World Neurosurgery: Case Reports

Surgical Management of Trigeminal Neuralgia in Children

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Number of tables and/or figures = 1 table; 2 figures
Number of videos = 1

Key Words: microvascular decompression in children; pediatric facial pain; trigeminal neuralgia

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This is the author's manuscript of the article published in final edited form as:

Chicoine, N. H., Yaacoub, A. P., Jea, A., & Raskin, J. S. (2018). World Neurosurgery: Case Reports Surgical Management of Trigeminal Neuralgia in Children. World Neurosurgery. https://doi.org/10.1016/j.wneu.2018.10.074

Abstract

BACKGROUND Trigeminal Neuralgia (TN) is a well-recognized facial pain syndrome. Discrete forms with disparate pain symptoms include TN1 and TN2, however, atypical facial pain includes neuralgiform pain along a spectrum. The majority of TN is diagnosed in the adult population. Case reports and series in children present TN as a similar diagnosis which can be similarly treated as in adults. This manuscript reviews pertinent literature and presents two pediatric TN cases successfully treated with microvascular decompression (MVD).

CASE DESCRIPTION Two pediatric patients (ages 12 and 15) were identified with TN that was refractory to previous medical therapy. The patients were both deemed appropriate surgical candidates and were treated with MVD to manage their TN. TN compression was arterial in both cases and involved portions of the anterior inferior cerebellar artery. Patient 1 was found to be pain-free 6 months following the procedure. Patient 2 was pain-free immediately following the procedure and has since been weaned off preoperative symptomatic management. The most recent follow-up is 12 and 8 months, respectively, with continued pain freedom.

CONCLUSIONS There are few reports on the effectiveness of MVD in the pediatric population for the management of TN. Supporting literature, as well as the cases presented, demonstrate that MVD is an effective treatment for pediatric patients in managing their TN. Furthermore, there appears to be minimal side effects, with excellent pain relief with MVD in this patient population.

Trigeminal neuralgia (TN) is a disorder characterized by recurrent paroxysmal attacks of intense unilateral facial pain occurring in the distribution of the trigeminal nerve.[1] The first detailed description of TN was provided by John Locke in 1677. He observed this condition while treating the countess of Northumberland, wife of the English Ambassador who experienced intermittent episodes of intense face and jaw pain. In 1756, Nicalaus André further described the condition giving the name "Tic Doulourex" to characterize the facial convulsions that accompany the severely painful episodes. It wasn't until 1779 that John Hunter first recognized the disease as a form of nervous disorder.[2] Present day descriptions of TN in adults have designated classic (TN1) and atypical (TN2) forms of the disorder.

Patients with TN1 complain of unilateral, episodic, lancinating pain in the one or more subdivisions of the trigeminal nerve. TN1 is caused by patient-specific somatosensory triggers. The etiology of TN1 remains unknown, although commonly there is compression of the root entry zone of the trigeminal nerve via an artery or vein, most commonly the superior cerebellar artery.[1,3] Over time, chronic compression leads to demyelination which can lead to ectopic impulses and ephaptic neurotransmission. It is believed that ephaptic neurotransmission alters afferent neurological input, resulting in the disinhibition of the pain pathways in the spinal trigeminal nucleus.[3-5]

Atypical TN (TN2) is characterized by similar symptoms to classic TN with persistent burning facial pain of moderate to severe intensity.[1] Etiologies of atypical TN include herpes zoster, posttraumatic neuropathy, multiple sclerosis, space occupying lesion such as a tumor, cyst, or lipoma[6] among others.[7-12]

TN occurs in 4–13/100,000 people,[13,14] and typically affects the older adult population, with a mean onset age of 63.[15] TN cases occur before age 20 in 1–1.5% of diagnoses, which may reflect a true lower incidence in children or may be confounded by a lack of diagnostic suspicion in young patients.[16,17] In a series of seven patients with mean age of symptom onset at age 19.6 years, patients were more likely to be female, were unlikely to have first division trigeminal pain, and had poorer pain outcome than adult populations.[17]

Initial management of TN is with symptomatic management; there are no disease modifying medical treatments. Carbamazepine is a common first-line option with oxcarbazepine, baclofen, and lamotrigine as alternatives.[18] Choice of medical therapy in pediatric patients include carbamazepine, phenytoin, baclofen, sodium valproate, sumatriptan, and acyclovir. [19] In patients who are refractory to medical therapy, surgical treatment can be a viable option.

The surgical interventions used to treat TN include microvascular decompression (MVD), stereotactic radiosurgery (SRS) [20-23] or rhizotomy with radiofrequency thermocoagulation, mechanical balloon compression, or glycerol injection. [24-28] In this paper we report our experience in two cases of pediatric TN that were successfully treated using MVD technique.

Case 1

The patient is a 15-year-old female who presents with medically intractable TN. She started developing episodes of facial pain at 3 years old which was refractory to medical management. Her symptoms were described as an episodic, right-sided, lancinating facial pain located in the V2 and V3 distributions. These episodes were exacerbated by head movement and brushing hair on the side of her face which prompted her to cut her hair short. Her pain episodes initially involved only the right side of the face. However, the pain episodes have recently progressed to involve the left side of the face. Interestingly, the left-sided facial pain will occasionally occur immediately following a right-sided episode and never occurs independently. The facial pain episodes were frequently followed by a headache and would awaken the patient from sleep. She has multiple episodes throughout the day. Occasionally, warm compresses or distraction provided temporary relief from the pain.

The patient was placed on carbamazepine but developed excessive somnolence with its use. Phenytoin provided some relief initially but lost effectiveness over time and caused an elevation in the patient's liver enzymes. She was started on oxcarbazepine and gabapentin, but these also have become ineffective over time.

The patient's past medical history includes type 2 diabetes, Asperger's syndrome, asthma, anxiety, depression, and obsessive-compulsive disorder (OCD). She did note that flares of anxiety, depression, or OCD would worsen her symptoms of TN. An MRI workup revealed a torturous anterior inferior cerebellar artery on the right side in the vicinity of the dorsal root entry zone (DREZ) of the trigeminal nerve (Figure 1). The patient was determined to be a good candidate for MVD.

A right retromastoid craniectomy was performed with the patient in a left lateral decubitus position. The cerebellum was retracted medially and inferiorly to access the cerebellopontine angle. The petrosal vein was identified which was then coagulated and divided. The facial and vestibulocochlear nerves were identified, and the trigeminal nerve located just medial to that within the field of view. We carefully dissected circumferentially around the trigeminal nerve. An indentation was found on the inferior aspect of the nerve away from the DREZ. This was associated with a loop of the anterior inferior cerebellar artery, which was as the radiology suggested throughout the surgery. Once the trigeminal nerve was dissected away from the artery, Teflon balls were placed between the artery and nerve.

The patient had an uncomplicated postoperative course, except for occasional migraines and mild muscle stiffness around the incisional site. She had no further episodes of facial pain and was weaned from preoperative TN medications by 5 months following the procedure.

Case 2

A 12-year-old female presented with several episodes of severe facial pain that occurred over the course of three weeks. The patient's pain was typical of left type 1 TN that was described as intermittent, explosive attacks of lancinating pain distributed primarily along the V3 branch of the left trigeminal nerve with some involvement of V1 and V2. The affected areas included the jaw, cheek, ear, temporal, and preauricular regions. The trigger mechanism for these episodes included chewing, light touch from slight breeze or raindrops. She also experienced pain with bumps or vibration while riding in a car.

She was initially treated with a course of prednisone that provided only minimal relief. She also received several combinations of therapies including indomethacin prochlorperazine, and chlorpromazine all were ineffective in reducing the frequency and intensity of the pain episodes. Carbamazepine, naproxen, gabapentin or valacyclovir also did not yield any benefit.

These episodes of TN were severely limiting, preventing her from attending school or participating in cheerleading. Her past medical history was unremarkable. Physical examination did not reveal any significant neurological findings other than notable allodynia around the left temporal region extending into the mandibular and maxillary region. MRI with and without contrast revealed an area of possible trigeminal nerve compression at the dorsal root entry zone involving a loop of blood vessels, likely the Labyrinthine artery, a named branch of the anterior inferior cerebellar artery (AICA) (Figure 2). The patient was referred to neurosurgery for a left retrosigmoid craniectomy and microvascular decompression.

The patient was placed in a three-quarter prone position, facilitating a retrosigmoid approach to access the cerebellopontine angle. The retractor system was incrementally advanced until the superior petrosal vein was identified. This was coagulated and cut in two places as there were two tributaries. The trigeminal, facial, and vestibulocochlear nerves were then identified. There was no arterial compression of the left trigeminal nerve; however, a circumferential venous complex that was compressing the nerve was identified which was not identifiable on imaging. This was dissected from the trigeminal nerve. The shoulder and axilla of the trigeminal nerve were inspected, and no other compressive areas were identified. Since a compressive vascular lesion was identified neurolysis was not performed. At this point, the nerve was completely decompressed and Teflon pledgets were placed to prevent further vascular compression (video).

Following the procedure, the patient had uneventful recovery and she experienced no further episodes of neuralgia. She was able to discontinue all medications and all provocative maneuvers that previously elicited pain would no longer trigger pain episodes. The patient returned to school, resumed previous extracurricular activities, and continued to remainpain-free at 6 months postoperatively.

Discussion

TN is an uncommon diagnosis and can be challenging to treat in the pediatric population. Most descriptions of pediatric TN in the literature are consistent with Type 1 TN which is similar to the experience at our institution. Treatment is crucial as the scarcity of pain-free episodes may cause performance decreases in essential activities.[19] Medical management of patients with TN begins with pharmacological intervention. In a study by Railei, an 8-year-old patient was treated for pediatric trigeminal neuralgia with 15mg/kg/day of carbamazepine.[29] Combination treatment with carbamazepine has also been utilized. In a study by Lopez, a 12-year-old female patient was treated with 1200mg/day of gabapentin in combination with 800mg/day of carbamazepine.[3] High doses of amitriptyline, phenytoin, baclofen, sodium valproate, sumatriptan, and acyclovir with a 3-

day dose of methylprednisolone have also be used for treatment in pediatric cases.[3]

Surgical intervention is indicated when pain is refractory to medical therapy. Approximately one-third of patients do not achieve adequate symptom reduction with medical therapy alone.[30] It should be determined as soon as possible if surgical intervention is necessary as prolonged symptoms of TN can eventually lead to disinhibition of pain pathways in the spinal trigeminal nucleus causing persistent background pain and possible progression from TN1 to TN2. Earlier decision for surgery is even more critical in the pediatric population in which low suspicion of the disease may lead to delays in diagnosis and application of appropriate therapies. Moreover, in the pediatric population with TN1 and documented compression by imaging, MVD may be indicated before medical treatment according to Bender et al.[20]

Table 1 tabulates eight manuscripts reporting pediatric TN; four of five manuscripts commenting on offending vessels report venous compression. This suggests a high proportion of venous compression in pediatric TN compared to the adult literature. Our series demonstrates one of two cases with clear circumferential venous constriction at the trigeminal root entry zone with video demonstrating surgical decompression. Surgical procedures used to treat TN include MVD, SRS, rhizotomy with radiofrequency thermocoagulation, mechanical balloon compression, or chemical (glycerol) injection. [24-28] The American Academy of Neurology and the European Federation of Neurological Societies concluded that MVD, rhizotomy, and gamma knife radiosurgery are potentially effective procedures for this condition.[18] In the pediatric population, Solth found success utilizing MVD in an 11-year-old patient, whose symptoms were caused by nerve compression via the superior cerebellar artery and anterior inferior cerebellar artery.[31] In a study of female pediatric patients at John Hopkins, 5 of the 6 cases also experienced complete pain relief via MVD.[16] Complications that have been associated with MVD include CSF leaks, infection, diplopia, meningitis, cerebellar lesions, and cranial nerve deficits.[16,18] Resnick et al. expressed some hesitancy for this procedure due to the involvement of varying vessels and increased recovery time.[15]Although the success of MVD in our series was observed over relatively short follow-up periods, the durability of the results following MVD have been generally favorable in the literature. Of 52 pediatric patients among 9 series, complete response to MVD was observed in 33 cases while partial response was seen in 17 cases at follow-up times ranging from 6 months to 18 years.[4,12,15,16,19,20,23,28,31]

Other surgical procedures used in the treatment of pediatric trigeminal neuralgia include percutaneous balloon compression, and peripheral glycerol injection.[19] The percutaneous balloon compression performed by Baabor demonstrated high success rate, low recurrence rate, low cost, and a low morbidity rate.[32] Peripheral glycerol injection has been previously tried for the relief of facial neuralgia, and is a less complex procedure, as it can easily be performed and repeated as necessary. Furthermore, there is no risk of facial sensory loss as compared to other procedures.[19]

Conclusions

MVD is a useful treatment option for those diagnosed with TN. It is a widely accepted first line surgical therapy for adult patients and has been reported in the pediatric population as an effective treatment modality for facial pain reduction. Earlier decision for surgical intervention should be considered for pediatric patients with TN1 and documented neurovascular conflict. The enclosed video demonstrates the surgical technique of an MVD in one of the cases with neurovascular conflict herein presented. Our cases, as well as those reviewed in this paper, support the use of MVD in pediatric populations suffering from TN. The procedure shows promising success rates of pain reduction and treatment of the disease, with minimal side effects.

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Figure Legends:

Figure 1: Axial T1 post gadolinium demonstrating a branch of anterior inferior cerebellar artery (AICA) (white arrow) coursing superiorly along the dorsal root entry zone of the right trigeminal nerve (black arrow).

Figure 2: Coronal and axial T2 FIESTA demonstrating a redundant course of the Labyrinthine artery medial and inferior to the left Trigeminal Nerve (arrows point to vessel in plane, dotted line represents vessel course out of plane).

Video: The video commences following a left retrosigmoidectomy and dural opening. The cerebellum is retracted. The superficial petrosal venous complex is coagulated and divided. The trigeminal nerve is identified. A venous complex is identified circumferentially around the trigeminal nerve, which is released with microsurgery. Teflon plegets are placed to decompress the neurovascular structure.

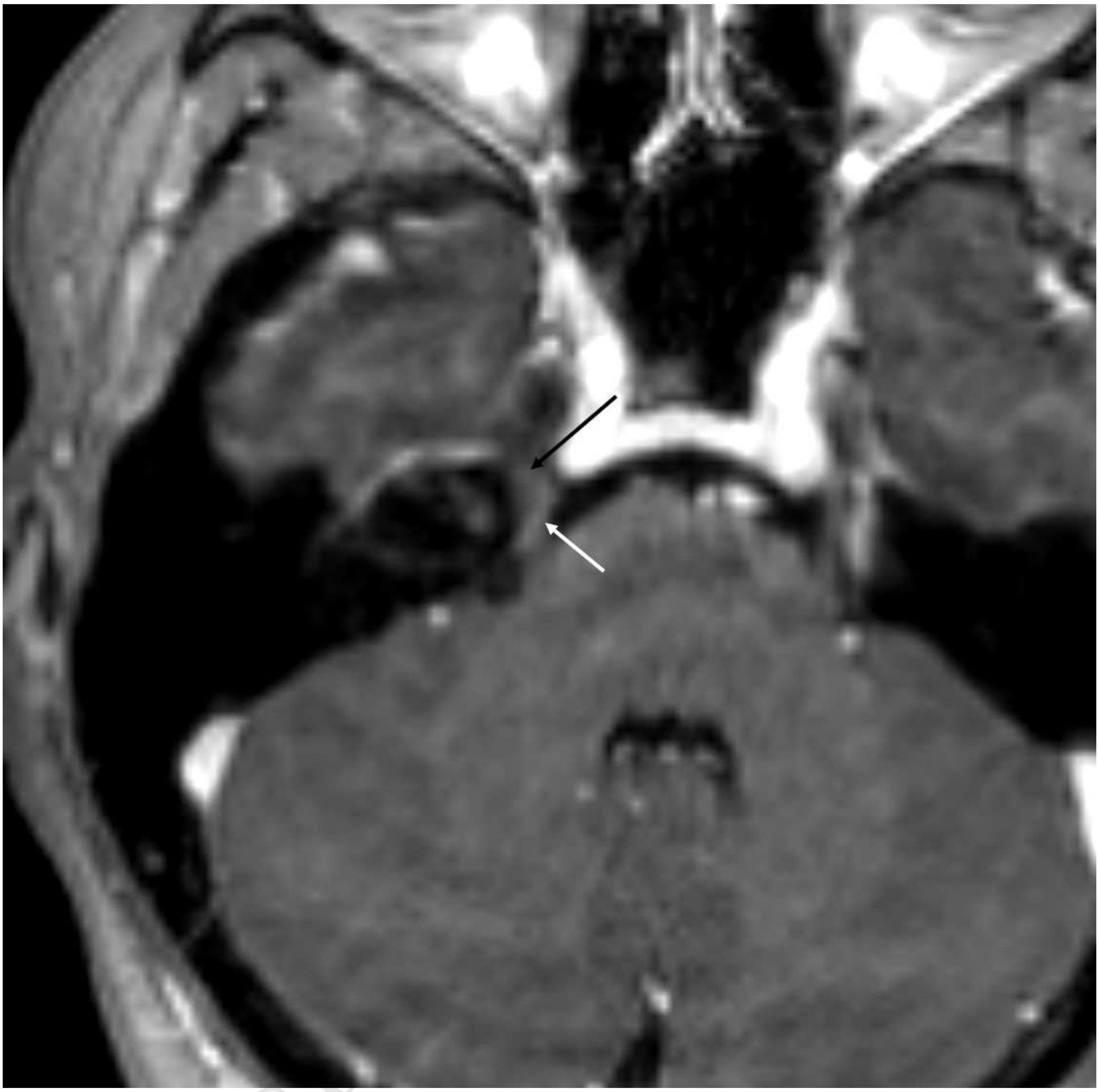
Table 1. Previous Reported Cases of Pediatric Trigeminal Neuralgia Treated via Surgery

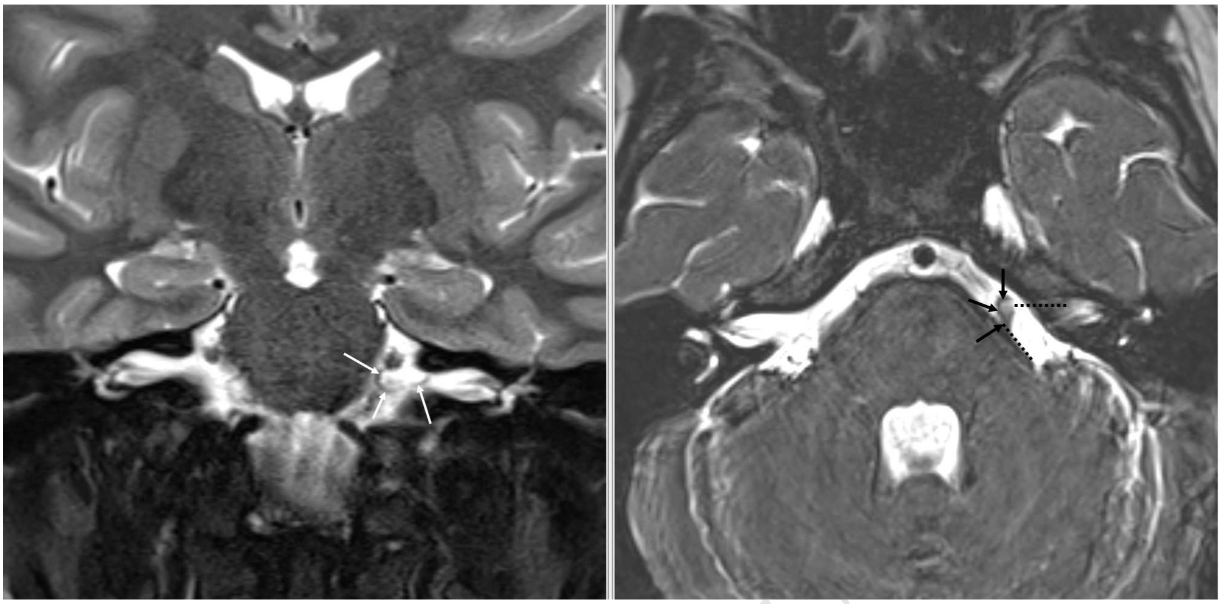
Author	Number of Patients	Age of Treatment	Sex	Treatment Modality	Response	Duration of relief
Barclay	1	11	M	Division of V2 and V3	Success	17m, ongoing (follow up duration after this is unknown)
Bender	5	3-18 (mean age 11.7)	F	MVD	Complete Success (5 of 6 cases; partial in 1 case= >50% reduction in attack frequency + severity)	Ongoing (Follow up duration: 9.1-24.8m)
Childs	2	9,12	M	MVD	Success	13m, Ongoing (Follow up duration after this is unknown)
Harris	1	4.5	F	Glycerol alcohol injection	Success	7m, ongoing (Follow up duration after this is unknown)
Mason	1	Aprx 7 (dx= 13 month)	F	MVD	Success	6m, ongoing

						(Follow up: 2 years)
Resnick	22	29.1 +/- 12.8 yr (onset at 13.6 +/- 4.6 yr)	7M, 15F	MVD	Initial: 16 complete success; 3 partial; 3 operative complications Follow Up: 9 complete success,2 partial, 10 <75% reduction in pain	12-225m, ongoing (Follow up duration after this is unknown)
Roski	1	10	M	MVD	Success	2 years, Ongoing (Follow up duration after this is unknown)
Solth	1	11	M	MVD	Success	6m, Ongoing (Follow up duration after this is unknown)
Yue	18	10–14	11M,7F	peripheral glycerol injection	72% Complete relief w/o medication 11.1% complete relief but use medication 11.1% pain relief <	3 years, ongoing (Follow

					50% 22.2% recurrence 2-13m	up duration after this is unknown)

MVD = microvascular decompression.





TN = Trigeminal neuralgia

MVD = microvascular decompression

SRS = stereotactic radiosurgery

OCD = obsessive-compulsive disorder

DREZ = dorsal root entry zone

AICA = anterior inferior cerebellar artery