

# Facial Pain in Childhood and Adolescence

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## ABSTRACT

**Background:** Facial pain syndromes are very disabling conditions. They are uncommon in children and adolescents and a lot of paediatric tests do not address them; also in literature only few cases are reported. A practitioner's appropriate knowledge is very important in order to avoid misdiagnosis. Also there are symptomatic forms that are secondary to other causes like tumors or vascular malformation.

**Methods:** This review updates the trigeminal neuralgia, nervous intermedius neuralgia, glossopharyngeal neuralgia, occipital nerve neuralgia, Bell's palsy, red ear syndrome, neck-tongue syndrome, trigeminal autonomic cephalalgias, cluster headache, paroxysmal hemicrania, SUNA and SUNCT and the persistent and idiopathic facial pain.

**Conclusion:** Knowledge of the clinical features of these facial pain syndromes in children allows physicians to establish the correct diagnosis and develop the optimal treatment plan.

**KEYWORDS:** Red Ear Syndrome; Facial Pain; Childhood; Adolescence; Neuralgias.

## INTRODUCTION

Facial pain syndromes are a heterogeneous group of conditions characterized by pain in craniofacial district. On the basis of pain features (expression, duration, localization, etc.), we distinguish typical and atypical facial pain syndromes. Even if they are rare conditions in patients of young age, it is important to know them and their clinical features in order to put early and correct diagnosis and to start up an appropriate treatment without delay. Also these facial pain syndromes may be secondary to systemic diseases or lesions in central nervous system. Therefore, it is necessary to perform an accurate general and neurological examination. Sometimes imaging should be required.

## METHODS

Facial pain syndromes have been the subject of few studies and reviews probably because they are uncommon conditions.

In this review we focused our attention on original articles reporting clinical features, etiology and treatment in pediatric population.

## Typical facial pain

Very infrequent in children and adolescents, usually it is secondary or symptomatic. Typical facial pain syndromes include trigeminal neuralgia, glossopharyngeal neuralgia, occipital nerve neuralgia, cluster headache and other forms.

Trigeminal neuralgia (TGN): It is a very disabling condition, especially if untreated. Fortunately it is infrequent in children: according to Bahgat D, Ray DK, Raslan A, et al. [1] only 1% of cases occurs in under 20-year-old patients with only rare cases under 13 years of age. Trigeminal neuralgia is defined by the International Association for the Study of Pain (IASP) as "sudden usually unilateral severe, brief, stabbing, recurrent episodes of pain in the distribution of one or more branches

of the trigeminal nerve". The mandibular branch is more commonly affected than the other two trigeminal divisions. Usually episodes have sudden onset and abrupt end and patients have not pain between attacks. Painful paroxysms may be isolated or in burst. The single one may last from few seconds to some minutes. The pain may arise naturally or be triggered by stimulation of a specific cutaneous or mucosal area (trigger zone), which is almost always ipsilateral to the affected side, localized in the same or a different trigeminal division. The presence of a trigger zone is reported in the fifty percent of TGN patients. Also in a small percent of patients is described a sensational aura (parasthesia, dysesthesia, or anesthesia) within the trigger area before the onset of painful paroxysm [2]. A lot of triggers were identified: eating, drinking, brushing the teeth or washing or combing the hair can trigger the pain; furthermore even tonsillitis has been reported as a trigger in children [3]. A muscle twitch or contraction (tic douloureux) may associate with painful paroxysm. A lot of searches have been detected in children with trigeminal neuralgia (TGN) to identify an underlying etiology. Most cases of TGN are idiopathic and neurovascular compression seems to have an important etiological role: a lot of surgical studies document the presence of neurovascular compression in TGN patients and the High-resolution MRA demonstrates the exact relation of the vessels in the pons pressing on the adjacent trigeminal nerves. Symptomatic TGN may be secondary to multiple sclerosis (sometimes it is the first manifestation of the disease): in this case TGN may be bilateral. Other causes of symptomatic TGN may be Chiari malformation [4], brain tumors, vascular malformation. and asymptomatic Dandy-Walker syndrome [5]. Also typical TGN may represent the evolution of the Idiopathic trigeminal sensory neuropathy, an uncommon condition that causes sensory deficits in all three divisions of the trigeminal nerve [6]. While idiopathic trigeminal neuralgia is rare in children, symptomatic one may arise at all ages. So, when a child has a trigeminal neuralgia, it is important to exclude the possibility of an underlying cause. The treatment of TGN may be medical or surgical. Idiopathic TGN can be effectively treated with carbamazepine or oxcarbazepine, even if some studies report good response and others no response to medical therapies [7]. The treatment of symptomatic TGN is represented by the specific treatment of the primary disorder. In childhood surgical treatment is less effective than in adulthood.

**Nervus Intermedius Neuralgia:** Also called geniculate neuralgia, it is a rare condition in childhood. It is characterized by short paroxysms of pain in external auditory canal that can

radiate to ipsilateral parieto-occipital region. A sensory branch of facial nerve is affected. Painful paroxysms are triggered by stimulation of periauricular area. Riederer F, Sandor PS, Linnebank M, et al. have proposed a genetic transmission[8]. Usually secondary NIN is a complication of a herpetic infection. However other causes have been reported: a schwannoma in a 1-year-old child [9] and a pontine pilocytic astrocytoma in a 3-year-old girl [10].

**Glossopharyngeal neuralgia (GPN):** It is a very uncommon condition in children. It is characterized by paroxysmal and intense pain localized in the tonsillar fossa, in the base of the tongue, in the external ear canal or in the angle of the jaw. The painful paroxysm may be triggered by eating, swallowing, talking, chewing, yawning and laughing. The idiopathic form may be provoked by a neurovascular compression as the nerve leaves the brain stem. The secondary forms can be caused by tumor, multiple sclerosis, trauma, amygdalotomy, tonsillectomy or Chiari malformation [11]. Medical treatment includes antiepileptic drugs and antidepressant medications. Surgical treatment is suitable when GPN is provoked by neurovascular compression or pain is refractory. This condition can induces secondary depression, suicidal tendencies, fear of swallowing, loss of weight, and malnutrition.

**Occipital nerve neuralgia (ONN):** It is a rare condition in children, characterized by the occurrence of painful paroxysms in the distribution of the greater, lesser or third occipital nerves. These paroxysms can be unilateral or bilateral and duration can be seconds to minutes. In some occasions, tenderness over the nerve and altered sensitivity (ipoesthesia, dysesthesia or anesthesia) may be associated with painful paroxysm. In children ONN can be associated with stenosis of the foramen magnum in achondroplasia and traumatic injuries. Treatment includes anti-inflammatory and antiepileptic drugs, local anesthetic, blocks of nerve and occipital nerve stimulation [12,13].

**Bell's palsy:** It is an acute idiopathic paralysis of the facial nerve. First symptom is pain in the ear or in the face ipsilateral to the palsy. Paraesthesia and lacrimation may be associated to pain. Many patients loose taste sensation. The treatment is symptomatic: it is very important to use eye drops and eye patch to protect the cornea; there is no consensus about the use of corticosteroid in children. Usually all children undergo complete recovery.

**Red ear syndrome (RES):** It is an uncommon condition, characterized by unilateral/bilateral episodes of burning

sensation and pain in the ear, in association with ipsilateral erythema. Sometimes pain is not accompanied by color changes [14]. The episodes lasts from seconds to hours (generally from 30 to 60 minutes). Frequency is variable: from multiple attacks in a day to a few per year. Burning sensation and pain can occur spontaneously or can be triggered by touch, temperature changes, chewing, brushing the hair, exercise, stress, or neck movements [15,16]. RES episodes can be isolated or occur in association with primary headaches. RES can be idiopathic or symptomatic. Secondary forms are associated with vascular problems or cervical spine disorders. A vasculitic thalamic infarct was reported in a child with systemic lupus and red ear syndrome [17]. Treatment is pharmacological and non pharmacological. The pharmacological one consists in NSAIDs, gabapentin, amitriptyline and indomethacin; the non pharmacological treatment consists in prescription of a dental plate, local anesthetic block, surgical section or an application of ice-pack to the ear. Three years ago, Raieli et al. proposed a review of the literature about RES and evidenced that in 5/13 cases treated with indomethacin an improvement was reported [18]. Red Ear Syndrome may be a disabling condition, especially when it is not responding to medical treatments.

Neck-tongue syndrome: It is a rare syndrome characterized by unilateral cervical or occipital pain and ipsilateral tongue numbness. The pain is stabbing and paroxysmal. Painful paroxysms last seconds to minutes and sometimes are associated with tongue movements, paralysis, spasm, and dysarthria [19,20]. Triggers are sudden movements of the head. Neck-tongue syndrome may be secondary to disorders of the cervical spine, prolonged poor posture during sitting [21], and Chiari 1 malformation [22]. Treatment is conservative and consists in analgesics and muscle relaxants.

Trigeminal autonomic cephalalgias (TAC): Trigeminal autonomic cephalalgias (TAC) constitute a group of headaches that are characterized by unilateral severe pain along the distribution of the trigeminal nerve with corresponding activation of the autonomic nervous system. The autonomic characteristics consist of at least one of the following: conjunctival injection or lacrimation; nasal congestion or rhinorrhea; eyelid edema; facial or forehead sweating; redness of the face or forehead; sense of ear muffling; and miosis or ptosis [23]. TACs include cluster headache (CH), paroxysmal migraine (PH), continuous migraine (HC) and short-lasting unilateral neuralgiform headache attacks with autonomic cranial symptoms (SUNA), and short-lasting unilateral neuralgiform headaches with conjunctival injection and tearing (SUNCT) [24]. These conditions are distinguished

by the duration and frequency of the attacks, as well as the response to treatment [25]. TACs are rarely reported in pediatric age. The early-onset and adolescent forms of TACs are similar to the adult form both in the characteristics of pain, associated symptoms and response to treatment. Despite this, pediatric-onset TACs are poorly recognized and there is often a delay of several years before the diagnosis is made [26]. In most cases the diagnosis can be made with a careful history and with a normal MRI; Video testimonials of the autonomic characteristics can facilitate the diagnosis. Unlike in migraine there is no familiarity - given the age, many of the treatments used in adults are not used in the pediatric population, making the treatment of these conditions difficult [24].

Cluster headache: Cluster headache (CH) is a primary headache that can occur in children and adolescents and belongs to the diagnostic group of trigeminal autonomic cephalalgias (TACs). It is characterized by repeated attacks that typically last from 15 to 180 minutes of severe unilateral headache with autonomic cranial characteristics [27]. In the general population, cluster headache has a prevalence below 1% and its peak frequency involves the second and third decades of life (mean age 28-30 years), with a clear male prevalence. It is extremely rare in pediatric age. The estimated prevalence of the pediatric population (age 0-18) varies from 0.09 to 0.1% and few cases have been reported with onset in the first decade [28]. Based on a study of 18-year-old Swedish men the prevalence in children and adolescents was estimated to be 0.1% [29]. As in adults, children may experience severe restlessness and agitation. When bouts are separated by a month the disorder is labeled as an episodic cluster headache, while without a break the term that is applied is chronic cluster headache [27]. Potential differences from adults include the following: lower male to females ratio in children and children may have a lower frequency and shorter duration of attacks. There are several effective treatments such as oxygen, triptans, aspirin, steroids, and verapamil. Melatonin and topiramate are also valid in alternative children. However, the long interval between the clusters in children makes decisions about long-term treatment more difficult [30].

Paroxysmal hemicrania: This headache syndrome is rare in children, with an estimated prevalence of 1.3% in children with chronic headache [31]. The main feature of this type of headache is that it almost always affects the same side of the head. Another characteristic of paroxysmal migraine is indomethacin responsiveness. The main differential diagnosis must be placed with the cluster headache, although the

latter is distinguishable from the absence of response, in general, to indomethacin. The pain is typically located in the frontal, orbital, supraorbital or temporal regions. If there is not complete well-being with indomethacin, other drugs can be used such as verapamil, topiramate and acetazolamide [32]. Autonomic symptoms are usually present but may not occur with each attack. The episodes occur several times during the day and typically last from minutes to about half an hour [26]. The ICHD-3 criteria indicate that the pain is severe. In the described pediatric patients, the characteristics of the headache syndrome seem similar to those in adults. Long-term prognosis in children is not known, but long-term treatment may be required [12].

**SUNA and SUNCT:** Short-lasting unilateral neuralgiform headache attacks with autonomic cranial symptoms (SUNA), and short-lasting unilateral neuralgiform headaches with conjunctival injection and tearing (SUNCT) are rare headaches that are characterized by severe and short-lived pain. These headaches are often refractory to treatments and may have secondary etiology [33]. Daily episodes are described and can last from a week up to a year with pain-free intervals greater than or equal to one month. Treatment can be difficult; Triptans and some antiepileptic drugs, especially lamotrigine, have proven to be successful drugs [34]. Spontaneous resolution was observed in one child after 5 months. A posterior fossa lesion can mimic SUNCT and has been reported in one pediatric patient [12].

### **Atypical facial pain**

Persistent and idiopathic facial pain, formerly called atypical facial pain, is a pain that does not meet any other diagnostic criteria. It is a facial pain that occurs at a daily frequency of more than two hours a day for more than three months, in absence of neurological deficits [35]. In adults, the incidence is about 1-2% of the general population. The condition is most commonly reported in middle-aged women and is very rare in children and young people. In a significant percentage of cases, facial algia is attributed to a psychopathological condition or is an expression of a different pathology [36]. The diagnosis is basically based on the clinical manifestations. Pain typically extends beyond the borders of cranial or peripheral nerve patterns. Objective signs are not documented: the physical examination is normal. It is a deep pain in the soft tissues or in the bone, burning, painful or severe and throbbing; sometimes all these features can be found, also associated with paraesthesia. Features may vary over time. The site of pain tends to vary, alternately affecting the two

sides of the face [37]. Atypical facial pain may present in comorbidity with other pain conditions such as chronic widespread pain and/or irritable bowel syndrome. Odontogenic causes of pain should be excluded through appropriate investigations. The intensity of pain generally fluctuates during the day, resulting greater during daylight hours and absent in night-time ones, so patients usually have a regular sleep waking rhythm. It may persist for days or months. The etiopathogenesis of these pains is not known, although it has been hypothesized that it could be caused by muscular hyperactivity, such as in bruxism, or related to vascular problems, demyelinating or infectious causes or could have psychogenic origin, especially in those patients with depression and pre-existing anxiety. And indeed, these diagnoses are often associated with significant psychiatric comorbidities and psychosocial disabilities. Drugs such as dotiepine and fluoxetine are proven, although modest, to be beneficial in the treatment of idiopathic chronic orofacial algias, although originally developed as antidepressant drugs [38]. Much more than in adults, children's paradigms emphasize that a strategic program may vary according to the patient's age, family, culture, diagnosis and associated disability. Pharmacological treatments are recommended only in the most severe cases. A multidisciplinary approach could be the best option to treat these patients as this treatment may be more effective than therapy with a single pharmacological agent in reducing the intensity and frequency of algia and related disability [36].

### **DISCUSSION AND CONCLUSION**

Despite facial pain syndromes are very disabling conditions, a lot of clinicians do not fully know them. A practitioner's appropriate knowledge of their clinical features in pediatric age is very important in order to establish the correct diagnosis and develop the optimal treatment plan. The differential diagnosis of these syndromes is extremely difficult, because some clinical features are common in the different forms and children often are incapable of describing important symptoms. Therefore, it requires a careful and systematic approach. Sometimes neuroimaging is necessary, especially to exclude symptomatic forms, secondary to other causes like tumors or vascular malformations. The aim of this review was to focus on the pediatric age highlighting the clinical features of facial pain syndromes in children and adolescent in order to avoid misdiagnosis and start up an appropriate treatment without delay.

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