Cranial Neuralgias in Children and Adolescents A review of the literature

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Abstract:

Cranial neuralgias are a well-established cause of headache-related morbidity in the adult population. These disorders are poorly studied in general due to their relative rarity, particularly in children and adolescents, and they are likely underdiagnosed in these populations. Recognizing these disorders and differentiating them from more common headache disorders, such as migraine, is important, as secondary disease is common. This review will cover the basic epidemiology, diagnosis, and treatment of trigeminal, occipital, glossopharyngeal and other, less common, cranial neuralgias. We have reviewed pediatric case reports of these conditions. For trigeminal neuralgia, the most common of these disorders, we have compiled the clinical features and treatment response of previous reports.

Keywords: Cranial neuralgia, Trigeminal neuralgia, occipital neuralgia, glossopharyngeal neuralgia, nervus intermedius neuralgia, supraorbital neuralgia, infratrochlear neuralgia, nasociliary neuralgia pediatric, child, adolescent, microvascular decompression

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Introduction

Cranial neuralgias are well-established headache disorders in the adult population, accounting for significant morbidity and loss of productivity in this population. These disorders, of which trigeminal neuralgia is the most common, are uncommon but not rare, and they have well defined pathways of diagnosis, evaluation, and treatment.

In the pediatric population, cranial neuralgias are considered rare. The prevalence is unknown, although widely assumed to be less than older patients. While this is likely true, these disorders are probably under recognized and underdiagnosed in this population. A presumption of rarity may decrease a provider's diagnostic consideration of these disorders, and they may present differently in children than adults. In the pediatric population, migraine is by far the most common disabling headache disorder and is often the default headache diagnosis. This diagnostic presumption will lead to misdiagnosis of these rare disorders that can have overlapping clinical features. The purpose of this review is to increase awareness, provide diagnostic criteria, incorporate data from pediatric cases, and suggest reasonable therapies.

Trigeminal Neuralgia

Clinical Features

Trigeminal Neuralgia (TN), also known as "tic douloureux," is the most common of the cranial neuralgias. The International Classification of Headache Disorder 3rd edition (ICHD-3) has defined diagnosis and classification of TN (Table 1). Trigeminal neuralgia is characterized by attacks of shooting, stabbing, or sharp pain, with duration of seconds to minutes, occurring in the distribution of the trigeminal nerve. These headaches are often triggered by sensory stimulation of the head, neck or mouth, such as chewing, eating, brushing teeth, cold wind, or a light touch to the face. The distribution may be more than one branch, occurs most commonly in V2 or V3, and is typically unilateral, although there are rare cases of bilateral disease [1]. Severe pain may cause muscle contractions to the ipsilateral side, hence the name "tic douloureux.' There is often a refractory period in which attack cannot be re-triggered [2].

Differential Diagnosis

Trigeminal neuralgia may have mild autonomic symptoms such as lacrimation, eye redness, or swelling [2]. This needs to be differentiated from other primary headache disorders with autonomic features, such as paroxysmal hemicrania, short-lasting unilateral neuralgiform headache attacks with conjunctival injection and tearing (SUNCT), and short-lasting neuralgiform headaches with cranial autonomic symptoms (SUNA). In TN, pain does not tend to radiate outside of the specific nerve branch distribution [6, 7, 8, 9]. Bilateral symptoms are rare and should lead to consideration of other disorders, such as demyelinating disease.

Pain triggered by mouth movements, chewing, or certain temperatures should be differentiated from dental related pain or temporomandibular joint disorder. Other considerations should be made for sialadenitis, salivary syndrome, giant cell arteritis, primary stabbing headache, trigeminal neuropathy, multiple sclerosis, postherpetic neuralgia, or mass lesion. Given the frequency of secondary disease, imaging of the brain with and without contrast is indicated in all cases of suspected TN.

Classification

Classification is divided in classical, secondary, and idiopathic (Figure 1). Classical TN is due to neurovascular compression at the trigeminal nerve root. Secondary TN is caused by a structural distortion or injury such as a space occupying lesion (SOL), arteriovenous malformation (AVM), or multiple sclerosis (MS). It is considered idiopathic when diagnostic testing had not revealed a secondary or classical cause [4].

Prior to 2018, idiopathic cases were categorized as classical, as many idiopathic cases were assumed to be due to vascular compression, largely due to limitations of early magnetic resonance imaging (MRI) in identifying this abnormality [3]. Diffusion tensor imaging has now also shown increased ability to detect demyelination and has led to earlier imaging and improved detection of symptomatic lesions [5].

Further classification can divide classical or idiopathic TN into the paroxysmal form, characterized by pain remission between attacks, and the concomitant continuous form, with persistent interictal pain [2].

Epidemiology

Trigeminal neuralgia incidence ranges from 4 to 29 per 100,000, increasing with age, and the prevalence is estimated to be 1.5 to 7 per 10,000 [6, 10, 11, 12, 13, 14, 15]. TN is slightly more common in females (60-69%), on the right side (56-60%), and the majority is in the V2 or V-3 distribution (69-84%) [6, 16] which coincides with the pooled pediatric case studies (table 2).

There are no dedicated studies on incidence and prevalence in a pediatric population. Less than 1.5% of classical TN experience their first symptom before the age of 18 [18].

Pediatric case reports were largely skewed toward secondary causes (table 2). However, when looking at retrospective studies or large case series, classical and idiopathic types are most common. A retrospective study of a pediatric headache clinic showed TN in 0.5% of patients in a 5-year period, all cases idiopathic [17]. Another retrospective study of adults with symptoms beginning in childhood found 22 of 23 (96%) adults with classical TN and only one (4%) secondary TN case [18]. It is likely that classical and idiopathic TN, which account for over 80% of cases, are underreported [20, 21].

The most common secondary causes of TN in adults are SOL, AVM, and MS. Many of the pooled pediatric case reports are caused by a SOL, most commonly cerebellopontine angle (CPA) lipomas, followed by epidermoid cysts [20, 23, 24, 25, 26, 27, 28]. CPA lesions are overall uncommon, as seen in one retrospective report of 36 CPA lesions out of 819 cases of TN (4.4%) with only one under age 18 (0.12%) [22]. Additional SOL cases include Burkitt lymphoma [29], CPA arachnoid cyst [30], choristoma [31], capillary telangiectasia [9], embryonal rhabdosarcoma [32], ruptured dermoid cyst [33], anterior inferior cerebellar artery loop [34], and pilomyxoid astrocytoma [35]. There have been two reported cases of AVM [36, 37]. There are no reported childhood cases of MS-associated TN in childhood [38]. Additional secondary cases seen with pediatric onset are Chiari 1 Malformation [39], sinusitis [40], maxilla-mandibular hypoplasia [41], hydrocephalus [42], extracranial hamartoma [43], pierced tongue [44], and tonsillitis [45].

Pathophysiology

The ultimate pathophysiology of TN is unknown. The classification of classical TN is largely based on the "ignition hypothesis". The inciting lesion is vascular compression at the trigeminal root where the nerve exits the brainstem. Compression leads to demyelination and ectopic firing as the nerve becomes hyperexcitable and susceptible to cross excitation. Cases of secondary TN may result from similar pathophysiology, as many SOLs compress the nerve root. Microvascular decompression (MVD) and symptomatic lesion resection often resolve symptoms, further supporting this theory [5, 6, 46].

The root cause in many cases of TN remains elusive. Voltage gated sodium channels seem to play an important role in this ectopic activity, as evidenced by dysregulation and mutation of these sodium channels in certain patients. This finding, in addition to clinical response to sodium channel blockade, argues for sodium channel pathology [5,46]. Other etiologic theories include include increased sensitization of trigeminal nociceptive systems [4,10], genetic predisposition with rare reports of familial disease [21], subclinical herpes simplex reactivation [6], or neuronal inflammation [5,10].

Management

Much of the evidence for treatment in pediatrics is based on adult studies. A summation of treatments in the collected studies is provided (Table 3). There is a skew towards failure of medical management as these tend to be surgical retrospective studies or secondary causes of TN. There are no prospective studies for medical management in adults and no studies for medical or surgical management in pediatrics.

Medical Management

First line therapy for TN is traditionally carbamazepine or oxcarbazepine, resulting in pain control in about 75-80% of adult patients [3, 10, 47]. Other medications considered for TN include baclofen, lamotrigine, levetiracetam, gabapentin, pregabalin, topiramate, and botulinum toxin-A [4, 5, 21]. Botulinum toxin-A has shown significant response in 70-100% of patients, resulting in reduction of pain and attack frequency by 50-100% at 4 weeks and may be used to bridge to surgical management [6, 20].

Acute treatment for exacerbation of TN includes local injection to trigger area with lidocaine or ropivacaine, intranasal lidocaine, intravenous fosphenytoin or valproic acid, and rapid titration of anti-epileptics [3, 5, 10, 16].

Surgical and Procedural Management

Surgical or procedural treatments should be considered with failure of medical treatment, insufficient pain control from medical treatment, or when medical treatment is not tolerated. Referral to neurosurgery may be considered early in the course while medical management is trialed, particularly when a causative lesion is identified [48]. Current accepted options include microvascular decompression (MVD), percutaneous rhizotomy, peripheral surgical techniques, and stereotactic radiosurgery. Of these, in pediatric classical or idiopathic TN, there are only reports of MVD and percutaneous glycerol rhizotomy with promising results (Table 3).

Microvascular decompression is considered the first choice in classical TN in adults and has proven most effective, providing the longest duration of pain freedom: 90% pain relief post-operatively, over 80% at one year, 75% at 3 year, 73% at 5 years [3, 5]. A pooled analysis shows 3-to-11-year pain freedom in 62-

89% [5]. It is the most invasive procedure but has generally low risks of morbidity and mortality [3, 5, 16]. Combined collected pediatric cases showed successful outcomes in 13 of 17 (76%). Typical complications include cerebrospinal fluid leak, infection, diplopia, stroke, cerebellar lesions, and cranial nerve deficits [3, 48]. Ipsilateral hearing loss is the most common long-term complication [3].

Percutaneous rhizotomy involving radiofrequency thermocoagulation, glycerol injection, or balloon compression is often preferred for idiopathic TN. For glycerol injections, pediatric patients had greater than 50% improvement in pain, of which 85% was sustained at 1 year and 72% at 3 years. Transient paresthesia was the most common complication [49]. Pooled analyses show 4-to-11-year pain freedom 19-58% after glycerol injection in adults [5].

Gamma knife is usually reserved for elderly or debilitated patients who are poor surgical candidates [16]. There are other peripheral surgical techniques which have low morbidity but high reoccurrence, including cryotherapy, neurectomy, alcohol or phenol injection, peripheral acupuncture, and thermocoagulation [3]. Emerging therapies include neuromodulation and deep brain stimulation [4, 10].

Psychologic management

Like most pain syndromes, TN has a high rate of comorbid depression and anxiety [50]. These patients should be screened psychologic comorbidities and referred for psychological evaluation and support groups when appropriate. Psychologic comorbidity in children has not been well defined, but there has been a report of comorbidity with a somatoform disorder [51].

Glossopharyngeal Neuralgia

GN is characterized by severe, electric, stabbing, or sharp painful attacks that last from seconds to minutes in the distribution of the glossopharyngeal nerve: tongue, tonsillar fossa, pharynx, angle of the lower jaw, or ear. These attacks may be precipitated by swallowing, coughing, talking or yawning (Table 4). Patients can have weeks to months of abnormal sensory symptoms before the pain begins.

Glossopharyngeal Neuralgia (GN) incidence has been reported to be 0.2 to 0.8 per 100,000 in the general population [11]. No studies have studied the prevalence or incidence in the pediatric population.

Involvement of vagal branches can precipitate additional symptoms of cough, hoarseness, syncope, bradycardia, and even asystole, which may warrant more urgent surgery. One quarter of patients are bilateral. To confirm diagnosis, local anesthetic can be applied to the pharynx and tonsils for transient relief [6].

Like TN, GN is classified into classical, secondary, and idiopathic. A retrospective study found 8 cases of idiopathic GN successfully treated with peripheral glycerol injections [19]. Secondary GN in children has been attributed to vagus nerve stimulator [52], Eagle Syndrome [53, 54, 55], Chiari 1 malformation [56], and schwannoma [51]. Two of the cases were initially attributed to acute otitis media, the diagnosis made following treatment failure [53, 57].

Eagle syndrome, an elongated styloid process or calcified stylohyoid ligament that causes impingement on the glossopharyngeal nerve, has been associated with pediatric GN, and has also been proposed as evidence of a genetic component [55]. There is one report of a 13-year-old boy with 6 years of

refractory GN symptoms and thought to be idiopathic. During the adolescent's second ablation surgery, they found an elongated styloid that was removed with resolution of symptoms [53].

Initial medical treatment mirrors that of TN, with carbamazepine and oxcarbazepine as first line, although studies are lacking for all ages. One case of classical GN has been reported in which carbamazepine failed, but gabapentin brought relief [57].

If refractory to medical management, surgery may be considered. Options include radiofrequency ablation, gamma knife treatment, rhizotomy, or MVD. A summary of pediatric cases involving procedural management is included in table 5. Operative mortality might be as high as 5% and complications can include dysphagia and hoarseness [58].

Occipital Neuralgia

Epidemiology

The International Headache Society describes occipital neuralgia (ON), or "Arnold's Neuralgia", as a paroxysmal, unilateral or bilateral pain of severe intensity that is shooting, stabbing, or sharp in quality located over the distribution of the greater (GON), lesser (LON), or third (TON) occipital nerves. It may be associated with dysesthesia, allodynia, and/or tenderness over the affected branches. There have been no dedicated studies to assess the incidence and prevalence of ON in the pediatric population, although one Dutch study found no cases in children and an incidence of 3.2 cases per 100,000 person years (PY) for all ages, as compared to 12.6 in TN for all-ages [13].

Clinical Features

The ICHD-3 diagnostic criteria for ON are reviewed in Table 6. In addition to those diagnostic features, patient may show a positive "Tinel's sign": tingling evoked by light pressure or percussion over the distribution of the nerve, and the "pillow sign": pain on hyperextension or rotation of the neck [59]. Some patients may also present with referred pain and headache in the occipitofrontal or hemi-cranial regions [60]. Children may present with a restricted range of motion of the neck or a head tilt [61].

Pathophysiology

The majority of data around the pathophysiology of ON implicates compression or irritation of the GON along its anatomical course. Studies have shown that 90% of cases involve GON pathology, followed by LON in 10% of cases, and only rarely TON. [62]. Multiple compression points have been identified and described [59, 63, 64].

ON-associated headache may occur due to abnormalities in the first three cervical nerve roots, with C2 being the predominant pain pathway. The C2 nerve root is vulnerable to injury due to its anatomical positioning in the atlantoaxial interlaminar disc space, which is prone to degenerative changes, instability, and congenital abnormalities [64]. ON has also been attributed to muscular hypertrophy and muscular spasms which have been relieved by surgical sectioning of muscles in close approximation with the GON [65]. In some rarer cases, the formation of fibrocartilage calluses, structural bone-related changes to the skull or spine, Arnold Chiari malformations, and AV malformations have been a cause of nerve compression [59, 66].

Diagnostic Evaluation and Management

Occipital headache has traditionally been considered an indication for neuroimaging to exclude intracranial pathology such as Chiari I malformation and vascular or structural lesions of the posterior fossa or upper cervical cord [67]. Later studies suggest this may not necessarily be true [68, 69]. It is important to clinically recognize occipital neuralgia and differentiate it from occipital predominant migraine, as the former requires neuroimaging. Please see the article “Approach to the Diagnosis of Headache” in this issue for further information.

Post traumatic headaches (PTH) have an annual incidence of 1.6 to 3.8 million in the United States. Patients with persistent headaches in the occipital and posterior cervical region following traumatic injury should be evaluated for ON, which may present acutely or sub-acutely following a traumatic event [70]. This may be similar to the pain experienced from injury of the atlantoaxial or upper zygopophyseal joints [71]. For persistent post-traumatic headache, occipital nerve blocks have found to produce satisfactory pain relief in most cases after a failed trial of conservative management [72, 73].

Patients presenting with persistent headaches and tenderness over the occipital region following ventricular shunt placement should be evaluated for ON [74]. This may be due to mechanical irritation of the occipital artery or direct irritation of the greater or lesser occipital nerves by the shunt tubing or reservoir. These headaches may respond to cautious occipital nerve blockade [75].

Occipital nerve block may be both diagnostic and therapeutic. Several studies have demonstrated the safety and efficacy of this procedure, whether performed with anesthetic alone or in combination with steroid [76, 77, 78, 79]. Conservative treatment options such as NSAIDs, neck immobilization, physiotherapy, and massage are often used, although good studies are lacking. Medications used for other neuralgias, such as carbamazepine, gabapentin, and tricyclics, are often used although suffer from a similar lack of evidence in both adults and children. Those patients who fail initial therapeutic interventions should be thoroughly evaluated for other causes of primary and secondary headache.

Other treatment modalities have been attempted for refractory cases, with some success in the adult population, including the use of botulinum toxin A injections, thermal and pulsed radiofrequency ablation, and peripheral nerve stimulation [80, 81, 82]. For patients with third occipital nerve headaches in particular, radiofrequency neurotomy has been performed with a success rate of 88% [83].

Surgery has been reserved as a last resort once minimally invasive treatments have failed, and it has shown a 68-70% success rate [84, 85, 86, 87, 88]. A study done on adult patients who underwent micro-surgical decompression of the GON in China concluded that 89.5% patients had complete resolution of symptoms. This seemed especially favorable in cases where the ON was caused by compression from surrounding structures, such a swollen lymph node or proximity to a muscle or tendon [89]. Although microvascular decompression has been successfully used in several pediatric cases, scientific data in this population is lacking, and its use remains controversial, particularly in children.

Other Cranial Nerve Neuralgias

It stands to reason than any cranial nerve or branch can be insulted and cause neuropathic pain in its sensory distribution. Known examples are nervus intermedius (geniculate) neuralgia (NIN), supraorbital neuralgia (SON), infratrochlear neuralgia (ITN), and nasociliary neuralgia (NCN). The latter three

neuralgias are branches of the V1 trigeminal nerve. Treatments for these rarer neuralgias are largely uninvestigated, and invasive procedures should be considered experimental.

The nervus intermedius (NI) is a somatic afferent branch of cranial nerve VII enervating the internal auditory canal. NI neuralgia classification is clinically similar to TN and GPN and presents with pain and sensory changes in the posterior wall of the ear canal and periauricular areas. There are 3 reported cases refractory to standard neuralgia medications, all receiving MVD and NI sectioning with relief [90, 91, 92].

Supraorbital neuralgia has been defined as a separate entity from trigeminal neuralgia in a few post-traumatic cases, and responds to supraorbital nerve block [16, 93]. Only two pediatric cases were found, and one self-resolved at follow-up [92, 95]. If refractory to medical treatment, cryoablation or surgical nerve decompression have been reported [16].

Infratrochlear neuralgia causes a paroxysmal pain in the internal angle of the orbit and generally will respond to nerve block, which was a successful treatment to the only reported pediatric case [16, 94]. A common etiology may be posttraumatic entrapment syndrome, possibly requiring reduction of zygomatic fracture [16]. Gabapentin was also effective in a case series of adults [94]. NCN is a similar syndrome with pain on the unilateral nose with no reported pediatric cases.

Conclusion

Cranial neuralgias are rare in children, creating a risk of under- and misdiagnosis, which can result in inadequate diagnostic evaluation and ineffective treatment. These disorders are often secondary, and require, at minimum, brain and neurovascular imaging to evaluate for structural abnormalities. Research in pediatric cranial neuralgias is limited. A variety of symptomatic lesions have been described. Case studies show that many proven treatments in adults can be used in children, although the true efficacy of these therapies is unknown and requires further study.

Table 1: The International Headache Society diagnostic criteria for Trigeminal Neuralgia and subforms

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. Additionally, there may be concomitant continuous pain of moderate intensity within the distribution(s) of the affected nerve division(s).

Diagnostic Criteria:

Recurrent paroxysms of unilateral facial pain in the distribution(s) of one or more divisions of the trigeminal nerve, with no radiation beyond, and fulfilling criteria B and C

A. Pain has all of the following characteristics:
    1. lasting from a fraction of a second to 2 minutes
    2. severe intensity
    3. electric shock-like, shooting, stabbing or sharp in quality
B. Precipitated by innocuous stimuli within the affected trigeminal distribution
C. Not better accounted for by another ICHD-3 diagnosis.

Classical: Trigeminal neuralgia developing without apparent cause other than neurovascular compression and demonstration on MRI or during surgery of neurovascular compression (not simply contact), with morphological changes in the trigeminal nerve root.

Secondary: Trigeminal neuralgia caused by an underlying disease. Clinical examination shows sensory changes in a significant proportion of these patients. Can either purely paroxysmal or associated with concomitant continuous or near-continuous pain. An underlying disease has been demonstrated that is known to be able to cause, and explaining, the neuralgia.

Idiopathic: Trigeminal neuralgia with neither electrophysiological tests nor MRI showing significant abnormalities. Can either purely paroxysmal or associated with concomitant continuous or near-continuous pain. Neither classical nor secondary has been confirmed by adequate investigation including electrophysiological tests and MRI.

Table 2: Statistical Findings of the case studies, case series, and retrospective studies for pediatric trigeminal neuralgia [1, 7, 9, 17, 19, 23-37, 40-45, 48, 96, 97, 99-101, 103, 105, 106]

Patient Total	63	%			%
Male	26	41	**Classical**	13	21
Female	37	59	**Secondary**	24	38
Average Age (yr)	11.6 (1.1 to 18)		*> SOL*	15	24
Average duration (yr) until presentation	1.7 (0 to 12)		- Lipoma	6	10
Right	27	43	- Epidermoid Cyst	2	3
Left	17	27	*> AVM*	2	3
Bilateral	1	2	*> Other Secondary*	7	11
Unknown side (Yue)	18	29	**Idiopathic**	26	41
V1	7	11			
V2	26	41			
V3	15	24			
V1-2	5	8			
V2-3	6	10			
V1-3	5	8			

Table 3: Treatment outcomes found in case studies, case series, and retrospective studies for pediatric trigeminal neuralgia [1, 7, 9, 17, 19, 23-37, 40-45, 48, 96, 97, 99-101, 103, 105, 106]

	Total patients	Successful Treatment	Notes
Carbamazepine	43	8	18 of these from Yue 2004 which indicated that TGN was refractory to medications such as carbamazepine, but said no more than that
Oxcarbazepine	3	1	
Pregabalin	1	0	
Gabapentin	17	4	
Topiramate	2	0	
Clonazepam	2	1	
Amitriptyline	4	0	
Phenytoin	2	0	
Levetiracetam	1	0	
Baclofen	3	0	
Valproic acid	1	0	
Lamotrigine	2	2	
Combination therapy	5	5	Nerve block + gabapentin, nerve block + pregabalin, Carbamazepine + Clonazepam, Carbamazepine + Gabapentin, Lamotrigine + Oxcarbazepine,
Glycerol rhizotomy	19	16	2 only after 2nd injection
Nerve block	5	3	Lidocaine + methylprednisolone
MVD	17	13	1 successful only after 4th MVD, another after 2nd MVD, 1 remained on carbamazepine
Secondary cause removal	10	10	SOL removal, tonsillectomy, stud of pierced tongue removed
Stereotactic radiation	2	2	Gamma knife, proton beam radiation for secondary cause
Other	5	5	Ventriculoperitoneal shunt, tincture of Belladonna, embolization of AVM, clipping and coagulation of AVM, oral antibiotics and decongestants for sinusitis

Table 4: The International Headache Society diagnostic criteria for Glossopharyngeal Neuralgia and subforms

A disorder characterized by unilateral brief stabbing pain, abrupt in onset and termination, in the distributions not only of the glossopharyngeal nerve but also of the auricular and pharyngeal branches of the vagus nerve. Pain is 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 or coughing and may remit and relapse in the fashion of trigeminal neuralgia.

Diagnostic criteria:

A. Recurring paroxysmal attacks of unilateral pain in the distribution of the glossopharyngeal nerve and fulfilling criterion B
B. Pain has all of the following characteristics:
    1. lasting from a few seconds to 2 minutes
    2. severe intensity
    3. electric shock-like, shooting, stabbing or sharp in quality
    4. precipitated by swallowing, coughing, talking or yawning
C. Not better accounted for by another ICHD-3 diagnosis.

Classical: Glossopharyngeal neuralgia developing without apparent cause other than neurovascular compression and with demonstration on MRI or during surgery of neurovascular compression of the glossopharyngeal nerve root.

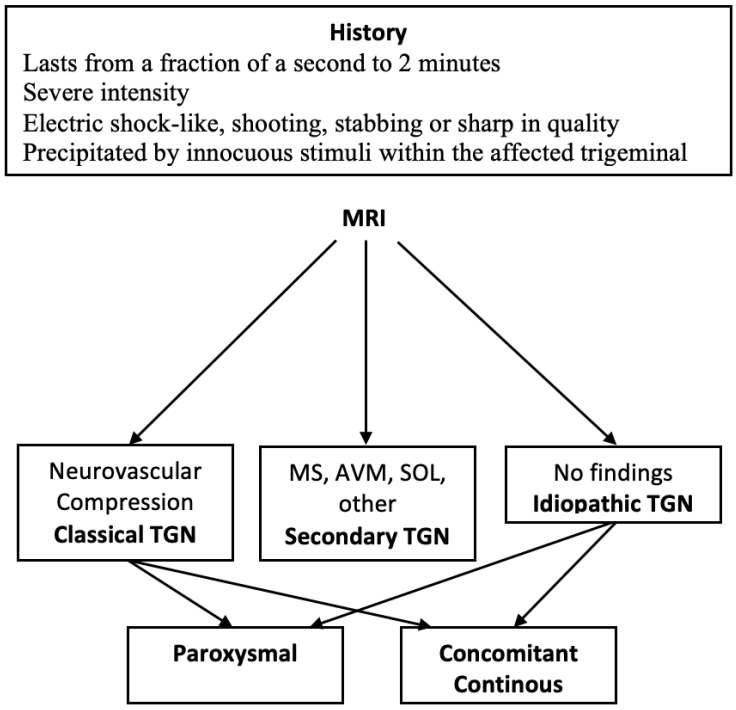
Secondary: Glossopharyngeal neuralgia caused by an underlying disease.

Idiopathic: Glossopharyngeal neuralgia with no evidence either of neurovascular compression or of causative underlying disease.

Table 5: Glossopharyngeal case studies with one retrospective study								
Reference	**Age**	**Duration (yr)**	**Sex**	**Side**	**Classification**	**Etiology**	**Successful Treatment**	**Prognosis**
Childs et al, 2000	13	12	F	R	Classical	Posterior inferior cerebellar artery	Gabapentin (Failed Carbamazepine)	Excellent, unknown follow up
Duhaime et al, 2000	16	0	F	L	Secondary	VNS	Decreased VNS output current & pulse width	Excellent at 10 months
Garriz-Luis et al, 2017	12	3	M	R	Secondary	Eagle Syndrome	Styloid shortening, stylohyoid shortening	Excellent at 7 years. Had 4 months anxiety and 1 year residual discomfort.
Kandt et al, 1986	13	7	M	R	Idiopathic vs Secondary	Eagle Syndrome?	3 total nerve ablations with 2 surgeries, styloid removal	Excellent at 13 months. Continuing mild right ptosis and miosis. Prior to surgeries, refractory to many medications.
Sarikaya-Sweiwert et al, 2013	14	0.3	M	L	Secondary, Vagal involvement	Schwannoma	Resection	Excellent 6 months, multiple complications requiring rehabilitation
Yglesias et al, 1996	8	2	M	?	Secondary	Chiari 1 Malformation	Surgical Decompression	Excellent at 2 years
Yue 2014	9 to 14 (mean 12.5)	0.25 to 1 (mean 0.4)	5 M 3 F	?	All 8 Idiopathic		Peripheral Glycerol Injections	5 Excellent, 2 Good, 1 Poor. Follow up ranged 10 to 32 months (mean 12.5). No long-term complications.

Table 6: ICHD-III Diagnostic criteria for Occipital Neuralgia
A. Unilateral or bilateral pain fulfilling criteria B - D B. Pain is located in the distribution of the greater, lesser and/or third occipital nerves C. Pain has two of the following three characteristics: 1. Recurring in paroxysmal attacks lasting from a few seconds to minutes 2. Severe in intensity 3. Shooting, stabbing or sharp in quality D. Pain is associated with both of the following: 1. Dysesthesia 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 distribution of C2 E. Pain is eased temporarily by local anesthetic block of the affected nerve(s) Not better accounted for by another ICHD-3 diagnosis.

Figure 1: Flow Diagram of the ICHD-3 Classification



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Appendix: all tabulated pediatric TN reports

Appendix of TGN studies								
Reference	**Age**	**Duration (yr)**	**Sex**	**Distribution**	**Classification**	**Etiology**	**Successful Treatment**	**Prognosis**
Akhaddar et al, 2012	13	0.1	F	R V2	Secondary	Burkitt Lymphoma		Died at 3 months
Alafaci et al, 2001	16	2	F	R V2-3	Secondary	CPA Lipoma	Resection + MVD	Excellent few weeks, hearing loss
Balasubramaniam et al, 2008	14	2	M	R V3	Secondary	CPA Lipoma	Resection	Good at 1 year, added anti-epileptic
Bender et al, 2011	13	0.19	F	R V1-2	Classical	Medullary and petrosal veins	MVD	Excellent at 24.8 months
	18	1.16	F	R V2	Classical	Petrosal and unnamed veins	MVD	Excellent at 20.6 months
	3	2.32	F	R V2	Classical	Medullary and petrosal veins	MVD	Excellent at 13.3 months
	4	0.29	F	L V1	Classical	Superior cerebellar artery, unnamed veins	MVD	Good at 9.1 months
Bilateral TN	12	R 0.16 L 5.02	F	R V2 L V3	Classical	Superior cerebellar artery, two unnamed veins	Separate MVD for each side	Excellent at 12 months
Bhoi et al, 2020	10	0	M	R V2	Secondary	CPA Lipoma	Carbamazepine	Excellent at 3 months
Brameli et al, 2020	9.5		F	R V1-2	Idiopathic		Nerve block	Excellent, unknown follow up
	18		F	L V1-2	Idiopathic		Nerve block x3 + Gabapentin	Good, had 3 relapses, transferred adult care
	14		F	R V1	Idiopathic		Nerve block +Pregabalin	Excellent at 1 year, lost to follow up
	16		F	L V1	Idiopathic		Carbamazepine	Partial, transferred adult care at 2 years
	17		F	L V1	Idiopathic		Gabapentin	Excellent at 1.5 years
Chicoine et al, 2019	15	12	F	R V2-3	Classical	Anterior inferior cerebellar artery	MVD	Excellent at 5 months, occasional migraines
	12	0.1	F	L V3	Classical	Venous complex	MVD	Excellent at 6 months

Childs et al, 2000	9	0.8	M	R V1	Classical	Mix of petrosal vein, unnamed artery, vein, & venule	4th MVD, 2nd MVD	Excellent at 13 months after 4th MVD
	12	0.2	M	L V1-2	Classical	Unnamed vein		Excellent after 2nd MVD, unknown follow up
Chun et al, 2017	13		M	L V2-3	Secondary	Sinusitis	Oral antibiotics +decongestants	Excellent, unknown follow up
Egemen et al, 2012	15	4	F	L V3	Secondary	CPA Lipoma	Carbamazepine	Excellent, unknown follow up
Figueiredo et al, 1989	9	0.25	M	L V1	Secondary	AVM	Clipped, Coagulated, Divided	Excellent, unknown follow up
Gazzeri et al, 2006	5	0.2	F	R V2-3	Secondary	Tongue Piercing	Removal of stud	Excellent at 1 year
Grande-Martin et al, 2015	1.2	0.2	M	R V2	Secondary	CPA arachnoid cyst	Open fenestration of cyst	Excellent, unknown follow up
Guttall et al, 2009	9	2	F	L V2	Secondary	Epidermoid cyst	Resection	Excellent at 1 year
Hung et al, 2013	12	0.1	F	R V3	Secondary	Epidermoid cyst	Resection	Excellent at 1 year
Kato et al, 1995	8	0.5	F	R V3	Secondary	CPA Lipoma	Resection	Excellent at 3 months, continuing sensory loss, transient hearing loss, tinnitus, horizontal nystagmus
Koul et al, 2009	8	0.33	F	R V2	Classical	Anterior inferior cerebellar artery	Gabapentin	Partial - Referred to NSG
Lena et al, 1994	10	5	F	L V2	Secondary	Choristoma	Resection	Excellent at 1 year, transient hypesthesia
Levitt et al, 2011	16		F	L V2	Secondary	AVM (dx age 15 months)	Embolization	Excellent at 8 months, occasional dysesthesias
Liapounova et al, 2017	11		M	R V1	Secondary	Capillary telangiectasia	Carbamazepine	Excellent at 1 year
Lopes et al, 2002	6	1.5	M	L V2	Secondary	Tonsilitis	Tonsillectomy	Excellent at 6 months
	12	0	F	L V2	Idiopathic		Carbamazepine + Gabapentin	Excellent at 4 months
Marshall et al, 1977	5	0	F	L V1-3	Secondary	Embryonal rhabdomyosarcoma		Died 3 months later
Mason et al, 1991	17		F	L V2-3	Classical	Veins	MVD at age 6	Excellent at 2 years, gradual improvement
Matoth et al, 2001	14		F	R V1-3	Idiopathic		Tincture of Belladonna	Excellent at 10 weeks
Ochoa et al, 2020	11		M	R V2-3	Secondary	Dermoid cyst	Resection	Excellent at 2 years

Pillay Smiley et al, 2016	1.2		F	R V1-3	Secondary	Pilomyxoid Astrocytoma	Proton beam radiation	Excellent at 4 months
Raieli et al, 2001	8	2	M	R V3	Secondary	CPA Lipoma	Carbamazepine	Good at 2 years, few low intense episodes
Ramanathan et al, 2007	8	2	F	R V1-3	Secondary	Maxilla-mandibular hypoplasia, reoccurrence after distraction	Carbamazepine and Clonazepam	Excellent at 3 months
Roski et al, 1982	10	4	M	R V1-2	Classical	Petrosal vein	MVD	Excellent at 2 years
Rossi et al, 2013	9		M	R V2	Idiopathic	Associated SUNA & Norrie disease	Lamotrigine + Oxcarbazepine	Excellent, unknown follow up
Sandyk, 1980	16	4	M	L V3	Secondary	Hydrocephalus secondary to idiopathic aqueduct stenosis	VPS	Excellent at 3 months
Solth et al, 2008	11	3	M	R V2	Classical	Superior cerebellar artery	MVD	Excellent at 6 months
Watanabe et al, 2002	17		F	R V2	Secondary	Lymphoma	Gamma Knife	Excellent, unknown follow up
White et al, 2015	14	0.5	F	R V2	Secondary	Extracranial Rhabdomyomatous Mesenchymal Hamartoma	Resection	Excellent at 3 months
Yue, 2004	10 to 14		11 M 7 F	10 V2 8 V3	All 18 Idiopathic		Percutaneous Glycerol Rhizotomy. 14 after 1st, 2 after 2nd	Greater than 50% relief achieved and maintained in 85% at 1 year, 77% at 2 years, 72% at 3 years. 22.2% recurrence rate at 13 months. 2 had paresthesias, resolved at 19 months