Overview of craniofacial pain

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INTRODUCTION

The diagnosis and management of patients with craniofacial pain can prove daunting even to experienced clinicians. The causes are myriad, and misdiagnosis and mismanagement are common.

This topic will provide a brief overview of craniofacial pain. Primary headache syndromes are discussed elsewhere. (See "Tension-type headache in adults: Etiology, clinical features, and diagnosis" and "Pathophysiology, clinical manifestations, and diagnosis of migraine in adults" and "Cluster headache: Epidemiology, clinical features, and diagnosis".)

NEURALGIAS AND PAINFUL CRANIAL NEUROPATHIES

Pain from neuralgia occurs in the distribution of a specific nerve or nerves that otherwise are normal in function, while neuropathy is defined as a disturbance of function or pathologic change in a nerve or nerves [1]. Neuropathic pain can be caused by a lesion of the central or peripheral somatosensory nervous system.

Head and neck pain is mediated by sensory fibers carried by the following nerves [1]:

- Trigeminal
- Nervus intermedius
- Glossopharyngeal
- Vagus
- Upper cervical spinal cord roots via the occipital nerves and great auricular nerve

Neuralgic pain has a paroxysmal quality, which is typically maximal at onset and is often described as lancinating, electrical shock–like, or jabbing. There may be a single sharp pain or repetitive pains in succession. The pain can last a fraction of a second or endure for several seconds. There may be a refractory period after the severe pain during which pain will not occur. Some neuralgic conditions have trigger zones (areas that when stimulated provoke an attack) or other triggers.

Careful history often reveals nonparoxysmal types of pain, such as continuous aching, burning, or throbbing pain. Inquiries must be made about all the sensations that occur, as some patients mention only the severe exacerbations. The response to treatment may provide a clue to diagnosis but can also prove misleading; "diagnostic blocks" in the setting of facial pain do not necessarily define the site from which the pain arises because of the overlap of cranial nerves V, VII, IX, X, which converge on the spinal trigeminal nucleus.

The mechanisms of neuropathic pain are complex. A nerve injury can induce peripheral and central changes that contribute to persistent pain and abnormal sensation. Through different mechanisms (inflammation, nociceptor activation, tissue injury), primary afferent fibers and central structures become sensitized. These processes are terminated under normal circumstances as tissues heal and inflammation declines. However, they can persist if primary afferent function is modified by disease, even in the absence of inflammation. While injury of primary afferent fibers results in peripheral neuropathic pain, direct damage to central nervous system structures results in central pain [2].

Trigeminal neuralgia — Trigeminal neuralgia is one of the most well-defined and common causes of facial pain. The pain of trigeminal neuralgia tends to occur in paroxysms and is maximal at or near onset. The pain has been described as electric shock–like or stabbing. Trigeminal neuralgia is discussed in detail separately. (See "Trigeminal neuralgia".)

Painful trigeminal neuropathy — Painful trigeminal neuropathy, also known as anesthesia dolorosa, is defined by head and/or facial pain in an area of reduced or absent sensation occurring in the distribution of one or more branches of the trigeminal nerve [1]. This condition is an example of how difficult central pain can be to describe since the patient has sensory loss but, at the same time, can feel pain. Potential causes include acute herpes zoster, postherpetic neuralgia (PHN), trauma, and others (eg, neural/perineural neoplasm). Painful trigeminal neuropathy can also be classified as idiopathic when clinical evaluation and thorough diagnostic testing identifies no underlying cause [1,3].

Postherpetic neuralgia — PHN is a type of painful cranial neuropathy associated with an acute herpes zoster infection. PHN is diagnosed as neuropathic pain that occurs in a distribution of a preceding herpes zoster attack and persists beyond three months after onset of the acute herpes zoster rash. PHN most often represents a continuum of pain that never resolved following an acute episode of herpes zoster. However, in rare cases, PHN can

occur months to years after resolution of the initial event. The epidemiology, pathogenesis, diagnosis, and treatment of PHN are discussed in detail separately. (See "Postherpetic neuralgia".)

Painful post-traumatic trigeminal neuropathy — Painful post-traumatic trigeminal neuropathy is characterized by unilateral facial or oral pain that occurs after traumatic injury of the trigeminal nerve, accompanied by additional symptoms or signs of trigeminal nerve dysfunction [1].

Diagnostic criteria for painful post-traumatic trigeminal neuropathy, according to the International Classification of Headache Disorders, 3rd edition (ICHD-3), require all of the following [1]:

- Facial and/or oral pain in the distribution of one or both trigeminal nerves
- History of an identifiable traumatic event to the trigeminal nerve, with clinically evident positive (hyperalgesia, allodynia) and/or negative (hypoesthesia, hypoalgesia) signs of trigeminal nerve dysfunction
- Evidence of causation demonstrated by both of the following:
  - Pain is located in the distribution(s) of the trigeminal nerve(s) affected by the traumatic event
  - Pain has developed within six months of the traumatic event
- Not better accounted for by another ICHD-3 diagnosis

The causative traumatic event may be mechanical, chemical, thermal, or radiation induced. Although sometimes related to surgical trauma, painful post-traumatic trigeminal neuropathy most frequently occurs as a complication of rhizotomy or thermocoagulation done to treat trigeminal neuralgia. In case series of patients treated for trigeminal neuralgia, painful post-traumatic trigeminal neuropathy has developed after glycerol rhizotomy in 0 to 1.6 percent of cases [4-8], after radiofrequency rhizotomy in 0.8 to 2 percent [9,10], and following percutaneous controlled thermocoagulation in 3 percent [11].

Painful post-traumatic trigeminal neuropathy can be more intolerable than the pain from trigeminal neuralgia itself [12]. This risk warrants careful decision making when considering surgical treatment for trigeminal neuralgia. (See "Trigeminal neuralgia", section on 'Surgery for medically refractory TN'.)

For treatment of non-neuralgic painful post-traumatic trigeminal neuropathy, we suggest tricyclic antidepressants (eg, amitriptyline) as initial pharmacologic therapy. When tricyclic medications are contraindicated or poorly tolerated, gabapentin or pregabalin are preferred

alternative choices. For superimposed neuralgiform pain, carbamazepine, oxcarbazepine, and baclofen are often used in clinical practice as in other cranial neuralgias.

Trigeminal trophic syndrome — The trigeminal trophic syndrome is characterized by facial skin ulceration and dysesthesia related to damage in the trigeminal nerve pathway affecting either peripheral components such as the trigeminal nerve (including the trigeminal ganglion and sensory root) or its central sensory nuclei [13-17]. The most frequent causes are therapeutic trigeminal nerve ablation and ischemic medullary or pontine stroke; others include craniofacial surgery, trauma, and herpes zoster infection [15]. Higher trigeminal pathway injuries affecting the trigemino-thalamic or thalamo-cortical tracts have not been reported to cause trigeminal trophic syndrome.

The most common symptoms of trigeminal trophic syndrome are bothersome dysesthesia, including itching, tickling, crawling, burning, as well as ocular foreign body and nasal airflow obstruction sensations [13-17]. Approximately one-half of affected individuals report facial pain. Examination reveals facial hypoesthesia or anesthesia. Skin ulceration tends to occur in the distribution of the infraorbital nerve, although it can occur in other trigeminal distributions [15]. The exact mechanisms for ulcer development have not been determined; self-manipulation with rubbing and scratching of the dysesthetic regions appears to be a contributing factor and can lead to erosion of the nasal ala and corneal lesions.

There is no established treatment for trigeminal trophic syndrome, though gabapentin and carbamazepine are often used to control dysesthetic symptoms [16,18,19]. Associated neuralgic pain may respond better to carbamazepine, oxcarbazepine, or baclofen. Measures to protect the injured area and promote healing may be beneficial; these include protective facial prosthetics and dressings, fingernail trimming, use of nocturnal scratch mittens, and behavioral modification.

Cluster-tic syndrome — The cluster-tic syndrome is a combination of cluster headache with coexistent trigeminal neuralgia [1,20-22]. The term "tic" refers to tic douloureux, another term for trigeminal neuralgia. It is characterized by three types of pain [20,21]:

- One component resembles the pain of trigeminal neuralgia and is paroxysmal, short lasting, and severe. (See "Trigeminal neuralgia".)
- The second component is characterized by trigeminal autonomic cephalalgia more like a cluster headache, although of variable length, with autonomic phenomena (eg, ptosis, miosis, lacrimation, conjunctival injection, rhinorrhea, nasal congestion). (See "Cluster headache: Epidemiology, clinical features, and diagnosis".)
- The third type of pain is a mixture of the first two and may be provoked by trigger points or moving the neck.

Cluster-tic syndrome usually affects patients between 20 and 70 years of age. It may exist in chronic or episodic (remissions and recurrences) forms. In one series, trigeminal neuralgia co-occurred with cluster headache in 4.5 percent of patients [23]. Some patients with this multifeatured headache syndrome have brief and recurring trigeminal autonomic cephalalgia symptoms more like short-lasting unilateral neuralgiform headache attacks than cluster attacks (table 1) [24,25].

Patients with cluster-tic syndrome require treatment directed at both cluster headache and trigeminal neuralgia in order to achieve remission of headaches [1,26]. Medical therapy usually consists of a combination of drugs used to treat trigeminal neuralgia and cluster headache and is often unsuccessful. Treatment of underlying causes such as demyelination or interventional and surgical treatment for structural contributors to symptoms may be warranted [27-29]. It is reported that if surgical microvascular decompression relieves the neuralgia, the cluster-like pain may be lessened and become more responsive to medical therapy [30].

Glossopharyngeal neuralgia — Glossopharyngeal neuralgia is an uncommon cranial neuralgia characterized by paroxysmal, severe, stabbing pain involving the ear, tonsillar fossa, base of the tongue, or beneath the angle of the jaw. It is typically triggered by activities that activate the glossopharyngeal nerve such as swallowing, coughing, speaking, yawning, certain tastes, or touching the neck or external auditory canal (rarely the pre- or postauricular areas). The epidemiology, pathogenesis, diagnosis, and treatment of glossopharyngeal neuralgia are discussed in detail separately. (See "Glossopharyngeal neuralgia".)

Nervus intermedius neuralgia — Nervus intermedius (sensory branch of facial nerve) neuralgia is a rare disorder characterized by brief paroxysms of pain felt deeply in the auditory canal. Other terms used are geniculate neuralgia or Hunt neuralgia. The epidemiology, pathogenesis, diagnosis, and treatment of nervus intermedius neuralgia are discussed in detail separately. (See "Nervus intermedius neuralgia".)

Auricular neuralgias — Other uncommon neuralgias that primarily affect the ear include great auricular neuralgia and auriculotemporal neuralgia.

- Great auricular neuralgia is an uncommon cause of neuralgic pain in the preauricular region. Pain can be triggered by head turning, neck touch, change of neck position during sleep, and jaw movement [31,32]. The great auricular nerve arises from C2 and C3 nerve roots, emerges from the midpoint of the sternocleidomastoid muscle, and ascends as it splits into preauricular and posterior branches where it provides innervation over the skin in the preauricular region, jaw angle, posteroinferior pinna and mastoid [31]. Symptoms may be idiopathic or secondary to a structural process (eg, lymphoma). It can be injured during procedures such as parotidectomy, submandibular

gland resection, rhytidectomy, cervical lymph node dissection, trauma, carotid endarterectomy, and pacemaker placement.

- Auriculotemporal neuralgia presents with neuralgic pain in the ear, temple, and preauricular regions [33,34]. The auriculotemporal nerve is a terminal branch of the mandibular (V3) nerve that encircles the middle meningeal artery and passes posteromedially to the mandibular condyle where it supplies sensation to the anterolateral aspect of the temporomandibular joint (TMJ) capsule, external acoustic meatus, anterior auricle, tragus, parotid gland, and the skin along the temple [35]. Entrapment, compression, inflammatory conditions, middle meningeal artery aneurysm, mandibular condyle fracture, perineural tumor spread, and TMJ synovial cysts have all been reported as possible causes [33,35].

Occipital neuralgia — Occipital neuralgia can be a cause of headache in the occipital region. It is described as a paroxysmal jabbing pain in the greater, lesser, and/or third occipital nerve distribution, infrequently accompanied by diminished sensation or dysesthesia in the affected area. Tenderness overlying the nerve affected may be present. The epidemiology, pathogenesis, diagnosis, and treatment of occipital neuralgia are discussed in detail separately. (See "Occipital neuralgia".)

OTHER CAUSES OF FACIAL PAIN

Optic neuritis — Headache due to optic neuritis (painful optic neuritis) is characterized by pain behind one or both eyes caused by optic nerve demyelination with associated central vision impairment [1]. Optic neuritis is discussed in detail elsewhere. (See "Optic neuritis: Pathophysiology, clinical features, and diagnosis".)

Headache attributed to ischemic ocular motor nerve palsy — Headache caused by ischemic injury of the ipsilateral cranial nerves III, IV, or VI (ie, attributed to ischemic palsy of ocular motor nerves) is described as a unilateral frontal and/or periorbital pain caused by and associated with other symptoms and signs of ischemic injury of the ocular nerves that control eye movements [1]. This is most prevalent with ischemic injury to cranial nerve III and may occur regardless of the presence or absence of underlying diabetes. (See "Third cranial nerve (oculomotor nerve) palsy in adults".)

Patients with transient and recurrent ocular motor palsies that are not clearly associated with ischemia should be evaluated for recurrent painful ophthalmoplegia neuropathy. (See 'Recurrent painful ophthalmoplegic neuropathy' below.)

Tolosa-Hunt syndrome — Headache due to Tolosa-Hunt syndrome is described as unilateral orbital pain associated with paresis of one or more of cranial nerves III, IV, or VI caused by a

granulomatous inflammation in the cavernous sinus, superior orbital fissure, or orbit. This condition is reviewed in detail separately. (See "Tolosa-Hunt syndrome".)

Paratrigeminal oculosympathetic syndrome — The paratrigeminal oculosympathetic syndrome (also known as Raeder syndrome) consists of constant unilateral burning facial pain with hypesthesia and/or dysesthesia in the distribution of the ophthalmic division of the trigeminal nerve, along with ptosis and miosis [36]. This syndrome may be caused by trauma, middle cranial fossa mass lesion, syphilis, and sinusitis. As an example, a restricted lesion in the middle cranial fossa can directly compromise trigeminal nerve fibers and cause neuralgic pain or sensory change with ptosis and/or miosis but no anhidrosis [37].

Diagnostic criteria for the paratrigeminal oculosympathetic syndrome, according to the International Classification of Headache Disorders, 3rd edition (ICHD-3), require all of the following [1]:

- Constant, unilateral headache
- Ipsilateral Horner syndrome, with imaging evidence of underlying disease of either the middle cranial fossa or of the ipsilateral carotid artery
- Evidence of causation demonstrated by both of the following:
  - Headache has developed in temporal relation to the onset of the underlying disorder or led to its discovery
  - Headache has either or both of the following features:
    - Localized to the distribution of the ophthalmic division of the trigeminal nerve, with or without spread to the maxillary division
    - Aggravated by eye movement
- Not better accounted for by another ICHD-3 diagnosis

Recurrent painful ophthalmoplegic neuropathy — Recurrent painful ophthalmoplegic neuropathy (RPON), previously called ophthalmoplegic migraine, is a rare condition most often seen in children and young adults. It is characterized by repeated attacks of paralysis of one or more ocular cranial nerves, typically cranial nerve III, after the onset of an ipsilateral headache [1]. Headache can develop up to two weeks before the onset of eye muscle weakness. In one review of 165 cases, cranial nerve III dysfunction was seen in 54 percent, cranial nerve VI in 37 percent, cranial nerve IV in 4 percent, and multiple cranial nerve involvement in 4 percent [38]. In unusual cases, it may present with isolated pupillary paralysis (internal ophthalmoplegia) due to oculomotor parasympathetic paresis [39].

The former term for this condition (ophthalmoplegic migraine) has been rejected because the syndrome is not migrainous [1]. Brain magnetic resonance imaging (MRI) reveals asymmetric thickening or gadolinium enhancement of the cisternal segment of the affected cranial nerve in approximately 30 percent of all cases [38]. However, when restricted to childhood-onset cases or acute presentations, these MRI findings may be found in more than 60 percent [38,40]. RPON may be due to recurrent episodes of demyelination [40,41], but the underlying causes and classification remain somewhat controversial [42-46]. In rare cases, oculomotor nerve tumors may mimic RPON; clues to the presence of tumor include incomplete recovery of ophthalmoplegia between attacks and the presence of a contrast-enhancing nodule of the oculomotor nerve on brain MRI [47-51].

Diagnostic criteria for RPON, according to the ICHD-3, require all of the following [1]:

- At least two attacks
- Both of the following:
  - Unilateral headache
  - Ipsilateral paresis of one, two or all three oculomotor nerves
- Orbital, parasellar, or posterior fossa lesion has been excluded by appropriate investigation
- Not better accounted for by another ICHD-3 diagnosis

Observational data suggest that treatment with glucocorticoids is beneficial for some patients [1,38,40]. In the absence of data to guide therapy, a short course of prednisone at 60 mg daily decreasing by 10 mg every three days is reasonable. Patients should follow up clinically and to assess efficacy as symptoms dictate. Symptoms typically last from a few days to up to two months, but recovery may be partial in some patients with multiple severe episodes [38].

Burning mouth syndrome — Burning mouth syndrome is characterized by an intraoral burning sensation for which no medical or dental cause can be found [1]. Pain may be restricted to the tongue or just the tip of the tongue and may be associated with dysesthesia, altered taste, and/or a sensation of having a dry mouth. This uncommon condition predominantly affects postmenopausal females [52,53], and 30 to 50 percent of patients improve spontaneously [54]. In a population-based epidemiologic study from Olmsted County, burning mouth syndrome was most commonly diagnosed after age 80 years [55]. An etiologic role for psychologic factors such as anxiety and depression has been suggested [56-58].

Although no definitive etiology has been established, one study suggested that trigeminal small-fiber sensory neuropathy is the cause of so-called idiopathic burning mouth syndrome [59]. Other studies identified a significantly higher number of unoccupied D2 dopamine receptors in the putamen associated with painful clinical conditions [60]. In this regard, a report described a patient with burning mouth syndrome whose pain responded to pramipexole, a nonergot dopamine agonist with a high selectivity for dopaminergic D2 receptors [61]. A subsequent case series reported six patients who failed other therapies but improved after treatment with pramipexole [62].

Diagnostic criteria for burning mouth syndrome, according to the ICHD-3, require all of the following [1]:

- Oral pain
- Recurring daily for more than two hours per day for greater than three months
- Pain has both of the following characteristics:
  - Burning quality
  - Felt superficially in the oral mucosa
- Oral mucosa is of normal appearance, and clinical examination including sensory testing is normal
- Not better accounted for by another ICHD-3 diagnosis

Prior to making the diagnosis, it is important to rule out oral mucosal diseases, such as herpes simplex and aphthous stomatitis. Other common conditions associated with mouth pain are psychiatric disorders, xerostomia (from drugs, connective tissue disease, or age), nutritional deficiencies (vitamin B12, iron, folate, zinc, vitamin B6), and allergic contact stomatitis. More unusual causes of mouth pain include geographic tongue, candidiasis, diabetes, denture-related pain, thyroid abnormalities, laryngopharyngeal reflux, and menopause [63-65]. Treating the underlying cause of mouth pain, if found, usually results in the remission of the symptoms [54]. When no underlying cause of symptoms is found, the condition is considered idiopathic burning mouth syndrome.

We suggest gabapentin or pregabalin as initial pharmacologic therapy for idiopathic burning mouth syndrome. Other alternatives or adjunctive treatments include amitriptyline, clonazepam, oral or topical capsaicin, and alpha-lipoic acid. Systematic reviews of treatment trials for burning mouth syndrome found several medications may be effective, including oral and topical clonazepam, gabapentin, pregabalin, oral or topical capsaicin, and alpha lipoic acid [66,67]. The quality of evidence to support the efficacy of alpha lipoic acid is low, but it may have a role as adjunctive treatment in combination with other agents [66].

Red ear syndrome — The red ear syndrome (RES) is an uncommon entity characterized by unilateral erythema and burning pain of the ear and adjacent face or hemicranium [68,69]. Less commonly, both ears may be involved. Symptoms may occur spontaneously or be triggered by touch, heat, neck movement, eating, or other stimuli. Episodes typically last less than one hour, and recurrence varies widely from monthly to multiple times each day. Symptoms occasionally occur as part of migraine or another primary headache syndrome. (See "Paroxysmal hemicrania: Clinical features and diagnosis", section on 'Atypical features'.)

The underlying mechanisms are uncertain, but RES has been associated with upper cervical spine disorders in some cases and migraine or trigeminal autonomic cephalgias in others [68]. RES may be a variant form of erythromelalgia, a clinical syndrome characterized by intermittently red and painful extremities [70,71]. (See "Pathophysiology, clinical manifestations, and diagnosis of migraine in adults", section on 'Pathophysiology' and "Pathophysiology of the trigeminal autonomic cephalalgias" and "Erythromelalgia".)

Treatment options derived mostly from case reports and series. Indomethacin, propranolol, amitriptyline, and gabapentin have been beneficial in some cases [70,72,73]. Local therapies such as ice packs and nerve blockade have also been used [72,74-76].

Persistent idiopathic facial pain — Persistent idiopathic facial pain (previously known as atypical facial pain) is characterized by persistent facial and/or oral pain in the absence of a neurologic deficit [1]. In a study of Dutch primary care patients, the incidence was 39.5 per 100,000 person-years [77]. Most cases are found in female patients [78,79]. In a German cohort of 150 patients, the mean age of onset was 43 years [79]. The symptoms may follow minor surgery or mild injury to the face, teeth, or gums and persist after healing without a clear local cause [1]. The pain is commonly felt in the nasolabial fold or one side of the chin but can spread to wider areas of the face and neck.

Diagnostic criteria for persistent idiopathic facial pain, according to the ICHD-3, require all of the following [1]:

- Facial and/or oral pain
- Recurring daily for more than two hours per day for more than three months
- Pain has both of the following characteristics:
  - Poorly localized and not following the distribution of a peripheral nerve
  - Dull, aching, or nagging quality
- Clinical neurologic examination is normal
- A dental cause has been excluded by appropriate investigations

- Not better accounted for by another ICHD-3 diagnosis

Persistent idiopathic facial pain is a diagnosis of exclusion; potential structural lesions, such as craniofacial neoplasms or abscesses, among others, need to be ruled out. Because it is essentially a face pain without a clear cause, persistent idiopathic facial pain is sometimes attributed to a psychogenic etiology [80]. However, this is not appropriate since there is enough reason to believe that persistent idiopathic facial pain is a central pain syndrome [54,81]. Although depression may be present, there is no evidence to support depression as the etiology [80].

For persistent idiopathic facial pain, tricyclic antidepressants (eg, amitriptyline) are the preferred treatment [82]. When tricyclic medications are contraindicated or poorly tolerated, gabapentin or pregabalin are preferred alternative choices. In one case report, topiramate (titrated to 125 mg two times a day) was beneficial [83].

Central neuropathic facial pain — By definition, central neuropathic facial pain is caused by a lesion or dysfunction in the central nervous system. The ICHD-3 recognizes two entities that are central causes of facial pain [1]:

- Central neuropathic pain attributed to multiple sclerosis
- Central poststroke pain

These are discussed in detail separately. (See "Central neuropathic facial pain".)

Secondary causes — Secondary causes of craniofacial pain include the following conditions:

- **Cancer pain** – Cancer is a rare cause of facial pain. Extracranial bony or soft tissue metastases may impinge upon cranial and upper cervical nerves causing headache or facial pain. In addition, occult lung neoplasms may cause referred pain in the periauricular region. Facial pain due to cancer is discussed in detail separately. (See "Overview of cancer pain syndromes".)
- **Dental pain** – Dental pathology is a common cause of facial pain [84]. Specific inquiry regarding prior dental procedures should be made of all patients with facial pain. The presence of provocative factors such as chewing or heat or cold sensitivity may provide useful clues. Trigeminal neuralgia also has been associated with ipsilateral dental pathology.
- **Temporomandibular joint syndrome** – Temporomandibular joint (TMJ) syndrome is characterized by chronic or acute musculoskeletal pain with dysfunction of the masticatory system. Many patients complain of headache or facial pain; in some cases, the disorder presents only as a headache without the patient being aware of a TMJ disturbance. The typical headache associated with TMJ syndrome presents as unilateral

ear or preauricular pain that radiates to the jaw, temple, or neck. The pain is deep, dull, continuous, and usually worse in the morning. (See "Temporomandibular disorders in adults".)

- **Giant cell arteritis** – Giant cell arteritis (GCA) is a chronic vasculitis of large and medium-sized vessels. Although systemic illness is characteristic of GCA and vascular involvement may be widespread, blood vessel inflammation most frequently involves the cranial branches of the arteries that originate from the aortic arch. The most feared complication of GCA, visual loss, is one potential result of the cranial arteritis associated with this disease. A new type of headache occurs in at least two-thirds of individuals affected with GCA. The head pain tends to be located over the temporal areas but can be frontal or occipital in location. The headaches may be mild or severe. The course is also variable; the headaches may become progressively worse but can subside before treatment is started in some cases. Nearly one-half of GCA patients suffer from jaw claudication. In some cases, a trismus-like symptom occurs rather than fatigue of the chewing muscles. Tender temporal or occipital arteries are found in approximately one-third of patients. (See "Clinical manifestations of giant cell arteritis".)

- **Carotidynia** – The ICHD-3 recognizes a subtype of headache attributed to noninflammatory lesions affecting the cervical carotid or vertebral arteries [1]. This category includes pain from arterial dissection, postendarterectomy headache, and headache attributed to carotid or vertebral angioplasty. Carotidynia is an older term that was used for pain that appears to emanate from the carotid artery [85]. Rather than a distinct entity, carotidynia is now considered a syndrome that encompasses many varieties of pain, including carotid dissection. (See "Cerebral and cervical artery dissection: Clinical features and diagnosis", section on 'Local symptoms'.)

- **Post-traumatic and postoperative pain** – Facial pain can occur after trauma, including bullet wounds or other head injuries and surgery. Facial pain may occur after maxillofacial surgery, orbital enucleations, sinus, and dental procedures. Dental procedures can trigger a variety of syndromes, including neuralgia and facial migraine. In all cases of post-traumatic facial pain, a careful search must be conducted for underlying pathology, especially dental problems. Some patients manifest constant burning pain, occasionally with tingling and stabbing but without the trophic changes, edema, and redness that are characteristic of reflex sympathetic dystrophy (complex regional pain syndrome) [86]. Treatment of post-traumatic facial pain can be extremely difficult. Blockade of the stellate ganglion may be effective in patients who complain of significant burning pain. Brief lancinating pains may respond to agents useful for neuralgias. Amitriptyline, for example, can reduce pain and lessen associated depression. Other agents useful for treating migraine headaches have been employed

empirically for symptomatic benefit. Post-traumatic facial pain generally resolves spontaneously within several years.

Primary headaches — Finally, primary headaches including migraine, cluster, and other headaches (such as paroxysmal hemicrania) can present mainly in the face. Careful attention to details of the history and examination should clarify the diagnosis. Questions revealing a family history, trigger factors, or the presence of an aura point toward migraine as the etiology. Similarly, cluster headache and other short-lasting headaches may present with pain principally in the face rather than periorbital or retro-orbital regions. Characterizing the duration of the episodes, along with any associated autonomic features (ptosis, rhinorrhea, lacrimation), is diagnostically helpful. (See "Pathophysiology, clinical manifestations, and diagnosis of migraine in adults" and "Cluster headache: Epidemiology, clinical features, and diagnosis" and "Paroxysmal hemicrania: Clinical features and diagnosis" and "Short-lasting unilateral neuralgiform headache attacks: Clinical features and diagnosis".)

SOCIETY GUIDELINE LINKS

Links to society and government-sponsored guidelines from selected countries and regions around the world are provided separately. (See "Society guideline links: Neuropathic pain".)

INFORMATION FOR PATIENTS

UpToDate offers two types of patient education materials, "The Basics" and "Beyond the Basics." The Basics patient education pieces are written in plain language, at the 5th to 6th grade reading level, and they answer the four or five key questions a patient might have about a given condition. These articles are best for patients who want a general overview and who prefer short, easy-to-read materials. Beyond the Basics patient education pieces are longer, more sophisticated, and more detailed. These articles are written at the 10th to 12th grade reading level and are best for patients who want in-depth information and are comfortable with some medical jargon.

Here are the patient education articles that are relevant to this topic. We encourage you to print or email these topics to your patients. (You can also locate patient education articles on a variety of subjects by searching on "patient info" and the keyword(s) of interest.)

- Basics topic (see "Patient education: Trigeminal neuralgia (The Basics)")

SUMMARY

- **Neuralgias and cranial neuropathies** – Neuralgia is defined as paroxysmal pain in the distribution of a specific nerve, which otherwise has normal function, while neuropathy is defined as a disturbance of function or pathologic change in a nerve or nerves. Neuralgias and painful cranial neuropathies that cause craniofacial pain include the following (see 'Neuralgias and painful cranial neuropathies' above):

  - **Trigeminal neuralgia** is characterized by recurrent paroxysms of pain in the distribution of one or more branches of cranial nerve V. (See 'Trigeminal neuralgia' above.)

  - **Painful trigeminal neuropathy** is defined by head and/or facial pain in the distribution of one or more branches of the trigeminal nerve caused by another disorder and indicative of neural damage. (See 'Painful trigeminal neuropathy' above.)

  - **Cluster-tic syndrome** is a combination of cluster headache with coexistent trigeminal neuralgia. It is characterized by the pain of trigeminal neuralgia and a cluster headache with autonomic phenomena. (See 'Cluster-tic syndrome' above.)

  - **Glossopharyngeal neuralgia** is defined as paroxysmal pain in areas innervated by cranial nerves IX. (See 'Glossopharyngeal neuralgia' above.)

  - **Nervus intermedius neuralgia** is a rare disorder characterized by brief paroxysms of pain felt in the auditory canal. (See 'Nervus intermedius neuralgia' above.)

  - **Auricular neuralgias** include great auricular neuralgia and auriculotemporal neuralgia. They are rare disorders that cause paroxysms of pain in the ear and adjacent structures. (See 'Auricular neuralgias' above.)

  - **Occipital neuralgia** is characterized by paroxysmal jabbing pain in the greater, lesser, and/or third occipital nerve distribution, sometimes accompanied by diminished sensation or dysesthesia in the affected area. (See 'Occipital neuralgia' above.)

- **Other causes of facial pain** – Other causes of facial pain include the following conditions:

  - **Optic neuritis** is characterized by pain behind the eye that is secondary to demyelination of the optic nerve with associated central vision impairment. (See 'Optic neuritis' above.)

  - **Headache attributed to ischemic ocular motor nerve palsy** is a unilateral frontal and/or periorbital pain caused by ischemic injury of the ipsilateral cranial nerves III,

IV, or VI that control eye movements. (See 'Headache attributed to ischemic ocular motor nerve palsy' above.)

- **Tolosa-Hunt syndrome** manifests as unilateral orbital pain associated with paresis of one or more of cranial nerves III, IV, or VI caused by a granulomatous inflammation in the cavernous sinus, superior orbital fissure, or orbit. (See 'Tolosa-Hunt syndrome' above.)

- **Paratrigeminal oculosympathetic syndrome** consists of constant unilateral burning facial pain with hypesthesia and/or dysesthesia in the distribution of the ophthalmic division of the trigeminal nerve, along with ptosis and miosis. (See 'Paratrigeminal oculosympathetic syndrome' above.)

- **Recurrent painful ophthalmoplegic neuropathy** is characterized by repeated attacks of paralysis of one or more ocular cranial nerves, typically cranial nerve III, with ipsilateral headache. (See 'Recurrent painful ophthalmoplegic neuropathy' above.)

- **Burning mouth syndrome** is characterized by an intraoral burning sensation for which no medical or dental cause can be found. The pain may be restricted to the tongue or just the tip of the tongue and may be associated with dysesthesia, altered taste, and/or a sensation of having a dry mouth. (See 'Burning mouth syndrome' above.)

- **Red ear syndrome** is an uncommon entity characterized by unilateral erythema and burning pain of the ear and adjacent face or hemicranium. Symptoms may occur spontaneously or be triggered by touch, heat, neck movement, eating, or other stimuli. (See 'Red ear syndrome' above.)

- **Persistent idiopathic facial pain** is characterized by persistent facial and/or oral pain in the absence of a neurologic deficit. Minor surgery or injury to the face, teeth, or gums may initiate the symptoms, which persist after healing without a clear local cause. The pain is commonly felt in the nasolabial fold or one side of the chin but can spread to wider areas of the face and neck. (See 'Persistent idiopathic facial pain' above.)

- **Central neuropathic pain** is caused by a lesion or dysfunction in the central nervous system. The two main recognized causes are central neuropathic pain attributed to multiple sclerosis and central poststroke pain. (See 'Central neuropathic facial pain' above and "Central neuropathic facial pain".)

- **Other causes of craniofacial pain** include cancer, dental pain, temporomandibular joint syndrome, giant cell arteritis, noninflammatory lesions affecting the cervical

carotid or vertebral arteries, post-traumatic and postoperative pain, and primary headaches. (See 'Secondary causes' above.)

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Topic 3355 Version 36.0

GRAPHICS

Clinical features and treatment of the trigeminal autonomic cephalalgias

	Cluster headache	Paroxysmal hemicrania	SUNCT* and SUNA¶	Hemicrania continua
Sex predominance	Male (4:1)	No (1:1)	Female (1.7:1)	Female (2:1)
Pain				
Type	Stabbing	Stabbing or throbbing	Stabbing or burning	Stabbing, throbbing, burning, or achin
Severity	Excruciating	Excruciating	Severe to excruciating	Mild to severe
Site	Orbital or temporal	Orbital or temporal	Orbital or temporal	Orbital, frontal, and/or temporal
Typical attack frequency	1 every other day to 8 daily	5 to 40 daily	1 to 200 daily	Continuous (with exacerbations)
Duration of attack	15 to 180 minutes	2 to 30 minutes	1 second to 10 minutes	Months to years (untreated)
Autonomic features?Δ	Yes	Yes	Yes (conjunctival injection and lacrimation prominent with SUNCT)	Yes
Restlessness and/or agitation?	Yes	Yes	Sometimes	Yes
Associated migrainous features?◇	Yes	Yes	Rare	Frequent
Triggers	Alcohol	Stress, exercise, alcohol	Tactile stimuli (eg, touching face, shaving, brushing teeth)	Alcohol
Indomethacin responsive?	No	Yes	No	Yes
Abortive treatment	Triptans (intravenous or nasal) Oxygen	None	Lidocaine (intravenous) for frequent and debilitating symptoms	None

Prophylactic treatment	Verapamil Glucocorticoids Galcanezumab Lithium Topiramate	Indomethacin Verapamil NSAIDs	Lamotrigine Oxcarbazepine Topiramate Gabapentin	Indomethacin

SUNCT: short-lasting unilateral neuralgiform headache attacks with conjunctival injection and tearing; SUNA: short-lasting unilateral neuralgiform headache attacks with cranial autonomic symptoms; NSAIDs: nonsteroidal anti-inflammatory drugs.

* Both conjunctival injection and lacrimation are present.

¶ May have either conjunctival injection or lacrimation but not both.

Δ Cranial autonomic symptoms: conjunctival injection, lacrimation, nasal congestion or rhinorrhea, facial sweating, miosis and/or ptosis, palpebral edema; symptoms are ipsilateral to the headache.

◇ Migraine-like features may include associated nausea, vomiting, photophobia, or phonophobia.

Graphic 65541 Version 14.0

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