
Cervicogenic headache: Criteria, classification and epidemiology

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Clin Exp Rheumatol 2000; 18 (Suppl. 19): S3-S6.

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Key words:

Headache, cervicogenic headache, unilateral headache, migraine without aura.

ABSTRACT

The concept that headache might stem from the neck is old. The term “cervicogenic headache” was coined in 1983. A new content was then given to this concept: cervicogenic headache (CEH) is in principle a unilateral headache, generally starting in the neck and “spreading” forwards. A strict unilaterality - that is, absolutely no pain on the opposite side - is rather rare. Unilaterality in this context is defined as follows: the headache dominates on one side. When weak, the pain may be only on that side; when severe, it may also be felt on the contralateral side, but to a lesser extent. It never dominates on the contralateral side. These special features of CEH cannot be emphasised strongly enough. There are signs pertaining to the neck, such as reduced range of motion in the neck, mechanical precipitation mechanisms and ipsilateral shoulder/arm sensation (or even pain). Migraine without aura symptoms are less prominent than in migraine.

Introduction

It has been speculated, probably for more than a century, that headache might stem from the neck. The ideas as to how such a headache materialises have varied: short-lasting or long-lasting pain episodes; unilateral or bilateral pain; occipital pain only or pain also involving the anterior parts of the head. Stepping stones along the road were the works of Barré & Lieou (1, 2) and Bärtschi-Rochaix (3). They unfortunately emphasised a particular sympathetic/arthritis background (1) or migraine as crucial factors (3), probably in this way diverting attention from the main theme. The work of Hunter & Mayfield (4) brought our understanding of headache to another level in our view: the contours of a headache stemming from the neck started to become visible, although still dimly. The unilaterality of the pain was stressed. The major occipital nerve was cut - with ensuing relief of symptoms (but with only a short observation period). This work was belittled in that, for

instance, Lance (5) characterised their cases as being cases of cluster headache (sic!), in spite of the fact that there was no male preponderance, no excruciating severity of the attacks, no clear-cut, localised autonomic phenomena, and probably, no real cluster phenomenon. In other words, 4 of the 5 major criteria (6) for cluster headache seemed to be lacking. With this constellation of features, it is in our opinion difficult to uphold a diagnosis of cluster headache (7). This deprecating attitude contributed to a setback - and no breakthrough - for the idea of headache stemming from the neck. Later, sporadic reports seemed to deal with this topic, although using other terms such as occipital neuralgia (8) or atypical facial neuralgia (9) (Fig. 1).

The term “cervicogenic headache”

Our introduction of the term cervicogenic headache (CEH) in 1983 (10) - with a brief description of 22 cases - seems to have caused a renewed interest in this item in neurological circles.

In order to grasp the concept of cervicogenic headache, it is important to understand the background. The question that we in the mid-seventies asked ourselves was: Can a primary disorder in the neck give rise to a headache, and if so, how does it manifest itself? As headache physicians, we naturally started our search for such cases among patients in whom headache was the main complaint. We more or less had to start our search from scratch, and when one does not know what one is searching for, the search is going to be characterised by trial and error. A decisive feature in identifying the first case around 1980 was the mechanical precipitation mechanism. Once the first case was identified, other cases followed suit.

The principal and fundamental difficulties we were still facing can be illustrated by the saga of Sluder's sphenopalatine ganglion headache (11, 12). Sluder felt that the headache he described might stem from this ganglion. However, the viability of this hypothesis proved to be feeble.

The reasons why may be the following: (1) The sphenopalatine ganglion might well give rise to pain, as could all the other (?) nerves and ganglia in the region; (2) The headache Sluder described may even be a rather stereotyped, homogeneous one; (3) Nevertheless, it was never proven beyond doubt that "Sluder's headache" ("sphenopalatine headache") really stemmed from the sphenopalatine ganglion.

The essentials of the theory behind cervicogenic headache

By headache we understand a pain situated above a line from the nasion - and including the eye - and ending at the tendon insertions in the back of the head (See Fig. 1).

That an ache in the neck might stem from various structures within the neck is common knowledge. The question as to whether an abnormality in the neck can give rise to a headache is at quite another level of intricacy.

The implications of unilaterality in CEH are fundamental: A trauma or other pain-generating disorder in the skin on the one side of the head, e.g. a furuncle, may give rise to ipsilateral head pain. Even a rapidly growing intrahemispheric disorder, for example a brain abscess, generally gives rise to ipsilateral pain. If, however,

excitation or deficiency phenomena arise in connection with such an intrahemispheric process, these will appear on the side opposite the intrahemispheric process. And this also goes for - the rarely occurring - extremity/body pain (13, 14) combined with the hemispheric process. If shoulder/upper extremity pain (10) appears in CEH (present in 77%; ref. 15 and personal communication 1998), it will be on the same side as the head/neck pain. The situation, in other words, is diametrically opposite from the one in an intrahemispheric process. This combination of ipsilateral head/upper extremity pain is not consistent with a skull or scalp/cerebral origin of the pain syndrome. Probably only a process in the neck (including the lower occipital area ?) can explain the constellation with ipsilaterality of head and extremity pain observed in CEH.

Some researchers have used the term "occipital neuralgia" for similar (identical?) headaches (8, 16). Neuralgia is generally thought of as a short-lasting pain (a paroxysm), but neuralgia may - according to the International Association for the Study of Pain (IASP) - also be a more long-lasting pain (17). This condition may therefore be met by CEH. Next, the pain should be within the territory of the nerve(s). To be realistic,

therefore, an occipital neuralgia should be a unilateral pain in the innervation territory of the occipital nerve(s), of shorter or longer duration (Fig. 1). The typical case of CEH involves the entire hemicranium, by far exceeding the boundaries of the occipital nerve(s): CEH is by this criterion not an occipital nerve neuralgia (Fig 1). This is a major point for understanding CEH. Local anaesthetic blockades provide further evidence for an origin of CEH in the neck (18). When greater occipital nerve/C2 root blockades are effective in CEH, the pain will disappear, not only in these innervation zones, but also in the fronto-temporal areas, where blocks have not been made. This makes sense in the light of Kerr's studies (19) of frontal pain in "posterior" disorders.

The diagnostic criteria

We therefore need to familiarize ourselves with the criteria for CEH (20), keeping in mind its similarity to migraine. A "polished" version of the criteria has recently been published by Vincent & Luna (21).

The clinical picture will be dealt with in detail in the next chapter by M.V. Vincent. The items from X upward are by no means obligatory, but may be present (Table I). Some of them (X/XII) - to be found especially in severe cases - demonstrate the closeness of the symptomatology of the CEH to migraine. Many CEH patients described in the past have sustained neck/head injuries (IX), for example 24% in Vincent & Luna's series (21). Whether in isolated cases there is a cause-and-effect relationship between trauma and headache, or whether the injury represents an additional effect or merely a fortuitous event may be hard to assess. Cases of a clear-cut causal relationship between trauma and CEH seem to exist.

Two features need special attention (Table I); the unilaterality (point I) and the symptoms and signs involving the neck (points II a-c, and VI and VII). The unilaterality is without sideshift. From a classification point of view this is essential.

Migraine is in principle also a unilateral headache. The unilaterality in migraine is frequently of a different type, that is with a sideshift from one attack to an-

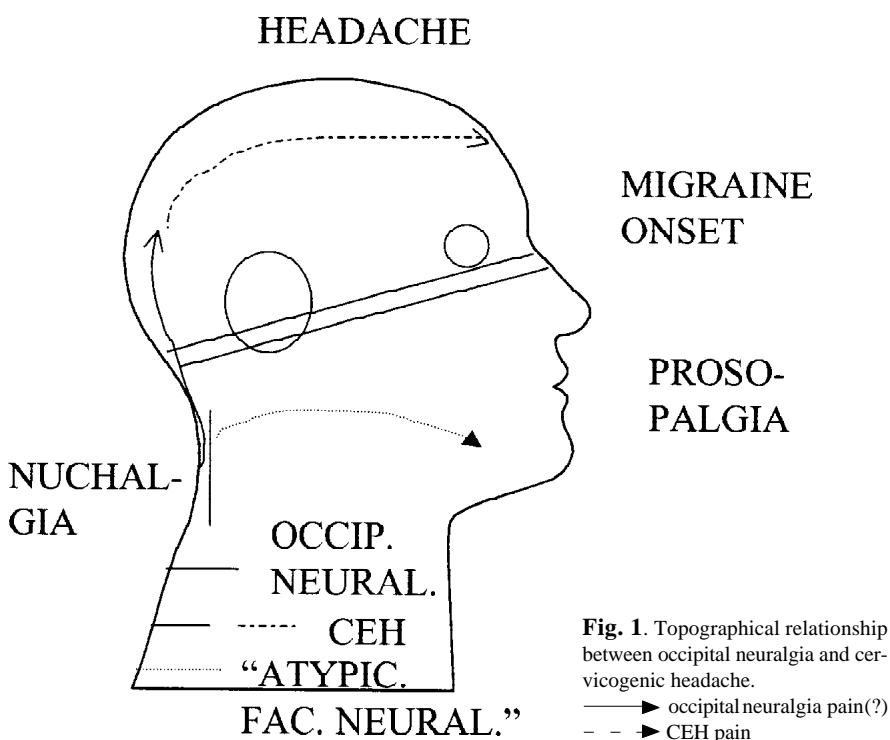


Fig. 1. Topographical relationship between occipital neuralgia and cervicogenic headache.
 —▶ occipital neuralgia pain(?)
 - - ▶ CEH pain

Table I. Criteria for cervicogenic headache.

I	Unilaterality without sideshift
Ila1	Pain triggered by neck movement and/or sustained awkward position
Ila2	Pain elicited by external pressure over the ipsilateral upper, posterior neck region or occipital region
Iib	Ipsilateral non-radicular neck, shoulder, and arm pain
Iic	Reduced range of motion in the cervical spine
III	Non-clustering pain episodes
IV	Pain episodes of varying duration or fluctuating, continuous pain
V	Moderate, non-excruciating pain, usually of a non-throbbing nature
VI	Pain starting in the neck, eventually spreading to oculo-fronto-temporal areas where the maximum pain is usually located
VII	Anaesthetic blockades of the major occipital nerve; C2 root or other appropriate structures on the symptomatic side abolish the pain transiently, provided anaesthesia is obtained
VIII	Female sex
IX	Head and/or neck trauma
Xa	Nausea
Xb	Vomiting
Xc	Ipsilateral edema, and - less frequently - flushing, mostly in the periocular area
XI	Dizziness
XII	Phono- and photophobia
XIII	Ipsilateral "blurred vision"
XIV	Difficulties on swallowing

other, or even within the same attack (22). Although this feature is hardly specific, it is typically "migrainous". Migraine may also be unilateral without sideshift, although rarely [i.e., in 16% of the cases in our series (22)], and it may also be bilateral. Since migraine is so prevalent, it is a possibility to be reckoned with also in cases of unilaterality without sideshift.

It is to be emphasised that even in typical, unilateral CEH cases, there may be a moderate - or more marked - spread of pain across the midline, mainly during maximal attacks, or at the peak of an attack, but still with a preponderance on the original side. There is never headache only on the contralateral side. This is what is understood by "unilaterality" in this context. Strict unilaterality, i.e. without any co-involvement of the other side, will be a rather rare phenomenon in CEH. These features cannot be stressed firmly enough.

Not infrequently, the bilaterality is of a more profound nature. The disease process can probably be duplicated on the contralateral side, like the "unilaterality on two sides" in tic douloureux, as described by Harris (23, 24). Since CEH is a syndrome, the pathological process on the two sides may even putatively differ in such a case. When this syndrome is fully understood, cases of bilateral headache may well be found to be as frequent as (or even more frequent than ?) cases

of unilateral headache (20). It should be emphasised that in any unilateral headache, there is a tendency to bilaterality; this tendency to bilaterality is probably more marked in CEH than in some other unilateral headaches, such as CPH.

At the very outset it was realised that if bilaterality was allowed as a criterion, the chances of including tension-type headache ("tension headache", TH) among CH cases would be greatly increased. More specifically, TH then would be the main differential diagnostic alternative, with all the implicit difficulties involved. There was, in other words, also a "political" reason for our strictness and stubbornness in this matter. Although a softening of the diagnostic criteria has been advocated recently (20), it is important to adhere to the concept of unilaterality until we have a clear picture of CEH. In clinical series, cases of unilaterality could be compared with bilateral cases (when such cases have been proven beyond a reasonable doubt to be "cervicogenic").

Equally important diagnostic features are the symptoms and signs relating to the neck. Such signs are diffuse (non-radicular) discomfort extending into the ipsilateral arm (perhaps an even stronger piece of evidence is radicular pain); reduced range of motion in the neck (25); and, probably even more importantly, the triggering of typical attacks by various awkward, sustained neck movements in

various directions (10, 20). The same procedure may reproduce the headache repeatedly. Particularly unfortunate may be a non-tolerated position of the head/neck during sleep. When the patient finally wakes up, the triggering event may already have passed the point of no return, since the patient has been unable to notice the initial warning during sleep. The pain - occasionally also an attack (if the pressure exerted is strong enough) can also be reproducibly provoked, iatrogenically, by external pressure applied to various tender, circumscribed areas of the neck, such as over the tendon insertions in the occipital area on the symptomatic side or over the occipital nerves. Since most attacks probably are mechanically precipitated, the temporal pattern can vary even within a single patient, and a non-continuous pattern may sometimes be present in the early stages. Eventually, a chronic-fluctuating pattern develops in most patients. The severity of the pain and the duration of the solitary episode/exacerbation may vary, depending upon the patient, the situation, and the duration of exposure, the pain ranging from mild to severe.

Classification

CEH is in principal one of the unilateral headaches. The special definition of unilaterality should be strictly observed (Table I). In a sense CEH has been separated out from migraine [cfr. "migraine cervicale" (3)]. "Migraine cervicale" was originally considered to be a migraine, but precipitated from the neck.

The similarity of CEH to common migraine must always be remembered, but also bearing in mind that 'similarity' does not imply 'identity'.

In recent years it has emerged that migraine without aura and CEH probably differ from a clinical point of view (7, 21, 22). It goes without saying, however, that since migraine is such a frequent disorder, there will probably occasionally be a coexistence of the two. The forms of coexistence will vary. These circumstances, however, should not detract attention from the fact that a "pure" CEH form exists, i.e. without migraine features. CEH and migraine without aura should accordingly be classified as separate disorders. This is more or less a must.

Epidemiology

Only a few studies on CEH have been carried out. Obviously, the criteria used will have a major impact on estimations of prevalence in the general population: Should one adhere to the most recent criteria (20) or to the International Headache Society (IHS) criteria, or to a modified version of these two sets of criteria - or to entirely "private" criteria? In the latter situation, one may end up with utter confusion (26). Is the standard to be unilaterality of headache? If bilaterality is allowed, high prevalence values may be obtained. The differential diagnostic difficulties versus tension headache may then be major ones, especially in studies carried out on the basis of questionnaires and/or by investigators not fully versed in the problems in this field. The latter are probably the source of the most misleading reports. Reproducibility of the results would necessitate standardised criteria.

The available studies that can tell something useful about the prevalence of CEH can be divided into two groups: (i) regular hospital/outpatient series that compare the prevalence of CEH with the prevalence of, for example, migraine and tension headache; and (ii) population studies.

Maciel *et al.* (27) found a 15% frequency of CEH in a headache clinic series (n = 1,229). This frequency is probably far above what can be expected for e.g. cluster headache. Their view that CEH is one of the three major headaches may be defensible. In Vincent & Luna's series (21), there were 33 CEH patients, 29 episodic tension headache patients, and 65 common migraine patients. Their figures are probably not widely different from our own outpatient headache series. In our series, however, there was probably a major selection bias, since episodic tension headache cases were rarely referred to us and, as importantly, our particular interest in CEH was known among referring physicians.

Population studies are rare. Monteiro (28) in a large study in Porto (n = 2,008) found a prevalence of 0.4% when using IHS criteria (29), 1% when using "6 of the most recent criteria", and 4.6% when using "5 or more of these criteria" (20). The 6 criteria used were not all among the major ones, however. This

study was carried out by means of questionnaires/proxies, which clearly limits its reproducibility. The variation in the value obtained, depending on the criteria used, is striking in the Portuguese study (i.e. > 10 times). Astonishingly, fewer cases of CEH were found (0.4%) using the IHS criteria, which allow bilaterality, than using our criteria (20), which at the time (1990) emphasised unilaterality. *A priori*, one would have expected that this should have been the other way around.

In Nilsson's study of 326 citizens in a Danish town (30), 17.8% were found to be CEH cases. The study utilised the IHS criteria (29). These were, however, modified: e.g. cervical x-rays, an obligatory diagnostic parameter according to the IHS were omitted. Unilaterality/anaesthetic blockade results do not form part of the IHS criteria. The combination of bilaterality and muscle tenderness, as in the IHS criteria for CEH, may lead the diagnostic search in the direction of tension headache rather than CEH. The modifications introduced by Nilsson will affect reproducibility.

It is remarkable that in two studies both claiming to have used the IHS criteria (28,30), one (30) found 44 times as many cases of CEH as the other (28). This indicates that the time is ripe for an appreciable upgrading of the IHS criteria (21, 28). In addition, proper population studies with proper criteria must be carried out as regards CEH.

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