Glossopharyngeal neuralgia, or vagoglossopharyngeal neuralgia, is a cranial nerve hyperactivity pain syndrome leading to severe, transient, sharp pain in the ear, base of the tongue, tonsillar fossa, or beneath the angle of the jaw corresponding to the distributions of the auricular and pharyngeal branches of cranial nerves IX and X. Swallowing, chewing, talking, coughing, or yawning commonly trigger this pain. These painful attacks are neuralgic and therefore paroxysmal, lasting for less than a second and as long as a few minutes. Recently the International Headache Society proposed a subclassification of GPN that includes classic and symptomatic forms. In the classic type, the pain is intermittent with no underlying cause or associated neurological deficit. The symptomatic type includes the same characteristics of the classic form; however, the aching pain can persist between neuralgic episodes, and sensory impairment can be found in the distribution of the above nerves due to structural lesions. This classification does not take into consideration associated syncopal events. Neoplastic processes causing symptomatic GPN are usually malignant and tend to affect the nerve at its extracranial segment, in contrast to TN caused by benign tumors in 5%–8% of cases. Infectious and inflammatory processes may also lead to GPN. Multiple sclerosis is rarely the cause of GPN, but is the cause in as many as 3% of cases of TN. The pain syndrome of GPN can be very similar to that of TN, leading to a misdiagnosis if a thorough history is not obtained. The tracts of trigeminal, glossopharyngeal, and vagus nerves have an intimal neuroanatomical and functional relationship, and overlapping symptoms are not uncommon. However, GPN and TN do not usually occur simultaneously.

Glossopharyngeal neuralgia is a rare entity, representing only 0.2%–1.3% of facial pain syndromes. The overall incidence of GPN in the population is estimated to be between 0.2 and 0.7 per 100,000 people per year. The current neurosurgical management of glossopharyngeal neuralgia and technical nuances for microvascular decompression surgery are discussed in this article.

Current neurosurgical management of glossopharyngeal neuralgia and technical nuances for microvascular decompression surgery

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Glossopharyngeal neuralgia (GPN) is an uncommon facial pain syndrome often misdiagnosed as trigeminal neuralgia. The rarity of this condition and its overlap with other cranial nerve hyperactivity syndromes often leads to a significant delay in diagnosis. The surgical procedures with the highest rates of pain relief for GPN are rhizotomy and microvascular decompression (MVD) of cranial nerves IX and X. Neurovascular conflict at the level of the root exit zone of these cranial nerves is believed to be the cause of this pain syndrome in most cases. Vagus nerve rhizotomy is usually reserved for cases in which vascular conflict is not evident. A review of the literature reveals that although the addition of cranial nerve X rhizotomy may improve the chances of long-term pain control, this maneuver also increases the risk of permanent dysphagia and vocal cord paralysis. The risks of this procedure have to be carefully weighed against its benefits. Based on the authors’ experience, careful patient selection with a thorough exploratory operation most often leads to identification of the site of vascular conflict, obviating the need for cranial nerve X rhizotomy.

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Key Words • glossopharyngeal neuralgia • microvascular decompression • vagus nerve • rhizotomy • cranial nerve

Abbreviations used in this paper: GPN = glossopharyngeal neuralgia; MVD = microvascular decompression; TN = trigeminal neuralgia.


year,29,36,44,54 This incidence is probably underestimated due to insufficient awareness of this condition. Glossopharyngeal neuralgia is more common on the left side (left:right ratio of 3:2), but TN is more common on the right (right:left ratio 5:3).29 Bilateral involvement (usually sequentially and not simultaneously) is more common in TN (4%) than in GPN (2%). GLOSSOGRAPHYNEAL neuralgia can lead to bradycardia and loss of sympathetic tone, and therefore syncopal episodes and even seizures occur in as many as 10% of the cases.1,3,12,14,15,58,63,65 In rare cases GPN can present as syncpne with no associated pain syndrome,29 making the diagnosis even more difficult. Like TN, GPN is treated using anticonvulsant medications, but this latter condition tends to be more refractory to medical therapy, especially in patients with vascular compression.11,13,32,42,51,53 PAIN refractory to medical therapy and poor tolerance of drug side effects are common indications for surgical intervention.

Historical Perspectives

Extracranial nerve ablation was one of the first procedures attempted for treatment of GPN.43,57 This approach was abandoned due to its high morbidity and pain recurrence due to a lack of supraganglionic ablation. Dandy performed some of the first intracranial rhizotomies of the glossopharyngeal nerve with good results. Even though short-term results were good after these procedures, long-term pain recurrence was frequent. RHIZOTOMY of the vagus sensory rootlets improved long-term outcomes.5,41 Intracranial rhizotomies of cranial nerves IX and X were the preferred surgical procedure until the 1970s. Based on his intraoperative observations, Dandy proposed vascular compression of the root entry/exit zones of the cranial nerves as a possible cause for cranial nerve hyperactivity syndromes. Jannetta further investigated this mechanism and published the first series of patients with GPN treated with MVD.39 Since that time, this operation has gained greater acceptance than the traditional rhizotomy procedures and many series have been published regarding its efficacy.15–17,19–26,132,34,39,45–47,50,56,60,65

Percutaneous procedures have been devised as an alternative to craniotomy; these procedures include radiofrequency rhizotomy,2,4,12,18,21,40,55 and trigeminal tractotomy.22,28,38 Most recently, stereotactic radiosurgery has also been explored.48,59,70 Although cranial nerve IX rhizotomy is low risk,66 and has been advocated during MVD for GPN,69 the efficacy of cranial nerve X rhizotomy, when considering the associated risks, remains less defined. We therefore attempted a comprehensive review of the literature to explore the role of cranial nerve X rhizotomy in a large group of patients. In addition, the senior author (A.A.C.G.) will describe his early personal experience and lessons learned from 15 such procedures.

Literature Search

We conducted a literature search to evaluate the impact of sectioning the upper rootlets of cranial nerve X on overall postoperative pain control and morbidity as compared with MVD of cranial nerve X alone. Our literature search (using search terms such as “glossopharyngeal neuralgia” along with “microvascular decompression” or “rhizotomy”) yielded 11 reports of MVD alone (427 patients), 3 series that included both procedures (38 patients), and 4 that included rhizotomy alone (146 patients). Overall, we found 454 patients who underwent MVD and 157 who underwent rhizotomy of the upper rootlets of cranial nerve X (Tables 1 and 2).

Results

The mean age of the patients in both series at the time of surgery was 54.5 years. Of these patients, 57.3% were female and 65% presented with left-sided pain. Only 5 patients (0.8%) were reported to have bilateral pain. In terms of the primary location of the pain, the throat type was the most common across the series (35.8%), followed by combined throat and ear pain (34%). Isolated ear pain was rare and averaged only 11.3% among patients. Only 7 patients (1.1%) presented with syncope. The mean duration of symptoms was 5.3 years. The overall reported surgical mortality rate was 2.1%. The mortality rates for MVD and rhizotomy procedures were 1.1% and 5%, respectively. However, these mortality rates correspond to those in the largest series available (Rushon et al.), which includes patients operated on before the current microsurgery era. There was no mortality reported for the rhizotomy operation in the other 6 most recent series.

Mean follow-up duration was 4.9 years for the MVD group (2 series did not report follow-up time) and 4.7 years for the rhizotomy group (3 series, including Rushion et al., did not report the length of follow-up). The rate of long-term pain freedom for patients who underwent MVD was 84.7% with recurrence in 7% of patients. Pain freedom for patients who underwent rhizotomy was 87.3% with pain recurrence in 8.2%. If the series from Rushion et al. is excluded, the rate of long-term relief after rhizotomy for more recent series is 96.4% with no long-term recurrence.

Transient cranial nerve X dysfunction (dysphagia, hoarseness, or both) occurred in 13.2% of MVD cases and 25% of rhizotomy cases. Permanent cranial nerve X deficits occurred in 5.5% of MVD operations on average (1 group did not report on permanent deficits) and 19.1% of rhizotomy cohorts. If the data from the era before microsurgery are excluded, the rate of permanent cranial nerve X dysfunction is 17.8%.

Discussion

Microvascular decompression is currently the most effective operation to treat GPN. If exploratory surgery does not identify an offending vessel, sectioning cranial nerve IX and the upper rootlets of cranial nerve X is an option.66 However, this maneuver can lead to dysphagia and vocal cord paralysis.34 Monitoring the motor vagus by placing electrodes directly on the false vocal cords or surface electrodes on the endotracheal tube can help the surgeon distinguish motor from sensory roots and thus potentially decrease the morbidity of the procedure.
Management of glossopharyngeal neuralgia

Sectioning the upper rootlets of cranial nerve X to improve pain control is based on clinical observations rather than careful, reliable anatomical/functional studies. The vagus nerve does not have a craniocaudal sensorimotor organization, and in fact, the sensory fibers might be located dorsally and the motor fibers ventrally. However, accumulated experience from older series (not considering MVD as an option) has demonstrated a high pain recurrence rate when cranial nerve IX and X rhizotomies are not performed simultaneously. Physiologically, sectioning the upper rootlets of the vagus nerve increases the pharyngeal sensory loss already caused by cranial nerve IX rhizotomy, potentially resulting in paralysis of the ipsilateral vocal cord and motor arc of the gag reflex. Most authors describe rhizotomy of the upper rootlets of cranial nerve X as benign, leading only to an irritative cough, foreign body sensation in the throat, and transient hoarseness or dysphagia.

Our review of the literature disclosed a 3-fold increase (18% vs 6%) in the risk of permanent postoperative cranial nerve X dysfunction during rhizotomy versus MVD. The rate of pain control increases slightly (85% vs 96%) for MVD and cranial nerve X rhizotomy, respectively. We therefore routinely avoided sectioning the upper rootlets of cranial nerve X in our 15 patients. In our series, all patients were found to have offending vessels without a need for cranial nerve X rhizotomy. We believe that careful patient selection and thorough inspection of the cranial nerve X root exit zone will minimize the rate of “negative” exploratory operations requiring cranial nerve X rhizotomy.

Our review of the literature disclosed consistent trends in reporting postoperative results. The number of patients in the reported series is most often small. The reporting paradigms are too heterogeneous and often do not include adequate information to draw meaningful conclusions. However, it appears that rhizotomy leads to slightly better pain control at the expense of higher postoperative permanent cranial nerve dysfunction. Based on our experience, careful patient selection with a thorough and safe exploratory operation most often leads to identification of the site of vascular compression, obviating the need for cranial nerve X rhizotomy, which is associated with a higher rate of cranial nerve dysfunction.

### TABLE 1: Summary of studies involving MVD of the upper rootlets of cranial nerve X for GPN*

<table>
<thead>
<tr>
<th>Authors &amp; Year</th>
<th>No. of Patients</th>
<th>Mean Age (yrs)</th>
<th>Deaths (%)</th>
<th>Mean Follow-Up (yrs)</th>
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* CN = cranial nerve; NR = not reported.
† Long-term follow-up available in only 50 patients in this series.
‡ Long-term follow-up available in only 29 patients in this series.

### TABLE 2: Summary of studies involving rhizotomy of the upper rootlets of cranial nerve X for GPN

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Disclosure
The authors report no conflict of interest concerning the materials or methods used in this study or the findings specified in this paper.

Author contributions to the study and manuscript preparation include the following: Conception and design: both authors. Acquisition of data: both authors. Analysis and interpretation of data: both authors. Drafting the article: both authors. Critically revising the article: both authors. Reviewed submitted version of manuscript: both authors.

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Neurosurg Focus / Volume 34 / March 2013
Management of glossopharyngeal neuralgia


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