

FACIAL PAIN: NEUROLOGICAL AND NON-NEUROLOGICAL

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The article will give the reader:

- ▶ An overview of facial pain in terms of epidemiology, classification, diagnosis and management
- ▶ Three case histories on which to try out diagnostic skills
- ▶ An overview of management of three types of facial pain.

▶ EPIDEMIOLOGY

Pain in the facial area may be due to neurological or vascular causes, but equally well may be dental in origin. The patient will often make the first attempt at diagnosis in that he or she chooses to consult either the dentist or the doctor. This may therefore lead to inappropriate diagnosis and treatment. Many patients with trigeminal neuralgia complain that their dentist treated them for dental causes of pain before finally they received the correct diagnosis. This is, however, highly understandable as dental pain is extremely common whereas trigeminal neuralgia is a rare condition and primary care medical and dental practitioners may only see three or four cases in their practising lifetime. All the neurological and vascular causes of facial pain (excluding headaches) are rare compared to the dental and temporomandibular causes. The risk factors for some of the conditions are known, but there is little information on natural history and prognosis. Further details of the epidemiology of facial pain can be found in *Epidemiology of pain*, which has been written using evidence based methodology.¹

CLASSIFICATION AND DIAGNOSIS

A classification system is useful when attempting to make a diagnosis, to facilitate treatment decisions, and to predict future outcome. Not only does the pain itself need to be classified but also its psychosocial effects. The International Association for the Study of Pain (IASP)² and the International Headache Society (IHS)³ have both classified these pains, and the fourth revision of the *Diagnostic and statistical manual* (DSM-IV) enables you to classify psychiatric disorders. Unfortunately only a handful of orofacial conditions have been well validated by case control studies.

I find the classification into musculoligamentous (group 1), dental (group 2), and neurological/vascular (group 3), as adapted from Hapak *et al*, to be a useful system (table 1).⁴ Group 1 patients are in general best referred to oral physicians or maxillofacial surgeons who work in the secondary care dental system, group 2 to dentists, and the last group to neurologists. The presence of psychosocial disability will also affect referrals as those with higher levels may require referral to liaison psychiatrists.

The diagnosis of the majority of these facial pains is based on a careful history and examination. The patient must be given time to “tell their story” in order to determine both the pain disease and the pain illness/suffering. This is greatly facilitated if patients are given questionnaires to complete before the formal consultation. There are several questionnaires we use as standard. The Brief Pain Inventory (BPI) uses a visual analogue scale (0–10) to determine severity of pain and the impact on quality of life issues such as mood, relationships, and work. The Hospital Anxiety and Depression scale (HAD) is used to determine if there is evidence for a diagnosis of depression or anxiety. We ask patients to complete a questionnaire about their treatment goals as it gives us an indication of which issues are important to the patient—for example, reassurance that their pain is not caused by a serious underlying disease, taking less medication, having a diagnosis, or being able to communicate about their pain. During the consultation we will use the McGill Pain Questionnaire (MPQ) to gauge not only the sensory component of the pain but also the affective and evaluative aspects. It has been validated in a variety of orofacial conditions. Further discussions on measurements can be found in the *Textbook of pain*.⁵ It is important to ascertain whether patients have other pain complaints and symptoms which might suggest a need to undertake an examination of their mental state.

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Table 1 Classification of orofacial pain

Musculoligamentous/soft tissue	Dentoalveolar	Neurological/vascular
Temporomandibular joint (TMJ) pain	Dentinal	Trigeminal neuralgia
Facial arthromyalgia, myofascial pain	Periodontal	Glossopharyngeal neuralgia
Atypical facial pain/idiopathic orofacial pain	Pulpal	Nerve compression
Salivary gland disease	Cracked tooth syndrome	Cluster headache
Optic neuritis	Maxillary sinusitis	Post-herpetic neuralgia
Internal derangements TMJ		Cranial arteritis
Burning mouth	Thermal sensitivities	Pre-trigeminal neuralgia
Candidiasis	Atypical odontalgia	SUNCT
Cancer, sinuses, nasopharynx, brain		Ramsay Hunt
		Tolosa Hunt syndrome

SUNCT, short lasting unilateral neuralgiform headache attacks with conjunctival injection and tearing.

MANAGEMENT PRINCIPLES

In many respects treatment of a patient with chronic facial pain is no different from treating any patient with chronic pain, as the psychosocial and behavioural response patterns are the same despite different medical and dental causes. However, you need to remember that the psychological and symbolic significance of the head in the development of self esteem, body image, and interpersonal relationships confers special meaning on pain in this area.

Time needs to be spent in educating the patient and coming to a negotiated treatment plan that places the patient in the centre. This process involves active patient participation, good communication skills, appropriate choice of treatment based on high quality evidence, increased patient information, and self support as well as an emphatic clinician. Chronic facial pain cannot always be totally abolished and so it is important to have in place long term strategies for its management. Regular reassessment is vital and may even include a possible change in diagnosis. A holistic approach is essential and patients' beliefs, expectations, and own goals must be explored using questions shown in the box below.

The aim of treatment can be summarised as below:

- ▶ Eliminate or minimise the facial pain
- ▶ Eliminate or minimise negative cognitive, behavioural, and emotional factors
- ▶ Increase efficacy of drug treatment by careful choice
- ▶ Improve adherence by ensuring you have a well informed patient
- ▶ Encourage self management which increases control over pain.

Treatments divide into medical, surgical, and alternative (ranging from acupuncture to cognitive behaviour therapy) and patients may need a variety of these.

SPECIFIC CONDITIONS

Three orofacial pains, their presentation, and management will be discussed. Read through the three case notes presented

Questions to ask patients about their expectations for management of their facial pain

- ▶ What are the most important results you hope to receive from this treatment?
- ▶ Are you expecting a complete cure or just relief of pain?
- ▶ How much reduction in suffering would be acceptable if complete reduction were not possible?
- ▶ Are you looking for rapid results or are you prepared to accept slow change?
- ▶ How important is your quality of life and how much can this be compromised to achieve better pain control?
- ▶ What help do you need to develop active coping skills?

below and then answer the following questions on each of the cases before reading the answers at the end of the article. Alternatively you could use these problem solving cases as a basis for a training session and invite an expert to facilitate the session.

1. List the main characteristics of each of the pains.
2. What measurements have you been given and how useful are they?
3. What other factors has the clinician given you that may help in management?
4. What is the differential diagnosis for each case, how would the list differ if you were in primary or secondary care sector?
5. Are there any diagnostic tests you could perform which would help?
6. What details in the history will help you determine whether the patient is going to adhere to treatment and whether it will be successful?
7. How would you start managing the patient?

Case 1

Mrs Willis is 40 years old and presents with a two year history of preauricular pain. The pain began gradually and she has had periods of weeks when the pain disappeared. The rest of the time she has pain which fluctuates in severity from 2–7 on a visual analogue scale (VAS) of 1–10, with an average score of 4. On the MPQ she chooses the following words: drilling, pressing, hurting, tender, annoying, nagging. The pain begins preauricular and then radiates behind and into the ear as well as partially down the muscles of the face. It is bilateral but worse on the left. It is worse when eating hard things or biting into food and is helped by rest and analgesics. It is associated with some limitation in opening, a clenching habit, and clicking of the left joint. She suffers from headaches, has occasional tinnitus, back pain, and premenstrual pain, and does not sleep well. On the HAD she shows evidence of mild depression and anxiety. The BPI shows mild impact of the pain on her quality of life. She was recently divorced and is now bringing up a 12 year old girl on her own. Her parents help her out while she goes out to work as a computer programmer. She thinks the pain is caused by a disease of the joint and would like some surgery for it. On examination there is some limitation in opening and an audible click on opening wide is heard. The muscles of mastication are tender on palpation. Intraorally there are signs of frictional keratosis (white lines) in the buccal mucosa in line with the occlusal plane.

Case 2

Mr Jones is 57 years old and presented complaining of a sharp, shooting pain on the left side of his face. The pain began two months ago but he had a similar episode one and a half years ago when it lasted for six weeks and disappeared completely

Table 2 Characteristics of temporomandibular joint (TMJ) pains

Site	TMJ and associated musculature
Radiation	Associated muscles, temple, neck
Character	Dull, aching
Severity	Mild to moderate
Duration	Weeks to years
Periodicity	Continuous, can be intermittent or worse on waking or at the end of the day
Provoking factors	Jaw movement, eating, stress
Relieving factors	Jaw rest, tricyclic drugs
Associated factors	Limited mouth opening, TMJ parafunction, anxiety

until recently. Each bout of pain lasts for a few seconds but the bouts seem to merge together and so it can seem like several minutes of pain. He may be free of pain for several hours. The pain begins in the left nasolabial fold and then spreads across the whole cheek up to the ear. He grades an average 7/10 on a VAS with a range of 4–10. The words chosen on the MPQ are shooting, stabbing, sharp, stinging, tender, exhausting, terrifying, gruelling, wretched, unbearable, and torturing. It appears to be made worse when eating, shaving and talking. He is terrified of brushing his teeth. He has lost weight and has stopped going out, as he cannot get through a meal. Sleep brings him relief. The HAD shows mild anxiety and severe depression. On the BPI five out of the seven quality of life parameters score above 5/10. Mr Jones has had time off work as he works as a telephone operator. He is married and has two grown up children. Mr Jones thinks he has a nerve pain and wants his nerve cutting. Examination shows that touching the skin in the area of the infraorbital nerve provokes pain. There are no other abnormalities.

Case 3

Mr Hinds is 30 years old and has had facial pain for four years. The pain began in his upper left jaw in his teeth and he had extensive dental treatment leading to root canal therapy and finally extraction of his upper molar. He had limited pain relief after each of the procedures. There have been periods of no pain after dental treatment, but since this finished a year ago the pain has been continuous. The pain scores on VAS range from 3–8 with an average of 4 out of 10. It does fluctuate in intensity and he may not be aware of it during the day, but it is worse in the evenings. The pain is described on the MPQ as boring, sharp, gnawing, tugging, aching, tender, tiring, wretched, miserable, radiating, and nagging. The pain is now located both externally and intraorally on the left maxilla and radiates as far as the forehead and down to the neck, remaining mainly left sided. Nothing seems to help and he has stopped using all analgesics as they do not help. Mr Hinds reports headaches, neck and back pains, as well as occasional pruritus. The HAD shows pronounced anxiety but no depression. The BPI shows five out of seven quality of life measures scoring above 5. Mr Hinds is married and has two children. He reported an unhappy childhood with frequent bullying. He had an authoritarian father whereas his mother was sympathetic towards his problems and ill health. He left school at 18 and did a course in social sciences. He has had a series of jobs in the civil service working on population surveys. He enjoys his job but finds it stressful at times. Four years ago a very close school friend died in an accident and he moved house five years ago. The lack of diagnosis has been very frustrating and he now wonders whether he may have a brain tumour. Examination reveals no abnormalities and cranial nerve testing is normal.

For answers to questions 1–7 on these cases, see box at the end of the article.

TEMPOROMANDIBULAR DISORDERS

Definitions, aetiology, diagnostic criteria, and investigations

The most common names are temporomandibular pain and dysfunction syndrome, oromandibular dysfunction, and facial arthromyalgia (FAM). The first two are by definition associated with some form of temporomandibular joint (TMJ) dysfunction whereas the third is a chronic pain condition not necessarily associated with dysfunction.

The aetiology is unknown. One theory suggests psychogenic factors such as depression, other bodily pains, and being female are risk factors. Other suggested factors include muscle hyperactivity, parafunctional habits, anxiety, stressful adverse life events, and vulnerable personality types (table 2). Occlusal factors and traumatic (meniscal displacement) have both been proposed, but the evidence is weak and there are no controlled studies.

The principal findings are tenderness of the TMJ and associated muscles of mastication, trismus, limited or jerky jaw movements, and evidence of bruxism—tooth wear, and frictional keratosis of the buccal mucosa and the tongue. Signs of internal derangement of the TMJ (subluxation) include clicking noises and lateral displacement of the meniscus.

Although radiography, arthroscopy, and magnetic resonance imaging are routinely used for investigation, none have been evaluated for sensitivity and specificity.

Evidence based evaluation of management

The American Dental Association recommends that only conservative, reversible treatments are used.⁶ There is no randomised controlled trial of surgical intervention. One systematic review has been done of the use of occlusal appliances and rehabilitation.⁷ Currently antidepressant therapy and psychological treatments seem the best approaches (table 3).

The prognosis for TMJ disorders is good, but pain is likely for 2–3 years.

TRIGEMINAL NEURALGIA

Definition, aetiology, diagnostic criteria, investigations

Trigeminal neuralgia is defined by the IASP as “a sudden, usually unilateral, severe, brief, stabbing, recurrent pain in the distribution of one or more branches of the fifth cranial nerve”. The aetiology of this neuropathic pain remains unknown although it is postulated that the pathophysiological mechanism is that of compression of the trigeminal nerve by blood vessels such as the cerebral arteries in the root entry zone of the pons. In a minority of cases the trigeminal neuralgia is secondary to benign or malignant tumours. Table 4 lists the IHS criteria for trigeminal neuralgia.

Table 3 Evidence based treatment of TMJ disorders

Treatment	Outcome	Type of evidence
Pharmacological		
Opioids	Unknown	No evidence
NSAIDs	Not effective	1 RCT
Minor tranquilisers	Effective	1 RCT
	Not effective	1 RCT
Antidepressants	Effective	1 Crossover
	Effective	3 RCTs
Physical		
Exercises		No controlled study
Heat		No controlled study
Ultrasound		No controlled study
Bite appliances	Not effective	1 RCT
Occlusal rehabilitation		No controlled study
Injection therapies		
Ringers solution	Ineffective	1 RCT
	Effective	1 uncontrolled
Steroids	Effective	1 uncontrolled
Anti-inflammatory (superoxide dismutase)	Effective	1 uncontrolled
Psychology		
Reassurance	Effective	Anecdotal
Cognitive behaviour therapy	Effective, increases control	2 RCTs, 1 uncontrolled

NSAIDs, non-steroidal anti-inflammatory drugs, RCT, randomised controlled trial

Table 4 International Headache Society criteria for trigeminal neuralgia

- A. Paroxysmal attacks of facial or frontal pain which last a few seconds to less than 2 minutes
- B. Pain has at least 4 of the following characteristics:
 1. Distribution along one or more divisions of the trigeminal nerve
 2. Sudden, intense sharp, superficial, stabbing or burning in quality
 3. Pain intensity severe
 4. Precipitation from trigger areas, or by certain daily activities such as eating, talking, washing the face or cleaning the teeth
 5. Between paroxysms the patient is entirely asymptomatic
- C. No neurological deficit
- D. Attacks are stereotyped in the individual patient
- E. Exclusion of other causes of facial pain by history, physical examination, and special investigation when necessary.

Computed tomography is useful to exclude tumours, whereas magnetic resonance imaging is used to assess whether compression of the trigeminal nerve has occurred. Although greater specificity and sensitivity is claimed for the latter, the evidence is not of high quality.

Evidence based evaluation of management

All patients are started on drug treatment. If this either fails to control pain or produces unacceptable side effects, surgery is then offered. The medical treatment of trigeminal neuralgia

has been published in *Clinical Evidence* and is kept updated; it includes two systematic reviews.⁸ The results are summarised in table 5. Many drugs used in trigeminal neuralgia have not been evaluated in randomised controlled trials (RCTs), and their use has been based on single case reports. Tocainide was used in an RCT but found to be too toxic to recommend its use. Proparacaine eye drops, when used in an RCT, proved to be ineffective. All antiepileptic drugs have side effects and a survey among patients has shown that all have side effects with a mean report of three. Drowsiness, inability to concentrate,

Table 5 Drugs used in the management of trigeminal neuralgia

Drug	Daily dose range	Efficacy	Side effects	Level of evidence
Baclofen	50–80 mg	Good	Few, beware rapid withdrawal	RCT
Carbamazepine	300–1000 mg	Excellent	Neurological side effects, introduce slowly, drug interactions	RCT
Clonazepam	4–8mg	Low	Severe drowsiness	Case report
Gabapentin	1800–3600 mg	Good	Neurological side effects	Case reports
Lamotrigine	200–400 mg	Good	Neurological side effects, rapid dose escalation leads to rashes	Small RCT
Oxcarbazepine	300–1200 mg	Good	Neurological side effects, hyponatraemia in high doses, no major drug interactions	Case reports
Phenytoin	200–300 mg	Good	Neurological side effects, easy to overdose	Case report
Pimozide	4–12 g	Excellent	Severe side effects, neurological	RCT
Tizanidine	6–18 mg	Poor	Neurological side effects	RCT
Topiramate	25–250 mg	May be good	Neurological side effects	Case report
Valproic acid	600–1200 mg	Poor	Neurological side effects	Case report

Table 6 Surgical management of trigeminal neuralgia

Type of surgery	Procedure	Mortality	Morbidity, recurrence rates
Peripheral therapies: cryotherapy, neurectomy, alcohol injections	The peripheral nerve branch is either directly exposed or injected under local anaesthesia	Nil	Low and local, mainly mild sensory loss. Recurrence rate mean 10 months
Gasserian ganglion	Short acting general anaesthesia used to penetrate foramen ovale		All result in varying degrees of sensory loss, which can lead to eye problems, temporary motor weakness
Radiofrequency thermocoagulation	The Gasserian ganglion is subjected to temperature varying from 60–80°C	Low	Highest risk of anaesthesia dolorosa. Recurrence rate at 5 years 60%
Glycerol injection	Meckel's cave is filled with glycerol	Very low	Less likely to cause eye problems or anaesthesia dolorosa. Recurrence rate at 5 years 65%
Microcompression	The Gasserian ganglion is compressed by a balloon for a few seconds	Very low	Peroperative hypotensive episodes. Recurrence rate at 2 years 20%
Posterior fossa	Full general anaesthesia used		7th and 8th cranial nerves may be affected
Microvascular decompression	Any vessels lying on the trigeminal nerve at the point of entry to the cranium are moved aside or removed	Up to 0.4 %	Minimal effects on sensory loss or eye problems, surgeon dependent. Recurrence rates at 5 years 25%
Gamma knife	Using stereotactic techniques the posterior fossa is identified and 75 G beam directed at the trigeminal nerve	Unknown at present	Long term effects of radiation unknown, takes up to 6 months for an effect. Recurrence rate at 2 years 25%

Table 7 Characteristics of idiopathic or atypical facial pain

1. Site	Deep non-muscular areas of face, unilateral or bilateral, does not follow nerve distribution
2. Radiation	Poor localisation
3. Character	Throbbing, deep, diffuse, boring, nagging
4. Severity	Moderate to severe
5. Duration	Weeks to years
6. Periodicity	Continuous, but can have prolonged pain free periods
7. Provoking factors	Stress, fatigue
8. Relieving factors	Rest, tricyclic drugs
9. Associated factors	Anxiety, depression, other bodily pains

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ataxia, and diplopia are the most commonly reported ones. I encourage patients to keep a pain diary to note how and when they took their medication and record the outcome, including side effects.

There is no high quality evidence in the surgical field and the only RCTs on peripheral treatments showed no improvement. Unfortunately most of the data have not even been analysed objectively and only a small percentage of the papers report their findings using Kaplan-Meier analysis. A summary is provided in table 6. A discussion of the techniques can be found in books on trigeminal neuralgia.⁹ Patient information and support is provided by the US and UK trigeminal neuralgia associations who also have excellent web sites (www.tna-support.org and www.tna-uk.org.uk).

IDIOPATHIC OR ATYPICAL FACIAL PAIN

Definitions, epidemiology, diagnostic criteria, investigations

The definition for this condition is very unclear and remains a subject of controversy according to the IASP. The IHS considers it a diagnosis of exclusion. Many patients suffer from facial pain as shown on epidemiological surveys, but because of lack of diagnostic criteria it is impossible to determine whether it is all the same condition. The characteristics as used in trials are listed in table 7 and the case described is very typical of those presenting in the secondary care system.

Examination shows no abnormalities. There are no appropriate investigations.

Evidence based management

Three trials, two of which were RCTs, have shown that antidepressants are effective in this condition. Treatment needs to be continued for between nine weeks and two years. Treatment of orofacial pain has been included in a systematic review on the use of antidepressants in pain.¹⁰

Reassurance that nothing is wrong does not help, and if the following factors are gone through during the consultation management is optimised:

- ▶ Clear statement that examination and investigations are negative
- ▶ Acknowledgment of the reality of pain
- ▶ Agreement that the patient is ill
- ▶ Allowing ventilation of patient's beliefs and attributions
- ▶ Educating the patient that stress plays a role, but introducing it gradually
- ▶ Non-illness behaviours and communications are reinforced with written material.

CONCLUSION

There remains a need for considerable research in this field if we are to improve our diagnosis and management of facial pain.

Answers to the three case studies: general comments that apply to all cases

1. Pain history should be taken using Ryle's classification.¹¹ You can look up the diagnostic criteria of all orofacial pain conditions in IASP² or IHS³ systems
2. Review section on diagnosis. Epidemiological studies have shown that up to 70% of patients with orofacial pain will report psychological impact
3. The histories show disability and widespread effects. They stress the importance of ascertaining whether the patient has whole body pains or not and a social history or life events that may contribute to initiating or potentiating the pain
4. Case 1: temporomandibular pain; case 2: trigeminal neuralgia; case 3: chronic idiopathic or atypical facial pain. In primary care, ear problems and TMJ are far more common. Patients with atypical facial pain are likely to have more disability among the secondary care sector; trigeminal neuralgia is the rarest.
5. There are no diagnostic tests. Computed tomography is helpful to eliminate a cancer.
6. Patients beliefs are fundamental and must be clarified before treatment is initiated. One of the most important ones is reassurance that there is no cancer. Severity of disease, impact on quality of life, insight, and chronicity will impact on adherence as well as how the family views it
7. Patients want reassurance that their pain is real and that it is not caused by mental illness. Showing patients the interaction between mental and somatic factors using the gate control theory of pain helps patients come to terms with their diagnosis and treatment

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