A New Classification for Facial Pain

Kim J. Burchiel, M.D.

Department of Neurological Surgery, Oregon Health and Science University, Portland, Oregon

Reprint requests:
Kim J. Burchiel, M.D., Department of Neurological Surgery, Mail Code L-472, Oregon Health and Science University, 3181 SW Sam Jackson Park Road, Portland, OR 97239.
Email: burchiek@ohsu.edu

Received, January 25, 2003.
Accepted, June 24, 2003.

PURPOSE: A patient-oriented classification scheme for facial pains commonly encountered in neurosurgical practice is proposed.

CONCEPT: This classification is driven principally by the patient's history.

RATIONALE: The scheme incorporates descriptions for so-called "atypical" trigeminal neuralgias and facial pains but minimizes the pejorative, accepting that the physiology of neuropathic pains could reasonably encompass a variety of pain sensations, both episodic and constant. Seven diagnostic labels result: trigeminal neuralgia Types 1 and 2 refer to patients with the spontaneous onset of facial pain and either predominant episodic or constant pain, respectively. Trigeminal neuropathic pain results from unintentional injury to the trigeminal nerve from trauma or surgery, whereas trigeminal deafferentation pain results from injury to the nerve by peripheral nerve ablation, gangliolysis, or rhizotomy in an intentional attempt to treat either trigeminal neuralgia or other facial pain. Postherpetic neuralgia follows a cutaneous herpes zoster outbreak (shingles) in the trigeminal distribution, and symptomatic trigeminal neuralgia results from multiple sclerosis. The final category, atypical facial pain, is synonymous with facial pain secondary to a somatoform pain disorder. Atypical facial pain can be suspected but not diagnosed by history and can be diagnosed only with detailed and objective psychological testing.

CONCLUSION: This diagnostic classification would allow more rigorous and objective natural history and outcome studies of facial pain in the future.

KEY WORDS: Classification, Facial pain, Trigeminal neuralgia

Neurosurgery 53:1164-1167, 2003 DOI: 10.1227/01.NEU.0000088806.11659.D8 www.neurosurgery-online.com

The surgeon, however, is chiefly concerned with the question: "What cases of neuralgia are suited for operative treatment, and what are the best methods to employ?" The answer, obviously, should depend upon a scientific classification, based solely upon the causes of neuralgia; at present such a classification is impossible.
—J. Hutchinson (1905) (10)

The facial pain syndrome known as trigeminal neuralgia was described more than 300 years ago (13). It is remarkable that despite the extensive literature, no natural history study has ever been published on the subject. Current outcome studies on the medical and surgical treatment of the disorder suffer from a lack of understanding of the course of the condition, ascertainment bias, "expert" opinion, and a perfusion of terminology describing the various subtypes of pain. I would submit that this confusion has substantively inhibited the study of trigeminal neuralgia and related facial pains. Hutchinson (10) recognized this almost a century ago, and relatively little has changed in the intervening years.

WHAT WE KNOW

Our current understanding holds that trigeminal neuralgia is a unique form of neuropathic pain (2). Most clinical neuroscientists recognize that the cause of the neuropathy is, in most cases, neurovascular compression (5, 7, 8, 11). We know that both central and peripheral demyelination, root injury, or both are important in the pathophysiology of trigeminal neuralgia (6, 9). Root entry zone pathology may be critical to the development of the typical syndrome (11). Ectopic action potential generation in the sensory root (portio major) of the nerve may either be directly responsible for or, at a minimum, "ignite" the typical episodic, electric, lancinating pains (4).

The spontaneous onset of constant aching, throbbing, or burning pain in the trigeminal distribution, with or without concurrent lancinating pains, has traditionally been viewed with suspicion by neurologists and neurosurgeons. Terms such as *atypical trigeminal neuralgia* and *atypical facial pain* (AFP) abound in the literature to describe these variants. However, the precise boundaries of, for example, "typical"

and "atypical" trigeminal neuralgia have never been reliably or reproducibly established. Likewise, the term *AFP* is a catch-all phrase, intended to warn the unwary of an origin of pain and potential outcome that is, at worst, murky and unpredictable and, at best, less favorable than in textbook cases of trigeminal neuralgia. In this system of diagnosis, there is a danger that patients with otherwise treatable facial pain will be discarded on the heap of the atypical.

As Hutchinson (10) so eloquently stated, a classification system of facial pain should be based on an understanding of the underlying pathophysiology of the neuralgia. Ironically, our current concept of the pathophysiology of trigeminal neuralgia may have been artificially constrained by our clinical classification of the disorder. We have been brought up on the idea that the clinical syndrome of trigeminal neuralgia is somewhat monolithic and that deviations of the classic syndrome are evidence of a different problem, not a variation on a theme. As noted above, clinical and experimental data indicate that hyperactivity in large myelinated fibers of the sensory root may be the fundamental trigger of the more typical pains. However, if the same type of hyperactivity occurred in the small myelinated axons (A-δ fibers) or unmyelinated axons (C fibers) of the main sensory root (portio major) or those in the motor root (portio minor) that are known to exist (15), the pains of trigeminal neuralgia could incorporate a variety of sensory experiences. Small myelinated axons mediate "fast" pain and could, and probably would, mediate a pricking or electrical sensation. Unmyelinated axons mediate "slow" pain and heat. Sensations described as aching, throbbing, and burning would be more likely from hyperactivity in these fiber types (1, 3, 12, 14, 16). Neurovascular compression of both the portio major at sites other than the root entry zone and of the portio minor clearly occurs (8). Whether or not neurovascular compression or injury to either the trigeminal nerve root at loci distal to the root entry zone or within the motor root (portio minor) produces more atypical pains has not been examined systematically in a surgically verified case series.

As opposed to the idiopathic nature of trigeminal neuralgia, an antecedent history of injury to the trigeminal system identifies a discrete subset of patients with facial pain. Although all of these patients could be classified as having a form of neuropathic pain and the pathophysiology of these patients may be, in fact, quite similar, this group of patients can be divided into two categories: trigeminal neuropathic pain would be the descriptor for pain resulting from unintentional injury to the trigeminal system from facial trauma; oral surgery; ear, nose, and throat surgery; root injury from posterior fossa or cranial base surgery; stroke; etc., and trigeminal deafferentation pain would be pain in a region of trigeminal numbness resulting from intentional injury to the trigeminal system from neurectomy, gangliolysis, rhizotomy, nucleotomy, tractotomy, or other denervating procedures.

Two other conditions are worthy of special consideration and are readily identified by history: 1) symptomatic trigeminal neuralgia resulting from multiple sclerosis, and 2) facial postherpetic neuralgia. Here again, both entities are fundamentally forms of neuropathic pain, but their natural histories differ substantially.

The last condition to discuss is a complaint of facial pain in a patient with a somatoform pain disorder. Experienced clinicians know that this is a distinct but relatively uncommon group of patients with facial pain. In the past, the term *AFP* has been used to describe any facial pain patient who had mostly constant facial pain, whatever the pathogenesis. Obviously, this categorization is inadequate, because it would encompass both patients with neuropathic pain and those with psychogenic pain. I would propose limiting the term *AFP* to only those patients with unequivocal evidence of a somatoform pain disorder that can be objectively diagnosed by psychological testing. This category is perhaps the most difficult for patients with facial pain and some clinicians to accept. However, to not acknowledge that some patients do suffer from predominantly psychogenic pain flies in the face of clinical experience. An unwillingness to recognize these patients also does them a great disservice, because the lack of appropriate referral for psychological support and counseling prolongs their suffering, and referral for inappropriate invasive procedures can make them worse. The category of AFP cannot be diagnosed strictly by history. It requires that patients undergo psychological evaluation, including standardized testing. Not all patients with facial pain need to have this testing, but certainly those with simultaneous bilateral facial pain, pain spreading well outside the trigeminal distribution, multiple pain complaints in other body regions, and diagnostic clustering with conditions such as fibromyalgia and chronic fatigue syndrome should be considered for psychological assessment.

CONCLUSION

The field of pain medicine is need of better means of communication and standardization of pain syndromes. Facial pain presents a unique opportunity, because the patient's history is the most powerful means of establishing a diagnosis. Furthermore, much of the facial pain seen in a neurologist's or neurosurgeon's practice is trigeminal neuralgia or one of its variants. In general, trigeminal neuralgia is a condition that is relatively easy to diagnose. Diagnosis of other conditions, such as trigeminal neuropathic pain, trigeminal deafferentation pain, symptomatic trigeminal neuralgia, and facial postherpetic neuralgia, should also be straightforward. AFP can be suspected but not diagnosed by history. Certainly, psychological testing can follow up suspicion of this condition. An appropriate level of suspicion for AFP does no harm, but underdiagnosis could adversely affect the patient and the treating clinician.

Although textbooks are full of other more exotic facial pain diagnoses, these seven diagnoses describe virtually all patients presenting to a clinical neuroscience practice with a primary complaint of facial pain (*Table 1*). By use of this simple and reductive process, patients can be accurately diagnosed and considered for therapies appropriate to their conditions.

TABLE 1. Classification scheme for facial pains commonly encountered in neurosurgical practice

Diagnosis	History
	Spontaneous onset
Trigeminal neuralgia, Type 1	*>50% episodic pain*
Trigeminal neuralgia, Type 2	*>50% constant pain*
	Trigeminal injury
Trigeminal neuropathic pain	*Unintentional, incidental trauma*
Trigeminal deafferentation pain	*Intentional deafferentation*
Symptomatic trigeminal neuralgia	Multiple sclerosis
Postherpetic neuralgia	Trigeminal *Herpes zoster* outbreak
Atypical facial pain[a]	Somatoform pain disorder

[a] Cannot be diagnosed by history alone.

A more objective classification will also facilitate both natural history studies and outcome studies of facial pain. It is my hope that this scheme will be used by others involved in the care of patients with facial pain or, at the very least, will serve as a catalyst for further discussion and better classification of these disorders.

REFERENCES

1. Boring E (ed): *Sensation and Perception in the History of Experimental Psychology*. New York, Irvington Publishers, 1942.
2. Burchiel KJ, Slavin KV: On the natural history of trigeminal neuralgia. **Neurosurgery** 46:152–155, 2000.
3. Collins WF, Nulsen FE, Randt CT: Relation of peripheral nerve fiber size and sensation in man. **Arch Neurol** 3:381–385, 1960.
4. Devor M, Govrin-Lippmann R, Rappaport ZH: Mechanism of trigeminal neuralgia: An ultrastructural analysis of trigeminal root specimens obtained during microvascular decompression surgery. **J Neurosurg** 96:532–543, 2002.
5. Fromm GH, Sessle BJ (eds): *Trigeminal Neuralgia: Current Concepts Regarding Pathogenesis and Treatment*. Boston, Heinemann Medical Books, 1990.
6. Golby AJ, Norbash A, Silverberg GD: Trigeminal neuralgia resulting from infarction of the root entry zone of the trigeminal nerve: Case report. **Neurosurgery** 43:620–623, 1998.
7. Haines SJ, Jannetta PJ, Zorub DS: Microvascular relations of the trigeminal nerve: An anatomical study with clinical correlation. **J Neurosurg** 52:381–386, 1980.
8. Hamlyn PJ, King TT: Neurovascular compression in trigeminal neuralgia: A clinical and anatomical study. **J Neurosurg** 76:948–954, 1992.
9. Hilton DA, Love S, Gradidge T, Coakham HB: Pathological findings associated with trigeminal neuralgia caused by vascular compression. **Neurosurgery** 35:299–303, 1994.
10. Hutchinson J: *The Surgical Treatment of Facial Neuralgia*. New York, Wm. Wood & Co., 1905, p 1.
11. Jannetta PJ: Arterial compression of the trigeminal nerve at the pons in patients with trigeminal neuralgia. **J Neurosurg** 26[Suppl]:159–162, 1967.
12. Lewis T, Pochin EE: The double pain response of the human skin to a single stimulus. **Clin Sci** 3:67–76, 1937.
13. Lewy FH: The first authentic case of major trigeminal neuralgia and some comments on the history of this disease. **Ann Med Hist** 10:247–250, 1938.
14. Torebjork HE, Hallin RG: Perceptual changes accompanying controlled, preferential blocking of A and C fibre responses in intact human skin nerves. **Exp Brain Res** 16:321–332, 1973.
15. Young RF, Stevens R: Unmyelinated axons in the trigeminal motor root of humans and cat. **J Comp Neurol** 205–214, 1979.
16. Zotterman Y: Studies in the peripheral nerve mechanisms of pain. **Acta Med Scand** 80:185–242, 1933.

COMMENTS

Burchiel has presented a new classification of facial pain. The rationale is that the current classification lumps together too many disparate groups of patients. In particular, the author points out the potential limiting effects of labeling a patient with atypical facial pain and the lack of understanding of the mechanisms that can lead to both typical episodic trigeminal neuralgia (TN) and the less common persistent face pain that may be, as the author thinks, an extension of the root abnormalities responsible for TN.

This article will generate controversy and will provoke thought. Both of these results, in regard to facial pain, will be of benefit. I disagree with the author's separation of pain on the basis of deliberate and accidental damage to the trigeminal nerve or root because the ultimate mechanisms are likely the same. This separation may serve to inhibit understanding of the basic mechanisms. Similarly, the separation of neurovascular causes of TN and multiple sclerosis-related causes of TN is artificial, given that presumed demyelination is the common thread and precipitant cause and that many, but not all, treatments are similar and often effective.

Charles J. Hodge, Jr.
Syracuse, New York

The classification described in this article is needed and highly important. The study provides a basis for the evaluation and treatment of facial pain, particularly neuralgia. Some surgeons who have an accumulation of knowledge and experience in certain applications have been naturally striving to treat every type of facial pain with only one technique of treatment. Even more disturbing are the attempts made to prove that this single technique is the best treatment for facial pain. However, each type of pain may require patient-specific evaluation and different techniques of treatment. Therefore, the classification provided by this study will contribute to a differentiation of various types of facial pain, so that treatment modalities can be selected accordingly.

Although I found Dr. Burchiel's study highly impressive and contributory, I emphasize that in the diagnostic phase of atypical facial pain, it is essential that an experienced physician evaluate whether psychological tests constitute the criteria, because pain surgery is not merely a technical issue. It should rely on expertise. An experienced team of physicians should evaluate each patient.

Yücel Kanpolat
Ankara, Turkey

This article is a proposal for a practical classification of facial pain that depends only on the patient to provide the information necessary for classification. The classification has reasonable justification, but no data to support its validity (ability to measure what is intended—i.e., to separate patients with distinct types of facial pain amenable to different types of treatment) or reliability (intra- and interobserver reproducibility) are presented. As such, it can only serve as the basis for discussion or empiric testing. My personal preference would be for the latter; the discussion would be much more useful if some data were available along with this first presentation of the concept.

The process that should be followed involves the testing of the classification for reliability and creating modifications to maximize the particular characteristics of the classification. Validity confirmation will be more difficult, because some of the diagnostic categories are new. However, the ability to agree with independent diagnoses of "typical TN," intentional deafferentation, and multiple sclerosis-associated and postherpetic neuralgia would ease concerns regarding the validity of a completely patient-generated diagnosis. The process of validation and reliability testing is arduous and too infrequently completed. However, when it is done properly, the contribution to neurosurgical research and practice can be profound, as is the case with the Glasgow Coma Scale.

Stephen J. Haines
Charleston, South Carolina

For many reasons, there is a great need for improved classifications of various forms of pain, and the taxonomy of pain has always been regarded as one of the most important tasks of the International Association for the Study of Pain. This applies not least to the many different pain conditions in the face and head, some of which exclusively affect that part of the body.

Dr. Burchiel has an extensive clinical experience of pain, in particular surgical treatment of pain. He presents a simple and condensed classification of some common forms of facial neuralgia. In particular, I strongly support the idea that "atypical facial pain" should be used exclusively to denote psychogenic forms of pain, whether or not they respect the trigeminal territory. Unfortunately, in the literature, that "diagnosis" has been applied even to denote painful trigeminal neuropathy. I agree that the phenomenon of facial pain not respecting the trigeminal territory is a reason to suspect psychogenic pain. However, this criterion should be applied with caution because in some cases of trigeminal neuropathy, sensory abnormalities, as well as pain, may spread along the neck and to the shoulder region. Presumably, such extraterritorial spreading represents central plasticity changes. In principle, I agree with the proposed list of diagnoses, but I am doubtful whether this classification really adds much to what is already applied by most clinicians in the field.

In clinical practice, it is important to differentiate between what is here referred to as TN 1 and TN 2, but it would have been of much interest and importance if these diagnoses could have been characterized not only in terms of the relative occurrence of paroxysmal and continuous pain components. In practice, this is not so easy because most of these patients present with an ongoing pain onto which intermittent, more or less typical, spells of pain are superimposed. I do not consider the proposed distinguishing features to be practically useful. Virtually nothing is known regarding the pathophysiological correlates to episodic versus constant pain components in TN. It might be that thorough quantitative sensory testing could help to further differentiate and characterize these components. The identification of "mixed" forms of TN is of considerable importance because they tend to benefit less from interventional treatments.

I remain unconvinced that it is rational to separate trigeminal neuropathic pain into two classes according to etiology. It is stated that deafferentation is a characteristic feature in pain resulting from interventions that aim to treat TN. In fact, different degrees of deafferentation are generally also present in neuropathic pain after facial surgery, trauma, and tumors, although *complete* denervation, as in anesthesia dolorosa, is more common as a result of rhizotomy, for example.

With regard to treatment options, it is important to differentiate between pre- and postganglionic lesions, which may present with identical clinical features. Painful neuropathy resulting from injury to the peripheral portions of the nerve may respond to stimulation of gasserian ganglion/rootlets, which is not possible in cases of complete deafferentation, e.g., that caused by rhizotomy. It is proposed that symptomatic TN is associated with multiple sclerosis. However, according to the taxonomy of the International Association for the Study of Pain (1), this type of TN also includes conditions associated with tumors, vascular malformations, and other forms of posterior fossa pathology.

It has been argued that a classification of facial pain should be based on the possible underlying pathophysiological mechanisms. This statement is fully in line with the present trend to try to base classifications of pain on "mechanism-based" diagnoses instead of on the description and history of the pain (2).

Björn Meyerson
Stockholm, Sweden

1. Merskey H, Bogduk N (eds): *Classification of Chronic Pain*. Seattle, IASP Press, 1994, ed 2.
2. Woolf CJ, Bennett GJ, Doherty M, Dubner R, Kidd B, Koltzenburg M, Lipton R, Loeser JD, Payne R, Torebjork E: Towards a mechanism-based classification of pain? **Pain** 77:227–229, 1998.