Headache and chronic facial pain

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Key points

Headache has been described as the most common medical complaint known to man.

The majority of patients presenting with headache and chronic facial pain have a normal neurological examination.

The *International Classification of Headache Disorders* (2005) classifies headache into 14 main categories and more than 300 headache disorders.

Migraine management includes correct diagnosis, explanation, reassurance, predisposing/trigger identification and avoidance, and drug/non-drug intervention.

Multidisciplinary management of headache and chronic facial pain is of major importance.

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This article explores headache and chronic facial pain disorders that commonly present to the chronic pain clinician.

Headache

Headache has been described as the most common medical complaint known to man. The lifetime prevalence for headache of any kind is 93% in men and 99% in women.[1]

Primary headache accounts for more than 90% of headache complaints. This includes migraine, tension-type headache (TTH), cluster headache (CH), and the trigeminal autonomic cephalalgias, none of which is associated with demonstrable organic disease or structural neurological abnormality.

Pain in the head and neck is mediated by afferent fibres in the trigeminal nerve, nervus intermedius, glossopharyngeal, and vagus nerves, and by the upper cervical roots via the occipital nerves. Lesions or dysfunction of these nerves or central pathways may produce pain localized to the innervated area.

Classification

In 1988, the International Headache Society (IHS) published a comprehensive system to classify headache, *The International Classification of Headache Disorders* (ICHD). This system attempted to reconcile the many complex aspects of headache and facial pain into one usable system. A second edition was produced in 2004 and a further revision took place in 2005 (ICHD-IIR1), dividing headache and facial pain into 14 main categories, comprising more than 300 headache disorders. Table 1 shows the ICHD-IIR1 hierarchical classification of the disorders discussed in this article.

Diagnosis

A careful history, examination, and in some cases diagnostic tests will permit a diagnosis in the majority of cases. Nevertheless, a precise diagnosis may be impossible due to a mixed presentation. The pain history should include details of onset, intensity, frequency, quality, location, duration, presence of a preceding aura, aggravating and precipitating factors, relation to menstruation, and response to treatments. The past medical history should enquire about head injuries, intracranial infections, and previous neurosurgery. A complete drug history should be sought, including the frequency of use of analgesia, herbal medicines, and oral contraceptives.

A physical examination of the head and neck evaluating the cranial nerves may assist in diagnosing focal secondary causes. Temporomandibular joint (TMJ) tenderness, clicking, crepitus, and reduced jaw opening may be observed when joint disease is the source. Teeth inspection may reveal dental disease or bruxism. Temporal region tenderness may suggest temporal arteritis. Craniofacial myofascial tender points maybe present.

Laboratory studies can reveal underlying systemic causes of chronic headache, for example, full blood count, renal profile, thyroid studies, C-reactive protein, and erythrocyte sedimentation rate. Neuroimaging is not helpful in the diagnosis of primary headache and is indicated only when history or examination suggest a secondary cause. Computed tomography or magnetic resonance imaging (MRI) will detect most causes of secondary headache, including sinusitis, otitis media, dental disease, temporal arteritis, cranial aneurysm, malignancy, intracranial haemorrhage or infarction, and meningococcal infection.

Secondary headaches are usually of recent onset and associated with abnormal clinical findings secondary to underlying pathology. The indications for neuroimaging are listed in Table 2.

Migraine

Migraine is a primary headache disorder characterized by various combinations of neurological, autonomic, and gastrointestinal symptoms.

The incidence of migraine is 8.1 per 1000 (male:female ratio 1:6) with onset commonly presenting in the second or third decade. About 70% of migraineurs have a positive family

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Table 1 ICHD-IIR1 hierarchical classification.[2] Reproduced with permission from Blackwell Publishing

Primary headaches
1. Migraine
1.1. Migraine without aura
1.2. Migraine with aura
2. Tension-type headache
2.2. Frequent episodic tension-type headache
2.3. Chronic tension-type headache
3. CH and other trigeminal autonomic cephalalgias
3.1. Cluster headache
3.2. Paroxysmal hemicrania
3.3. SUNCT
4. Other primary headaches
4.7. Hemicrania continua
8. Headache attributed to a substance or its withdrawal
8.2. Medication overuse headache
11. Headache or facial pain attributed to disorder of cranium, neck, eyes, ears, nose, sinuses, teeth, or mouth
11.7. Headache or facial pain attributed to TMJ disorder
13. Cranial neuralgias and central causes of facial pain
13.1. Trigeminal neuralgia
13.15. Head or facial pain attributable to herpes zoster

Table 2 Indications for neuroimaging

Sudden onset headache
History suggestive of focal neurological dysfunction
Headache associated with other symptoms such as fever and weight loss
Previous history of malignancy
Previous history of chronic infection including HIV
Examination revealed neurological abnormality
Course of headache not corresponding to presumed diagnosis
Repeated attendance with the same symptoms

Table 3 Treatment of migraine[3]

Predisposing factors
Stress: lifestyle change, relaxation therapy, coping strategies, biofeedback, hypnotherapy
Depression/anxiety: specific therapy
Menstruation: mefenamic acid, frovatriptan, transdermal oestrogen, combined oral contraceptive, oestrogen/progesterone dermal implants
Trigger factors
Relaxation: 'weekend migraine'—change life style
Strenuous unaccustomed exercise: regular exercise
Dietary: alcohol, cheese—avoid provoking foodstuffs
Changes in habit: poor sleep, missing meals, long distance travel—avoid if possible
Menstruation: see above
Acute treatment: stepped management
Step 1: Simple oral analgesic ± anti-emetic
Aspirin, ibuprofen, tolfenamic acid, or both. Acetaminophen alone is not efficacious. Prochlorperazine, domperidone, or metoclopromide as indicated.
Step 2: Rectal analgesic ± anti-emetic
Diclofenac suppository ± rectal domperidone
Step 3: Specific anti-migraine drugs ± anti-emetic
Sumatriptan p.o./s.c., zolmitrptan p.o./nasal, rizatriptan buccal, naratriptan p.o.
Ergotamine in cases of recurrent relapse
Step 4: Combination therapy of steps 1, 2, 3, or both
NB: Opiods provide small additional benefit
Drug prophylaxis
First line: atenolol, propanolol, bisoprolol, amitriptyline, nortripyline
Second line: topiramate, sodium valproate
Third line: gabapentin, methysergide
Other: pizotifen, clonidine, verapamil, and fluoxetine have unproven efficacy
Non-drug interventions: exercise, physiotherapy, relaxation, stress reduction, biofeedback

history. Migraine is divided into two types: migraine without aura (70%) and migraine with aura (30%). The ICHD-IIR1 has specific criteria for diagnosing migraine with or without aura.[2]

Migraine with aura, previously termed *classic migraine*, has a reversible preceding aura comprising one or more of the following visual disturbances: homonymous hemianopsia, tunnel vision, scotoma, and photopsia. Additionally, patients may experience photophobia, phonophobia, paraesthesia, hemiplegia, aphasia, nausea, vomiting, abdominal discomfort, and diarrhoea. Migraine headache can be unilateral (60%) or bilateral (40%), located anywhere about the head or neck and last for 4–72 h. It has a throbbing, pulsating quality with moderate to severe intensity, and numerous accompanying features including nausea (90%), vomiting (33%), vertigo, fatigue, confusion, ataxia, drowsiness, photophobia, phonophobia, and nasal congestion. Migraine is aggravated by postural change, activity, and raised intracranial pressure. The recovery phase is one of the irritability or malaise, although some feel refreshed or euphoric. Migraine attacks are triggered by stress, menses, pregnancy, dietary habit (e.g. red wine, cheese, chocolate, and nuts), odours, light, and poor sleep.

The treatment of migraine (Table 3) is either by active or by preventative means. A combination of techniques can be implemented depending on attack frequency and severity.[3]

Tension-type headache

TTH is the most common primary headache disorder, previously termed *muscle contraction headache* and *stress headache*. It has a lifetime prevalence of 69% in men and 88% in women.[4] The ICHD-IIR1 has specific diagnostic criteria for TTH, subdividing it into episodic (ETTH) and chronic (CTTH), with a further subdivision of ETTH into frequent and infrequent.[2]

These headaches are characterized by generalized pressure or tightness in the head. The discomfort is mild to moderate and is unaffected by activity. Nausea, photophobia, or phonophobia are not prominent features. Infrequent ETTH, in which episodes occur less than once a month, do not have a significant impact on the individual, responding well to simple analgesics. Frequent ETTH and CTTH are associated with considerable disability. Diagnosis can be difficult because as many as 94% of migraineurs are bothered by co-existing TTH.[5]

In patients with CTTH, effective prophylactic treatment is the primary treatment aim. The secondary goal is careful management of any residual headache to minimize the risk of *medication*

overuse headache. Patients should limit their use of analgesics to two times weekly to prevent its development. A more frequent analgesic requirement indicates the need for prophylaxis, which should be aimed at those with both frequent ETTH and CTTH. The drug of choice is amitriptyline, which is the only secondary analgesic that has shown consistent efficacy across a number of studies. Other potentially useful agents include mirtazapine, fluoxitene, and clomipramine. Botulinum toxin injection for TTH has produced inconsistent results. Acupuncture is not currently recommended.[4] Relaxation techniques, biofeedback training, and cognitive behaviour therapy are empirically validated behavioural treatments for TTH, with meta-analyses indicating that they produce similar results to amitriptyline prophylaxis.[6]

Cluster headaches and other trigeminal autonomic cephalagias

The trigeminal autonomic cephalagias comprise headache with signs of cranial autonomic hyperactivity. CH predominately affects males: male to female ratio 9:1, with a prevalence of 0.1–0.4%. Age of onset is usually 20–40 yr old. Attacks occur for a period of several weeks or months, then remit, leaving the patients pain free for several months or years, only for the attacks to recur. The pain is so intense that the sufferer becomes extremely agitated and restless. One symptom of localized autonomic hyperactivity occurring in about 20% of patients is a Horner's syndrome, affecting the ipsilateral side. Unlike migraine, nausea and vomiting are uncommon. Trigger factors, troublesome only during the cluster phase, include alcohol, vasodilator drugs, and sleep apnoea-induced hypoxaemia.

Treatment is aimed at cluster prevention; the attacks are too brief and frequent to treat easily. Drugs of known prophylactic efficacy include verapamil (sometimes required in doses of up to 960 mg per day), methysergide (significant potential long-term side-effects include pulmonary or retroperitoneal fibrosis), ergotamine (1–2 mg p.r. for short-term management of episodic cluster headaches), lithium (tolerance may be a problem), and prednisolone (60–100 mg per day for 2–5 days).[3] Evidence for anti-epileptics including gabapentin and sodium valproate is limited. Symptomatic treatment chiefly involves the serotonin agonists, sumatriptan (intranasal or s.c.) and zolmitriptan. Oxygen inhalation (100% at 6–10 litre min^{-1} for 10–20 min) appears effective in young patients. A subgroup of patients may respond to intranasal lidocaine and there is current interest in the use of occipital nerve stimulation, although there is, as yet, insufficient evidence of its efficacy.

The paroxysmal hemicranias (PH) are headaches that resemble CH but fail to respond to standard anti-cluster therapy. Attacks of PH have similar characteristics to CH, but are shorter and more frequent, lasting 8–25 min and occurring 5–24 times per day. A female predominance occurs (1:3). Diagnostically and therapeutically, PH exhibits an absolute responsiveness to indomethacin.

Short-lasting, unilateral neuralgiform headache attacks with conjunctival injection and tearing (SUNCT) syndrome is distinguished from CH and PH by the extremely short duration of attacks (20–120 s) and high frequency (30–100 times daily). Most occur during daylight hours and the pain distribution is confined to the ophthalmic division of the trigeminal nerve. For this latter reason, SUNCT is commonly misdiagnosed as trigeminal neuralgia (TGN). There is considerable autonomic hyperactivity during attacks, the most important of which is conjunctival tearing and injection. Preventative treatment is achievable with anticonvulsants: topiramate, valproate, gabapentin, and lamotrigine. Attacks are too short to treat.

Chronic daily headache

A number of patients complain of very frequent and almost continuous headaches, which are described under the term 'chronic daily headache' (CDH). This does not appear in the ICHD-IIR1 classification. The reported prevalence of CDH in the general population is 3–5%. CDH includes a heterogeneous group of primary headaches that present for more than 15 days per month, and for longer than 3 months. There are four main categories: transformed migraine (TM), CTTH, new daily persistent headache (NDPH), and hemicrania continua. Distinction between TM, CTTH, and NDPH is often difficult.

TM forms the largest group of CDH sufferers (70–80%); it is not included in the ICHD-IIR1 classification but bears strong similarities to the ICHD-IIR1 subform of *chronic migraine*. Specifically, TM refers to the presence of periodic migraine that 'transforms' to a more frequent and continuous headache over many years. Many headaches retain certain characteristics of migraine, whereas others are indistinguishable from CTTH. Frequent accompaniments include sleep disturbance, analgesic overuse, anxiety, depression, and impaired physical, social, and occupational functioning. Treatment of TM is managed no differently to regular migraine summarized in Table 2.[3]

Hemicrania continua is a rare form of unremitting CDH that presents unilaterally. Autonomic symptoms such as rhinorrhoea and conjuctival injection are often present. It frequently responds to indomethacin.

Medication-overuse headache (or analgesic rebound headache) describes the headache-precipitating tendency that may result if analgesics are used frequently or long term; the condition is widespread in the headache population. Daily headache can drive some sufferers to over-medicate on analgesia, further perpetuating their headache symptoms. It may be in part due to a physical and psychological dependency and the prime quest for pain control and symptom relief. Provoking agents include ergotamine, triptans, simple analgesics, opioids, and combination analgesics. Medication overuse is present in up to 40% of chronic migraineurs. Management of medication-overuse headache primarily involves education, explanation of the diagnosis, and follow-up. Gradual withdrawal of the causative agent improves symptoms (7–10 days in triptan overuse and 2–4 weeks in opioid overuse). The majority of patients revert to their original headache disorder within 2 months.

Chronic facial pain

A host of facial pain disorders can present to the chronic pain management clinic. These are classified in Groups 11, 13, and 14 of the ICHD-IIR1.[2] The most frequently presenting conditions are temporomandibular disorder (TMD), TGN, post-herpetic neuralgia (PHN), and persistent idiopathic facial pain.

Temporomandibular disorder

TMD, which is not included in the IHS classification, is used as a generic term to include the heterogeneous conditions, including muscle and joint disorders, which cause symptoms in and around the TMJ. In addition to pain, joint sounds at mouth opening, clunking, locking, and restriction of mouth opening may occur. The Research Diagnostic Criteria for TMD is the most widely used diagnostic system.[7] Pain from the TMJ or related tissues is common. TMD has a reported prevalence of 30–46% with pain occurring in 5–15%.[8]

The difficulty in diagnosing TMJ problems lies in determining whether pain in the area of the joint is due to a muscle, joint, or systemic disorder. Joint MRI, bone scintigraphy, or both remain the investigations of choice. Most TMJ arthralgias cause pain anterior to the ear and incorporate an element of myofascial pain. The management of TMJ disorders aims to reduce pain and restore normal joint function. A self-care programme is probably the most important component of overall management of TMD.[10] Programmes include: jaw relaxation techniques; softer diet; chewing techniques; use of non-steroidal anti-inflammatory drugs (NSAIDs); and avoidance of teeth grinding, prolonged jaw opening, prone sleeping, and caffeine. Other treatments include intra-oral splinting, primary and secondary analgesia, and physiotherapy. In patients with chronic TMD pain, tricyclic antidepressants can impact upon insomnia, depression, and pain. Cognitive-behavioural techniques (biofeedback, relaxation, and stress management) can be effective alone or in conjunction with other treatments.

Trigeminal neuralgia

TGN has an incidence of 3–5/100 000, with a peak onset in the fifth and sixth decades. The hallmark is agonizing, paroxysmal, lancinations confined strictly to one or more branches of the trigeminal nerve. Non-noxious stimuli trigger the pain, typically around the peri-oral region. Mild flushing in the corresponding division may occur during an attack. The pain is nearly always unilateral.

The diagnosis of TGN is made by careful history and examination. In classical TGN, neurological examination and standard neuroimaging are normal. High resolution MRI and magnetic resonance angiography provide detailed images of the cerebellopontine angle and of any nerve-vascular contacts that exist. About 80% of cases are associated with vascular compression of the trigeminal nerve. TGN is diagnosed in up to 5% of patients with multiple sclerosis. Other causes include schwannoma, meningioma, epidermoid cyst, pontine infarction, and Chiari malformation.

About 70% of patients with TGN can initially be controlled non-surgically. The drug of choice is carbamazepine; systematic review has demonstrated a number-needed-to-treat (NNT) of 1.8 for carbamazepine in trigeminal neuralgia.[9] Baclofen, gabapentin, lamotrigine, oxcarbazepine, and topiramate have all been used successfully. Those that fail medical treatment may respond to one of the surgical options available. Percutaneous neuroablative techniques, aimed at denervating the gasserian ganglion, include glycerol gangliolysis, radiofrequency thermocoagulation, microcompression, and stereotactic radiosurgery. All are safe and well-tolerated procedures. Microvascular decompression surgery through the posterior fossa is directly aimed at the proposed cause, the target area being the nerve–pons junction. It provides prolonged analgesia with pain relief in 90% of patients short-term, and 55–70% long-term. The procedure has an annual recurrence rate of 3.5%, mortality of 0.2–1%, and hearing loss occurs in 1%.

Post-herpetic neuralgia

The annual incidence of acute zoster infection is 400 in 100 000. Herpes zoster infection affects middle-aged to elderly individuals without gender bias. The immunocompromised are at greater risk. The acute infection is self-limiting, associated with a rash and lasting 1–2 weeks. PHN occurs in 15% of acute zoster infections.

PHN consists of pain persisting in the zoster affected dermatome at 3 months.[2] It typically affects the first division of the trigeminal nerve and may be associated with trophic changes such as scarring, loss of pigmentation and hair, and allodynia. Paroxysmal bouts of pain occur during the day, superimposed on burning pain or dysaesthesia. Individuals with PHN are often very difficult to treat. Fortunately, the severity of many cases subsides with time, with the majority resolving within the first year. Pharmacological treatment of PHN includes single or combination therapy using antidepressants and anticonvulsants. Amitriptyline is the agent of choice, with evidence that early institution of treatment shortens the duration and severity of symptoms. Gabapentin is useful if there is associated insomnia. Capsaicin cream, topical lidocaine, and opioids have all been used with varying degrees of success.

Persistent idiopathic facial pain

Persistent idiopathic facial pain, formerly known as *atypical facial pain*, is a persistent pain that does not have the characteristics of the cranial neuralgias, and is not attributed to another disorder.[2] The incidence is 1/100 000; both sexes are affected equally, but more women than men seek medical care. There is a strong association with functional disorders such as irritable bowel syndrome and psychological distress.

Pain may be initiated by surgery or trauma to the face, teeth, or gums. It is present daily for all or most of the day, without any identifiable local cause. The pain is often described as a continuous dull ache with intermittent severe episodes, affecting any area of the face, but sparing the joints and muscles of mastication. It is

typically deep and poorly localized, and will often have been present for several years. There is no associated sensory loss or other physical signs. Investigations including facial and jaw X-ray do not demonstrate any relevant abnormality. Primary analgesics are ineffective, and lack of response to diagnostic nerve block is a common feature. The analgesic response sometimes observed with tricyclic agents appears to be independent of their antidepressant effect. Long-term support is often necessary.

References

1. Rasmussen BK, Jensen R, Olesen JA. A population-based analysis of the diagnostic criteria of the International Headache Society. *Cephalalgia* 1991; **11**: 129–34
2. Headache Classification Subcommittee of the International Headache Society (IHS). *The International Classification of Headache Disorders*, 2nd Edn, 1st revision (May, 2005) (ICHD-IIR1). Available from http://www.ihs-classification.org
3. Steiner TJ, MacGregor EA, Davies PTG. Guidelines for All Healthcare Professionals in the Diagnosis and Management of Migraine, Tension-Type, Cluster and Medication-Overuse Headache, 3rd Edn. British Association for the Study of Headache, 2007; 1–52
4. Rasmussen BK. Migraine and tension-type headache in a general population: psychosocial factors. *Int J Epidemiol* 1992; **21**: 1138–43
5. Lyngberg AC, Rasmussen BK, Jørgensen T, Jensen R. Has the prevalence of migraine and tension-type headache changed over a 12-year period? A Danish population survey. *Eur J Epidemiol* 2005; **20**: 243–9
6. Penzien DB, Rains JC, Lipchik GL, Creer TL. Behavioural interventions for tension-type headache: overview of current therapies and recommendation for a self-management model for chronic headache. *Curr Pain Headache Rep* 2004; **8**: 489–99
7. International Consortium for Research. 2004. Available from http://www.rdctmdinternational.org
8. Wänman A. Temporomandibular disorders among smokers and non-smokers: a longitudinal cohort study. *J Orofac Pain* 2005; **19**: 209–17
9. Wiffen PJ, McQuay HJ, Moore RA. Carbamazepine for acute and chronic pain. *Cochrane Database Syst Rev* 2005: CD005451. doi:10.1002/14651858.CD005451
10. Fricton JR. Temporomandibular muscle and joint disorders. *Pain: Clinical Updates* 2004; XII, No. 2

Please see multiple choice questions 19–23