Preface

Summary
The IRSA® (International RadioSurgery Association) Radiosurgery Practice Guideline Initiative aims to improve outcomes for typical trigeminal neuralgia patients by assisting physicians and clinicians in applying research evidence to clinical decisions while promoting the responsible use of health care resources.

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Key Words trigeminal neuralgia tic douloureux stereotactic radiosurgery Gamma Knife® linear accelerator irradiation CyberKnife

Consensus Statement

Objective
To develop a consensus-based radiosurgery practice guideline for trigeminal neuralgia treatment recommendations to be used by medical and public health professionals following the diagnosis of trigeminal neuralgia.

Participants
The working group included physicians and physicists from the staff of major medical centers that provide radiosurgery.

Evidence
The first authors (LDL/AN) conducted a literature search in conjunction with the preparation of this document and development of other clinical guidelines. The literature identified was reviewed and opinions were sought from experts in the diagnosis and management of trigeminal neuralgia, including members of the working group.

Consensus Process
The initial draft of the consensus statement was a synthesis of research information obtained in the evidence gathering process. Members of the working group provided formal written comments that were incorporated into
the preliminary draft of the statement. No significant disagreements existed. The final statement incorporates all relevant evidence obtained by the literature search in conjunction with final consensus recommendations supported by all working group members.

**Group Composition**
The radiosurgery guidelines group is comprised of neurosurgeons, radiation oncologists and physicists. Community representatives did not participate in the development of this guideline.

**Names of Group Members:** L. Dade Lunsford, M.D., Neurosurgeon, Chair; Ajay Niranjan, M.B.B.S., M.Ch., Neurosurgeon; Ron Young, M.D., Neurosurgeon; Ronald Brisman, M.D., Neurosurgeon; David Cunningham, M.D., Neurosurgeon; Christer Lindquist, M.D., Neurosurgeon; David Newell, M.D., Neurosurgeon; John C. Flickinger, M.D., Radiation Oncologist; Ann Maitz, M.S., Medical Physicist; Tonya K. Ledbetter, M.S., M.F.S, Editor; Rebecca L. Emerick, M.S., M.B.A., C.P.A., ex officio.

**Conclusions**
Specific recommendations are made regarding target population, treatment alternatives, interventions and practices and additional research needs. Appropriate use of radiosurgery to those with intractable trigeminal neuralgia following medical management may be beneficial.

This guideline is intended to provide the scientific foundation and initial framework for patients who have been diagnosed with trigeminal neuralgia. The assessment and recommendations provided herein represent the best professional judgment of the working group at this time, based on clinical research data and expertise currently available. The conclusions and recommendations will be regularly reassessed as new information becomes available.

**Stereotactic Radiosurgery**
Brain Stereotactic Radiosurgery (SRS) involves the use of precisely directed, closed skull, single fraction (one session) of radiation to create a desired radiobiologic response within the brain with minimal effects to surrounding structures or tissues. In the case of trigeminal neuralgia a relatively high dose of focused radiation is delivered precisely to the trigeminal nerve under the direct supervision of a radiosurgery team. In Centers of Excellence, the radiosurgery team is composed of a neurosurgeon, radiation oncologist, physicist and registered nurse.

**Trigeminal Neuralgia: Overview and Surgical Management**

**Background**
Trigeminal neuralgia (TN), also known as tic douloureux, is a pain syndrome recognizable by patient history alone. The condition is characterized by intermittent unilateral facial pain. The pain follows the unilateral (>95%) sensory distribution of trigeminal nerve (V), typically radiating to the maxillary (V2) or mandibular (V3) area. Ophthalmic division (V1) pain alone occurs in <5% patients. Physical examination findings are typically normal, although mild light touch or pin perception loss has been described in the central area of the face. Significant sensory loss suggests that the pain syndrome is secondary to another process, and requires high-resolution neuroimaging to exclude other causes of facial pain.

**Epidemiologic Features**

**Sex**
Male-to-female ratio is 2:3.

**Age**
The average age of pain onset in idiopathic TN typically is the sixth decade of life, but it may occur at any age. Symptomatic or secondary TN tends to occur in younger patients. Trigeminal neuralgia caused by arterial
compression is more likely in older patients.

**Pathophysiology**

The mechanism of pain production remains controversial. One theory suggests that peripheral injury or disease of the trigeminal nerve increases afferent firing in the nerve, perhaps by ephaptic transmission between afferent unmyelinated axons and partially damaged myelinated axons; failure of central inhibitory mechanisms may also be involved. Pain is perceived when nociceptive neurons in a trigeminal nucleus involve thalamic relay neurons. Blood vessel-nerve cross compression, aneurysms, chronic meningial inflammation, tumors or other lesions may irritate trigeminal nerve roots along the pons. Uncommonly, an area of demyelination, such as may occur with multiple sclerosis, may be the precipitant. In some cases no vascular or other lesion is identified, rendering the etiology uncertain. Development of trigeminal neuralgia in a young person (<45 years) raises the possibility of multiple sclerosis, which should be investigated. Lesions of the entry zone of the trigeminal roots within the pons may cause a similar pain syndrome. Thus, although TN typically is caused by a dysfunction in the peripheral nervous system (the roots or trigeminal nerve itself), a lesion within the central nervous system (1–8%) may also cause similar problems.

**Causes**

Some patients’ conditions are idiopathic, but compression of the trigeminal root entry zone by blood vessels (especially branches of the superior cerebellar arteries or venous channels) or tumors may cause pain.

**Clinical Presentation**

**Nature of Pain**

Pain is stabbing or an electric shock-like sensation and is typically quite severe. Pain is brief (few seconds to 1–2 minutes) and paroxysmal, but it may occur in volleys of multiple attacks. Pain may occur several times a day; patients typically experience no pain between episodes.

**Distribution of Pain**

Pain is unilateral (rarely bilateral). One or more branches of the trigeminal nerve (usually maxillary or mandibular) are involved. It is rare that pain is simultaneously bilateral, but pain on the contralateral side is seen in about 5% of patients with TN and about 20% of patients with trigeminal neuralgia and multiple sclerosis.

**Trigger Points**

Various triggers may commonly precipitate a pain attack. Light touch or vibration is the most evocative. Activities such as shaving, face washing or chewing often trigger an episode. Over years periods of remission are less and less and severity of pain increases.

**Course**

The disease course is usually one of clusters of attacks that wax and wane in frequency. Over the years recurrences become sharper and exacerbations longer. Some patients report that their exacerbations commonly occur in the fall and spring.

**Imaging Studies**

Patients with characteristic history and normal neurologic examination may be treated without further workup. An MRI scan with and without contrast is essential to rule out the presence of a tumor, arteriovenous malformation, or multiple sclerosis, all of which can cause trigeminal neuralgia in a small number of patients. In addition, skull base conditions such as maxillary sinusitis may mimic symptoms of TN.

**Management Options**

**Medical Management**
The goal of pharmacologic therapy is to reduce pain. Carbamazepine is regarded as the most effective medical treatment. Oxcarbazepine (Trileptal) is often effective like carbamazepine, but has fewer side effects. Additional agents that may benefit selected patients include baclofen, gabapentin (Neurontin), and Klonazepin. The combination of carbamazepine and baclofen may provide relief from episodic pain. Other anticonvulsants, including phenytoin, reportedly are beneficial in some patients. Topiramate may be beneficial in refractory cases; however, controlled studies have not been performed. Some patients are not helped by these medications, experience breakthrough pain, or suffer with undesirable side effects. In such cases a referral to a neurosurgeon is indicated.\textsuperscript{57}

**Surgical Management**

Prior to considering surgery, all eligible patients should have an MRI, with close attention being paid to the posterior fossa. Imaging is performed to rule out other causes of compression of the trigeminal nerve such as mass lesions, large ectatic vessels, or other vascular malformations. Thin section MRI (with contrast) may be helpful (although not with 100% accuracy) in identifying blood vessel contact and/or compression of the trigeminal nerve. The surgical options for TN include peripheral nerve blocks or ablation, gasserian ganglion and retrogasserian ablative procedures, microvascular decompression (MVD) and stereotactic radiosurgery.\textsuperscript{8} Percutaneous transosseous techniques include radiofrequency trigeminal electrocoagulation, glycerol rhizotomy and balloon microcompression.\textsuperscript{8,36,61,64} Microvascular decompression is often preferred for younger patients with typical trigeminal neuralgia. High initial success rates (>90%) have led to the widespread use of this procedure. Pain control rate after single MVD at 10 years is 64% with good results in an additional 4%.\textsuperscript{5} This procedure provides treatment of the cause of TN in many patients. Percutaneous techniques are advocated for elderly patients, patients with multiple sclerosis, patients with recurrent pain after MVD, and patients with impaired contralateral hearing;\textsuperscript{4} however, some authors recommend percutaneous techniques as the first surgical treatment for most patients.\textsuperscript{60,64} In a comparative study of three techniques, Lee et al. noted initial success rates of 96.5%, 92% and 83% for MVD, radiofrequency trigeminal electrocoagulation and glycerol rhizolysis, respectively.\textsuperscript{31} Facial dysesthesias were reported for 6.3%, 5% and 3% of patients, respectively. In a review of different techniques, Lovely and Jannetta reported excellent or good pain control for 78%, 74% and 56% of patients using MVD, radiofrequency trigeminal electrocoagulation, and glycerol rhizolysis, respectively.\textsuperscript{37} Microvascular decompression was associated with a 0.5% mortality rate and 3–29% facial numbness rates. In a study of balloon microcompression treatment of trigeminal neuralgia, 62.5% of patients achieved excellent pain control, whereas 32.5% developed recurrences (after a median interval of 51 months). Sensory dysesthesia was noted for 3% and hearing impairment was observed for 11% of patients. It is generally agreed that MVD provides the longest duration of pain relief while preserving facial sensation. In experienced hands, MVD can be performed with low morbidity and mortality. Most authors offer MVD to young patients with TN.\textsuperscript{37,63}

**Radiosurgery**

Several reports have documented the efficacy of Gamma Knife\textsuperscript{®} stereotactic radiosurgery for TN.\textsuperscript{1,3,16,18,20,26,27,29,32,35,39-42,46,56-53,56,62,68} Because radiosurgery is the least invasive procedure for TN, it is a good treatment option for patients with co-morbidities, high-risk medical illness, or pain refractory to prior surgical procedures. Radiosurgery is a good alternative for most patients with medically refractory trigeminal neuralgia, especially those who do not want to accept the greater risk of an MVD for a greater chance of pain relief.

To date, the largest reported series are still characterized by a wide spectrum of success rates after radiosurgery with Grade I outcome in 21–76.8% of patients and Grade II outcome in 65–88% of patients.\textsuperscript{6,7,21,29,38,48,52,58,67} Regis et al. reported that 87% of patients were initially free of pain in their series of 57 patients treated with a maximum dose of 75–90 Gy.\textsuperscript{52,54} In many patients, they used the higher maximum dose of 90 Gy, and their target was placed at a more anterior site (closer to retrogasserian portion). In a series of 441 patients presented at the 2001 meeting of the International Stereotactic Radiosurgery Society, Young et al. noted that 87% of patients were free of pain after radiosurgery, with or without medication (median follow-up period, 4.8 years, including repeat procedures). Brisman et al. noted vascular contact with trigeminal nerve on thin section MRI in 59% of patients with TN. These authors reported a complete (100%) pain relief without medicines in 22% of patients, 90% or greater relief with or without small doses of medicines in 30% of patients, 75–89% relief in 11% of patients, 50–74% relief in 7% of patients, and less than 50% relief in 8% of patients. Recurrent pain requiring a second procedure occurred in 24% of patients.\textsuperscript{7}
In a study, Petit et al. assessed the safety, efficacy and quality of life associated with radiosurgical treatment for TN in 112 patients treated with Gamma Knife® radiosurgery using a standard questionnaire. Ninety-six patients completed questionnaires for a median follow-up of 30 months. Seventy-four patients (77%) reported pain relief at a median of three weeks after the procedure. A decrease in medication usage was noted in 66% of patients. Seventy (7.3%) patients reported new or increased trigeminal dysfunction; however, only 3.1% reported these symptoms as bothersome. Patients with sustained pain relief reported an average of 100% improvement in their quality of life as a direct result of pain relief after radiosurgery, and 100% believed that the procedure was successful. Furthermore, among those patients with temporary pain relief and subsequent recurrence, 65% felt their treatment was a success with an average of 80% improvement in their quality of life. Smith et al. recently published the results of trigeminal neuralgia radiosurgery using a dedicated linear accelerator. These investigators treated 60 patients with central doses of 70–90 Gy delivered to trigeminal nerve root entry zone using a 5-mm collimator. Pain relief was experienced at a mean of 2.7 months. Significant pain relief was obtained in 87.5% of the patients who had essential TN and in 58.3% of the patients who had secondary facial pain. In a recent article, Longhi et al. reported on the results of Gamma Knife® radiosurgery for treatment of medically and, in some instances, surgically refractory TN. These authors found 57% Grade I and 33% Grade II pain control after Gamma Knife® radiosurgery. These favorable results are similar to those reported by Pollock et al. and Kondziolka et al. Recurrence of pain occurred in 18% of patients at a mean interval of 14.2 months after radiosurgery. The side effects of trigeminal paresthesia or hypoesthesia were observed in 9.5% of patients; no cases of anesthesia dolorosa were observed. A higher radiosurgical dose and no previous neurosurgical intervention for TN were positive predictors of a pain-free outcome. The growing body of recent literature suggests that low rates of complications of Gamma Knife® radiosurgery, coupled with high success rates and patient satisfaction, allow it to be increasingly used as primary intervention for trigeminal neuralgia for appropriate patients.

**Predictive Factors**

Several factors have been associated with the positive results achieved by Gamma Knife® radiosurgery in treating TN: the absence of multiple sclerosis, greater radiation dose, no previous surgery, typical pain features, and proximity of the isocenters to the brainstem edge. In a population of 54 patients, Rogers et al. found a higher Grade I outcome in patients with idiopathic typical TN in comparison with those with atypical features: 49% (21 of 43) as opposed to 9% (1 of 11). In their series of 179 patients, Brisman et al. reported that 41% of their patients had a Grade I outcome and 17% had a Grade II outcome. Better results were achieved in patients with no previous surgery (60% [35 of 58]). Similar results were reported by Young et al. and Maesawa et al. who described the presence of atypical pain as the most important factor yielding a poor response to Gamma Knife® radiosurgery (84.4% versus 43.8%). Several authors have reported similar results using linear accelerator based radiosurgery.

**Recurrence After Radiosurgery**

Recurrence is still a major concern in the Gamma Knife® treatment of trigeminal neuralgia. The experience suggests that longer follow-up periods are needed before drawing any conclusion. Thirty-one patients underwent repeat radiosurgery at the University of Pittsburgh due to pain recurrence. The time to recurrence after the first radiosurgical procedure varied from 3 to 64 months. The mean time to recurrence was 18.2 months, and the mean interval between the first and second radiosurgical procedures was 22.3 months (range, 6–73 months). The duration of pain relief after the initial response was analyzed in all patients using the Kaplan-Meier product-limit method. Complete pain relief (excellent or good) was maintained in 63.6 ± 3.3% of patients at one year and in 56.6 ± 3.8% at three years. Greater than 50% pain relief was maintained in 75.8 ± 2.9% of patients at one year and in 67.2 ± 3.9% at three years. A history of no prior surgery was significantly associated with achieving and maintaining complete pain relief (p = 0.01). The absence of preradiosurgical paresthesia, which is strongly correlated with no prior surgery, was significantly associated with achieving and maintaining greater than 50% pain relief (p = 0.02). According to the other literatures, recurrence rates at the last follow-up after radiosurgery varied from 3.3–21%. Currently, the etiological basis and physiopathology of TN recurrences after Gamma Knife® radiosurgery still need to be clarified. The recent description of two distinct mechanisms in pain generation, ephaptic transmission and neuronal afterdischarge, will likely explain how glial processes stimulated by radiosurgery might be more efficient in blocking the ephaptic transmission, but not the neuronal afterdischarge, which is a nonsynaptic, nonephaptic transmission that could lead to painful relapses.

**Complications after Radiosurgery**
The main complication after radiosurgery was new facial sensory symptoms caused by partial trigeminal nerve injury. Seventeen patients (7.7%) in the University of Pittsburgh series developed increased facial paresthesia and/or facial numbness that lasted longer than six months.38 One patient (0.4%) developed deafferentation pain. No patient developed other neurological morbidities. Other reported risks include new facial sensory symptoms (2.7–10%). Brisman et al.7,9 reported a 5% complication rate in patients treated with doses ranging between 70 and 80 Gy. Pollock et al.49,50 reported a significant association between high radiation dose and increased risk of trigeminal neuropathy; 45% of the patients treated with a maximum dose over 90 Gy reported a trigeminal deficit compared with 15% of the lower-dose group. Increased risk of trigeminal nerve dysfunction has been reported with the use of high-dose (90 Gy) radiosurgery for TN.42,49 Smith et al., using a dedicated linear accelerator for TN radiosurgery, reported a 25% rate of new numbness.59 Results to date suggest that CyberKnife® SRS for trigeminal neuralgia has an increased risk of sensory dysfunction and other neurological complications (Table 1).

Table 1: Recent Outcomes of Trigeminal Neuralgia Radiosurgery

<table>
<thead>
<tr>
<th>First Author</th>
<th>Year</th>
<th>Technique</th>
<th>% of Patients with Excellent Pain Relief</th>
<th>% of Patients with New Facial Numbness and Other Side Effects</th>
</tr>
</thead>
<tbody>
<tr>
<td>Maesawa38</td>
<td>2002</td>
<td>Gamma Knife®</td>
<td>74.5%</td>
<td>10.2%</td>
</tr>
<tr>
<td>Longhi35</td>
<td>2007</td>
<td>Gamma Knife®</td>
<td>61%</td>
<td>9.5%</td>
</tr>
<tr>
<td>Chen10</td>
<td>2008</td>
<td>LINAC</td>
<td>49%</td>
<td>10.5%</td>
</tr>
<tr>
<td>Richards55</td>
<td>2005</td>
<td>LINAC</td>
<td>57%</td>
<td>14%</td>
</tr>
<tr>
<td>Lim33</td>
<td>2005</td>
<td>CyberKnife®</td>
<td>78%</td>
<td>51.2%</td>
</tr>
<tr>
<td>Villavicencio65</td>
<td>2008</td>
<td>CyberKnife®</td>
<td>50%</td>
<td>47%</td>
</tr>
</tbody>
</table>

Repeat Radiosurgery

Patients who experience recurrent pain during the long-term follow-up despite initial pain relief after radiosurgery can be treated with a second radiosurgery procedure.18,21,23,47,66 At the second procedure, the radiosurgical target and dose slightly differ from the first procedure. The target is placed anterior to the first target so that the radiosurgical volumes at the second procedure overlap with the first one by 50%. Dose selection is still controversial. We advocate a lesser radiation dose (50–70 Gy) for the second procedure, because we believe that a higher combined dose would lead to a higher risk of new facial sensory symptoms. On the other hand, Pollock et al.48 suggested a greater radiation dose to the same target at the second procedure than at the first. They updated their results in a later study and concluded that repeat trigeminal neuralgia radiosurgery at higher doses had better facial pain outcomes; however, the rate of bothersome numbness was relatively high (16%). Therefore, dose reduction is recommended to reduce the morbidity of repeat trigeminal neuralgia radiosurgery. Twenty-seven patients who had a second procedure at the University of Pittsburgh were evaluated at a median follow-up of 20 months. The mean time to recurrence was 18 months, and the mean interval between the first and second radiosurgical procedures was 22 months. After the second radiosurgery, 5 of 27 (18.5%) patients had an excellent outcome, 8 (29.6%) had a good outcome, 10 (37.0%) had a fair outcome and 4 (14.8%) had a poor outcome. The rates of excellent or good outcome (complete relief and complete or partial pain relief) were 48.1% and 85.2%, respectively. We believe that the second radiosurgery procedure is as effective as the first in terms of the initial response.

Follow-up and Medications

After radiosurgery, patients are followed to assess pain relief at three months and six months, and then at yearly intervals. Their pre-radiosurgery pain medications are continued at the same doses until pain relief is obtained. Medications then can be gradually tapered off if the patient remains pain free.

Indications for Radiosurgery

Many surgical treatments exist for trigeminal neuralgia, and long-term results have been reported. We have identified a high rate of imaging-defined pontine or cerebellar infarction (24%) in patients who had prior microvascular decompressions.34 We recommend Gamma Knife® radiosurgery for older or infirm patients because
it is the least invasive option. The lack of mortality and the low risk of facial sensory disturbance, even after a repeat procedure, argue for the use of primary or secondary radiosurgery in this setting. Repeat radiosurgery remains an acceptable treatment option for patients who have failed other therapeutic alternatives.

CLINICAL ALGORITHM(S):

A number of factors are considered in making a recommendation. These factors include:

1. Patient’s age
2. Patient’s medical condition
3. Presence or absence of multiple sclerosis
4. Presence or absence of vascular contact and/or compression on thin section MRI
5. Presence or absence of prior procedures
6. The type of prior procedure and its response
7. Severity of pain and how long the patient can reasonably wait for pain relief
8. Patient’s concern and risk tolerance for dysesthesias, recurrence or complications from surgery

A broad outline of management algorithm is shown below; however, the final recommendation is usually influenced by the recommending neurosurgeon’s experience.

![Trigeminal Neuralgia Management Algorithm](image)

RMC = Retro-Mastoid Craniotomy,
MVD = Microvascular Decompression,
PRR = Percutaneous Retrogasserian Rhizotomy (Glycerol / Radiofrequency / Balloon Compression)
SRS = Stereotactic Radiosurgery

References
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60. Sweet WH: Percutaneous methods for the treatment of trigeminal neuralgia and other faciocephalic pain; comparison with microvascular decompression. Semin Neurol 8:272-279, 1988
Complete Summary

TITLE:

Stereotactic Radiosurgery For Patients With Intractable Typical Trigeminal Neuralgia Who Have Failed Medical Management.

ADAPTATION:

Not applicable: the guideline was not adapted from another source.

DATE RELEASED:

September 2003

UPDATE DATE:

January 2009

DEVELOPER AND FUNDING SOURCE:

IRSA (International RadioSurgery Association)

DEVELOPER COMMENT:

IRSA (International RadioSurgery Association) is an independent entity dedicated to promoting the development of scientifically relevant practice guidelines for stereotactic radiosurgery. IRSA is a professional organization that works to educate and provide support for physicians, hospitals, insurers and patients.

COMMITTEE:

The IRSA Medical Advisory Board Guidelines Committee and representatives in the industry.

GROUP COMPOSITION:

The radiosurgery guidelines group is comprised of neurosurgeons, radiation oncologists and physicists. Community representatives did not participate in the development of this guideline.

Names of Group Members: L. Dade Lunsford, M.D., Neurosurgeon, Chair; Ajay Niranjan, M.B.B.S., M.Ch., Neurosurgeon; Ron Young, M.D., Neurosurgeon; Ronald Brisman, M.D., Neurosurgeon; David Cunningham, M.D., Neurosurgeon; Christer Lindquist, M.D., Neurosurgeon; David Newell, M.D., Neurosurgeon; John C. Flickinger, M.D., Radiation Oncologist; Ann Maitz, M.S., Medical Physicist; Tonya K. Ledbetter, M.S., M.F.S, Editor; Rebecca L. Emerick, M.S., M.B.A., C.P.A., ex officio.

DISEASE/CONDITION:

Trigeminal neuralgia, typical (tic douloureux)

NUMBER OF SOURCE DOCUMENTS:

68
CATEGORY:

Treatment, proposed surgical management, radiosurgery

CLINICAL SPECIALTY:

Neurological surgery
Neurology
Radiation oncology

INTENDED USERS:

Physicians
Health Care Providers
Hospitals
Managed Care Organizations
Medical Physicists
Nurses
Utilization Management

OBJECTIVES:

To develop an evidence and consensus-based stereotactic radiosurgery practice guideline to be used by medical and public health professionals to make radiosurgery treatment recommendations for patients with intractable typical trigeminal neuralgia who have failed (refractory to or intolerant of) medical management.

TARGET POPULATION:

Patients with intractable (medically refractory) typical trigeminal neuralgia, often those with concomitant medical co-morbidity or advanced age.

INTERVENTIONS AND PRACTICES:

Stereotactic radiosurgery of the trigeminal nerve is performed using the following dose schedules:

- 75 Gy in a single fraction to the trigeminal nerve
- 80 Gy in a single fraction to the trigeminal nerve
- 90 Gy in a single fraction to the trigeminal nerve
- 50–70 Gy in a single fraction for repeat radiosurgery for recurrent trigeminal neuralgia

OUTCOMES CONSIDERED:

Pain control is the primary endpoint of interest. Use of pain medications, quality of life (addressed through the adverse effects of radiosurgery) and neurological outcome are also considered.

METHODS TO COLLECT EVIDENCE:

Computerized searches of published literature (primary sources); hand-searches of published literature (secondary sources); searches of electronic databases: clinical experience

DESCRIPTION OF METHODS TO COLLECT EVIDENCE:
MEDLINE and PUBMED searches were completed for the years 1966 to January 2009. Search terms included: tic douloureux, trigeminal neuralgia, stereotactic radiosurgery, Gamma Knife®, irradiation, clinical trials, research design, practice guidelines and meta-analysis. Bibliographies from recent published reviews were reviewed and relevant articles were retrieved.

METHODS TO ASSESS THE QUALITY AND STRENGTH OF THE EVIDENCE:

Expert consensus (committee)

RATING SCHEME:

Not applicable

METHODS TO ANALYZE EVIDENCE:

Systematic review with evidence tables

DESCRIPTION OF METHODS TO ANALYZE EVIDENCE:

Not applicable

REVIEW METHODS:

External peer review; internal peer review

DESCRIPTION OF REVIEW METHODS:

The recommendations were originally suggested by a core group of two members (AN and LDL). These recommendations were mailed to all committee members. Feedback was obtained through this mailed survey consisting of proposed guidelines asking for comments on the guidelines and whether the recommendation should serve as a practice guideline.

IMPLEMENTATION TOOLS:

Clinical Algorithm
Patient Resources

MAJOR RECOMMENDATIONS:

♦ Patients with typical trigeminal neuralgia who have had an adequate trial of medications can be offered stereotactic radiosurgery. Radiosurgery is typically used in patients with medical co-morbidities, patients at risk for side effects from percutaneous ablative procedures, and those in more advanced age groups.

♦ The optimal dose range for trigeminal neuralgia has been established. A commonly used dose range of 75–90 Gy in a single fraction to the trigeminal nerve is suggested, using a 4 mm collimator radiation field. Most centers prefer 80 Gy as a central dose targeted to the trigeminal nerve a few millimeters proximal to its entry into the brain stem; however, 90 Gy as a central dose to the trigeminal nerve near the trigeminal ganglion has also been used routinely in some centers.

♦ Patients who have failed other surgical procedures for trigeminal neuralgia should also receive 75–90 Gy to the trigeminal nerve. A safe interval between the initial surgery and stereotactic radiosurgery is unknown, but it is reasonable to perform radiosurgery if there is no improvement or pain recurs following the initial surgical procedures.
♦ After radiosurgery, patients are followed to assess pain relief at three months and six months, then at yearly intervals. Their pre-radiosurgery pain medications are continued at the same doses until pain relief is obtained. Medications then can be gradually tapered off if the patient remains pain free.

♦ Patients who have recurrence of pain following trigeminal neuralgia radiosurgery or who had a partial initial response can undergo a second stereotactic radiosurgery using 50–70 Gy to the trigeminal nerve (depending on the elapsed time between treatments). A generally safe interval between first and second radiosurgeries is six months.

♦ At present, technology to deliver focal small-volume fields is limited to Gamma Knife® by the strength of published data. (Gamma Knife® is a registered trademark of Elekta Instruments, Inc.) Current data suggest that CyberKnife® SRS for trigeminal neuralgia has an increased risk of sensory dysfunction and other neurological complications (Table 1).

**TYPE OF EVIDENCE:**

Class II and III

**QUALIFYING STATEMENTS:**

None

**COST ANALYSIS:**

Not applicable

**POTENTIAL BENEFITS:**

All of the published studies have shown a significant decrease in pain (decreased severity and reduced need to take medications).

**SUBGROUP(S) MOST LIKELY TO BENEFIT:**

Patients with typical trigeminal neuralgia (intermittent lancinating pain typically relieved by carbamazepine) without prior surgical procedure.

**POTENTIAL HARMS:**

Major adverse effects of trigeminal neuralgia radiosurgery occur very infrequently with Gamma Knife® technology.

Facial numbness < 10%
Neuropathic pain < 1%
Motor weakness < 1%

**SUBGROUP(S) MOST LIKELY TO BE HARMED:**

Patients with atypical facial pain (non-paroxysmal pain), deafferentation facial pain, TMJ related facial pain or orofacial pain.

**GUIDELINE STATUS:**

This is the full current release of the guideline.

**GUIDELINE AVAILABILITY:**

Electronic copies: Available in Portable Document Format (PDF) from [www.IRSA.org](http://www.IRSA.org)
Print copies: Available from IRSA, 3005 Hoffman Street, Harrisburg, PA  17110
COMPANION DOCUMENTS:
None available

PATIENT RESOURCES:

Patient resources are available online at www.IRSA.org, by email at office1@IRSA.org or by calling +717-260-9808.

See “Publications” for patient resources for trigeminal neuralgia: http://IRSA.org/publications.html
Another Perspective® Volume 4, No. 3 and Brain Talk® Volume 8, No. 1.

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