scar and ligamentous and vascular elements that compressed the nerve. Fourteen of 31 patients were rendered pain-free. Details on the remaining patients are incomplete, but ostensibly, 51% gained what was called adequate relief and 11% suffered a recurrence.

Pulsed radiofrequency therapy applied to the greater occipital nerve is no more effective than an injection of methylprednisolone and bupivacaine onto the nerve, with 9 of 15 patients reporting 50% relief at 9 months after pulsed radiofrequency and 5 of 15 patients having the same outcome after injection.⁷⁰ Otherwise, the application of pulsed radiofrequency to the C2 ganglion has been described only in case reports.^{71,72}

A novel intervention has recently been reported. It involves the injection of 3 to 10 mL of processed, autologous adipose tissue onto the greater occipital nerve.⁷³ Nineteen of 24 patients were said to have had a good clinical response (not otherwise defined) at 3 months, with 7 suffering a recurrence by 6 months.

Lateral atlantoaxial joint

In 1 study, 32 patients with headache suspected to arise from a lateral atlantoaxial joint were treated with intra-articular injection of 1 mL of a mixture of bupivacaine and triamcinolone.⁷⁴ At 3 months, 25% had at least 50% relief of pain, and 16.6% had sustained relief at 9 months. In the absence of controls, a placebo effect cannot be excluded.

Another study did not indicate how patients were selected but 86 were treated with intra-articular pulsed radiofrequency.⁷⁵ At least 50% relief was reported by 43 patients at 6 months and by 38 patients at 12 months.

For patients whose headache can be relieved by lateral atlantoaxial joint blocks, an option for treatment is arthrodesis of the joint. The surgical literature attests to complete relief of pain being achieved, albeit in small numbers of patients, for more than 2 years. ^{76–78}

C2-3 disk

In those patients in whom the source of headache can be traced to the C2-3 intervertebral disk, disk excision and anterior cervical fusion reportedly can be effective. Because this intervention is surgical in nature and uncommonly used for headache, controlled trials are unlikely to be conducted. Anecdotal data, therefore, will remain the highest level of evidence available.

Zygapophysial joints

For headache stemming from the C2-3 zygapophysial joint, 1 study reported that at 19 months after an intra-articular injection of steroids into the joint, 11% of patients were free of pain. ⁷⁹ Otherwise, most studies of third occipital headache and headache from C3-4 have addressed the effectiveness of thermal radiofrequency neurotomy.

Radiofrequency Neurotomy

Thermal radiofrequency neurotomy is a procedure in which conduction along a nerve is blocked by coagulating it with an electrode inserted percutaneously. The rationale for the procedures is that if pain can be relieved temporarily by anesthetizing a nerve, the relief can be prolonged by coagulating the nerve. The critical indication for thermal radiofrequency neurotomy is, therefore, complete relief of pain after controlled, diagnostic blocks of the target nerve or nerves. In the context of cervicogenic headache, the target nerve is the third occipital nerve (**Fig. 4**) or the medial branches of the C3-4 dorsal rami, in patients whose headache can be relieved by anesthetizing one or other of these nerves. A randomized, placebo-controlled trial⁸⁰ has shown that responses to thermal

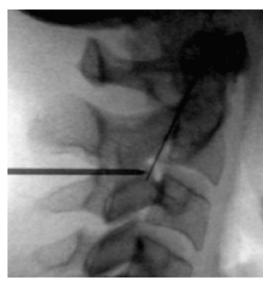


Fig. 4. A lateral radiograph of an electrode in place for performing thermal radiofrequency neurotomy of a right third occipital nerve.

radiofrequency neurotomy are not caused by placebo effects. Its success in the treatment of headache, therefore, cannot be dismissed as a placebo effect.

Three studies purport to show that radiofrequency neurotomy is not effective. 81–83 However, they do not provide evidence to this effect, for several reasons. In all of these studies, patients were selected on clinical criteria. So, it is not evident if all patients had cervicogenic headache, and if they did, what the source of pain was. Second, the neurotomy technique used has never been validated. So, it is not evident if treatment was technically adequate. Third, neurotomy was performed at segmental levels (C3-C6) that have never been incriminated as a common source of headache. So, even if technically adequate, the treatment may have been at a clinically irrelevant segmental level and, therefore, tantamount to a sham treatment. Jointly and several, these features preclude these studies from providing valid evidence on the treatment of cervicogenic headache.

Totally opposite results are obtained if a diagnosis is carefully established using controlled diagnostic blocks, and meticulous surgical technique is used. For patients in whom diagnostic blocks indicate that the C2-3 zygapophysial joint is the source of pain, it is possible to denervate that joint by radiofrequency neurotomy of the third occipital nerve. If the source of pain lies in the C3-4 joint, diagnostic blocks of the medial branches of C3 and C4 should relieve the headache, and those nerves can be coagulated. The procedure involves placing an electrode parallel to the target nerve (see Fig. 4) and using it to coagulate the nerve.

If patients are selected using controlled, third occipital nerve blocks, with the diagnostic criterion being complete relief of headache, and if meticulous technique is carefully used to coagulate the nerve, complete relief of headache can be achieved in 88% of patients after thermal radiofrequency neurotomy. ⁸⁴ The median duration of relief was 297 days, with some patients still having continuing relief at the time of review. These results have been corroborated by a second ⁸⁵ and third study. ⁸⁶

Similar outcomes have been reported in patients whose headache could be relieved by controlled blocks of the C3-4 medial branches.⁸⁷ After thermal radiofrequency

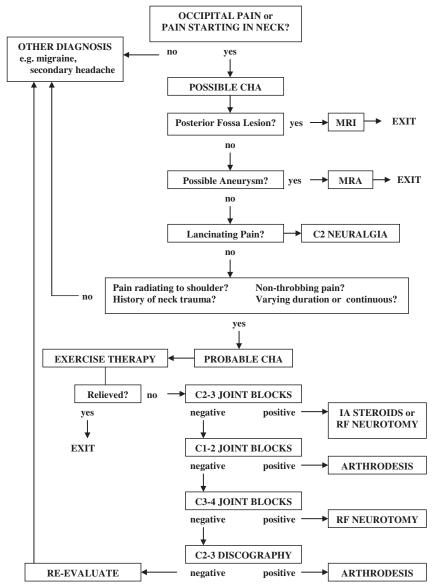


Fig. 5. A flow chart depicting a clinical pathway for the diagnosis and management of cervicogenic headache. CHA, cervicogenic headache; IA steroids, intra-articular steroids; MRA, magnetic resonance angiography; MRI, magnetic resonance imaging; RF neurotomy, radiofrequency neurotomy.

neurotomy of these nerves, more than 70% of patients maintained at lest 75% relief of their headache at 6 months and 12 months.

For patients in whom headaches recur, relief can be reinstated by repeating the neurotomy. By repeating neurotomy as required, some patients have been able to maintain relief of their headache for longer than 2 years.⁸⁴

CLINICAL PATHWAY

A clinical pathway has been recommended for the management of cervicogenic headache.^{3,4} The pathway embraces interventions for which there is at least reasonable evidence of efficacy coupled with safety but abjures interventions that are anecdotal or for which there is little or trivial evidence (Fig. 5).

The first half of the pathway pertains to formulating a diagnosis. Occipital pain or pain in the neck invites a diagnosis of possible cervicogenic headache. Thereafter, important differential diagnoses should be considered and excluded. Subsequently, additional clinical features can promote the diagnosis to one of probable cervicogenic headache, which is sufficient to pursue conservative therapy.

Of conservative therapies, there is strong evidence for only 1 intervention, which is exercise therapy, coupled or not with manual therapy. Most patients in primary care should benefit from this intervention.

Patients who do not benefit can be investigated more intensively. Because the pretest probability is highest for C2-3 zygapophysial joint pain, investigations should start with third occipital nerve blocks to test for this condition. If controlled blocks are positive, the patient can be treated with radiofrequency neurotomy. Intra-articular injection of steroids is a plausible, less destructive option, but lacks a definitive evidence-base.

If C2-3 blocks are negative, the next step is to test for lateral atlantoaxial joint pain with C1-2 blocks. If these blocks are positive, treatment by arthrodesis can be considered, because this is the only treatment of lateral atlantoaxial joint pain that has been found to provide complete relief of pain. Palliative treatment, in the form of intraarticular injections of steroids, or pulsed radiofrequency, might be an alternative, but with the understanding that these interventions may be no more than a placebo.

If C1-2 blocks are negative, the C3-4 zygapophysial joint should be tested. If blocks are positive, treatment is possible with C3-4 medial branch radiofrequency neurotomy. If C3-4 blocks are negative, the only remaining option is to test for C2-3 disk pain by diskography. If diskography is positive, treatment by arthrodesis can be considered.

If diskography and blocks of the joints of the upper 3 cervical segments all prove negative, there are no further, established investigations to pursue. The patient and their management should be revaluated. Either the diagnosis is not cervicogenic headache, or the patient has a cervical cause of pain that cannot be pinpointed using available technology. The options may be to treat the patient palliatively, by providing nonspecific pain relief, or to enroll them in whatever ethics-approved study is available of procedures that have experimental or investigational status. Ethics-approved studies guarantee patient safety and protect them from unwittingly being subjects to practitioners who are no more than experimenting with untested and unproven procedures, without disclosing to the patient that they are doing so.

REFERENCES

- Bogduk N. Distinguishing primary headache disorders from cervicogenic headache: clinical and therapeutic implications. Headache Current 2005;2:27–36.
- Bogduk N, Bartsch T. Headaches of cervical origin: focus on anatomy and physiology. In: Goadsby PJ, Silberstein SD, Dodick DW, editors. Chronic daily headache for clinicians. London: BC Decker; 2005. p. 369–81.
- 3. Bogduk N, Bartsch T. Cervicogenic headache. In: Silberstein SD, Lipton RB, Dodick DW, editors. Wolff's headache. 8th edition. New York: Oxford University Press; 2008. p. 551–70.

- 4. Bogduk N, Govind J. Cervicogenic headache: an assessment of the evidence on clinical diagnosis, invasive tests, and treatment. Lancet Neurol 2009;8:959–68.
- 5. Kerr FW. Structural relation of the trigeminal spinal tract to upper cervical roots and the solitary nucleus in the cat. Exp Neurol 1961;4:134–48.
- 6. Kerr FW. Trigeminal nerve volleys. Arch Neurol 1961;5:171-8.
- Angus-Leppan H, Lambert GA, Michalicek J. Convergence of occipital nerve and superior sagittal sinus input in the cervical spinal cord of the cat. Cephalalgia 1997;17:625–30.
- 8. Bartsch T, Goadsby PJ. Stimulation of the greater occipital nerve induces increased central excitability of dural afferent input. Brain 2002;125: 1496–509.
- Bartsch T, Goadsby PJ. Increased responses in trigeminocervical nociceptive neurons to cervical input after stimulation of the dura mater. Brain 2003;126: 1801–13.
- Kerr FW. A mechanism to account for frontal headache in cases of posterior fossa tumors. J Neurosurg 1962;18:605–9.
- 11. Piovesan EJ, Kowacs PA, Tatsui CE, et al. Referred pain after painful stimulation of the greater occipital nerve in humans: evidence of convergence of cervical afferences on trigeminal nuclei. Cephalalgia 2001;21:107–9.
- 12. Cyriax J. Rheumatic headache. Br Med J 1938;193(2):1367-8.
- 13. Campbell DG, Parsons CM. Referred head pain and its concomitants. J Nerv Ment Dis 1944;99:544–51.
- 14. Feinstein B, Langton JB, Jameson RM, et al. Experiments on referred pain from deep somatic tissues. J Bone Joint Surg Am 1954;36A:981–97.
- 15. Wolff HG. Headache and other head pain. 2nd edition. New York: Oxford University Press; 1963. p. 582–616.
- 16. Schellhas KP, Smith MD, Gundry CR, et al. Cervical discogenic pain: prospective correlation of magnetic resonance imaging and discography in asymptomatic subjects and pain sufferers. Spine 1996;21:300–12.
- 17. Grubb SA, Kelly CK. Cervical discography: clinical implications from 12 years of experience. Spine 2000;25:1382–9.
- **18.** Dwyer A, Aprill C, Bogduk N. Cervical zygapophysial joint pain patterns I: a study in normal volunteers. Spine 1990;15:453–7.
- 19. Dreyfuss P, Michaelsen M, Fletcher D. Atlanto-occipital and lateral atlanto-axial joint pain patterns. Spine 1994;19:1125–31.
- Bogduk N, Marsland A. On the concept of third occipital headache. J Neurol Neurosurg Psychiatr 1986;49:775–80.
- 21. Bogduk N, Marsland A. The cervical zygapophysial joints as a source of neck pain. Spine 1988;13:610–7.
- 22. Lord S, Barnsley L, Wallis B, et al. Third occipital headache: a prevalence study. J Neurol Neurosurg Psychiatr 1994;57:1187–90.
- 23. Ehni G, Benner B. Occipital neuralgia and the C1-2 arthrosis syndrome. J Neurosurg 1984;61:961–5.
- 24. McCormick CC. Arthrography of the atlanto-axial (C1-C2) joints: technique and results. J Intervent Radiol 1987;2:9–13.
- 25. Busch E, Wilson PR. Atlanto-occipital and atlanto-axial injections in the treatment of headache and neck pain. Reg Anesth 1989;14(Suppl 2):45.
- 26. Aprill C, Axinn MJ, Bogduk N. Occipital headaches stemming from the lateral atlanto-axial (C1-2) joint. Cephalalgia 2003;22:15–22.
- 27. Cooper G, Bailey B, Bogduk N. Cervical zygapophysial joint pain maps. Pain Med 2007;8:344–53.

- 28. Yin W, Bogduk N. The nature of neck pain in a private pain clinic in the United States. Pain Med 2008;9:196–203.
- 29. Lord SM, Bogduk N. The cervical synovial joints as sources of post-traumatic headache. J Musculoskel Pain 1986;4:81–94.
- 30. Govind J, King W, Giles P, et al. Headaches and the cervical zygapophysial joints: a prevalence study. Syllabus of the 14th Annual Scientific Meeting of the International Spine Intervention Society. Salt Lake City, July 13–15, 2006. p. 169–71.
- 31. Sjaastad O, Saunte C, Hovdahl H, et al. "Cervicogenic" headache. An hypothesis. Cephalalgia 1983;3:249–56.
- 32. Sjaastad O, Fredriksen TA, Pfaffenrath V. Cervicogenic headache: diagnostic criteria. Headache 1990;30:725–6.
- 33. Sjaastad O, Fredriksen TA, Pfaffenrath V. Cervicogenic headache: diagnostic criteria. Headache 1998;38:442–5.
- 34. International Headache Society. The international classification of headache disorders, 2nd edition. Cephalalgia 2004;24(Suppl 1):115–6.
- 35. Lance JW, Anthony M. Neck tongue syndrome on sudden turning of the head. J Neurol Neurosurg Psychiatr 1980;19(43):97–101.
- **36.** Bogduk N. An anatomical basis for neck tongue syndrome. J Neurol Neurosurg Psychiatr 1981;44:202–8.
- 37. Bogduk N. Pain of cranial nerve and cervical nerve origin other than primary neuralgias. In: Olesen J, Goadsby PJ, Ramadan NM, et al, editors. The headaches. 3rd edition. Philadelphia: Lippincott Williams & Wilkins; 2006. p. 1043–51.
- 38. Jansen J, Markakis E, Rama B, et al. Hemicranial attacks or permanent hemicrania—a sequel of upper cervical root compression. Cephalalgia 1989;9:123–30.
- **39.** Jansen J, Bardosi A, Hildebrandt J, et al. Cervicogenic, hemicranial attacks associated with vascular irritation or compression of the cervical nerve root C2. Clinical manifestations and morphological findings. Pain 1989;39:203–12.
- 40. Poletti CE, Sweet WH. Entrapment of the C2 root and ganglion by the atlantoepistrophic ligament: clinical syndrome and surgical anatomy. Neurosurgery 1990;27:288–91.
- 41. Kuritzky A. Cluster headache-like pain caused by an upper cervical meningioma. Cephalalgia 1984;4:185–6.
- 42. Sharma RR, Parekh HC, Prabhu S, et al. Compression of the C-2 root by a rare anomalous ectatic vertebral artery. J Neurosurg 1993;78:669–72.
- 43. Hildebrandt J, Jansen J. Vascular compression of the C2 and C3 roots-yet another cause of chronic intermittent hemicrania? Cephalalgia 1984;4:167-70.
- 44. Kim HH, Kim YC, Park YH, et al. Cervicogenic headache arising from hidden metastasis to cervical lymph node adjacent to the superficial cervical plexus—a case report. Korean J Anesthesiol 2011;60:134–7.
- 45. Schofferman J, Garges K, Goldthwaite N, et al. Upper cervical anterior diskectomy and fusion improves discogenic cervical headaches. Spine 2002;27: 2240–4.
- 46. Schonstrom N, Twomey L, Taylor J. The lateral atlanto-axial joints and their synovial folds: an in vitro study of soft tissue injuries and fractures. J Trauma 1993;35: 886–92.
- Bogduk N. On cervical zygapophysial joint pain after whiplash. Spine 2011;36: S194–9.
- 48. Curatolo M, Bogduk N, Ivancic PC, et al. The role of tissue damage in whiplash-associated disorders. Spine 2011;36:S309–15.

- 49. Antonaci F, Ghirmai S, Bono S, et al. Cervicogenic headache: evaluation of the original diagnostic criteria. Cephalalgia 2001;21:573–83.
- International Spine Intervention Society. Lateral atlanto-axial joint blocks. In: Bogduk N, editor. Practice guidelines for spinal diagnostic and treatment procedures. 2nd edition. San Francisco (CA): International Spinal Intervention Society; 2013. p. 35–62.
- International Spine Intervention Society. Third occipital nerve blocks. In: Bogduk N, editor. Practice guidelines for spinal diagnostic and treatment procedures. 2nd edition. San Francisco (CA): International Spinal Intervention Society; 2013. p. 141–63.
- International Spine Intervention Society. Cervical medial branch blocks. In: Bogduk N, editor. Practice guidelines for spinal diagnostic and treatment procedures. 2nd edition. San Francisco (CA): International Spinal Intervention Society; 2013. p. 101–39.
- 53. Biousse V, D'Anglejan-Chatillon J, Massiou H, et al. Head pain in non-traumatic carotid artery dissection: a series of 65 patients. Cephalalgia 1994;14:33–6.
- 54. Silbert PL, Makri B, Schievink WI. Headache and neck pain in spontaneous internal carotid and vertebral artery dissections. Neurology 1995;45:1517–22.
- 55. Sturzenegger M. Headache and neck pain: the warning symptoms of vertebral artery dissection. Headache 1994;34:187–93.
- 56. Mokri B, Houser W, Sandok BA, et al. Spontaneous dissections of the vertebral arteries. Neurology 1988;38:880–5.
- 57. Jull G, Bogduk N, Marsland A. The accuracy of manual diagnosis for cervical zygapophysial joint pain syndromes. Med J Aust 1988;148:233–6.
- 58. King W, Lau P, Lees R, et al. The validity of manual examination in assessing patients with neck pain. Spine J 2007;7:22–6.
- 59. Martelletti P, van Suijlekom H. Cervicogenic headache. Practical approaches to therapy. CNS Drugs 2004;18:793–805.
- 60. Farina S, Granella F, Malferrari G, et al. Headache and cervical spine disorders: classification and treatment with transcutaneous electrical nerve stimulation. Headache 1986;26:431–3.
- Linde M, Hagen K, Salvesen Ø, et al. Onabotulinum toxin A treatment of cervicogenic headache: a randomised, double-blind, placebo-controlled crossover study. Cephalalgia 2011;31:797–807.
- 62. Haldeman S, Dagenais S. Cervicogenic headaches: a critical review. Spine J 2001;1:31–46.
- 63. Posadzki P, Ernst E. Systematic reviews of spinal manipulations for headaches: an attempt to clear up the confusion. Headache 2011;51:1419–25.
- 64. Chaibi A, Russell MB. Manual therapies for cervicogenic headache: a systematic review. J Headache Pain 2012;13:351–9.
- 65. Jull G, Trott P, Potter H, et al. A randomized controlled trial of exercise and manipulative therapy for cervicogenic headache. Spine 2002;27:1835–43.
- 66. Anthony M. Cervicogenic headache: prevalence and response to local steroid therapy. Clin Exp Rheumatol 2000;18(Suppl 19):S59–64.
- 67. Bovim G, Fredriksen TA, Stolt-Nielsen A, et al. Neurolysis of the greater occipital nerve in cervicogenic headache. A follow up study. Headache 1992;32:175–9.
- **68.** Anthony M. Headache and the greater occipital nerve. Clin Neurol Neurosurg 1992;94:297–301.
- 69. Pikus HJ, Phillips JM. Characteristics of patients successfully treated for cervicogenic headache by surgical decompression of the second cervical root. Headache 1995;35:621–9.

- Garbhelink T, Michálek P, Adamus M. Pulsed radiofrequency therapy versus greater occipital nerve block in the management of refractory cervicogenic headache—a pilot study. Prague Med Rep 2011:112:279–87.
- 71. Zhang J, Shi DS, Wang R. Pulsed radiofrequency of the second cervical ganglion (C2) for the treatment of cervicogenic headache. J Headache Pain 2011;12:569–71.
- 72. Bovaira M, Peñarrocha M, Peñarrocha M, et al. Radiofrequency treatment of cervicogenic headache. Med Oral Patol Oral Cir Bucal 2013;18:e293–7.
- 73. Gaetani P, Klinger M, Levi D, et al. Treatment of chronic headache of cervical origin with lipostructure: an observational study. Headache 2013;53:507–13.
- Narouze SN, Casanova J, Maekhail N. The longitudinal effectiveness of lateral atlantoaxial intra-articular steroid injection in the treatment of cervicogenic headache. Pain Med 2007;8:184–8.
- 75. Halim W, Chua NH, Vissers KC. Long-term pain relief of patients with cervicogenic headaches after pulsed radiofrequency application into the lateral atlantoaxial (C1-2) joint using an anterolateral approach. Pain Pract 2010:10:267–71.
- Ghanayem AJ, Leventhal M, Bohlman HH. Osteoarthrosis of the atlanto-axial joints-long-term follow-up after treatment with arthrodesis. J Bone Joint Surg Am 1996;78A:1300-7.
- 77. Joseph B, Kumar B. Gallie's fusion for atlantoaxial arthrosis with occipital neuralgia. Spine 1994;19:454–5.
- 78. Schaeren S, Jeanneret B. Atlantoaxial osteoarthritis: case series and review of the literature. Eur Spine J 2005;14:501–6.
- Slipman CW, Lipetz JS, Plastara CT, et al. Therapeutic zygapophyseal joint injections for headache emanating from the C2-3 joint. Am J Phys Med Rehabil 2001;80:182–8.
- 80. Haspeslagh SR, van Suijlekom HA, Lame IE, et al. Randomised controlled trial of cervical radiofrequency lesions as a treatment for cervicogenic headache. BMC Anesthesiol 2006;6:1.
- 81. Stovner LJ, Kolstad F, Helde G. Radiofrequency denervation of facet joints C2-C6 in cervicogenic headache: a randomised, double-blind, sham-controlled study. Cephalalgia 2004;24:821–30.
- 82. van Suijlekom HA, van Kleef M, Barendse GA, et al. Radiofrequency cervical zygapophyseal joint neurotomy for cervicogenic headaches: a prospective study of 15 patients. Funct Neurol 1998;13:297–303.
- 83. Govind J, King W, Bailey B, et al. Radiofrequency neurotomy for the treatment of third occipital headache. J Neurol Neurosurg Psychiatr 2003;74:88–93.
- 84. Barnsley L. Percutaneous radiofrequency neurotomy for chronic neck pain: outcomes in a series of consecutive patients. Pain Med 2005;6:282–6.
- 85. MacVicar J, Borowczyk J, MacVicar AM, et al. Cervical medial branch radiofrequency neurotomy in New Zealand. Pain Med 2012;13:647–54.
- 86. Lord SM, Barnsley L, Wallis BJ, et al. Percutaneous radio-frequency neurotomy for chronic cervical zygapophysial-joint pain. N Engl J Med 1996;335:1721–6.
- 87. Lee JB, Park JY, Park J, et al. Clinical efficacy of radiofrequency cervical zygapophyseal neurotomy in patients with chronic cervicogenic headache. J Korean Med Sci 2007;22:326–9.