

Surgical Management of Trigeminal Neuralgia (Course Notes)

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Half-day course: *Medical and Surgical Management of Trigeminal Neuralgia and Cluster Headache.*

Learning Objectives

- 1) *Gain knowledge of the surgical management options for Trigeminal Neuralgia.*
- 2) *Gain knowledge of timing and choice of surgical options.*

Disclosure Statement

The author has no commercial or financial interests in products or services included in this course outline or presentation.

Surgical Management of Trigeminal Neuralgia

The neurosurgical management of trigeminal neuralgia must incorporate a strong working knowledge of the disease diagnosis and course, effective use of medications and all available surgical procedures. What follows is the general framework we consider in approaching patients referred for neurosurgical treatment of trigeminal neuralgia at our centre.

Clinical History

The clinical features of trigeminal neuralgia have been well defined, and details should be determined for each patient. Approximately half of the patients will have a distinctly memorable onset of trigeminal neuralgia, vividly recalling their first attack. One third will have experienced preceding “prodrome” symptoms of tingling or aching in the affected area weeks or months before. The patient should also be questioned about the nature of their original pain, including its distribution, character, duration, initiating triggers and associated symptoms during or between pain attacks (eg. numbness, tingling, aching, ear pain). The subsequent disease course progresses with exacerbations that increase in severity, frequency and duration, while remissions become less frequent and shorter. It should be established whether the trigeminal neuralgia attacks have changed over time, with the development of advanced, atypical or clustered attacks. A complete history of treatment interventions and effects should be determined. Included with these, details of original diagnosis may reveal delays or errors as warned against in the original descriptions of the disease over 200 years ago. Still today, at least half of all patients we see with a clear diagnosis of trigeminal neuralgia had been subjected to earlier diagnostic errors and misguided, ineffective treatments ranging from dental extractions and root canals to tongue biopsies and psychiatric hospital admission.

The diagnosis of trigeminal neuralgia is confirmed by the clinical history and the absence of other neurological or craniofacial disorders. Diagnostic imaging is not necessary for diagnosis and we do not rely on this to identify the causative neurovascular compression. However, 1 to 4% of trigeminal neuralgia patients will have relevant co-pathology such as benign tumors. MRI is mandatory whenever



there are findings of neurological deficits such as facial numbness or question of related multiple sclerosis.

Nomenclature and Pathophysiology

The appropriate treatment of trigeminal neuralgia, or *tic douloureux*, relies on a correct diagnosis. Unfortunately, there is often imprecision applied in the definitions of facial neuralgic pains. Typical trigeminal neuralgia has been defined in the preceding section, and has overlapping clinical and pathophysiological features with atypical trigeminal neuralgia. This latter condition is defined as typical trigeminal neuralgia with additional associated feature of some constant, aching, burning pain in the same distribution. Medical and surgical treatment for both conditions are the same, although the use of rhizotomy procedures may be more inclined to aggravate the “neuropathy” features of atypical trigeminal neuralgia. These conditions must be differentiated from trigeminal neuropathy pain secondary to tumors, nerve injury, stroke, or of unknown etiology (i.e. idiopathic trigeminal neuropathy). Unfortunately, the term trigeminal neuralgia may be inappropriately applied to all of these neuropathic conditions. Idiopathic or atypical facial pain should not raise diagnostic confusion, although these patients often seek diagnosis of trigeminal neuralgia to gain access to the surgical treatment options that would however prove ineffective or hazardous to their condition. Trigeminal neuralgia due to multiple sclerosis deserves a special label such as multiple sclerosis-related trigeminal neuralgia, although it has the same symptomatology as typical trigeminal neuralgia.

The etiology of trigeminal neuralgia has been established over the last century. Observations by early neurosurgeons Cushing, Dandy and Gardner introduced the idea that trigeminal neuralgia may be caused by blood vessels contacting with the trigeminal nerve root, as seen at operation seeking to relieve trigeminal neuralgia pain by cutting the nerve. The formalization of this neurovascular compression concept and introduction of effective neurosurgical alleviation of the culprit irritation by microvascular decompression surgery was pioneered by Peter J. Janetta.

Several surgical series by experienced neurosurgeons have demonstrated the universal relationship between typical trigeminal neuralgia and pulsatile neurovascular compression upon the



proximal trigeminal nerve root. Anatomical analyses of cadavers has further supported the neurovascular compression concept as have high-definition special MRI studies. The pathophysiological mechanism underlying the development and course of trigeminal neuralgia appears to involve peripheral pathology (i.e. neurovascular compression) leading to secondary changes centrally (i.e. trigeminal nucleus hyperactivity). This is supported by the effectiveness of centrally acting anti-convulsant medications as well as electrophysiological studies in patients with the analogous neurovascular compression disorder hemifacial spasm. Alleviation of the culprit vascular compression by surgically repositioning the vessel has the greatest success rate of all surgical procedures for trigeminal neuralgia. Conversely, multiple sclerosis-related trigeminal neuralgia is caused by demyelinating plaques involving central pathways of the trigeminal nerve, and therefore not amenable to microvascular decompression surgery.

Management Strategies

Treatment of trigeminal neuralgia over the last 3 centuries has included an assortment of ineffective and bizarre interventions, such as hemlock, arsenic, tooth extractions, purging, bleeding, carotid ligation, abdominal steam, and others. The "success" ascribed to these remedies was likely due to spontaneous remissions, which are frequent in the early course of disease. References to the seizure-like nature of the trigeminal neuralgia led Bergouignan to prescribe the anticonvulsant phenytoin in 1942, which became the first effective medical treatment for trigeminal neuralgia. This was followed by the breakthrough introduction of carbamazepine by Blom in 1962. Over 80% of patients initially respond to medical treatment, although between 10% and 20% develop early intolerance to these drugs. Among those maintaining long-term treatment, at least one third experience serious dose-related side effects.

Trigeminal neuralgia pain becomes refractory to any combination of medical treatments in over 50% of patients, and neurosurgical interventions are required. In general, the four most common rhizotomy procedures (glycerol, balloon-compression radiofrequency and radiosurgery) offer similar results. Long-term control is achieved in approximately 50%, but may be associated with sensory loss, and risk of deafferentation complications such as paresthesias, dysesthesias, or *anesthesia dolorosa*. Alternatively, microvascular decompression surgery offers a greater potential to cure trigeminal



neuralgia while maintaining normal nerve function, although it entails a more invasive procedure. If trigeminal neuralgia recurs after initially successful surgical intervention of any kind, medical treatment may prove more effective than preoperatively. However, some patients do undergo multiple procedures for the treatment of tic, and rare cases become refractory to any standard medical and surgical interventions.

The timing of interventions is tailored to each patient based on their own disease experience and comparative assessment of treatment options. Physicians must not underestimate the intensity of trigeminal neuralgia pain, widely described as the most painful of conditions, nor disregard a “pain free” patient’s fear of the inevitable next exacerbation. Even patients newly presenting with medically controllable trigeminal neuralgia should be introduced to all the treatment options and the potential for severe and sudden future exacerbations. Furthermore, patients with trigeminal neuralgia should have a “game plan” regarding treatment for future attacks, including emergency medical and/or surgical treatments.

Medical Management

The first line of treatment are anti-convulsants such as carbamazepine or gabapentin, in a slowly escalating dose. Pain relief can usually be achieved early in the disease course. The lowest effective drug dose is then maintained until the patient is completely pain free for at least two weeks, and then a trial of gradual dose reduction and discontinuation. Advanced trigeminal neuralgia cases will require higher drug doses and many patients experience dose-related side effects of fatigue, concentration and memory decline and balance incoordination. Other anti-convulsants may be tried, including phenytoin, oxcarbazepine and pregabalin. Medical treatment of severe trigeminal neuralgia often requires a combination of two or more medications, including the synergistically acting baclofen. Many patients however will strongly consider the option of surgery rather than embark on lifelong polypharmacotherapy. It must be appreciated that over 50% of trigeminal neuralgia sufferers will eventually require a surgical procedure, when their medical treatment fails to provide adequate pain relief or causes intolerable dose-related drug side effects. We consider the surgical options for any



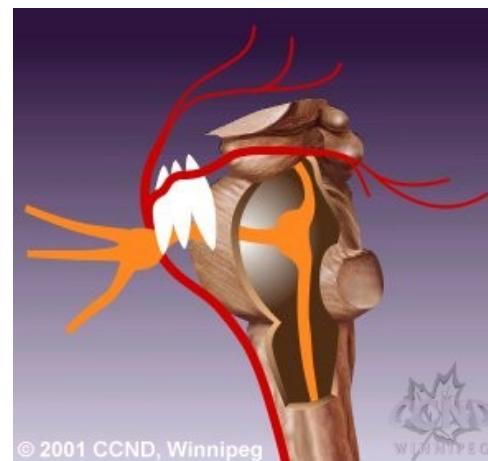
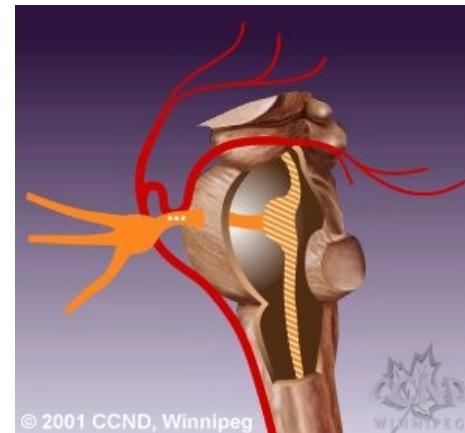
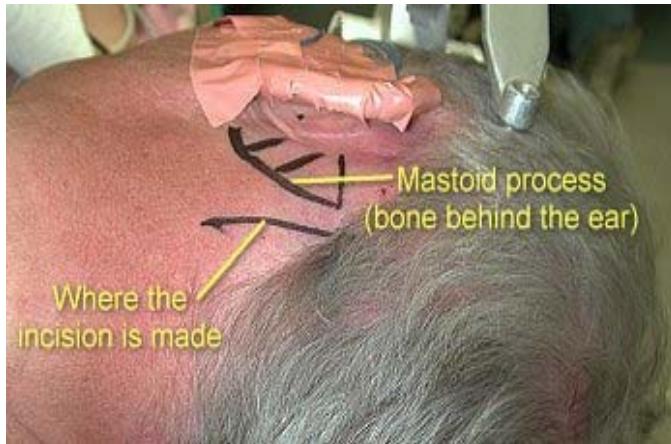
patient unsatisfied with their medical treatment for reasons of continuing pain, drug side effects or aversion to ongoing medication use. A point of caution however, is the need to prescribe these medications carefully. Starting at a low dose and gradually increasing allows for enhanced drug metabolism and reduced side effects, improving the chances a patient will reach an effective dose before giving up on the drug.

Emergency treatment of trigeminal neuralgia deserves special mention. Severe exacerbations may not permit the patient opportunity to sufficiently increase their medication intake to control the pain. Emergency room administration of intravenous phenytoin can be very effective, followed by a maintenance dose. Many emergency room patients with trigeminal neuralgia are often given opiate analgesics, which typically are not effective in reducing trigeminal neuralgia pain and should not be prescribed for outpatient trigeminal neuralgia treatment. If emergency room interventions are ineffective, hospital administration for additional intravenous medications such as the combination of lidocaine, ketamine and fentanyl is warranted. Local anesthetic blocks can be administered to buy time, but temptations to perform peripheral ablative procedures (e.g. alcohol injection) should be avoided, and more definitive treatment measures pursued. We perform a few emergency percutaneous rhizotomies and emergency microvascular decompression surgeries each year, with most gratifying results.

Microvascular Decompression

Most patients with typical trigeminal neuralgia who become dissatisfied with medical treatment are candidates for microvascular decompression surgery; if they can withstand the pain attacks, they can probably get through surgery. The surgery is well tolerated by the elderly, although frail individuals or those with significant medical co-morbidities are directed to destructive procedures under local anesthetic as described in the following sections. Microvascular decompression surgery offers the advantages of attacking the presumed etiology, preserving the trigeminal nerve function, and providing the best chance for permanent pain relief. This surgery is performed through a small thumbprint-sized craniectomy behind the ipsilateral ear. Microsurgical techniques are employed to mobilize the offending

vessels off the symptomatic nerve and maintain their new position with placement of small permanent implants, such as shredded Teflon felt.



Initial pain control following surgery is achieved in over 90% of patients, with large series reporting 70% "cure" rates at 10-year followup. Success and complication rates are closely related to surgical experience and expertise. Major morbidity and death risk should be less than 0.5% and risk of hearing loss less than 1%. Intraoperative monitoring of auditory evoked potentials should be routinely employed. Permanent trigeminal numbness is also rare, and deafferentation pain including *anaesthesia dolorosa* is virtually never encountered. Failures and significant recurrence are seen in 10% to 20% of patients, often in association with severe or long-standing vascular compression, previous destructive procedures, and isolated venous neurovascular compression identified at surgery.

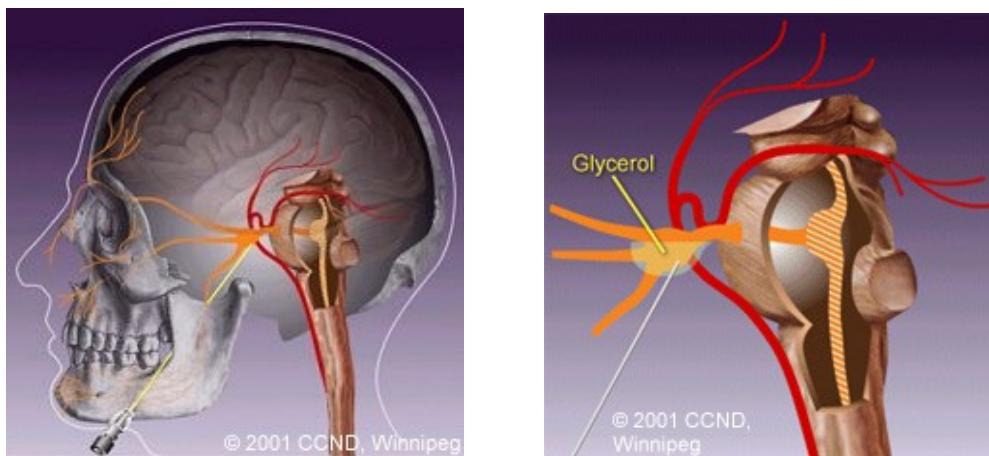
Microvascular decompression surgery aims to achieve atraumatic alleviation of the causative pathology (i.e. pulsatile neurovascular compression) resulting in reversal/correction of the neural-pain generating pathophysiology (i.e. trigeminal nucleus hyperactivity). In series by experienced neurosurgeons, the neurovascular compression is universally present and outcomes with microvascular decompression surgery are significantly superior to other trigeminal neuralgia surgeries including the incidence of “life-alternating complications”. On the other hand, results of microvascular decompression surgery have the greatest inter-surgeon variability of all trigeminal neuralgia procedures. A recent review of microvascular decompression in the USA indicated that approximately 1,500 operations are performed annually, 1300 for trigeminal neuralgia. More than half the neurosurgeons did only 1 to 3 operations per year and approximately one dozen did more than 30 procedures per year. Although overall morbidity/mortality rates were extremely small, a significant difference between high and low volume centers and surgeons was demonstrated. Other reports have also pointed out that outcomes and complication rates are closely related to surgeon experience and expertise. Similarly, failure to identify culprit neurovascular compression in patients with typical trigeminal neuralgia should be considered a failure of surgical technique and is associated with suboptimal outcome results. We have seen many examples of “failed” microvascular decompression where the culprit neurovascular compression was readily apparent and correctible at re-do surgery.

In my own series of over 400 microvascular decompressions performed over the last ten years, there have been no deaths or permanent major morbidities. One patient suffered a postoperative cerebellar venous infarct requiring surgical decompression but without permanent sequelae. One patient had intraoperative venous bleeding from a bridging vein that interrupted surgery; the microvascular decompression was successfully completed a few weeks later without difficulty. The incidence of hearing loss has been less than 1% and the postoperative rates of cerebrospinal fluid leak, infection or aseptic meningitis have been 1%, 1% and 3% respectively. Over 90% of patients experienced immediate trigeminal pain relief that has continued without need for medications in over 70% to date. The 2-hour operation is followed by an average length of hospitalization less than 3 days.

The patients are not restricted in their activity following surgery and those living far away are flying back home within one week of surgery. The convalescence time ranges between two weeks and two months.

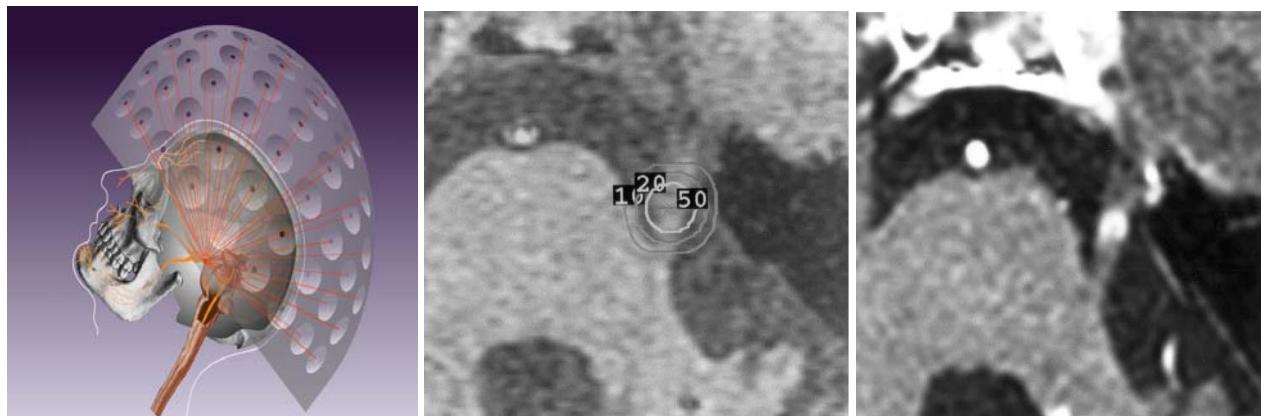
Controlled Destructive Procedures

The surgeon to Louis XIV, Maréchal, pioneered the surgical treatment for facial pain by making a wide intraoral incision to section the infraorbital nerve. In 1756, André reported superior results by destroying the nerve with caustics. Neurectomies were introduced in the 1800s, although surgical and percutaneous rhizotomies of the ganglion and trigeminal root were subsequently found to provide more long-lasting relief and preservation of some facial sensation. The most widely employed “ablative” techniques today include the percutaneous needle approaches to the gasserian ganglion and stereotactic radiosurgery.



These minor procedures avoid the risks of craniotomy, have low morbidity, and nearly no mortality risk. However, patients often experience facial numbness postoperatively, and are at risk to develop corneal anesthesia and keratitis, trigeminal motor weakness, or deafferentation pain. Permanent painful numbness (*anesthesia dolorosa*) or facial dysesthesia develops in 1% to 10% of patients treated with destructive interventions. Long-term control of trigeminal neuralgia pain is directly related to the degree of permanent postoperative numbness, as are risks of deafferentation

complications. Glycerol and GammaKnife rhizotomies performed with local anesthetic, have the lowest rate of postoperative numbness although recurrence rates are high, approximately 50% at 2 years. Radiofrequency rhizotomy, requiring conscious sedation, has the advantage of intraoperative localization to selectively lesion trigeminal divisions, although it produces greater nerve injury and associated hypalgesia, hypesthesia, and risk of deafferentation complications. Long-term pain control or reduction is reported in approximately 60 to 70% of patients. This is similar to results with balloon compression of the gasserian ganglion that is performed under general anesthesia. This injury procedure is preferred for ophthalmic division trigeminal neuralgia, as V1 injury can be achieved with corneal sensation sparing. Most recently, focused radiation to the trigeminal nerve root has become widely utilized, most commonly with GammaKnife, delivering 80 Gy maximal dose. Radiosurgery treats trigeminal neuralgia pain by partial nerve injury just like the other destructive interventions. Unlike the percutaneous needle procedures, however, pain relief usually follows a latency period of 3 weeks to 3 months. The procedure avoids needle-related complications. While a large number of publications report favourable results, only a few have provided long-term follow up and indicate one third of patients will be pain-free and off medications at 5 year followup.



A partial microsurgical rhizotomy of the trigeminal nerve root in the posterior fossa is sometimes performed and is advocated by some surgeons when they cannot find the neurovascular compression during attempted microvascular decompression surgery. The rostral third to half of the nerve root is

spared to preserve corneal sensation. The pain relief may be long lasting; postoperative deafferentiation pain is rare. Other destructive techniques include peripheral nerve injections, avulsions, and sectioning. These simple peripheral nerve procedures provide relatively short-term relief ranging from months to several years and are associated with severe sensory loss and risk for deafferentation pain. They are generally reserved for those who have sufficient medical comorbidity the other surgical procedures, such as bed-ridden palliative care patients.

Choosing The Right Surgery

The best choice for any individual patient should provide the maximum potential for long-term pain relief with the lowest risk of procedure-related side effects and complications. Unfortunately, these variables are not easily quantified so there is difficulty in weighing the merits of each option. The medical literature does provide outcome results, usually from centres with the high volumes of expertise and experience, a factor to be especially considered for technique-dependent procedures such as microvascular decompression surgery. Unfortunately, published series may be difficult to interpret and compare, in part because of inconsistency of outcome grading scores. In the recent review of a 175 studies on the four most common ablative neurosurgical techniques for trigeminal neuralgia, only 9 were found to meet adequate data quality criteria. Furthermore, no conclusions about the “best treatment” option could be reached. Physician and surgeon bias clearly plays a key role and are based on their individual experience.

The experience at our centre supports the utilization of the microvascular decompression surgery for a majority of the patients with unsatisfactory results from medical treatment for several reasons; highest long-term pain relief or cure rates, lowest risk of facial numbness and lowest risk of major morbidity. This latter item deserves further elaboration as we consider *anesthesia dolorosa* or life-long dysesthesia seen in 1 to 10% of rhizotomy patients as a major morbidity, in addition to the very small risks of needle-related complications such as intracranial hemorrhage, vascular injury and myocardial infarction reported in the literature. Conversely, risk of major morbidity with microvascular decompression

surgery in our experience has been exceptionally small. This operation, however, does require hospitalization and a period of convalescence.

Among the nerve injury procedures, we first offer GammaKnife or percutaneous glycerol rhizotomies, as these are associated with the lowest incidence of facial numbness. The GammaKnife rhizotomy is preferred if immediate pain relief is not required or if there are medical contraindications to more invasive surgical procedures (e.g. anti-coagulation, unstable ischemic heart disease). Balloon compression is the preferred rhizotomy technique for urgent relief of ophthalmic division pain.

In summary, a patient with trigeminal neuralgia should be thoroughly informed of the anticipated course of their disease and all the management options available.

Website Reference: http://www.umanitoba.ca/cranial_nerves/ccndhome.htm

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