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Painful Cranial Neuropathies and other Facial Pain

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ABSTRACT

Head pain is caused by activation of the trigeminocervical complex, which is common final pathway for all headaches. Due to the overlapping innervation of the trigeminal nerve and the terminal branches of the first cervical nerves, the innervation area of the trigeminocervical complex covers the entire head. In response to various, endogenous or exogenous, potential or real causes of damage, activation of the trigeminocervical complex can also trigger vegetative and vasomotor reactions. Irradiation of pain to adjacent regions of the head and face reduces the localization significance of pain, which represents a significant differential diagnostic difficulty. It is known that the following structures of the head are sensitive to pain: skin, subcutaneous tissue, muscles, arteries, periosteum, structures of the eye, ear and nasal cavities, intracranial venous sinuses, parts of the dura mater at the base of the brain, arteries in the dura and soft meninges and cranial nerves (nn.V, IX, X and nn. cervicales). On the other hand, the bones of the skull, most of the dura and soft meninges, the brain parenchyma, the ependyma of the ventricles and the choroid plexus do not represent pain-sensitive structures.

Analysis and interpretation of data obtained from a structured clinical interview, and clinical and neurological examination of the patient is usually sufficient to establish an accurate diagnosis. Although the diagnosis of head pain syndromes is most often clinical, in some conditions additional diagnostics are necessary. When examining the patient and during the clinical interview, special attention should be paid to data related to the pain syndrome itself, such as pain localization, lateralization, frequency and duration of pain, intensity and character of pain, the presence of associated symptoms, the presence of precipitating and relaxing factors, the degree of disability during and after the attack, the presence of complaints before the painful attack itself, the sensitivity of pain to the applied pain therapy, etc.

Data from personal health history and family history are especially important. Clinically, it is significant that chronic pain syndromes of the head and face arise from the transformation of primary, recurrent episodic syndromes.

Timely diagnosis and therapeutic treatment of episodic conditions is important for preventing their chronicity. At the end of the clinical evaluation of a patient with a painful syndrome of the head and face, it is necessary to consider whether the presented clinical condition is a consequence of some other pathological process; what is its clinical course so far (episodic - rare/frequent attacks or chronic course); whether additional diagnostics are necessary and which diagnostic procedure should be performed; which therapy is optimal for an acute approach, whether and with which therapy preventive intervention should be used, etc.

- Data from personal health history and family history are especially important. Clinically, it is significant that chronic pain syndromes of the head and face arise from the transformation of primary, recurrent episodic syndromes.
- Timely diagnosis and therapeutic treatment of episodic conditions is important for preventing their chronicity.
- Given that the chronicity/transformation of this pain conditions result over medication their clinical knowledge is crucial.

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Painful cranial neuropathies and other facial pain classified as a separate part of headache disorders [1-3].

It includes Painful cranial neuropathies and other facial pain, Trigeminal neuralgia, Classic trigeminal neuralgia, Classic trigeminal neuralgia, purely paroxysmal, Classic trigeminal neuralgia with associated persistent facial pain, Painful trigeminal neuropathy, Painful trigeminal neuropathy attributed to acute herpes zoster, Postherpetic trigeminal neuropathy, Painful posttraumatic trigeminal neuropathy, Painful trigeminal neuropathy attributed to plaque multiple sclerosis (MS), Painful trigeminal neuropathy attributed to expansive lesion, Painful trigeminal neuropathy attributed to other disorders, Glossopharyngeal neuralgia, N. intermedius (facial nerve) neuralgia, Classic intermedial neuralgia, Neuropathy n. intermedius attributed to Herpes zoster, Occipital neuralgia, Optic neuritis and Headache attributed to ischemic paresis of the ocular motor nerves, Tolosa-Hunt syndrome, Paratrigeminal oculosympathetic (Raeder) syndrome, Recurrent painful ophthalmoplegic neuropathy, Burning mouth syndrome (BMS), Persistent idiopathic facial pain (PIFP), Central neuropathic pain, Central neuropathic pain attributed to multiple sclerosis (MS), Central post-stroke pain (CPSP) [4,5].

13. Painful cranial neuropathies and other facial pains, 13.1 Trigeminal neuralgia, 13.1.1 Classical trigeminal neuralgia 13.1.1.1 Classical trigeminal neuralgia, purely paroxysmal, 13.1.1.2 Classical trigeminal neuralgia with concomitant persistent facial pain, 13.1.2 Painful trigeminal neuropathy, 13.1.2.1 Painful trigeminal neuropathy attributed to acute Herpes zoster, 13.1.2.2 Post-herpetic trigeminal neuropathy, 13.1.2.3 Painful post-traumatic trigeminal neuropathy, 13.1.2.4 Painful trigeminal neuropathy attributed to multiple sclerosis (MS) plaque, 13.1.2.5 Painful trigeminal neuropathy attributed to space-occupying lesion, 13.1.2.6 Painful trigeminal neuropathy attributed to other disorder, 13.2 Glossopharyngeal neuralgia, 13.3 Nervus intermedius (facial nerve) neuralgia, 13.3.1 Classical nervus intermedius neuralgia, 13.3.2 Nervus intermedius neuropathy attributed to Herpes zoster 13.4 Occipital neuralgia 13.5 Optic neuritis 13.6 Headache attributed to ischaemic ocular motor nerve palsy, 13.7 Tolosa-Hunt syndrome 13.8 Paratrigeminal oculosympathetic (Raeder's) syndrome 13.9 Recurrent painful ophthalmoplegic neuropathy 13.10 Burning mouth syndrome (BMS) 13.11 Persistent idiopathic facial pain (PIFP) 13.12 Central neuropathic pain 13.12.1 Central neuropathic pain attributed to multiple sclerosis (MS) 13.12.2 Central post-stroke pain (CPSP) [6,7].

13.1 Trigeminal neuralgia is a disorder characterized by recurrent unilateral brief electric shock-like pains, abrupt in onset and termination, limited to the distribution of one or more divisions of the trigeminal nerve and triggered by innocuous stimuli. It may develop without apparent cause or be a result of another diagnosed disorder. There may or may not be, additionally, persistent background facial pain of moderate intensity. 13.1.1 Classical trigeminal neuralgia previously used called as tic douloureux. Trigeminal neuralgia developing without apparent cause other than neurovascular compression. Diagnostic criteria: A. At least three attacks of 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: 1. recurring in paroxysmal attacks lasting from a fraction of a second to 2 minutes 2. severe intensity 3. electric shock- like, shooting, stabbing or sharp in quality 4. precipitated by innocuous stimuli to the affected side of the face1 D. No clinically evident neurological deficit2 E. Not better accounted for by another ICHD-3 diagnosis [1].

Classical Trigeminal Neuralgia - Purely Paroxysmal

Trigeminal neuralgia without persistent background facial pain. Diagnostic criteria: A. Recurrent attacks of unilateral facial pain fulfilling criteria for 13.1.1 Classical trigeminal neuralgia B. No persistent facial pain between attacks C. Not better accounted for by another ICHD-3 diagnosis. Comment: 13.1.1.1 Classical trigeminal neuralgia, purely paroxysmal is usually responsive, at least initially, to pharmacotherapy (especially carbamazepine or oxcarbazepine). 13.1.1.2 Classical trigeminal neuralgia with concomitant persistent facial pain Previously used terms: Atypical trigeminal neuralgia; trigeminal neuralgia type 2 [1-4].

Trigeminal neuralgia with persistent background facial pain. Diagnostic criteria: A. Recurrent attacks of unilateral facial pain fulfilling criteria for 13.1.1 Classical trigeminal neuralgia B. Persistent facial pain of moderate intensity in the affected area C. Not better accounted for by another ICHD-3 diagnosis. Comments: 13.1.1.2 Classical trigeminal neuralgia with concomitant persistent facial pain has been referred to as atypical trigeminal neuralgia or, recently, as trigeminal neuralgia type 2. Central sensitization may account for the persistent facial pain. Neurovascular compression on MRI is less likely to be demonstrated. 13.1.1.2 Classical trigeminal neuralgia with concomitant persistent facial pain responds poorly to conservative treatment and to neurosurgical interventions. It is less likely to be triggered by innocuous stimuli [5-7].

Painful trigeminal neuropathy 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. The pain is highly variable in quality and intensity according to the cause. Painful trigeminal neuropathy attributed to acute Herpes zoster Description: Unilateral head and/or facial pain of less than 3 months' duration in the distribution of one or more branches of the trigeminal nerve, caused by and associated with other symptoms and/or clinical signs of acute Herpes zoster. Diagnostic criteria: A. Unilateral head and/or facial pain lasting C. Evidence of causation demonstrated by both of the following: 1. pain preceded the herpetic eruption by Following acute Herpes zoster, post-herpetic neuralgia is more prevalent in the elderly. The first division of the trigeminal nerve is most commonly affected in post-herpetic trigeminal neuropathy, but the second and third divisions can be involved also. Typically, the pain is burning and itching. Itching of affected areas may be very prominent and extremely bothersome. Sensory abnormalities and allodynia are usually present in the territory involved. Pale or light purple scars may be present as sequelae of the herpetic eruption. Painful post-traumatic trigeminal neuropathy Previously used term: Anaesthesia dolorosa. Coded elsewhere: Here are described painful post-traumatic neuropathies; most trigeminal nerve injuries do not result in pain and therefore have no place in ICHD-4 beta. Description: Unilateral facial or oral pain following trauma to the trigeminal nerve, with other symptoms and/or clinical signs of trigeminal nerve dysfunction. Diagnostic criteria: A. Unilateral facial and/or oral pain fulfilling criterion C B. History of an identifiable traumatic event1 to the trigeminal nerve, with clinically evident positive (hyperalgesia, allodynia) and/or negative (hypoaesthesia, hypoalgesia) signs of trigeminal nerve dysfunction C. Evidence of causation demonstrated by both of the following: 1. pain is located in the distribution of the same trigeminal nerve 2. pain has developed within 3-6 months of the traumatic event D. Not better accounted for by another ICHD-3 diagnosis [8-10].

The traumatic event may be mechanical, chemical, thermal or caused by radiation. Pain duration ranges widely from paroxysmal to constant, and may be mixed. Specifically following radiation-induced postganglionic injury, neuropathy may appear after more than 3 months. 13.1.2.4 Painful trigeminal neuropathy attributed to multiple sclerosis (MS) plaque Unilateral head and/or facial pain in the distribution of a trigeminal nerve and with the characteristics of classical trigeminal neuralgia, induced by a multiple sclerosis plaque affecting the trigeminal nerve root and associated with other symptoms and/or clinical signs of multiple sclerosis. Diagnostic criteria: A. Head and/or facial pain with the characteristics of Classical trigeminal neuralgia with or without concomitant persistent facial pain, but not necessarily unilateral B. Multiple sclerosis (MS) has been diagnosed C. An MS plaque affecting the trigeminal nerve root has been demonstrated by MRI or by routine electrophysiological studies (blink reflex or trigeminal evoked potentials) indicating impairment of the affected trigeminal nerve(s) D. Not better accounted for by another ICHD-4 diagnosis. Current studies indicate that about 7% of MS patients have a syndrome that is similar to Classical trigeminal neuralgia. However, symptoms of trigeminal neuralgia are very rarely a presenting feature of MS. Symptoms of Painful trigeminal neuropathy attributed to multiple sclerosis (MS) plaque are more likely to be bilateral than those of Classical trigeminal neuralgia [11-13].

Patients with Painful trigeminal neuropathy attributed to multiple sclerosis (MS) plaque benefit less from pharmacological interventions than those with Classical trigeminal neuralgia. Painful trigeminal neuropathy attributed to space-occupying lesion Description: Unilateral head and/or facial pain in the distribution of a trigeminal nerve and with the characteristics of classical trigeminal neuralgia, induced by contact between the affected trigeminal nerve and a space-occupying lesion Diagnostic criteria: A. Unilateral head and/or facial pain with the characteristics of Classical trigeminal neuralgia with or without concomitant persistent facial pain and fulfilling criterion C B. A space-occupying lesion, and contact between the lesion and the affected trigeminal nerve, have been demonstrated by imaging C. Pain has developed after contact occurred between the lesion and the trigeminal nerve, or led to its discovery D. Not better accounted for by another ICHD-3 diagnosis. Patients with Painful trigeminal neuropathy attributed to space-occupying lesion have clinically detectable sensory signs or electrophysiological abnormalities. Painful trigeminal neuropathy attributed to other disorder Diagnostic criteria: A. Head and/or facial pain with the characteristics of Classical trigeminal neuralgia with or without concomitant persistent facial pain, but not necessarily unilateral B. A disorder, other than those described above but known to be capable of causing painful trigeminal neuropathy, has been diagnosed C. Pain has developed after onset of the disorder, or led to its discovery D. Not better accounted for by another ICHD-4 diagnosis [14,15].

Glossopharyngeal neuralgia Previously used term is Vagoglossopharyngeal neuralgia. A severe, transient, stabbing, unilateral pain experienced in the ear, base of the tongue, tonsillar fossa and/or beneath the angle of the jaw. It is commonly provoked by swallowing, talking and/or coughing, and may remit and relapse in the fashion of classical trigeminal neuralgia. Diagnostic criteria: A. At least three attacks of unilateral pain fulfilling criteria B and C B. Pain is located in the posterior part of the tongue, tonsillar fossa, pharynx, beneath the angle of the lower jaw and/or in the ear C. Pain has at least three of the following four characteristics: 1. recurring in paroxysmal attacks lasting from a few seconds to 2 minutes 2. severe intensity 3. shooting, stabbing or sharp in quality 4. precipitated by swallowing, coughing, talking or yawning D. No clinically evident neurological deficit E. Not better accounted for by another ICHD-4 diagnosis (1.15).

Glossopharyngeal neuralgia is felt in the distributions of the auricular and pharyngeal branches of the vagus nerve as well as branches of the glossopharyngeal nerve. Prior to its development, unpleasant sensations can be experienced in affected areas for weeks to several months. Glossopharyngeal neuralgia is less severe than Classical trigeminal neuralgia but can be bad enough for patients to lose weight. These two disorders can occur together. In rare cases, attacks of pain are associated with vagal symptoms such as cough, hoarseness, syncope and/or bradycardia. Some authors have proposed distinguishing between pharyngeal, otalgic and vagal subtypes of neuralgia, and suggested using the term vagoglossopharyngeal neuralgia when pain is accompanied by asystole, convulsions and syncope. Imaging may show neurovascular compression of the glossopharyngeal nerve. There are single reports of secondary glossopharyngeal neuropathy caused by neck trauma, multiple sclerosis, tonsillar or regional tumours, cerebello-pontine angle tumours and Arnold-Chiari malformation.

Glossopharyngeal neuralgia is usually responsive, at least initially, to pharmacotherapy, especially antiepileptics. It has been suggested that application of local anaesthetic to the tonsil and pharyngeal wall can prevent attacks for a few hours. Nervus Nervus intermedius (facial nerve) neuralgia Description: A rare disorder characterized by brief paroxysms of pain felt deeply in the auditory canal, sometimes radiating to the parieto-occipital region. It may develop without apparent cause or as a complication of Herpes zoster [16,17].

Classical nervus intermedius neuralgia Description: Nervus intermedius neuralgia developing without apparent cause. Diagnostic criteria: A. At least three attacks of unilateral pain fulfilling criteria B and C B. Pain is located in the auditory canal, sometimes radiating to the parieto-occipital region C. Pain has at least three of the following four characteristics: 1. recurring in paroxysmal attacks lasting from a few seconds to minutes 2. severe intensity 3. shooting, stabbing or sharp in quality 4. precipitated by stimulation of a trigger area in the posterior wall of the auditory canal and/or periauricular region D. No clinically evident neurological deficit E. Not better accounted for by another ICHD-3 diagnosis. Comments: Disorders of lacrimation, salivation and/or taste sometimes accompany the pain of Classical nervus intermedius neuralgia. In view of the complex and overlapping innervation of the external ear, deriving from the trigeminal (auriculotemporal nerve), facial (nervus intermedius), glossopharyngeal, vagus and second cranial nerves, attribution of neuralgias to a single nerve may not be easy in this body region if a specific neurovascular contact cannot be visualized. The pain of Classical nervus intermedius neuralgia can result in psychological effects and significantly impair quality of life. Secondary nervus intermedius neuropathy attributed to acute Herpes zoster Previously used term: Ramsay Hunt syndrome. Description: Unilateral pain felt deeply in the auditory canal, sometimes radiating to the parieto-occipital region, associated with facial paresis and caused by Herpes zoster of the nervus intermedius. Diagnostic criteria: A. Unilateral facial pain fulfilling criterion C B. Herpetic eruption has occurred in the ear and/or oral mucosa, in the territory of the nervus intermedius C. Evidence of causation demonstrated by both of the following: 1. recurring in paroxysmal attacks lasting from a few seconds to minutes 2. severe intensity 3. shooting, stabbing or sharp in quality D. Pain is associated with both of the following: 1. dysaesthesia and/or allodynia apparent during innocuous stimulation of the scalp and/or hair 2. either or both of the following: a) tenderness over the affected nerve branches b) trigger points at the emergence of the greater occipital nerve or in the area of distribution of C2 E. Pain is eased temporarily by local anaesthetic block of the affected nerve F. Not better accounted for by another ICHD-4 diagnosis [17,18].

Post-Herpetic Trigeminal Neuropathy
Previously used Term is Post-Herpetic Trigeminal Neuralgia
Unilateral head and/or facial pain persisting or recurring for at least 3 months in the distribution of one or more branches of the trigeminal nerve, with variable sensory changes, caused by Herpes zoster. Diagnostic criteria: A. Unilateral head and/or facial pain persisting or recurring for 3 months and fulfilling criterion C B. History of acute Herpes zoster affecting a trigeminal nerve branch or branches C. Evidence of causation demonstrated by both of the following: 1. pain developed in temporal relation to the acute Herpes zoster 2. pain is located in the distribution of the same trigeminal nerve branch or branches D. Not better accounted for by another ICHD-4 diagnosis [1,17].

11. Headache or facial pain attributed to disorder of the cranium, neck, eyes, ears, nose, sinuses, teeth, mouth or other facial or cervical structure 11.1 Headache attributed to disorder of

cranial bone 11.2 Headache attributed to disorder of the neck 11.2.1 Cervicogenic headache 11.2.2 Headache attributed to retropharyngeal tendonitis 11.2.3 Headache attributed to craniocervical dystonia 11.3 Headache attributed to disorder of the eyes 11.3.1 Headache attributed to acute glaucoma 11.3.2 Headache attributed to refractive error 11.3.3 Headache attributed to heterophoria or heterotropia (latent or persistent squint) 11.3.4 Headache attributed to ocular inflammatory disorder 11.3.5 Headache attributed to trochleitis 11.4 Headache attributed to disorder of the ears 11.5 Headache attributed to disorder of the nose or paranasal sinuses 11.5.1 Headache attributed to acute rhinosinusitis 11.5.2 Headache attributed to chronic or recurring rhinosinusitis 11.6 Headache attributed to disorder of the teeth or jaw 11.7 Headache attributed to temporomandibular disorder (TMD) 11.8 Head or facial pain attributed to inflammation of the stylohyoid ligament 11.9 Headache or facial pain attributed to other disorder of cranium, neck, eyes, ears, nose, sinuses, teeth, mouth or other facial or cervical structure [16-18].

Bullet Points

- Data from personal health history and family history are especially important. Clinically, it is significant that chronic pain syndromes of the head and face arise from the transformation of primary, recurrent episodic syndromes.
- Timely diagnosis and therapeutic treatment of episodic conditions is important for preventing their chronicity.
- Given that the chronicity/transformation of this pain conditions result over medication their clinical knowledge is crucial.

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