

Chapter 4

Nomenclature and Differential Diagnosis



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Introduction

Facial pain is a debilitating pain condition which can affect at one point or another up to 26% of the general population [1]. As a matter of fact, it may be safe to assume that every living person has experienced facial pain at some point of her/his life due to dental issues, sinus problems, diseases, and injuries of the eyes, face, etc. In general, the facial pain may be a sign of various pathologies of facial structures, central nervous system (i.e., brain stem, thalamus, etc.) lesions, and cranial nerves or, in many of the cases, originate from an unknown source.

The general category of facial pain disorders includes diverse number of pathological conditions; these may be divided into neuropathic and nociceptive, odontogenic and non-odontogenic, neurogenic and non-neurogenic, etc. The differentiation and subcategorization vary among the disciplines and with different grouping strategies.

Neuropathic facial pain is a common term used for facial pain associated with nerve lesions or injuries [2]. The sensory innervation of the face is provided by both spinal nerves (C2–C4) and cranial nerves (V, VII, IX, X), and the trigeminal nerve is the source of the facial pain in the majority of the anatomically obvious neuralgias and neuropathies. Trigeminal neuralgia (TN) is worth particular attention as a distinct subtype of the facial pain as this potentially devastating condition can be successfully managed with multiple well-established interventions. While its diagnosis is quite straightforward, the exact terminology of its variants remains a common issue in publications.

According to a recent systematic review, the prevalence of trigeminal neuralgia (TN) ranges from 0.03 to 0.3% [3]. This tenfold discrepancy may be explained by a relatively small number of studies that fulfill the commonly accepted criteria of

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quality but also by heterogeneity of TN characteristics and subtypes [4–6]. The situation with confusing terminology gets even more complicated when it comes to more general facial pain epidemiology studies. There are only few meta-analyses in this field as multiple studies that use different classification strategies and criteria get discarded.

The general understanding of common facial pain terminology and differential diagnosis is the purpose of this chapter. Emphasis is placed on neuropathic facial pain.

Nomenclature

The terms used in this field are based on classifications and taxonomies proposed by study groups, author-based consensus, and individual expert opinions. In some classification systems, facial pain is grouped separately or together with the headache and orofacial pain. Five current major classifications that are used worldwide include facial pain disorders:

1. International Classification of Headache Disorders (ICHD), by the International Headache Society (IHS)
2. Classification of Chronic Pain, by the International Association for the Study of Pain (IASP)
3. International Classification of Diseases (ICD) coding, by the World Health Organization (WHO)
4. Classification from the American Association of Orofacial Pain (AAOP)
5. Burchiel classification

Among these, only IASP, ICHD, and Burchiel classifications contain descriptive information and diagnostic criteria for neuropathic facial pain [7–9].

Nonetheless, there are many publications do not follow the terms used in these classifications. The need for a terminological clearout for the confusing pool of interpretations is therefore quite evident [10]. The misinterpretations of pain conditions create a burden for decision-making, prescribing of treatments, evaluation of treatment efficacy, and planning of research and communication [11].

The attempt of Nixdorf et al. for orofacial pain taxonomy was promising. They have used persistent dentoalveolar pain disorder as an example to show how ontological principles can be used to improve related taxonomy [10]. However, the problem is not always the terms. Different stages of disease progression or overlapping conditions may have an impact on confusing nomenclature in literature [12, 13]. One example is the term “atypical odontalgia” which can also refer to phantom tooth pain, deafferentation pain, trigeminal neuropathy, or atypical facial pain [10, 13]. Although it was advised not to group these disorders for the aim of improving treatment selections, the confusing terms are still being used [14, 15].

Table 4.1 Comparative table of facial pain classifications

Facial pain classifications	Year	Extent of classification	Structure	Comments
The International Classification of Diseases (ICD) coding-ICD 10	2015	All diseases	Coding system Not diagnostic, not categorized Etiologically based	ICD-11 will be integrated with ICHD
International Headache Society (IHS) International Classification of Headache Disorders (ICHD-3 beta)	2013	Headache and facial pain	Diagnostic, theoretical, etiological, unidimensional Consensus of experts Criteria for each disorder	Validation problem for atypical pain Insufficient evidence
International Society for the Study of Pain (IASP)-Classification of Chronic Pain	1994	Pain disorders	Multiaxial, descriptive theoretical No criteria	Validation problem for atypical pain
American Association of Orofacial Pain (AAOP)	2005	Orofacial pain	Based on symptomatology Multiaxial No criteria	Two axes (physical and psycho); first axis: six subgroups

Despite the recent attempts to combine and integrate some of them, current terminology remains problematic and will be mentioned in context of relevant classifications under the section of differential diagnosis.

Diagnostic Classifications (Table 4.1)

The first consensus on the classification of headache disorders was pursued by an ad hoc committee formed by the US National Institutes of Health in 1962 [16]. In 1986, it was followed by IASP task force classification which was revised in 1994 and updated twice since then [17]. Recently, it was announced that ICD-11 will be integrated with new classification of IASP task force [18]. One out of seven main chronic pain topics, the “chronic headache and orofacial pain,” contains four subgroups, and chronic orofacial pain is among them:

1. Chronic primary headaches
2. Chronic secondary headaches
3. Chronic orofacial pains
4. Headache and orofacial pain not otherwise specified

ICHD is another general diagnostic classification that is used in most epidemiological studies; it was first published in 1988. Two revisions later, its latest version, so-called beta-edition, was published to collect evidence and provide field testing opportunities [19, 20] (Table 4.2). All physicians working in the related fields were

Table 4.2 ICHD-3 beta classification with brief descriptions

Painful cranial neuropathies and other facial pains
1. Trigeminal neuralgia
A. <i>Classical trigeminal neuralgia</i>
(a) Classical TN, purely paroxysmal
(b) Classical TN with concomitant persistent facial pain
B. Painful trigeminal neuropathy
(a) Painful trigeminal neuropathy attributed to acute herpes zoster
(b) Postherpetic trigeminal neuropathy
(c) Painful post-traumatic trigeminal neuropathy
(d) Painful trigeminal neuropathy attributed to multiple sclerosis (MS) plaque
(e) Painful trigeminal neuropathy attributed to other disorder
2. Glossopharyngeal neuralgia
3. Nervus intermedius (facial nerve) neuralgia
A. Classical nervus intermedius neuralgia
B. Nervus intermedius neuropathy attributed to herpes zoster
4. Occipital neuralgia
5. Optic neuritis
6. Headache attributed to ischemic ocular motor nerve palsy
7. Tolosa-Hunt syndrome
8. Paratrigeminal oculosympathetic (Raeder's) syndrome
9. Recurrent painful ophthalmoplegic neuropathy
10. Burning mouth syndrome (BMS)
11. Persistent idiopathic facial pain (PIFP)
12. Central neuropathic pain
A. Central neuropathic pain attributed to MS
B. Central poststroke pain (CPSP)

invited to submit evidence to the chair persons of the relevant chapter for the next ICHD revision [19, 20]. ICHD-3 is expected to be published in 2018.

Neuropathic pain is classified in most pertinent fashion by Burchiel in 2003 with partial subsequent revision in 2005 (Table 4.3) [8, 21]. Here, we use Burchiel classification as a diagnostic guide (Table 4.4).

Questionnaires

While clinically based differential diagnosis is achieved by the classifications' diagnostic criteria and physicians' clinical assessments, instrument-based differential diagnosis was tested by various authors as well (Table 4.5).

Table 4.3 Trigeminal neuralgia types in different classifications

Classification, year	# of sub-categories for TN	TN type							Other	
		TN1 Idiopathic sharp, shooting, electric shock-like episode pain	TN2 Idiopathic aching, throbbing, burning, >50% constant pain	TNP Unintentional trigeminal injury	Trigeminal deafferentation pain Intentional trigeminal injury	Resulting from herpes outbreak	ATN Somatoform pain disorder	Associated with MS		
Burchiel classification, 2005	7									
International Classification of Diseases (ICD), 2015	6	G50.0 TN: syndrome of paroxysmal facial pain	-	-	S04.3 Injury of trigeminal nerve	B02.22 Postherpetic TN	G50.1 Atypical facial pain	-	G.50.8 Other disorders of TN G50.9 Disorders of TN, unspecified	
IASP, 1994 (updated 2011, 2012)	5	TN	-	-	Secondary trigeminal neuralgia from facial trauma	Acute HZV (trigeminal) Postherpetic neuralgia	-	Secondary TN from CNS lesions		
ICHD, 2013	8	Classical TN	Classical TN with concomitant persistent facial pain (previously ATN/TN2)	-	Painful post-traumatic trigeminal neuropathy (previously anesthesia dolorosa)	Painful trigeminal neuropathy attributed to acute herpes zoster Postherpetic trigeminal neuropathy	Persistent idiopathic facial pain (PIFP)	-		

(continued)

Table 4.3 (continued)

Classification, year	# of sub-categories for TN	TN type					Other
		Classical TN (MR proved)	TN with continuous pain	-	-	-	
AAN-EFNS guidelines, 2016	4						Idiopathic TN when no etiology found Grading system added Secondary TN from lesions including MS

Table 4.4 Burchiel TN classification (2003)

1. Trigeminal neuralgia type 1 (TN1): Idiopathic, sharp shooting, electrical shock-like, episodic pain
2. Trigeminal neuralgia type 2 (TN2): Idiopathic, aching, throbbing, burning, >50% constant pain
3. Trigeminal neuropathic pain (TNP): It is caused by unintentional injury to the trigeminal system from trauma; oral surgery, root injury from posterior fossa, or cranial base surgery, stroke, etc. can be causes of this type of pain
4. Trigeminal deafferentation pain: The presence of numbness as a result of intentional injury from denervating pain procedures
5. Symptomatic trigeminal neuralgia: Associated with multiple sclerosis
6. Facial postherpetic neuralgia: Resulting from outbreak of facial herpes zoster
7. Atypical trigeminal neuralgia: Somatoform disorder

Table 4.5 Diagnostic questionnaires

Author/s	Year	Topic	Predictability rate (%)	# of groups, subgroups
Hapak et al.	1994	Craniofacial pain	74.3	Three groups and nine subgroups
Limonadi et al.	2006	Trigeminal neuralgia	95	Seven groups
Aggarwal et al.	2007	Orofacial pain	94	Three groups
McCartney et al.	2014	Facial pain	87.1	10
MacFarlane et al.	2004	Orofacial pain	71, 71, 57	Three groups; 23 subgroups

In 1975, Melzack first published a pain questionnaire that can measure pain and provide quantitative information at the same time [22]. It has been found that McGill's questionnaire may help differentiate between TN and atypical facial pain with 90% correct prediction [23].

In 1994, a self-administered questionnaire based on diagnostic classification was proposed by Hapak et al. [24]. Three categories were obtained: musculoligamentous, neurologic, and dentoalveolar. Neurologically based conditions included atypical facial pain, TN, migraine, cluster headache, and muscular contraction headache. The findings indicated that the sensitivity and specificity of this diagnostic questionnaire were 78.7%, 78.9%, and 37.5% and 81.5%, 78.2%, and 97% for the musculoligamentous, neurologic, and dentoalveolar groups, respectively. They used questionnaire and digital pain scales for undiagnosed patients and then classified them according to the most probable diagnosis.

In 2004, Macfarlane et al. presented a new classification questionnaire for orofacial pain and classified a total of 125 patients into three groups: musculoligamentous with 71%, dentoalveolar with 71%, and neurological/vascular with 57% good prediction [25].

Another questionnaire-based tool for classifying self-reported orofacial pain was developed and validated in population-based studies by Aggarwal et al. [26].

However, this tool grouped together the orofacial pain conditions that were likely to have an underlying pathology and those likely to be idiopathic. In a second study, these authors aimed to develop and validate a questionnaire-based tool that would enable classification of idiopathic orofacial pain in the general population [27]. They classified three categories, idiopathic, dentoalveolar, and muscoligamentous, based on distinct characteristics reported in a self-administered questionnaire. Ninety-four percent of the cases were successfully classified.

In 2006, Limonadi et al. presented the artificial neural network (ANN) which provided a tool for self-diagnosis based on a computerized questionnaire for TN patients [28]. Patients were classified according to Burchiel classifications. Predictability rates were higher than 95%. However, the study population was skewed due to a high percentage of typical TN patients. In 2014, an update was published by the same group [29]. They added four more questions and turned the questionnaire into a web-based diagnostic tool for ten different facial pain diagnoses. The sensitivity and specificity of new ANN were reported to be 92.4% and 87.8% for TN1, respectively, which was an improvement compared to the previous results of 84 and 83% of the earlier version. The ANN was still less sensitive at determining an accurate TN2 diagnosis (62.5% sensitive) but also better than the previous version. They mentioned that in forthcoming data sets, they are determined to improve temporomandibular joint disorders, nervus intermedius neuralgia, and glossopharyngeal neuralgia diagnoses as well.

Pain is a uniquely personal experience, and measuring it is highly perceptual. The controversy in the literature about the validation of the pain questionnaires is therefore almost inevitable.

Differential Diagnosis

Diagnostic criteria offered in classifications and/or extensive clinical experience draw the path to an accurate diagnosis. The first step is the clinical data gathering. History-taking stands as a gold standard method for facial pain diagnostic process. It should be integrated with the physical examination and imaging, with particular emphasis on the relevant differential diagnosis. The differential diagnosis of facial pain is detailed in (Table 4.2).

Neuralgia

The term neuralgia generally refers to a painful condition of a named nerve; it is considered a true example of painful mononeuropathy. When it comes to classification of neuralgias, the terminology varies, but in general, they are divided into primary and secondary, typical, or classic and atypical, central and peripheral,

idiopathic, or unknown, and these terms are used freely in multiple publications making nomenclature definition very complex.

Use of the term “secondary” is expected to be limited to those cases where the painful nerve is affected by a known distinct pathological process such as tumor, vascular malformation, trauma, infection, or demyelination. Such straightforward distinction becomes somewhat controversial when IHS classification suggests naming neuralgia “secondary” if a vascular compression is identified during surgical intervention and “primary” if surgical intervention never took place. In surgeon’s mind, however, the neuralgia that occurs in the absence of conditions listed above (tumor, etc.) is considered “primary” whether or not there is a documented vascular compression.

Trigeminal Neuralgia/Trigeminal Pain

Trigeminal neuralgia is a heterogeneous group of disorders presenting with neuro-pathic pain in one or more branches of the trigeminal nerve.

Pre-trigeminal Neuralgia

This term refers to a continuous dull pain in the upper or lower jaw that later develops into classic TN [30].

Idiopathic Trigeminal Neuralgia: Type I

(ICD: G50.0 trigeminal neuralgia, syndrome of paroxysmal facial pain, tic douloureux; ICHD, 13.1.1.1 classical TN, purely paroxysmal; IASP, 006.X8a trigeminal neuralgia, tic douloureux) (Table 4.6)

Table 4.6 ICHD-3 beta diagnostic criteria for classical TN

A. At least three attacks unilateral facial pain fulfilling criteria B and C
B. Occurring in one or more divisions of the trigeminal nerve, with no radiation beyond the trigeminal distribution
C. Pain has at least three of the following four characteristics: <ol style="list-style-type: none"> 1. Recurring in paroxysmal attacks lasting from a fraction of a second to 2 min 2. Severe intensity 3. Electric shock-like, shooting, stabbing, or sharp in quality 4. Precipitated by innocuous stimuli to the affected side
D. No clinically evident neurologic deficit
E. Not better accounted for by another ICHD-3 diagnosis

Idiopathic TN, previously known as classical or typical TN, is characterized by unilateral severe brief sharp pain attacks in one or more branches of the trigeminal nerve with spontaneous onset or triggered by non-painful stimuli. In between these electric shock-like pain attacks, there are refractory periods when triggering no longer produces pain. It may be bilateral in some cases.

It has been hypothesized that this condition may further advance to the type of TN formerly known as atypical, characterized by more than 50% constant pain [31]. The vascular compression of the trigeminal nerve root has been accepted as an etiopathogenetic factor for TN1 as the surgical decompression may cure this condition. However, multiple anatomical and radiological studies showed that vascular compression does not always result in TN symptoms [31–33]. As a matter of fact, up to 32% of people without pain have been found to have neurovascular compromise on high resolution imaging [31].

Idiopathic Trigeminal Neuralgia: Type II

(ICD: G50.0 syndrome of TN with concomitant persistent pain; IASP, 006.X8a trigeminal neuralgia, tic douloureux)

This type of facial pain is characterized by an aching, throbbing, or burning constant pain that is present at least 50% of the time. Sharp/episodic pain may accompany TN2 but is not required for diagnosis. Both ICHD-2 and ICHD-3 beta describe a new term of “TN with concomitant persistent pain” [34, 35]. In Burchiel classification, this condition is referred to as TN2 [8].

Postherpetic Neuralgia

(ICD, B02.22 postherpetic trigeminal neuralgia; ICHD, 13.1.2.2 postherpetic trigeminal neuropathy; and 13.1.2.1 painful trigeminal neuropathy attributed to acute herpes zoster; IASP, 003.X2b postherpetic neuralgia [trigeminal])

Pain in the trigeminal nerve territory that developed after a herpetic infection in the same area is considered postherpetic neuralgia. There are theories that pain in acute and late periods of viral infection with herpes zoster is caused by different mechanisms. The pain is burning and dysesthetic and may be associated with sensory loss and some degree of allodynia; trophic changes may also occur.

Trigeminal Deafferentation Pain

This condition is characterized by burning crawling itching tearing pain and is seen after intentional injury of the trigeminal system, usually as a result of neuroablative intervention (neurectomy, gangliolysis, rhizotomy, nucleotomy, tractotomy, etc.) Anesthesia dolorosa is an advanced form of this kind of pain; in addition to

neurodestructive procedures, it may also be seen in brainstem and mesencephalic infarctions.

Post-traumatic Trigeminal Neuropathy

(ICHD: 13.1.2.3 painful post-traumatic trigeminal neuropathy)

This type of neuralgia develops as a result of unintentional direct insult of to the trigeminal system, such as trauma, maxillofacial surgery, skull base surgical procedures, and posterior fossa surgery.

Constant, dull, throbbing, or burning pain with or without sharp pain paroxysms may be observed in the affected area. Nerve involvement in post-traumatic pain is distal to the trigeminal root and ganglion, and the character of pain differs from deafferentation pain and idiopathic types of TN.

Symptomatic Trigeminal Neuralgia

(ICHD: 13.1.2.4 painful trigeminal neuropathy attributed to multiple sclerosis (MS) plaque)

The term *symptomatic* is not present in the latest versions of HIS and IASP classifications. The neurosurgeons and neurologists continue using this term [8, 36, 37]. TN is seen in up to 10% of MS patients. The associated neuropathic pain can be either constant or episodic.

Secondary Trigeminal Neuralgia

(ICHD: 13.1.2.5–13.1.2.6 painful trigeminal neuropathy attributed to space-occupying lesion or other disorder)

Secondary TN is caused by other primary conditions that are summarized in Box 2. The underlying condition should be treated first to relieve this type of pain. According to the American Academy of Neurology—European Federation of Neurological Societies (AAN-EFNS) guidelines, “secondary” TN is caused by a major neurological disease such as tumor of the cerebellopontine angle or multiple sclerosis [38]; these guidelines group secondary and symptomatic TN into one category.

Geniculate Neuralgia

(ICD, G51.1 geniculate ganglionitis, disorder of geniculate ganglion; ICHD, 13.3 geniculate (facial nerve) neuralgia; IASP, 006.X2 geniculate neuralgia (VII cranial nerve), Ramsay Hunt syndrome)

Geniculate neuralgia presents with sharp and lancinating unilateral pain that is localized to an area behind the ear and/or the external auditory canal. It may be accompanied by other symptoms such as salivation, tinnitus, and bitter taste. ICHD-3 beta criteria require at least three attacks of brief paroxysms of pain felt deeply in the auditory canal, sometimes radiating to the parieto-occipital region. The term “geniculate neuralgia” is frequently used interchangeably with “neuralgia of nervus intermedius” (see below).

Glossopharyngeal Neuralgia

(ICD, G52.1 disorders of glossopharyngeal nerve, glossopharyngeal neuralgia; ICHD, 13.2 glossopharyngeal neuralgia; IASP, 006.X8b glossopharyngeal neuralgia)

Glossopharyngeal neuralgia is also called vagoglossopharyngeal neuralgia. It presents with sharp shooting pain in the posterior part of the tongue, pharynx, tonsils, and ear, sometimes with trigger zones located in the ipsilateral half of the tongue and throat. The pain can be provoked by chewing, swallowing, and talking. Vascular compromise can be seen on imaging and during surgery. ICHD criteria require the number of attacks to be three or more.

Sphenopalatine Neuralgia (Sluder)

Sluder neuralgia is a rare condition that presents with infraorbital or retro-orbital pain radiating toward the neck. It may be accompanied by lacrimation and conjunctival injection.

Superior Laryngeal Neuralgia

(ICD, G52.2 disorders of vagus nerve, superior laryngeal neuralgia; IASP, 006.X8e neuralgia of superior laryngeal nerve [vagus nerve neuralgia])

Superior laryngeal neuralgia presents with severe paroxysmal pain felt in the throat, in the mandibular region, or under the ear. Pain is triggered by swallowing, head turning, and straining the voice.

Paratrigeminal Neuralgia (Raeder)

(ICHD, 13.8 paratrigeminal oculosympathetic (Raeder’s) syndrome; IASP, 002.X4 [tumor]/002.X1a [trauma]/002.X3b [inflammatory, etc.]/002X8 [unknown] Raeder’s syndrome (Raeder’s paratrigeminal syndrome) Type I–Type II)

Paratrigeminal neuropathic pain presents in the frontotemporal region with associated partial Horner syndrome.

Occipital Neuralgia (Arnold)

(ICD, M53.82 other specified dorsopathies in cervical region, occipital neuralgia; ICHD, 13.4 occipital neuralgia; IASP: 004.X8–004.X1 [trauma] occipital neuralgia)

Occipital neuralgia is localized primarily in the occipital area and radiates to ear and retromandibular region.

Nervus Intermedius Neuralgia

(ICD, G52.9 disorders of other cranial nerves; ICHD, 13.3 nervus intermedius [facial nerve] neuralgia, [13.3.1 classical; 13.3.2 attributed to herpes zoster]; IASP, 006.X8c neuralgia of the nervus intermedius)

Nervus intermedius neuralgia presents with unilateral paroxysmal attacks deep in the ear. This term is used interchangeably with geniculate neuralgia, and this commonality is reflected in ICHD but not clear in other classifications.

Atypical Facial Pain

Frazier and Russell first used the term “atypical” in 1924 for facial pain that did not respond to surgical therapy [39]. Since that time terms “atypical facial pain” and “atypical trigeminal neuralgia” have often been used interchangeably even though these two terms describe very different conditions. In general, atypical facial pain refers to a poorly localized, vaguely described facial pain, nonanatomical in distribution, and with no evidence of a defined organic cause [3, 16]. Atypical trigeminal neuralgia, on the other hand, refers to condition that stems from trigeminal dysfunction but differs from “typical” TN by the presence of constant pain in addition to the classical electric shock-like attacks (typical TN is expected to be episodic only), by the presence of sensory deficits (typical TN patients have normal neurological examination), by the absence of trigger zones, by lack of response to anticonvulsants primarily carbamazepine (response to carbamazepine is considered pathognomonic for typical TN), etc. The pain in atypical TN, however, remains very anatomically defined and does not cross midline. To avoid this confusion, it is now recommended to stop using the term “atypical TN” and instead refer to it as TN type 2, or TN2, based on Burchiel classification.

According to one published concept, the main difference in symptomatology of “typical” TN and “atypical” TN is the severity and/or duration of the vascular compression of the trigeminal nerve root [12]. This concept is supported by clinical experience; it postulates that typical TN, atypical TN, and trigeminal neuropathic pain may not be separate conditions but just different degrees (or successive stages) of progressive injury of the trigeminal nerve. Well-known observations of vascular compression by nearby arterial or venous vessels in patients with atypical TN sup-

port this theory. As a further proof of this concept, Miller et al. in 2009 reported six patients with TN1 who subsequently progressed to TN2 [31]. They suggested that TN type at the onset is more meaningful than its ultimate presentation at the time of clinical evaluation.

With this, the term “atypical facial pain” (AFP) is currently reserved for patients with unequivocal evidence of a somatoform pain disorder that can be objectively diagnosed by psychological testing [21]. This term should not be used for those patients who are refractory to treatment or those “not-completely-typical” pain conditions that may be included in any other diagnostic category. Similarly, the other terms that were used to describe the atypical facial pain, such as dental causalgia, atypical facial neuralgia, and phantom orofacial pain, are no longer recommended for use [40].

Although both IASP and IHS excluded the term “atypical facial pain” from their lists and suggested the terms “other and unspecified pain in the jaws” or “facial pain not fulfilling other criteria,” this term remains widely used by various authors [41]. A recent survey of UK clinicians from all specialties who treat facial pain showed that 89% of them still use the term [41]. One hundred forty-three randomly selected specialists (oral and maxillofacial surgeons, oral medical experts, ear nose and throat surgeons, anesthetists, psychiatrists, and neurologists) completed questionnaires; 127 of them used the term atypical facial pain, and the rest used various other terms.

Persistent Idiopathic Facial Pain (PIFP)

In general, “idiopathic” refers to conditions with no identifiable cause. In case of trigeminal neuralgia, “idiopathic” is usually the same as “primary” TN, but in case of the persistent idiopathic facial pain (PIFP), idiopathic refers to the absence of identifiable organic disease and serves as substitute to the term “psychogenic” [11].

In ICHD PIFP, formerly known as atypical facial pain, describes chronic facial pain without evidence of structural or other specific causes of pain [9, 42]. Alternatively, PIFP is defined as a pain along the territory of the trigeminal nerve, which does not fit criteria for other cranial neuralgias [2, 43, 44]. ICHD suggests that “atypical odontalgia,” based on the history, can either be PIFP or a painful post-traumatic trigeminal neuropathy.

Headache Disorders

Trigeminal autonomic cephalgias (TACs) and migraine have been linked to the ophthalmic division of the trigeminal nerve. Based on a study of prevalence of facial pain in the migraine population, only 9% out of 517 migraine patients were found

to experience pain in the lower half of the face [45]. This was attributed to the anatomical overlap of the trigeminal and cervical afferents throughout the trigemino-cervical complex causing a referral of pain with otherwise typical clinical symptoms of a migraine attack. TACs are normally perceived in the upper part of the person's face but every now and then radiate to the face and teeth, and in turn, orofacial structures may give rise to headaches [46]. These conditions are characterized by short-lasting pain, some in the facial region and some in the head, and are accompanied by different autonomic features. As a matter of fact, there was a suggestion that short-lasting unilateral neuralgiform headache with cranial autonomic features (SUNA), short-lasting unilateral neuralgiform headaches occurring with conjunctival injection and tearing (SUNCT), TN may represent different stages of a single continuum [47].

In general, most causes of headache and craniofacial pain, including SUNCT, cluster-tic syndrome, paroxysmal hemicrania, and primary stabbing headache, should be considered in the differential diagnosis of TN.

Non-neurogenic Orofacial Pain and Temporomandibular Causes

Classic TN or secondary TN may sometimes be confused with dental causes of pain. Dental pain is usually continuous, intraoral pain that is dull or throbbing, whereas classic TN is typically intermittent and sharp pain. Furthermore, some patients describe a phase of “pre-trigeminal neuralgia” characterized by atypical (for TN) symptoms (e.g., jaw or tooth pain) that might mimic dental pain [30].

Non-neurogenic oral cavity diseases can present with stimulus-evoked, sharp, throbbing, or continuous pain; they may be easily confused with neurogenic pain, but oral examination and radiographs would help in establishing correct diagnosis.

Differential Diagnosis Algorithm

Our facial pain diagnosis and management algorithm was published in 2007 [48] (Table 4.7). Determination of anatomical distribution of pain and its correlation with representation of neurological structure(s) is the first step. Then, to differentiate secondary and primary causes, radiological evaluation—usually brain MRI with contrast—is performed. Pain nature and significant history information (previous surgery, infection, trauma, vascular formation, etc.) help to establish correct diagnosis and choose proper medical and surgical management.

If the pain does not follow anatomical distribution, a psychological evaluation may be in order to establish diagnosis of AFP/PIFP.

Table 4.7 Differential diagnosis of facial pain

Trigeminal neuralgia
Trigeminal neuralgia Type I: Idiopathic typical trigeminal neuralgia
Trigeminal neuralgia Type II: Idiopathic atypical trigeminal neuralgia
Symptomatic/secondary trigeminal neuralgia
Trigeminal neuropathic pain
Post-traumatic trigeminal pain
Trigeminal deafferentation pain
Anesthesia dolorosa
Central deafferentation syndrome
Postherpetic neuralgia
Glossopharyngeal neuralgia
Geniculate neuralgia
Sphenopalatine (Sluder) neuralgia
Paratrigeminal (Raeder) syndrome
Pain ophthalmoplegia (Tolosa-Hunt syndrome)
Petrous apex syndrome (Gradenigo syndrome)
Cancer-related pain
Atypical facial pain
Non-neurogenic orofacial pain and temporomandibular joint-related pain
Headache and other conditions

Table 4.8 Comparison of IASP and IHS classification

IASP	IHS
Central pain (if confined to the head and face)	12.7.2 Thalamic pain
Trigeminal neuralgia (tic douloureux)	12.2.1 Trigeminal neuralgia
Secondary neuralgia (trigeminal) from central nervous system lesions (tumor or aneurysm)	12.2.2.2 Symptomatic trigeminal neuralgia
Acute herpes zoster (trigeminal)	12.1.4.1 Herpes zoster
Postherpetic neuralgia (trigeminal)	12.1.4.2 Chronic postherpetic neuralgia
Geniculate neuralgia (seventh cranial nerve): Ramsay Hunt syndrome	12.1.4.1 Herpes zoster
Glossopharyngeal neuralgia (ninth cranial nerve)	12.3.1 Idiopathic glossopharyngeal neuralgia 12.3.2 Symptomatic glossopharyngeal neuralgia
Neuralgia of the superior laryngeal nerve (vagus nerve neuralgia)	12.5 Superior laryngeal neuralgia
Occipital neuralgia	12.6 Occipital neuralgia
Hypoglossal neuralgia	12.1.7 Other causes of persistent pain of cranial nerve origin
Glossopharyngeal pain from trauma	12.3.2 Symptomatic glossopharyngeal neuralgia
Hypoglossal pain from trauma	12.1.7 Other causes of persistent pain of cranial nerve origin
Tolosa-Hunt syndrome (painful ophthalmoplegia)	12.1.5 Tolosa-Hunt syndrome

Conclusion

Since the time of Hippocrates (circa 400 BC), there are some challenges related to diagnosis and treatment of facial pain [49]. Both surgical and nonsurgical approaches are used in facial pain patients; the choice of approach is generally guided by practical treatment algorithms.

Most challenge remains with so-called atypical pain patients, as their diagnosis and management require advanced multidisciplinary expertise. Meanwhile, the lack of universally accepted classification makes therapeutic decisions more difficult [27]. This is further complicated by a fact that somewhere between 7 and 44% of cases may be unclassifiable in view multiple diagnostic classifications [20] (Table 4.8). Even when symptoms point toward involvement of a specific nerve(s), there is a general lack of imaging tools that may help in establishing clinical diagnosis. The key to a proper diagnostic approach is to evaluate the symptoms as a whole and present the patient with treatment options in a goal-oriented manner. Since the duration of chronic pain may affect the treatment results, a timely and accurate diagnosis remains a cornerstone of efficient management.

The use of a grading system such as “definite,” “probable,” or “possible” has been suggested for use in case of diagnosing neuropathic pain. It has been suggested that this type of grading may be extended to various orofacial pain diagnoses as a means of managing the uncertainty in providing diagnoses for conditions that have varied clinical presentations [37].

Although the head and face are closely related, diagnostic classifications of headache and orofacial pain are not properly integrated. It does appear, however, that chronic orofacial pain and headaches can be classified together as they may be sharing similar underlying pathophysiology, clinical characteristics, and neurovascular issues. To test this concept, the headache definitions have been applied in a cohort of chronic orofacial pain patients. The researchers concluded that both headaches and OFP should own their own subclassifications.

In general, the use of comprehensive classification systems does not guarantee better outcomes. But discrepancy in terminology and existence of conflicting classification systems may delay diagnosis, negatively affect interprofessional communication, or result in inaccurate labeling. In addition to this, use of different classifications may make meta-analyses difficult if not impossible.

Summary

Chronic facial pain often requires multidisciplinary and multi-interventional therapy. Pain medicine, neurology, neurosurgery, otolaryngology, ophthalmology, dentistry, and maxillofacial surgery are those disciplines that deal with diagnosis and treatment of facial pain.

Major classification guidelines lack the common language for diagnosis. It is therefore not surprising that most studies and publications include unclassified, overlapping, and mixed (combined) cases making it problematic for further analy-

sis. The approach to gather evidence as used in the last version of ICHD and publish a beta version for field testing is a promising step in creating a unified and comprehensive classification. Inclusion of orofacial pain groups in this collaboration is expected to strengthen the future versions of classifications.

The unfortunate part of classifications that are not validated or supported by evidence/validation is that they not only result in the wrong selection of patients for treatment but also facilitate collection of uncategorizable data that cannot be properly analyzed, making it all but impossible to create the evidence-based approach that is desperately needed for management of this complicated group of patients.

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