Cervicogenic headache: Clinical aspects

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ABSTRACT
Cervicogenic headache (CEH) is a relatively common but often overlooked disorder. There is sufficient evidence to support this category and the existing diagnostic criteria are adequate. Subgroups may exist and the clinical picture sometimes may be similar to that of other headache disorders, however. The pathophysiology of this condition and its relationship with other headache syndromes remain to be determined. Migrainous features may occur in some patients.

Introduction
It is now clear that some headaches may originate in the neck. In spite of some lingering controversy (1-4), the era of doubting whether cervicogenic headache (CEH) actually exists is now over. The questions now are: How can the limits of CEH be established more precisely? Where does it end and where do other primary headaches begin? When and how do different clinical pictures intermingle? What are the subtypes of CEH? What is its pathophysiology? Does it share common pathways with migraine and, if so, to what extent? How can it be treated? In spite of the lack of objective diagnostic markers and, consequently, the diversity of the reported clinical data, these are the issues that must be comprehensively addressed from now on. CEH is a syndrome characterized by intermittent or continuous headache of cervical origin (5-8). The pain is mostly unilateral, does not change to the contralateral side from one attack to another as does migraine, and usually spreads up from the posterior part of the head to the frontal area, where it is more intense. When bilateral, one side generally predominates in terms of intensity. In some patients, vague pain may also irradiate to the ipsilateral arm. Cervical bursting is the essential feature of CEH. Attacks may be precipitated by neck movements or awkward positions, or by digital pressure over trigger areas at the posterior part of the neck, such as the greater occipital nerve (GON) or the C2 area (9, 10). The presence of a side-locked headache together with evidence of neck triggering, either by movement, positioning or digital pressure, should raise the possibility of CEH.

As with other head pain disorders, CEH patients may suffer from other concomitant types of headache. They can usually differentiate the CEH attack from other eventual co-morbid entities, such as migraine, however. Females are more often affected than males, and a trauma may precede the onset of the syndrome, although many patients do not report any preceding neck or head injury.

Clinical aspects
The most important clinical aspect of CEH is the strict unilaterality of the pain, a headache that does not change from one side to the other. Pain in the nuchal area or occipital region is not a sufficient criterion for CEH. This generally corresponds to non-specific occipitalgia. In CEH the pain occurs in the trigeminal areas. Although some patients may complain of bilateral headache, when carefully asked they often admit that the intensity typically predominates on one side. The intensity is usually greater at the forehead. If not specifically asked about, subjects may neglect to mention the fact that the pain starts at the back of the head and irradiate to the front, simply because the intensity at the occipital and nuchal areas may be comparatively lower. Irradiation downward to the ipsilateral arm may be radicular as well as less precise, vague and diffuse. Some individuals may have pain in the arm not concomitant with the headache on every occasion. Some individuals may consider disease onset as the point in time when the pain became troublesome. Special attention must be paid to trauma, which may be relatively mild and therefore not remembered by the patient. Whiplash-like trauma could contribute to cervicogenic headache, but many patients will not report any trauma at all. The role of whip-
lash in CEH remains rather confusing, partially due to obvious methodological difficulties with such studies. In cervicogenic headache the pain may be throbbing. A deep, pressing soreness of the head may turn into pulsatile pain if the appropriate triggering factor comes into play. The description of trigger factors may vary largely. Some patients cannot tolerate sitting side by side with a friend in a restaurant because talking would require much head turning. Others state that they have to sit always on the same side of an aircraft because looking outside the window contralaterally would evoke pain. Backing a car, sitting in a dentist’s chair for a long period of time, or unexpectedly falling asleep in an uncomfortable position while reading or watching TV are other examples. Asking about the patient’s profession, habits and main occupation are mandatory. The author once treated a woman patient who used to work as a telemarketing sales representative. Not being equipped with headsets, she had to hold the telephone earpiece between her head and shoulder while talking and using a computer keyboard. Similarly, eliciting clues as to what time of the day the headache tends to worsen may provide some indication of eventual triggering factors. If the headache is present when the patient wakes up but improves during the morning hours, his or her sleeping position may play an essential role.

Attention should be given to the interictal phase. People tend to minimize or forget the relatively mild symptoms that may occur between the (comparatively) stronger attacks. Some patients may report that the head remains slightly painful between attacks, pointing to a fluctuating pattern that never reaches a pain-free level. Sensitivity of the scalp to light touch or wind may be present between attacks. Females usually prefer having their hair styled in simple ways, as hypersensitive scalps are not compatible with a complex coiffure.

CEH may start early or late in life. It is interesting to note that the ‘age at onset’ distribution seems to be rather scattered during adulthood, with the frequency tending to diminish with age and increase again during the 7th decade. This may indicate that the mechanisms generating CEH vary with age. Theoretically, older patients could experience the syndrome because of degenerative disorders, while in younger subjects CEH could be related to trauma or other non-age-dependent predisposing factors. Other types of headache may occur in a given subject for a long period of time before the cervicogenic pattern develops. In a migraine patient, trauma may trigger CEH-like headaches that will be recognized as such by the patient, who usually differentiates it from the previous migrainous attacks. Comparing the age at onset of migraine (M), tension-type headache (TTH) and CEH the picture starts earlier for M than for the other two types of headache (11, 12) (CH: 35.06 ± 17.53; M: 18.4 ± 10.3; TTH: 29.0 ± 13.8; p = 0.0001). Concerning the age at first visit, CH patients were comparatively older than M patients (CH: 45.21 ± 13.67; M:34.35 ± 13.44; p = 0.003, ANOVA), but not significantly older than TTH patients (38.2 ± 17.5).

Migrainous traits may occur in CEH, but to a lesser extent (Table I).

The differential diagnosis may sometimes be difficult, as not all cases will present with a clear-cut picture. The author believes the existing diagnostic criteria to be appropriate for differentiating CEH from the most frequent types of headache - migraine and tension-type headache. However, the exact borderline between such disorders remain to be established from the pathophysiological point of view. The following are examples of CEH patients with particular differential diagnosis concerns.

### Case 1
AB, a 23-year-old female patient (born December 20, 1975), suffered the onset of CER at the age of 15, with left peri-auricular pain. With time, the pain became right-sided, over the TMJ/temporal area. She sometimes had diffuse pain in the right arm. Attacks occurred once a week, starting at any time and lasting 1 to 4 days. She complained of a sensation of edema over the zygomatic region, with her face appearing asymmetric when she looked in a mirror. Her friends observed the same phenomenon. No ocular autonomic disorders were reported by the patient. She sometimes experienced nausea, vomiting, and photo- and phonophobia during these attacks. Particular triggering factors were chewing and remaining for a long period of time in the same position, mostly while writing, when she tended to bend her head forward and to the right. She would improve when she wore the occlusion plaques prescribed by her dentist. A physical examination was normal, except for diffuse tenderness on palpation of the back of the head on the right side, the mandible ligaments, and face.

This patient meets the following important diagnostic criteria for CEH (13):

<table>
<thead>
<tr>
<th>Number</th>
<th>Criterion</th>
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<tbody>
<tr>
<td>Ia1</td>
<td>Precipitation by awkward head positioning;</td>
</tr>
<tr>
<td>Ib</td>
<td>Ipsilateral arm pain;</td>
</tr>
<tr>
<td>III</td>
<td>Unilateral without sideshift.</td>
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</tbody>
</table>

It is interesting to note that various forms of CEH may include TMJ complaints. The stylo-mandibular and temporo-mandibular ligaments may be tender on palpation, as seen in this case. It remains speculative whether trauma could have triggered both the CEH and the TMJ disorders in this patient.

### Case 2
MAGC, a 56-year-old male patient (born August 8, 1942) suffered from frequent frontal, bilateral migrainous headache during adolescence. He became asymptomatic but, many years later, occipital pain became frequent. A clinician diagnosed this pain as being secondary to hypertension, which disappeared with oral Metoprolol.

The patient was again asymptomatic for many years until September 1998, when he noticed left hemianopia in the nuchal, parietal, temporal and frontal areas. The pain could start in the back, with a needling or throbbing sensation. No trauma was reported. More recently ipsilateral arm pain was noticed. There was con-

### Table I. Comparison between migraine and CEH patients (11, 12).

<table>
<thead>
<tr>
<th></th>
<th>Cervicogenic headache</th>
<th>Migraine</th>
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<tbody>
<tr>
<td>Nausea</td>
<td>53%</td>
<td>89%</td>
</tr>
<tr>
<td>Vomiting</td>
<td>22%</td>
<td>61%</td>
</tr>
<tr>
<td>Photo- and phonophobia</td>
<td>53%</td>
<td>86%</td>
</tr>
<tr>
<td>Pulsating pain</td>
<td>50%</td>
<td>89%</td>
</tr>
</tbody>
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P < 0.001 for all items.
junctival injection and ptosis during the attacks, but no lacrimation or nasal symptoms. The duration and frequency of the attacks was highly variable. The main triggers were lying on the symptomatic side or extending the head; the patient reporting that he could feel the exact path of the pain. CEH triggered by getting into and out of the car, coughing, sneezing, or on right rotation with flexion of the neck were reported. During examination there was impressive limitation of the range of motion (ROM), mostly in extension, but a remarkable tolerance to digital pressure over the greater occipital nerve (GON) and the nuchal area. Three GON blockades performed elsewhere were ineffective, but we do not know the appropriateness of the technique used. Indomethacin 75 mg for 1 week was ineffective. CT and MR scans were unremarkable. This patient was referred for physiotherapy with very good subjective improvement.

The CEH diagnostic criteria (13) present in this case were:

Ia1 Precipitation by awkward head positioning
Ib Restricted ROM
Ic Ipsilateral arm pain

III Unilaterality without sideshift

The ROM was so reduced in this patient that he could turn his head only a few degrees in either direction. Surprisingly enough, the response to strong digital pressure was normal in terms of pain threshold asymmetry and attack precipitation. This shows that clinical diversity exists in CEH. In addition, the patient had a marked autonomic ocular disorder, indicating some overlapping with a cluster headache-like disorder.

Case 3

RK, a 31-year-old male patient born on 25 May 1964 suffered pain onset at the age of 16. He had 3 attacks per week, mostly between 15:00 and the evening hours. The pain started at the back of the head, and then spread to the temporal area, the patient describing it as throbbing and intense, severe enough to interfere with daily activities. 20% of the attacks were bilateral and 80% were unilateral (either left or right). There was no nausea, vomiting, photophobia or phonophobia. Moving the head too much or talking to someone at his side would induce pain within 15 minutes. Other triggering factors reported were lack of food, or drinking beer (but not distilled beverages). A brother and a sister were reported to have frequent headaches. When the GON was pressed he said, “I feel quite clearly that a pipe connects the posterior part of the head to the frontal area: You press there, it dilates here” and “I am certain the back is linked with the front”. A cervical MR was unremarkable. Atenolol 50 mg/day was prescribed and the CEH attacks stopped completely. From December 20 to January 5, 1997 the patient stayed in a hotel where he claimed that the bed was particularly hard. He was certain this was the cause of a new picture- diffuse, bilateral head pain, mostly on the left. Pain and paresthesia were present in the 4th and 5th left fingers. This pain was comparatively intense, non-throbbing, and without nausea, vomiting, or photo- or phonophobia. The symptoms were most severe just after waking and lasted all day long. Sumatriptan 100 mg was ineffective, while Toltenamic acid 200 mg was marginally effective. After bed rest the pain disappeared, but the fingers remained affected for some days.

The following CEH diagnostic criteria (13) were present in this case:

Ia1 Precipitation by neck movement
Ia2 Precipitation by GON pressure
Ic Ipsilateral finger symptoms

III Unilaterality without sideshift

(2nd picture)

Worth noting is the migraine “contamination” in this case. The first clinical picture was of a unilateral, alternating, throbbing pain responsive to 8-blockers and resembling ordinary migraine. There was, however, a back-to-front irradiation and a particular sensitivity to GON examination. The second form that the patient’s headache took was much more suggestive of CEH. This case underlines the possibility of CEH syndrome occurring in individuals with a clear history of migraine. The CEH profile in patients carrying a strong genetic predisposition to migraine remains to be established (14).

Case 4

MMC, a 25-year-old female patient (born June 14, 1971) suffered paroxysmal headache since she was 16 years old, usually 1 attack per month just before menstruation, with a duration of 2 days. The pain was exclusively right-sided, and “like a wire from the temporal area to the back”, according to the patient’s own description. The pain was pulsating, intense, and would get worse with routine activity. There was nausea, and photo- and phonophobia. Prodromal phenomena included emotional distress and tachycardia. Aphasia was present during the attacks. An MR scan and physical examination were normal. Treatment with sumatriptan 100 mg was effective. Substantial improvement with Atenolol 50 mg/day was obtained, and therefore she interrupted the prophylaxis 7 months later. After 9 months the pain recurred and a combination of 25 mg Amitryptiline and 50 mg Atenolol was prescribed.

In March 1999 a new pattern began. The pain, still right-sided, was much more intense and occurred 6 times per month. Head positioning, especially to the right, would trigger an attack. This had never occurred before. There was also irradiation to the right shoulder. When asked about trauma, she admitted that in December 98 she was hit from behind while riding her motorcycle, with immediate, intense neck pain. One month later, her car was hit from behind. The main CEH criteria (13) present in this case were:

Ia1 Precipitation by awkward neck positioning
Ic Ipsilateral shoulder pain

III Unilaterality without sideshift

(both the 1st and 2nd pictures)

As in the previous case, here the CEH seemed to occur in a patient with a predisposition to migraine. The initial picture (migraine) was also marked by a strict unilaterality. When the patient suffered her accidents, it is possible that the new superimposed picture of CEH occurred on the right side because of her lowered right threshold since the start of her migraines.
Final remarks

The neck may induce chronic, remitting head pain, perhaps more often than hitherto supposed. On studying cases of CEH, it becomes clear that distinct subtypes of the syndrome exist. Furthermore, CEH may occur in patients suffering from other headaches and/or in those with predisposition for head pain of a different type. The interplay between CEH and, in particular, migraine deserves study from the pathophysiological point of view, since clinically the two disorders do not seem to belong to the same category.

References

1. EDMEADS J: The cervical spine and headache.