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Diagnosis and management of chronic facial pain

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Learning objectives

By reading this article, you should be able to:

- Classify the different causes of facial pain.
- Assess patients with facial pain.
- Formulate treatment plans for patients with facial pain.

The diagnosis of facial pain is challenging because of interdisciplinary differences in defining both anatomical boundaries and the diagnoses themselves.[1] The International Classification of Headache Disorders-3 (ICHD-3) provides a comprehensive systematic approach for clinicians to diagnose head pain. This review focuses on the diagnosis of orofacial pain associated with disorders of the cranial nerves using ICHD-3 nomenclature. Recently, there has been a movement within this group to classify facial pain further. This project has become known as the International Classification of Orofacial Pain (ICOP) .[2] They have recently launched their classification paradigm as a 'beta' version for review, which is worthwhile reading into the other broader differential diagnosis of facial pain. For completeness, we have outlined the differential diagnosis of non-neuralgic orofacial pain in Table 1, but full discussion of all these entities is outside the scope of this article.

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Key points

- Facial pain/neuralgia is challenging to localise and to diagnose; history, physical examination and knowledge of the different peripheral nerves help with diagnosis and guide treatment.
- Trigeminal neuralgia (TN) is the most common and well-known facial neuralgia.
- Treatment of trigeminal neuralgia is done conservatively, commonly with good response to carbamazepine or oxcarbazepine. If no benefit is reached with pharmacologic means, a surgical referral is appropriate.
- Pharmacological treatment of other facial neuralgias is approached similarly as trigeminal neuralgia.

Neuralgic facial pain

Trigeminal neuralgia

Trigeminal neuralgia (TN) is perhaps the most well-known neuralgia. However, in the case of this nerve, the definition of classic TN (CTN) has come to be associated with a suspected cause reflecting a fundamental shift in our understanding of the disease and the definition of it. Our knowledge about the pathophysiology of CTN has spilled over to our understanding and treatment of other neuralgias as well.

TN is divided into three categories[1]

(i) CTN.
(ii) Secondary TN, resulting from neurological disease such as multiple sclerosis (MS), cerebellopontine angle tumours and arteriovenous malformations.
(iii) Idiopathic TN.

TN is defined as unilateral, recurrent paroxysms of facial pain in the distribution(s) of one or more divisions of the trigeminal nerve, with no radiation beyond.[1] The pain is brief (fractions of a second to 2 min), and severe, stabbing, shooting and lancinating or electrical in quality. It is precipitated by

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Table 1 Non-neuralgic aetiologies of orofacial pain.[1,2] List of differential diagnoses for orofacial pain adapted from ICHD-3 and from the ICOP classification criteria. These are from causes other than cranial neuralgias. This table may guide clinicians on the appropriate specialty referral needed for individuals with facial pain.

1. Orofacial pain associated with disorders of dentoalveolar and associated structures
Dental pain
Pulpal pain
Periodontal pain
Gingival pain
Non-dental pain
Oral mucosal pain
Salivary gland pain
Jaw bone pain
2. Orofacial pain associated with regional muscles
Primary myofascial pain
Secondary myofascial pain
3. Orofacial pain associated with disorders of the temporomandibular joint (TMJ)
Primary TMJ arthralgia
Secondary TMJ arthralgia
4. Orofacial pain associated with lesion/disorders
5. Orofacial pain resembling presentations of primary headaches
Orofacial migraine
Tension-type orofacial pain
Trigeminal autonomic orofacial
6. Idiopathic orofacial pain
Burning mouth syndrome (BMS)
Persistent idiopathic facial pain (PIFP)
Persistent idiopathic dentoalveolar pain

Table 2 Differential diagnoses for trigeminal neuralgia.[1] There are peripheral and central diagnoses with similarities to TN. The list of these is in this table, as adapted from ICHD-3.

Peripheral aetiologies
Trigeminal neuropathic pain
Glossopharyngeal neuralgia/glossopharyngeal neuropathic pain
Nervus intermedius neuralgia/painful nervus intermedius neuropathy
Painful optic neuritis
Persistent idiopathic facial pain
Burning mouth syndrome
Dental aetiologies (see Table 1)
Temporal mandibular joint arthralgia
Temporal mandibular joint dysfunction/myofascial pain
Central aetiologies
Short-lasting unilateral neuralgiform headache attacks with autonomic symptoms (SUNA), short-lasting unilateral neuralgiform headache attacks with conjunctival injection and tearing (SUNCT)
Paroxysmal hemicrania
Cluster headache
Primary stabbing headache
Tolosa–Hunt syndrome
Central pain attributed to multiple sclerosis
Central post-stroke syndrome

innocuous stimuli within the affected trigeminal distribution.[1] The most common distribution of the maxillary (V_2) or mandibular division (V_3) of the TN. There is often high variability in the number and duration of paroxysms. Stimulus dependence is a striking and key feature evoked by multiple triggers including light touch, chewing, talking, face washing, tooth brushing or even wind or cold air. Extreme pain can evoke contraction of the muscles of the face on the affected side ('tic douloureux'). Autonomic symptoms such as conjunctival tearing and redness, rhinorrhoea may arise. It is common to be asymptomatic between attacks, and there are frustratingly few predictors as to the onset or emergence from an attack.[1,2] The full differential diagnosis for TN is listed in Table 2.

Classical trigeminal neuralgia

Classical trigeminal neuralgia appears to have a unique pathophysiology. It is suspected to result from neurovascular compression (NVC) of the trigeminal nerve root at the root entry zone (REZ). In order to make the diagnosis, evidence of compression must be apparent on MRI.[1,2]

Although rare, classsical TN is the most common form. It is three times more common in women than men. The average age of onset is 50–60 yrs, and it more often located on the right side, and more commonly affects V_2, V_3 or both than the ophthalmic division of the TN (V_1).[3]

CTN may be diagnosed by history and examination. The key elements of the history include onset of pain, location of pain, paroxysmal duration of pain and triggers. Patients often have a memorable onset of pain. Although half of patients are asymptomatic between paroxysms, almost 50% of patients experience a lower intensity background pain in the same distribution. This background pain can be continuous or nearly continuous.[3] More than 90% of patients report triggers.[3] Patients report the intensity of their pain as quite severe, and they can have nighttime pain as well and autonomic symptoms, although not as prevalent.[3] Examination of the trigeminal territories is imperative, looking for allodynia, hyperalgesia, and hypoalgesia.[3] Most patients with CTN will *not* have motor weakness of the muscles of mastication. If motor deficits are present, then a secondary cause should be explored through prompt neuroimaging studies and other diagnostic tests.

Neurovascular compression, not just contact, is considered to cause CTN. The ICHD-3 criteria for CTN list demonstration of NVC, not contact, with morphological changes seen on MRI or during surgery.[1] The most common vasculature involved are the superior cerebellar artery, anterior inferior cerebellar artery, and the basilar artery.[3] The most common morphological change is nerve root atrophy, but displacement, distortion, dislocation, distension, indentation and flattening can also be seen on MRI or during surgery.[1]

MRI is the first-choice diagnostic procedure, providing a clear view of cranial nerves and cerebral vasculature. Other causes of TN may also be excluded. Sensitivity and specificity may range from 67% to 100% and from 50% to 100%, respectively, depending on the MRI protocols used.[4]

The usual MRI techniques and sequences are:

(i) Three-dimensional (3D) MRI for detailed imaging of the cisternal and cavernous segments of the nerve [3D constructive interference in steady state (3D CISS), or Fast Imaging Employing Steady-sTate Acquisition (FIESTA), driven equilibrium (DRIVE) sequences].
(ii) 3D MRA for good visualisation of arteries T2-weighted MRI to depict the nerve and nearby blood vessels [Fast

Low Angle Shot (FLASH), Fast Imaging with Steady-state Progression (FISP), or SPoiled Gradient Recalled Imaging (SPGR) sequences].

(iii) 3D T1-gadolinium for nerve visualisation and characterisation in relation to cerebrospinal fluid.[3,4]

Ideally, all three should be performed.[4]

If MRI is inconclusive or contraindicated, the trigeminal blink reflex test allows for side-to-side comparison of the V_1, V_2 and V_3 divisions. It is normal in CTN and abnormal in secondary pathologies. Testing of the trigeminal reflex includes electrical stimulation of the supraorbital nerve (V_1) to obtain the blink reflex, and stimulation of the infraorbital (V_2) and mental (V_3) nerves to produce the masseter inhibitory reflex. Laser-evoked potentials are not clinically useful to distinguish classical from secondary TN as they can be abnormal in both.[4]

Pharmacological therapy is the first-line treatment for CTN. The European Academy of Neurology (EAN) issued guidelines on TN in 2019. They rate the evidence for the use of carbamazepine in CTN as moderate and gave a strong recommendation for the use of long-term treatment in CTN. Although there is only low-quality evidence for oxcarbazepine, based on clinical experience, a strong recommendation was also given.[4] The sodium channel blockers carbamazepine and oxcarbazepine are considered first line for CTN. These work in a frequency-dependent manner leading to stabilisation of hyperexcited neural membranes and inhibition of repetitive firing. Either carbamazepine 200–1200 mg day^{-1} or oxcarbazepine 300–1800 mg day^{-1} may be used first for long-term treatment.[4]

Carbamazepine has a number-needed-to-treat (NNT) of 1.7–1.8 for TN. However, its numbers-needed-to-harm (NNHs) are 3.4 for minor and 24 for severe adverse events. Adverse effects include drowsiness, dizziness, rash, liver damage and ataxia and the potential for multiple drug interactions. These make medication titration challenging and are the usual cause of treatment discontinuation. Hyponatraemia and blood dyscrasias must be watched for with serial blood testing.[5]

Lamotrigine acts at the level of voltage-sensitive sodium channels, stabilises neural membranes and inhibits the release of excitatory neurotransmitters. There is evidence for using lamotrigine in combination with carbamazepine. The maximum dose is 400 mg day^{-1}. Slow titration is recommended because of the possibility of CNS adverse effects, a rash, or worse, Stevens–Johnson syndrome.[5] However, the EAN guidelines gave it a weak recommendation based on the low quality of evidence.[4]

Gabapentinoids reduce the release of excitatory neurotransmitter through modulation of voltage-gated calcium channels. Gabapentin is considered a second-line agent for CTN and evidence is of low quality. Dosage ranges from 300 to a maximum of 3600 mg day^{-1}.[4]

Other second-line agents include pregabalin, baclofen and levetiracetam. Pregabalin has not been tested in an RCT. An open-label study of 53 patients with TN notes a 50% reduction in pain in 74% of patients. Dosage ranged from 150 to 600 mg day^{-1}. An open-label cross-over trial of 22 patients with refractory TN comparing carbamazepine/pregabalin and carbamazepine/lamotrigine showed comparable efficacy and better patient tolerance in the pregabalin group.[6] Baclofen is a gamma aminobutyric acid B (GABA B) receptor agonist and depresses excitatory neurotransmission. It may have an analgesic effect in TN and can be used as add-on therapy.[4]

Levetiracetam acts by binding to the synaptic vesicle glycoprotein SV2 and modulating synaptic neurotransmitter release in the brain. There are two studies of small sample sizes and short treatment durations that report improvement in severity or frequency of painful paroxysms. The dosage ranged from 3 to 4 g day^{-1}.[7]

OnabotulinumtoxinA (BTX), a serotype of botulinum neurotoxin derived from *Clostridium botulinum*, is an option for those in whom treatment has failed or need an adjuvant to medical treatment is needed. Its analgesic effect has been investigated in a number of open-label studies and RCTs, and one systematic review. The pain relief may last several months, adverse effects potentially include temporary facial muscle weakness in the injected area and dysphagia.[8] The EAN guidelines gave a weak recommendation based on low-quality evidence.[4]

When first-line agents are not efficacious, and second-line agents either alone or as adjuvant therapy have not helped within 3–6 months, a surgical referral is appropriate. Surgical options for CTN can be categorised into four groups: posterior fossa intervention by microvascular decompression (MVD), lesion of the root by stereotactic radiosurgery (SRS), lesions at the ganglion level, and lesions distal to the ganglion. Long-term outcomes of all treatments have been studied, but the relapsing and remitting nature of this disorder make them difficult to evaluate.

MVD is the only surgery requiring hospital admission given the requirement for general anaesthesia with tracheal intubation, craniotomy, disentangling the looping artery by the nerves in the cerebellopontine angle and leaving a small sponge to separate the vessel from the trigeminal root. It is the most efficacious of surgical options.[4] Ninety percent of patients obtain pain relief, and more than 80% will still be pain-free at 1 yr, 75% at 3 yrs and 73% at 5 yrs.[9] Oesman and colleagues demonstrated that of their 156 CTN patient cohort 82% experienced good long-term outcome at a median follow-up period of 9.7 yrs.[10]

SRS applies targeted radiation to the trigeminal nerve root. Pain relief can take time; often 6–8 weeks before initial relief is seen. More than half of patients have complete pain relief 1 yr after SRS, but the probability of maintaining pain relief after 7–10 yrs is significantly lower. Long-term outcome studies with SRS (specifically gamma knife) for CTN show an initial pain free rate of 91.75% with actuarial rate of development of hypoaesthesia in 21.1%.[11] With repeated interventions, toxicity increases and facial hypoaesthesia or paraesthesia may persist in 50% of patients at 1 yr follow-up. The EAN guidelines were based on low-quality evidence, but using clinical experience, they issued a strong preference for MVD over SRS in patients with CTN who are able and willing to undergo posterior fossa surgery.[4]

Fluoroscopy-guided interventions for lesions of the trigeminal (or Gasserian) ganglion can be performed percutaneously, and the ganglion can be ablated with thermocoagulation by radiofrequency, or by injection of high-concentration glycerol to chemically ablate, or by mechanical compression by balloon inflation. There is a lack of robust clinical evidence for these procedures, although they are still considered options should medication or MVD prove ineffective or inappropriate.[4]

Thermocoagulation requires an awake patient initially to report where they feel paraesthesia evoked by electric impulses through the tip of the cannula. This prevents damaging the wrong trigeminal division. Importantly, the first division

should not be a target because of corneal deafferentation and keratitis. An awake patient also allows for slight adjustment until evoked paresthesia is concordant with the pain. The patient is then anaesthetised before actual thermocoagulation. Glycerol injection and balloon decompression can be performed under general anaesthesia. There is no risk of corneal keratitis with these two interventions. Risks include damage to the maxillary artery or dura mater, and a wet tap. Trigeminal sensory deficits are unavoidable and usually transient with glycerol injection and balloon compression.[4]

Procedures that have not been supported by adequate trials include neurectomy, alcohol injections, radiofrequency lesions or cryoablative lesions of the peripheral trigeminal nerves. Lesioning procedures may cause painful trigeminal neuropathy of traumatic origin or anaesthesia dolorosa, which is discussed in a separate section.

Overall, MVD has superior long-term outcomes compared with all the other interventional options such as SRS, thermocoagulation or neurolysis.[4]

Secondary TN

Patients with secondary TN are younger, and more likely to have bilateral pain and sensory deficits.[4] The most common cause of secondary TN is MS.[12] Space-occupying lesions at the cerebellopontine angle such as acoustic neuromas, epidermoid tumours and meningiomas are also responsible for secondary TN, as are arteriovenous malformations.[13] Demyelinating plaques are seen in the pontine trigeminal REZ, whereas brain tumours cause nerve compression in the same pontine region.[13]

On neurological examination patients with secondary TN have trigeminal nerve findings. Abnormal trigeminal reflexes, including the blink reflex, and evoked potentials are highly accurate in identifying MS-related TN.[12] The EAN guidelines note that patients with secondary TN respond less well to medications and procedures. However, Gasserian ganglion blocks can be considered, and MVD is an option if NVC is noted.[4]

Idiopathic TN

Should clinical symptoms fit the diagnosis of TN without NVC on imaging or secondary cause is found, then it is idiopathic. Pharmacological treatments are similar to those for CTN, and percutaneous interventions may also be considered.[1,4]

Painful trigeminal neuropathy or trigeminal neuropathic pain other than TN

Trigeminal neuropathic pain is distinct from TN. It is a continuous facial pain in the distribution of one or more branches of the trigeminal nerve because of another disorder causing neural damage.[1] The primary neuropathic pain is continuous or near-continuous, described as burning, squeezing, aching or like pins and needles. There may be pain paroxysms, but this is not a predominant symptom. There are clinically detectable sensory changes within the trigeminal nerve distribution, mechanical allodynia and cold hyperalgesia indicative of neuropathic pain.

There are three major groups of trigeminal neuropathic pain:

(i) Trigeminal neuropathic pain attributed to herpes zoster infection.
(ii) Trigeminal post-herpetic neuralgia (PHN).
(iii) Post-traumatic trigeminal neuropathic pain (previously known as anaesthesia dolorosa or painful post-traumatic trigeminal neuropathy).

Trigeminal neuropathic pain attributed to herpes zoster is an acute condition of <3 months' duration with clinical signs or symptoms of acute herpes zoster in the same trigeminal nerve division distribution. The annual incidence of herpes zoster infection from more than 60 studies in North America, Europe, Asia, South America and the Middle East is 3–5 per 1,000 person years, and 10–15% of all zoster infections involve the ophthalmic division of TN.[14]

Patients have notable cutaneous allodynia. Third, fourth and fifth cranial nerve palsies may be present. There are rare cases of pain not followed by a herpetic eruption ('zoster sine herpete') thus requiring confirmation of the disease through polymerase chain reaction (PCR) of varicella zoster virus DNA in the cerebrospinal fluid, or in cells from the base of lesions by PCR or direct immunofluorescence assay.[1]

Treatment with acyclovir, famciclovir or valacyclovir should be started within 72 h after onset of symptoms. A 7-day course is usual. To manage the pain, systemic analgesics may be supplemented with neuropathic agents such as amitriptyline, gabapentin or pregabalin.[15]

Beyond 3 months, about 25% of all acute herpes zoster infections involving the trigeminal nerve will develop into trigeminal PHN. This was previously termed post-herpetic trigeminal neuropathy. The incidence of trigeminal PHN is 3.3/100,000 per year (20/100,000 per year after age 60 yrs).[15] The pain is similar to an acute infection but may be accompanied by itching.

Reactivation of latent varicella zoster housed in the dorsal root ganglion of neural cells causes acute herpes zoster. The virus replicates and spreads down sensory nerves to skin. This process brings about cytopathic and ischaemic damage to nerve cells, sensory ganglions, blood vessels and mucocutaneous damage of epithelial cells. This acute process can lead to downregulation of central pain inhibitory pathways, alterations in gene expression of neuropeptides and activation of wide dynamic range neurons resulting in hyperexcitability of dorsal horn neurones and thus sensory functional alterations leading to allodynia, hyperalgesia, burning and electric shock-like sensations characteristic of PHN.[16]

The strongest evidence for effective pain control in PHN is for capsaicin 8% patch, gabapentin, gabapentin extended-release (ER), lidocaine plasters, opioids, pregabalin and tricyclic antidepressants. The capsaicin patch is not approved for use on the face, thus making it second line.[15,16] A multidisciplinary approach to PHN is most effective, which includes medications, and psychological and social support.[15]

Post-traumatic trigeminal neuropathic pain is pain caused by an identifiable traumatic event to the nerve. Previously termed anaesthesia dolorosa, it can stem from an external trauma, iatrogenic injuries from dental treatments or neuroablative procedures for TN; this pain may coexist with TN if the latter recurs.[17]

Neuropathies of traumatic origin initiate from peripheral nervous system alterations owing to inflammation. As peripheral nerves heal, disorganised sprouting ensues and may

lead to neuroma formation that can be an ectopic centre for neurophysiological activity. Over time, central changes are induced by hyperactivity of afferent fibers, resulting in central sensitisation.[18]

Patients have sensory loss in the injured region but can feel pain that is more severe than TN, with evident signs of trigeminal nerve dysfunction: hypoaesthesia/hypoalgesia or hyperalgesia/allodynia. Trigeminal reflex and laser-evoked potentials are abnormal on the affected side.[17]

With early intervention, central sensitisation and chronicity may be prevented. Early management includes controlling inflammation with steroids or NSAIDs.

If a larger nerve trunk is damaged, surgical repair may be considered and performed within the first 12–18 months. Treatment paradigms are similar to those previously discussed for PHN.[18]

Pain attributed to a lesion or disease of glossopharyngeal nerve

Glossopharyngeal neuralgia (GN) and glossopharyngeal neuropathic pain (GNP) are similar in presentation to CTN and trigeminal neuropathic pain, respectively.

Patients with GN present with unilateral, brief, stabbing pain in the ear, as well as the base of the tongue, tonsillar fossa, back of the throat, or beneath the angle of the jaw. It is triggered by chewing, swallowing, talking, yawning, drinking cold liquids or coughing. The onset is abrupt, and the condition may remit and relapse in the fashion of CTN. There may also be vagal symptoms of bradycardia, life-threatening syncopal episodes, hypotension or cardiac arrest considering the anatomy of the glossopharyngeal and vagus nerves.[1] Episodes of pain may occur multiple times throughout the day, lasting from seconds to 2 min, with pain freedom between attacks. Importantly, overall incidence is much lower than that of TN, estimated to be 0.2–0.7 cases per 100,000 individuals.[19] It accounts for 1.3% or less of the cranial neuralgias.[19] In the majority of patients, the cause of pain is an artery compressing the GN near the brainstem. This is termed classical GN. If an underlying disease can explain the neuralgia, it is a secondary type of GN. This can be attributable to neck trauma, MS, tonsillar or regional tumours, cerebellopontine angle tumours and Arnold–Chiari malformation. These aetiologies and iatrogenic injury can also lead to GNP.[1]

Diagnosis is primarily clinical and based on the patient's history and symptoms. Despite cranial nerve involvement, the neurological examination of patients with GN is normal. Similar to CTN, triggers are present and can include swallowing, chewing, talking, yelling and coughing.[19]

GNP differs from GN in that it has a continuous rather than paroxysmal pain, and the pain quality is described as burning, squeezing, pins and needles, with superimposed paroxysms. On examination there might be a weak or absent gag reflex, hearing loss and decreased or absent sensation to touch in the posterior ipsilateral tongue, tonsillar fossa and middle ear.[1]

For classical GN, first-line treatment is pharmacological and mirrors that of CTN. Refractory GN is amenable to surgery. The three major interventional treatments include direct nerve section (NS), MVD and SRS. A recent systematic review of 792 cases demonstrated that the short-term pain relief rate was highest after NS after surgery with an incidence rate (IR) of 94% [confidence interval (CI), 88–98%] and lowest after SRS at 3 months after operation (IR, 80%; CI, 68–96%). The rate of postoperative complication was highest in MVD (IR, 26%; CI, 16–38%) and lowest after SRS (IR, 0%; 95% CI, 0–4%). Long-term pain relief rate was highest after NS (IR, 96%; CI, 91–99%) and lowest after SRS (IR, 82%; CI 67–94%). The review supports the assessment that NS should be considered the most favourable treatment, in terms of short- and long-term pain relief and postoperative outcomes.[19]

Pain attributed to a lesion or disease of nervus intermedius

Nervus intermedius neuralgia

In 1907, Hunt described 'geniculate neuralgia', caused by the sensory affarents of the seventh cranial nerve located in the geniculate ganglion. Now known as nervus intermedius neuralgia (NIN), it is characterised a similar manner to the other previously described neuralgias in that it is unilateral, lancinating, paroxysmal and lasting seconds to minutes. Importantly, the pain is localised to the auditory canal and retroauricular regions, but can also spread to the temporal regions.[1] Triggers include sensory or mechanical stimuli at the posterior wall of the auditory canal. The pain is often accompanied by disorders of lacrimation, salivation and taste, as the nervus intermedius travels with the greater superior petrosal nerve and chorda tympani nerve.[1] It is rarer than TN or GN, and in a systematic review of literature from 1932 to 2012, fewer than 150 cases were noted.[20]

Similar to CTN, classical NIN is also suspected to be caused by NVC either by the anterior inferior cerebellar artery (AICA) or posterior inferior cerebellar artery (PICA), but this is yet to be definitively established. To be categorised as secondary, an underlying disease must be identified. If no compression or causative underlying disease is discovered, it is considered idiopathic.[1] NIN is difficult to detect on MRI because of its small diameter and proximity to the vestibular nerve. Fine-cut MRI of the cerebellopontine angle may identify a vascular loop compressing the nervus intermedius. This has limited specificity as asymptomatic individuals may have similar imaging findings.[20]

Given its location, pathology in the ear should be fully investigated. First-line therapy for NIN is pharmacological, medications similar to management of TN.[20] In patients who have exhausted conservative therapy, surgical treatments have been attempted in the past. Surgical resection of the nervus intermedius, geniculate ganglion, or chorda tympani may help to relieve symptoms. MVD may be an effective treatment.[20]

Painful nervus intermedius neuropathy

The painful neuropathy associated with nervus intermedius neuropathy was previously known as Ramsay Hunt syndrome and attributed to acute herpes zoster infection.[1] Similar to NIN, the pain is inside the auditory canal, auricle or region of the mastoid process. The pain is dull, deep in the ear and continuous or near-continuous. Brief paroxysms may be superimposed, but they do not predominate.[1] Additional cranial nerves (VIII, IX, X, XI) may also be affected because of viral spread leading to tinnitus, hearing loss, vertigo, nausea, hoarseness or dysphagia. Apart from herpes zoster, there are rare reports of facial tumours causing injuries to the

geniculate ganglion and thus painful nervus intermedius neuropathy.

The pathophysiology, diagnosis and treatment are similar to those of acute herpes zoster infection of the fifth cranial nerve, as described above under the section on trigeminal neuropathic pain. The diagnosis should be attributed to herpes zoster if the neuropathy is temporally related to a herpes zoster infection. Vesicles may be visible on the tympanic membrane, auditory canal, auricle and skin overlying the mastoid process. However, vesicles may be isolated to the tympanic membrane and difficult to see, and so the absence of lesions does not exclude it. If vesicles are seen, one should treat for herpes zoster infection. Treatment may also be considered if vesicles are not seen but are reported by the patient. The pain may continue for more than 3 months and evolve into PHN of nervus intermedius and should be treated similarly to herpetic and PHN. If no other secondary causes are found, the painful nervus intermedius neuropathy is idiopathic.[1]

Conclusions

Orofacial pain is challenging both for the breadth of diagnoses and rarity of particularly challenging entities. Owing to the difficulty in diagnosing these patients correctly, patients with a suspected diagnosis may benefit from a referral to a specialised centre. This may also help steer patients towards centres that participate in clinical trials, which may help improve our treatment algorithms.

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Declaration of interest

The authors declare that they have no conflicts of interest.

MCQs

The associated MCQs (to support CME/CPD activity) will be accessible at www.bjaed.org/cme/home by subscribers to *BJA Education*.

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