Orofacial Pain Syndromes
Evaluation and Management

Ramesh Balasubramaniam, BDSc, MSa,*, Gary D. Klasser, DMDb

INTRODUCTION

It is important to recognize and understand various orofacial pain syndromes, as the first health practitioner visited by patients is often the primary care physician. Therefore, having knowledge in the evaluation and management of orofacial pain syndromes is beneficial. Unfortunately, these conditions may be rather complex and this often leads to misdiagnosis and/or incomplete diagnosis, resulting in misdirected and/or incomplete treatment, despite well-intentioned efforts.

In this article, the evaluation and management of various neuropathic, neurovascular, and vascular pains are discussed.

NEUROPATHIC PAIN

Neuropathic pain (NP) is currently defined by the International Association for the Study of Pain (IASP)1 as “pain caused by a lesion or disease of the somatosensory nervous..."
In spite of this definition, clinically in many cases, there may be no demonstrable lesion or disease. Costigan and colleagues designate NP as “dysfunctional pain.” Dysfunctional pain is considered a malfunction (which can be considered a disease unto itself) of the somatosensory nervous system, involving both spontaneous and stimulus-dependent pain (evoked by both low-intensity and high-intensity stimuli), but without known structural nervous system lesions or active peripheral inflammation.

Classification of NP based on a temporal component may be divided into episodic and continuous. Episodic NP is characterized by sudden episodes of electriclike, severe, shooting pain that lasts only a few seconds to several minutes and is referred to as neuralgia. Often there exists a perioral and/or intraoral trigger zone whereby nontraumatic stimuli, such as light touch, provoke severe paroxysmal pain. Continuous NPs are pain disorders originating in neural structures and present as constant, ongoing, and unremitting pain. Patients usually experience varying and fluctuating intensities of pain, often without total remission.

Episodic

Trigeminal neuralgia

Trigeminal neuralgia (TN) is mainly a unilateral painful (moderate to severe) disorder characterized by brief, electric-shocklike pains, which are abrupt in onset and termination, and limited to the distribution of one or more divisions of the trigeminal nerve (usually the maxillary [V2] and mandibular [V3] divisions are affected). Often there are pain-free (refractory) periods between attacks. The International Classification of Headache Disorders-3 (ICHD-3) suggests 3 main variants (Table 1) and presents diagnostic criteria for each one, including its many subvariants. The prevalence of TN in the general population is between 0.01% and 0.3%. The gender ratio of women to men is approximately 2:1. Disease onset usually occurs after the age of 40 years with peak age of onset between the ages of 50 and 60 years.

Evaluation involves a thorough history and comprehensive examination (applicable for all neuropathic disorders), which includes a cranial nerve examination, mainly to rule out other possible causes for symptom presentation. Magnetic resonance imaging (MRI) of the brain and associated structures is the most useful imaging technique to determine the presence of other conditions that may mimic the symptoms of TN.

Medical management typically consists of the following:

- First-line therapy: carbamazepine (200–1200 mg/d) or oxcarbazepine (600–1800 mg/d).
- Second-line therapy: combination of first-line therapy with lamotrigine (400 mg/d) or a switch to lamotrigine or baclofen (40–80 mg/d).
- Third-line therapy: phenytoin, gabapentin, pregabalin, valproate, tizanidine, and tocainide.

If a medical approach is unsuccessful or results in marked deterioration in activities of daily living due to medication side effects, surgical procedures (not without serious adverse effects) should be considered.

- Peripheral surgical procedures: cryotherapy, neurectomy or alcohol injection, microvascular decompression (nondestructive surgical technique) of the nerve/ vessel contact or percutaneous ablative techniques (radiofrequency thermocoagulation, balloon compression, and glycerol rhizotomy) of the Gasserian ganglion.
- Stereotactic radiosurgery: gamma knife surgery (a focused beam of radiation aimed at the trigeminal root in the posterior fossa).
The American Academy of Neurology and the European Federation of Neurological Societies have published guidelines regarding TN management (medical and surgical).10

Other neuralgias
Glossopharyngeal neuralgia (GPN), known as vagoglossopharyngeal neuralgia, is typically a unilateral painful (mild to moderate) disorder characterized by brief, electric-shocklike pains, is abrupt in onset and termination, and is localized to the ear, the base of the tongue, posterior aspects of the throat (especially the tonsillar fossa), or beneath the angle of the jaw (distributions of the auricular and pharyngeal branches of the vagus nerve and branches of the glossopharyngeal nerve). Bilaterality of presentation may occur in up to 25% of patients.11 GPN is commonly provoked by swallowing, talking, coughing, and/or yawning; has similar characteristics as TN; and may coexist with TN in 10% to 12% of patients with GPN.12,13 Other common triggers of attack are sneezing, clearing the throat, touching the gums or oral mucosa, blowing the nose, or rubbing the ear.11 Cardiac dysrhythmias and syncope may occur due to stimulation of the vagus nerve. GPN incidence in the general population has been reported to be 0.2 per 100,000 persons per year.11,12 As observed in TN, a significant association between symptoms of GPN and multiple sclerosis has been reported.13 Computed tomography or MRI may reveal lesions, as well as neurovascular compression.

### Table 1
**Description of TN variants**

<table>
<thead>
<tr>
<th>Features</th>
<th>Classical TN</th>
<th>Classical TN with Concomitant Persistent Facial Pain</th>
<th>Symptomatic TN</th>
</tr>
</thead>
<tbody>
<tr>
<td>Previously used term</td>
<td>Tic douloureux</td>
<td>Atypical TN; TN type 2</td>
<td>—</td>
</tr>
<tr>
<td>Description</td>
<td>TN developing without apparent cause other than neurovascular compression (usually located at the trigeminal root entry to the brainstem and most frequently by the superior cerebellar artery)</td>
<td>TN with persistent background facial pain</td>
<td>TN attributed to a structural lesion other than vascular compression such as multiple sclerosis</td>
</tr>
<tr>
<td>Note</td>
<td>Imaging (preferably MRI) is recommended to exclude secondary causes and, in most patients, to demonstrate neurovascular compression of the trigeminal nerve</td>
<td>Neurovascular compression on MRI is less likely to be demonstrated. Responds poorly to conservative treatment and to neurosurgical interventions. Less likely to be triggered by innocuous stimuli.</td>
<td>—</td>
</tr>
</tbody>
</table>

**Abbreviations:** MRI, magnetic resonance imaging; TN, trigeminal neuralgia.


The American Academy of Neurology and the European Federation of Neurological Societies have published guidelines regarding TN management (medical and surgical).10

Orofacial Pain Syndromes
Management is similar to that of TN:

- First-line therapy: carbamazepine (200–1200 mg/d) or oxcarbazepine (600–
  1800 mg/d).\textsuperscript{14}
- Second-line therapy: local anesthetic to the tonsil and pharyngeal wall can pre-
  vent attacks for a few hours.\textsuperscript{12}

If medical treatment is unsuccessful, surgical procedures include microvascular
de compression, intracranial sectioning of the glossopharyngeal nerve and the upper
rootlets of the vagus nerve, or gamma-knife surgery. The major complications include
dysphagia, hoarseness, and facial paresis.\textsuperscript{4,15–17}

Occipital neuralgia (ON) is a unilateral or bilateral paroxysmal, shooting or stabbing
severe pain in the posterior part of the scalp, in the distribution of the greater, lesser, or
third occipital nerves. It is important to distinguish ON from occipital pain referred from
the atlantoaxial or upper zygapophyseal joints or from tender trigger points in neck
muscles or their insertions.\textsuperscript{18,18} Imaging studies are necessary to exclude underlying
pathologic conditions.

Management with injection of local anesthetics and corticosteroids may provide
temporary and even long-lasting pain relief.\textsuperscript{20}

Continuous

\textbf{Peripheral painful trigeminal traumatic neuropathy}

Peripheral painful trigeminal traumatic neuropathy (PPTTN), also known as anesthesia
dolorosa, is a unilateral (bilateral in 10\% of cases) facial or oral pain (moderate to se-
vere intensity and usually burning and/or stabbing) following trauma (within 3–6 months
of the event) to the trigeminal nerve.\textsuperscript{5,21} Often, there is clinical evidence of either pos-
itive (hyperalgesia, allodynia) and/or negative (hypoesthesia, hypoalgesia) signs of tri-
geminal nerve dysfunction.\textsuperscript{21} The traumatic event may be mechanical, chemical,
thermal, or caused by radiation.\textsuperscript{5} In orofacial pain clinics, reported onset most often
has a clear association with craniofacial or oral trauma,\textsuperscript{22,23} but pain may begin after
minor dental trauma, such as root canal therapy.\textsuperscript{24,25} The duration of the pain ranges
widely from paroxysmal to constant (most cases), and may be mixed.\textsuperscript{5,21} It has been
reported that after considerable injury to trigeminal nerve branches, chronic pain de-
vlops in approximately 3\% to 5\% of patients,\textsuperscript{22,26} as compared to approximately 5\% to
17\% in other body regions.\textsuperscript{27,28} Diagnostic testing may involve quantitative sensory
testing or advanced neurophysiological testing; however, this is not always possible to
do intraorally.\textsuperscript{21}

Management consists of antidepressants and anticonvulsants to modulate pain.\textsuperscript{21}
Microsurgical repair has shown to be effective in only 1 of 7 patients.\textsuperscript{29} Dorsal root en-
try zone lesioning (DREZ) has shown some promise,\textsuperscript{30,31} as has sensory thalamic neu-
rostimulation.\textsuperscript{32} It has been reported that most cases that have undergone peripheral
surgical procedures, such as exploratory procedures or apicoectomies, have resulted
in pain escalation.\textsuperscript{33}

\textbf{Persistent idiopathic facial pain}

Persistent idiopathic facial pain (PIFP), previously termed atypical facial pain, is persis-
tent facial and/or oral pain (mild to severe; superficial or deep), with varying presenta-
tions but recurring daily for more than 2 hours per day over more than 3 months, in the
absence of clinical neurologic deficit.\textsuperscript{5,34,35} However, sensory deficits have been re-
ported in up to 60\% of cases.\textsuperscript{36,37} PIPF has been characterized as being poorly local-
ized, not following the distribution of a peripheral nerve, while having a dull, aching, or
nagging quality (may have sharp exacerbations); is aggravated by stress; and, with
time, may spread to a wider area of the craniocervical region.\textsuperscript{5} PIPF may originate from a minor operation or injury to the orofacial region but persists after healing of the initial noxious event or presents without any demonstrable local cause.\textsuperscript{38} Patients experiencing PIPF often report multiple ineffective dental interventions in the area of complaint.\textsuperscript{39,40}

Before definitive diagnosis, all other local or systemic causes must be excluded. Imaging studies of the brain and skull base may be necessary in ruling out underlying pathology. Psychophysical or neurophysiological tests may demonstrate sensory abnormalities; however, this may not be clinically practical.\textsuperscript{41}

The Canadian Pain Society, the Neuropathic Pain Special Interest Group of the IASP, and the European Federation of Neurological Societies Task Force developed guidelines regarding the pharmacologic management of neuropathic pain.\textsuperscript{42–44} Medications commonly used for neuropathic pain management are tricyclic antidepressants (amitriptyline, desipramine, nortriptyline), anticonvulsants (gabapentin, pregabalin), serotonin noradrenaline reuptake inhibitors (venlafaxine, duloxetine, milnacipran), topical lidocaine, and analgesics (opioids, tramadol). Surgical procedures have been trialed; however, these techniques are not currently approved by the Food and Drug Administration (FDA) for chronic pain.\textsuperscript{45–48}

**Neuritis**

**Peripheral neuritis** Currently, peripheral neuritis is used to describe localized nerve pathologies secondary to inflammation. Inflammation may affect the nerve either by direct effects of mediator secretion, mainly cytokines or secondary to pressure induced by the accompanying edema.\textsuperscript{49,50} Temporary perineural inflammation in the orofacial region is most likely due to dental and other invasive procedures, but this is usually asymptomatic. However, misplaced dental implants or periapical inflammation close to a nerve trunk can produce chronic symptoms. Other conditions, such as temporomandibular joint pathologies,\textsuperscript{51} paranasal sinusitis,\textsuperscript{52} or early malignancies\textsuperscript{53} can induce symptomatic perineural inflammation, pain, and other aberrant sensations.

Early administration of anti-inflammatory medication (corticosteroids or nonsteroidal anti-inflammatory drugs) can be beneficial.\textsuperscript{54}

**Herpes zoster/postherpetic neuralgia** Acute herpes zoster (HZ) or shingles is a reactivation of latent varicella-zoster virus (VZV) infection that may occur decades after the primary infection. HZ is a disease of the dorsal root and cranial nerve ganglion and therefore induces a dermatomal vesicular eruption. HZ affects trigeminal nerves in approximately 10\% to 15\% of all cases. The ophthalmic branch is affected in more than 80\% of the trigeminal cases, particularly in elderly men, and may cause sight-threatening keratitis.\textsuperscript{5} The vesicles and pain are dermatomal and unilateral and will appear intraorally when the maxillary or mandibular branches of the trigeminal nerve are affected. The incidence of HZ is higher among people aged 60 to 70 (6–7 cases per 1000 person-years) and older than 80 years (>10 cases per 1000 person-years).\textsuperscript{55} HZ typically begins with prodromal symptoms, such as malaise, headache, photophobia, abnormal skin sensations, and occasionally fever. These symptoms may occur 1 to 5 days before the appearance of the rash.\textsuperscript{56} Diagnosis may be obtained by analyzing fluid from vesicles with the use of polymerase chain reaction testing, viral culture, or direct immunofluorescence antigen staining.\textsuperscript{57}

Management for HZ should be directed at controlling pain, accelerating healing, and reducing the risk of complications, such as dissemination, postherpetic neuralgia (PHN), and local secondary infection.\textsuperscript{58}
- Antivirals (acyclovir, valacyclovir, famciclovir) initiated less than 72 hours from onset of rash, particularly in patients older than 50 years, decrease rash duration, pain severity, and the incidence of PHN.59,60
- Nonopioid analgesics: acetaminophen or nonsteroidal anti-inflammatory drugs used to control fever and mild to moderate pain.
- Opioids: used for severe pain.
- Corticosteroids added to antivirals: decreases the pain of HZ; however, a systematic review found no significant difference between corticosteroids and placebo in preventing PHN 6 months after onset of the rash.61
- Adjunctive therapies: antidepressants (amitriptyline, desipramine, venlafaxine, bupropion) or gabapentinoids (gabapentin, pregabalin) provide analgesia, shorten illness duration, and reduce the risk of PHN.63,64

PHN, a neuropathic pain syndrome due to replication of the varicella-zoster virus in the basal ganglia causing nerve injury and manifesting as pain in the affected dermatome, is the most common complication of HZ. It occurs in approximately 30% of patients older than 80 years and in approximately 20% of patients 60 to 65 years old; it is rare in patients younger than 50 years.65 Postherpetic pain may include allodynia, hyperpathia, and dysesthesia.66 Women are at greater risk of PHN with additional risk factors, including older age, moderate to severe rash, moderate to severe acute pain during the rash, ophthalmic involvement, and history of prodromal pain.66,67 PHN may persist from 30 days to more than 6 months after the lesions have healed, and most cases resolve spontaneously.35

Management of established PHN should be immediate, as it improves prognosis, with ophthalmic PHN having the worst prognosis.68 Management options include antidepressants, gabapentinoids, opioids, and topical lidocaine patches.69 Invasive modalities include epidural and intrathecal steroids and a variety of neurosurgical techniques.70

HZ and PHN are relatively preventable conditions. The Shingles Prevention Study found the HZ vaccine to be 51.3% effective in preventing HZ and 66.5% effective in preventing PHN (when defined as pain rated at least a 3 of 10 on a severity scale that persisted for at least 90 days after rash onset).71 Vaccination also has been shown to reduce the incidence of PHN by 39% among patients who develop HZ.72 Currently, the Centers for Disease Control and Prevention recommends that patients aged 60 years and older should be vaccinated regardless of their prior exposure to VZV.72 However, the vaccine is approved for patients as young as 50 years of age as well.

**Burning mouth syndrome**

Burning mouth syndrome (BMS), also known as stomatodynia, glossodynia, oral dysesthesia, or stomatopyrosis, is a poorly understood pain condition that is most probably neuropathic. BMS is most common in postmenopausal women with reported prevalence rates in the general population varying from 0.7% to 15%,73 with individuals younger than 30 years rarely affected.75 For clinical utility, BMS may be classified into “primary BMS” or essential/idiopathic BMS for which a neuropathological cause is likely; and “secondary BMS” resulting from local or systemic pathologic conditions.76 By definition, primary BMS cannot be attributed to any systemic or local cause and therefore is essentially a diagnosis by exclusion (Table 2).77,78 The condition is characterized by a burning (mild to severe) sensation in the mucosa devoid of clinical findings and without abnormalities in laboratory testing or imaging, often accompanied by dysgeusia and xerostomia. Burning pain commonly presents with a bilateral symmetric distribution, most frequently involving the anterior two-thirds of the tongue,
the dorsum and lateral borders of the tongue, the anterior hard palate, and the mucosa of the lower lip, and often presenting in multiple sites.\textsuperscript{79,80} BMS is typically of spontaneous onset and lasts from months to several years,\textsuperscript{78,81} with spontaneous remission reported in 3% of patients approximately 5 years after onset.\textsuperscript{82} Minimal symptoms are reported by most patients on awakening, with gradual increase in intensity of symptoms as the day progresses, climaxing in the evening. Most patients report an intensification of the burning sensation while experiencing personal stressors and fatigue, with aggravation on eating acidic/hot/spicy foods. However, in about half the patients, oral intake/stimulation and distraction reduce or alleviate the symptoms.\textsuperscript{83} A possible association with anxiety, depression, and personality disorders is described in the literature, particularly in postmenopausal women,\textsuperscript{75,76} but it is unclear if pain initiated the psychological disorder or vice versa.\textsuperscript{84–86}

Management approaches include 3 strategies, which may be used singularly or in combination.

- Behavioral strategies: cognitive behavioral approaches and/or group psychotherapy.
- Topical medications: anxiolytics (clonazepam), atypical analgesics (capsaicin), antimicrobials (lysozyme-lactoperoxidase), artificial sweeteners (sucralose), and low-level laser therapy.
- Systemic medications: antidepressants (amitriptyline, trazodone, paroxetine, milnacipran), anxiolytics (clonazepam, diazepam, chlordiazepoxide), anticonvulsants (gabapentin, topiramate), antioxidants (alpha lipoic acid), atypical

| Table 2 |
|---|---|
| Factors needed to be ruled out before a diagnosis of burning mouth syndrome |
| Local | Systemic |
| Poorly fitting dental prostheses/mechanical irritations | Nutritional deficiencies (iron, B complex vitamins, zinc) |
| Dental anomalies | Endocrine disorders (diabetes, thyroid disorders, hormone deficiencies, menopause) |
| Parafunctional habits (clenching, bruxism, tongue posturing) | Anemia |
| Allergic contact stomatitis (dental restorations, denture materials, oral care products, foods, preservatives, additives, flavorings) | Gastrointestinal disorders (esophageal reflux) |
| Oral/perioral infections (bacteria, fungal, viral) | Medication (angiotensin-converting enzyme inhibitors, antihyperglycemics, chemotherapeutic agents) |
| Hyposalivation (radiation therapy, salivary gland disorders, medications) | Connective tissue/autoimmune diseases |
| Xerostomia | Neuropathy/neuralgia |
| Oral mucosal lesions (lichen planus, benign migratory glossitis) | |
| Tongue alterations (scalloped and/or fissured tongue) | |
| Chemical irritants | |
| Taste alteration/dysfunction | |
| Neurologic alterations | |
| Myofascial pain | |
analgesics/antipsychotics (capsaicin, olanzapine), histamine receptor antagonists (lafutidine, which is not FDA approved for use in the United States), monoamine oxidase inhibitors (moclobemide, which is not FDA approved for use in the United States), salivary stimulants (pilocarpine), dopamine agonists (pramipexole), and herbal supplements (hypericum perforatum or St. John’s wort).

Recently, a randomized controlled trial indicated that systemic use of clonazepam should be considered as first-line treatment.87

NEUROVASCULAR PAIN

Primary headache disorders are often associated with orofacial pain.88–91 These include migraine, tension-type headache, and trigeminal autonomic cephalalgias (TACs). The reader is encouraged to review the ICHD-3 for an extensive overview of all the headache disorders.5

Migraine

The 2 most common types of migraine are migraine without aura (80% of cases) and migraine with aura (20% of cases).92 Fifteen percent of migraineurs will report daily or chronic (near daily) headache.93 Migraine affects 6% of men and 18% of women in the adult population.94,95 Its prevalence peaks between the ages of 35 and 45.96 Migraine is associated with significant burden and decreased quality of life.95,97

Clinical features

Patients with migraine often report a trigger(s) for their migraine. Potential triggers include stress; altered sleep patterns; certain foods, such as cheese and chocolate; alcohol (wine); bright or flashing lights; menstruation; or changes in barometric pressure.98,99 The clinical presentation of migraine may occur in phases.

- Phase 1 (prodrome): occurs hours or days before the headache onset. Associated with cravings, lethargy, tiredness, stiff neck, and difficulty concentrating.100
- Phase 2 (aura): occurs in patients with migraine with aura. Auras may be visual, such as scotomas or fortification spectrum; sensory, such as numbness, and pins and needles; or motor, such as dysarthria.101 The aura develops within 5 to 20 minutes after the trigger and may last up to 60 minutes.
- Phase 3 (headache): headache is typically a unilateral pain localized around the ocular, temporal, and frontal regions. Occipital and neck area also may be involved.102–104 Patients report throbbing or pulsating pain at moderate to severe intensity.93,101,102,105 Aggravation of the pain with physical activities and sudden head movements is often reported.101,102 Duration of the headache is usually between 4 and 72 hours, although it can last longer.105 Most patients report fewer than 1 headache monthly, although some patients may suffer up to 4 migraines a month.94,105 Migraine also may be chronic, whereby the headache may occur more than 15 days per month.5 Many migraine sufferers report nausea (80%), vomiting (50%), and photophobia and/or phonophobia (>50%).102,103
- Phase 4 (postdrome): feeling of being washed out, irritable, depressed, and tired. Migraine is associated with anxiety, depression, allergies, stroke, and other pain conditions.106–109

Facial migraine

There are reports in the literature of patients who present with lower facial pain associated with nausea, photophobia, or phonophobia and autonomic symptoms consistent with migraine or TACs except it is a headache of the lower face.110–113 Benoliel
and colleagues\textsuperscript{114} proposed the term “neurovascular orofacial pain” along with criteria for its diagnosis (\textbf{Table 3}).

\textbf{Management}

Management for migraine is divided into 3 categories: behavioral interventions, preventive medications, and abortive medications. Behavioral changes may be adopted by the patient to prevent an attack. Patient education, information regarding sleep hygiene, diet, and stress may be used to prevent a migraine. In some cases, patients may have regular migraine and will need to use a preventive medication (\textbf{Table 4}). In the event of a migraine episode, an abortive medication may be used (\textbf{Table 5}).

\textbf{Tension-type Headache}

Tension-type headaches (TTH) are a common headache with a 1-year prevalence of greater than 80%.\textsuperscript{115,116} The age of onset is 20 to 30 years, with peak prevalence in the third and fifth decades.\textsuperscript{117} The ICHD-3 classifies TTH into infrequent (<12 episodes/year) or frequent (>12 and <180 headache days per year), episodic, chronic, or probable. The headache may be associated with or without pericranial tenderness.\textsuperscript{5} The clinical features of episodic and chronic TTH are summarized in \textbf{Table 6}.

Episodic TTH may be precipitated by stress, fatigue, lack of sleep, disturbed meals, menstruation, and alcohol.\textsuperscript{118,119} Patients with chronic TTH usually report a long

\begin{table}
\centering
\begin{tabular}{|l|l|}
\hline
\textbf{Table 3} & \textbf{Proposed criteria for neurovascular orofacial pain} \\
\hline
\textbf{Diagnostic Criteria} & \textbf{Notes} \\
\hline
A & At least 5 attacks of facial pain fulfilling criteria B–E \\
\hline
B & Severe, unilateral oral and/or perioral \\
& May refer to orbital and/or temporal regions. Side shift may occur; rarely are bilateral cases reported. \\
\hline
C & At least 1 of the following characteristics: \\
1. Toothache with no local pathology \\
2. Throbbing \\
3. Awakens from sleep \\
& Frequently painful vital teeth will be hypersensitive to cold stimuli. Some of the teeth in the painful region may have undergone root canal therapy with no long-lasting pain relief. \\
\hline
D & Episodic attacks lasting 60 min to >24 h \\
& Chronic unremitting cases that may result in subclassification into episodic and chronic forms have been observed. \\
\hline
E & Accompanied by at least 1 of the following: \\
1. Ipsilateral lacrimation and/or conjunctival injection \\
2. Ipsilateral rhinorrhea and/or nasal congestion \\
3. Ipsilateral cheek swelling \\
4. Photophobia and/or phonophobia \\
5. Nausea and/or vomiting \\
& Dental pathology may be very difficult to differentiate and needs careful assessment. \\
\hline
F & Not attributed to another disorder \\
\hline
\end{tabular}
\caption{Proposed criteria for neurovascular orofacial pain}
\end{table}

history of episodic headaches that evolved into the chronic form, which is associated with depression, anxiety, and lack of sleep.\textsuperscript{120} Management for TTH is summarized in Table 7.

\textbf{Trigeminal Autonomic Cephalalgias}

A collective group of headaches, trigeminal autonomic cephalalgias (TACs), are characterized by head and facial pain with accompanying autonomic features. The ICHD-3 classifies TACs as follows\textsuperscript{5}: cluster headache (CH), episodic or chronic; paroxysmal hemicranias,\textsuperscript{121} episodic or chronic; short-lasting unilateral neuralgiform headache attacks; and hemicrania continua (HC).

\textbf{Cluster headache}

CH affects 120 to 300 individuals per 100,000 in the general population.\textsuperscript{122–124} It is a severe unilateral, short-lasting pain overlying the orbital, supraorbital, or temporal sites accompanied by ipsilateral autonomic features, such as conjunctival injections,

\begin{table}[h]
\centering
\begin{tabular}{|c|c|}
\hline
\textbf{Clinical Phenotype} & \textbf{Strategy} \\
\hline
Mild – moderate pain & 1.a. Acetaminophen \& 1.b. NSAID \\
& Acetaminophen ± metoclopramide \\
& Ibuprofen, diclofenac potassium, naproxen sodium, ASA, all ± metoclopramide \\
& Moderate – severe pain/ \& NSAID failure & 2.a. NSAID with triptan \& rescue \\
& NSAID ± metoclopramide + a triptan \& later for rescue if necessary \\
\hline
\end{tabular}
\caption{Abortive medications for migraine}
\end{table}

\textbf{Table 5}

\textbf{Abortive medications for migraine}

<table>
<thead>
<tr>
<th>Clinical Phenotype</th>
<th>Strategy</th>
<th>Medications</th>
</tr>
</thead>
<tbody>
<tr>
<td>Mild – moderate pain</td>
<td>1.a. Acetaminophen &amp; 1.b. NSAID</td>
<td>Acetaminophen ± metoclopramide, Ibuprofen, diclofenac potassium, naproxen sodium, ASA, all ± metoclopramide</td>
</tr>
<tr>
<td>Moderate – severe pain/NSAID failure</td>
<td>2.a. NSAID with triptan rescue</td>
<td>NSAID ± metoclopramide + a triptan later for rescue if necessary</td>
</tr>
</tbody>
</table>

\textit{Abbreviations: ASA, acetylsalicylic acid; NSAID, nonsteroidal anti-inflammatory drugs.}  
lacrimation, nasal congestion, rhinorrhea, forehead and facial sweating, miosis, and ptosis. Occasionally, the pain may radiate to involve the maxilla, nostril, gingiva, palate, mandible, teeth, and neck. The pain is so severe that patients may be restless, pace, and bang their head. The autonomic features typically cease after an attack, which may last between 15 minutes and 3 hours. The features of CH are reviewed in Table 8. Chronic CH affects 10% of patients. An MRI of the brain and skull base is necessary to exclude central pathology.

Management for CH may include avoiding triggers, such as nitrates, alcohol, and smoking, and treating obstructive sleep apnea. There are also abortive and preventive interventions for CH (Table 9).

**Paroxysmal hemicrania**
PH affects 1 individual per 50,000 in the general population. It is a severe, short-lasting, strictly unilateral pain localized to the orbital, supraorbital, frontal, temporal, and occipital sites associated with ipsilateral autonomic features, such as lacrimation,
conjunctival injection, nasal congestion, rhinorrhea, ptosis, and facial flushing.\textsuperscript{5,138,139} It may confuse the clinician, as the pain can present in the facial structures.\textsuperscript{110,140,141} It is an excruciating pain of boring or stabbing quality and patients may be agitated, restless, or aggressive.\textsuperscript{5,138,139} The pain may occur spontaneously or be triggered by glyceryl trinitrate, alcohol, or manipulation of the head and neck.\textsuperscript{142,143} The clinical presentation of PH is summarized in Table 8. There is a chronic and episodic form of the pain with the latter having bouts of pain ranging from 2 weeks to 4.5 months with periods of remission between 1 and 36 months.\textsuperscript{5,139,142,144}

<table>
<thead>
<tr>
<th>Features</th>
<th>Cluster Headache</th>
<th>Paroxysmal Hemicrania</th>
<th>Short-Lasting Unilateral Neuralgiform Headache Attacks</th>
<th>Hemicrania Continua</th>
</tr>
</thead>
<tbody>
<tr>
<td>Sex, male:female</td>
<td>5:1</td>
<td>1:2</td>
<td>2:1</td>
<td>1:2</td>
</tr>
<tr>
<td>Age, y</td>
<td>20–40</td>
<td>30</td>
<td>40–70</td>
<td>30</td>
</tr>
<tr>
<td>Pain</td>
<td></td>
<td></td>
<td></td>
<td></td>
</tr>
<tr>
<td>Type</td>
<td>Boring</td>
<td>Boring</td>
<td>Electriclike</td>
<td>Throbbing or sharp</td>
</tr>
<tr>
<td>Severity</td>
<td>Very severe</td>
<td>Very severe</td>
<td>Severe</td>
<td>Varying severity</td>
</tr>
<tr>
<td>Location</td>
<td>Orbital</td>
<td>Orbital</td>
<td>Orbital</td>
<td>Temporal/ frontal</td>
</tr>
<tr>
<td>Duration</td>
<td>15–180 min</td>
<td>2–30 min</td>
<td>15–240 s</td>
<td>Continuous</td>
</tr>
<tr>
<td>Frequency</td>
<td>1–8/d</td>
<td>2–40/d</td>
<td>3–200/d</td>
<td>Constant</td>
</tr>
<tr>
<td>Autonomic</td>
<td>Yes</td>
<td>Yes</td>
<td>Yes</td>
<td>Yes</td>
</tr>
<tr>
<td>Trigger</td>
<td>Alcohol, nitrates</td>
<td>Mechanical</td>
<td>Cutaneous</td>
<td></td>
</tr>
</tbody>
</table>


Table 8
Clinical features of trigeminal autonomic cephalalgias

<table>
<thead>
<tr>
<th>Features</th>
<th>Cluster Headache</th>
<th>Paroxysmal Hemicrania</th>
<th>Short-Lasting Unilateral Neuralgiform Headache Attacks</th>
<th>Hemicrania Continua</th>
</tr>
</thead>
<tbody>
<tr>
<td>Sex, male:female</td>
<td>5:1</td>
<td>1:2</td>
<td>2:1</td>
<td>1:2</td>
</tr>
<tr>
<td>Age, y</td>
<td>20–40</td>
<td>30</td>
<td>40–70</td>
<td>30</td>
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<tr>
<td>Pain</td>
<td></td>
<td></td>
<td></td>
<td></td>
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<tr>
<td>Type</td>
<td>Boring</td>
<td>Boring</td>
<td>Electriclike</td>
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Management for PH is indomethacin and resolution of the pain within 24 hours is considered pathognomonic. The recommended dose for indomethacin is 75 to 150 mg 3 times daily.5,138,139,145

**Short-lasting unilateral neuralgiform headache attacks**
The prevalence of this extremely rare headache is 6.6 individuals per 100,000 in the general population.146 There are 2 forms of headache attacks: (1) short-lasting unilateral neuralgiform headache attacks with conjunctival injections and tearing (SUNCT), when no other autonomic features are apparent; (2) Short-lasting unilateral neuralgiform headache attacks with cranial autonomic symptoms (SUNA), when not limited to conjunctival injections and tearing.5 This is a moderate or severe unilateral head pain localized to the orbital, supraorbital, temporal, and frontal areas.147,148 The pain occurs at least once and up to 200 times a day. Common autonomic features are conjunctival injections and tearing and occur 1 to 2 seconds after the attack that last between 2 and 600 seconds.5,137,147 Attacks may occur spontaneously or with innocuous stimulation similar to TN. Unlike TN there are no refractory periods between attacks (see Table 8).147

First-line therapy for SUNCT is lamotrigine 100–300 mg per day or intravenous lidocaine 1.5 to 3.5 mg/kg per hour. Failing a therapeutic response, gabapentin 800 to 2700 mg per day or topiramate 50 to 300 mg per day may be used.145,149

**Hemicrania continua**
Hemicrania continua (HC) affects approximately 900 individuals per 100,000 in the general population.150 HC is a continuous unilateral headache localized to the temporal and frontal areas and, to a lesser extent, the orbital and retroorbital of varying intensity without side change.5,151–154 Cases of bilateral pain are rare.155 It is associated with mild autonomic features, namely lacrimation, conjunctival injection, nasal symptoms, and ptosis or miosis.155 Typically, the quality of pain is described as throbbing, stabbing, or sharp. Similar to PH, HC responds to indomethacin.5,156

**VASCULAR HEADACHE**

**Giant Cell Arteritis**
Giant cell arteritis, also known as temporal arteritis, is the result of granulomatous inflammation of the temporal artery, typically occurring in individuals older than 50 years. Patients may complain of a swollen, tender temporal artery, headache, hip and shoulder pain, fatigue, and malaise. Jaw claudication, aching, and cramping of the masseter and temporal muscles are common complaints and easily confused with temporomandibular disorders.157 A delayed diagnosis may result in blindness due to anterior ischemic optic neuropathy.158

Investigation typically reveals an elevated erythrocyte sedimentation rate of greater than 50 mm per hour. The diagnosis may be confirmed by temporal artery biopsy. Prompt treatment with corticosteroids is necessary to avoid blindness.159

**SUMMARY**
Patients will often visit their primary medical practitioner with orofacial pain complaints. Hence, it is important to recognize and have an understanding of these conditions to properly evaluate and potentially manage these disorders. If the practitioner is uncertain or uncomfortable with these conditions, then patient referral to a knowledgeable health care practitioner should be considered for further evaluation and management.
REFERENCES


