Vagal Paraganglioma

A Review of 46 Patients Treated During a 20-Year Period

James L. Netterville, MD; C. Gary Jackson, MD; Frank R. Miller, MD; John R. Wanamaker, MD; Michael E. Glasscock, MD

Background: Vagal paragangliomas (VPs) arise from paraganglia associated with the vagus nerve. Approximately 200 cases have been reported in the medical literature. Because of their rarity, most information regarding these tumors has arisen from case reports and small clinical series.

Objective: To detail the clinicopathologic features of 46 patients with VP with an emphasis on the role of a multidisciplinary skull base team in both the successful extirpation and rehabilitation.

Design: Retrospective review of 46 patients with VP managed by a single skull base team.

Setting: An academic tertiary medical center.

Results: Forty-six patients were treated over a 20-year period (1978-1998). Ten (22%) demonstrated intracranial extension. There was a history of familial paragangliomas in 9 (20%) of the patients. The incidence of multicentric paragangliomas was 78% in patients with familial paragangliomas vs 23% in patients with nonfamilial paragangliomas. Management of this group of 46 patients consisted of surgery (n = 40), radiation therapy (n = 4), and observation (n = 2). The operative approach consisted of a transcervical excision often combined with a transtemporal or lateral skull base approach as dictated by the tumor extent. Postoperative cranial nerve deficits were common, and, as such, aggressive rehabilitation was a vital component in the management of these tumors.

Conclusions: The management of VP and its associated cranial nerve deficits remains a difficult clinical problem. Options for treatment include surgical resection, radiation therapy, and, in selected cases, observation. Surgical extirpation requires a multidisciplinary skull base team to achieve complete tumor resection. Radiation therapy is reserved for elderly patients and patients at risk for bilateral cranial nerve deficits. Rehabilitation of cranial nerve deficits is an integral part of the management of VP.


T he vagal paraganglioma (VP) arises from paraganglionic tissue located along the vagus nerve (Figure 1). This neoplasm represents less than 5% of all head and neck paragangliomas, and since its original description in 1935, fewer than 200 cases have been reported in the literature.1

Because of their rarity, most of the information about these tumors is from case reports and small series.2-18 In many cases, they are “lumped” together with other vagal nerve tumors or intermixed with other head and neck paragangliomas.10-15 Biller et al16 presented the Mount Sinai experience in 1989 with 18 patients from a single institution, reviewing the diagnosis and management of glomus vagale tumors. Recently, Johnson et al17 presented an experience with 19 patients with glomus vagale tumors treated at 2 universities, and Van Der Mey et al18 reported on the largest series to date in which 32 patients were treated over a 32-year period.

Although many authors have advocated surgical excision as the primary modality of therapy for paragangliomas of the head and neck, the relative effectiveness of surgery vs radiation therapy remains controversial.1,19-28 Literature can be cited to support either modality. Radiation therapy, while it may significantly slow tumor growth and provide palliation in selected patients, has not been shown to cure paragangliomas. In most cases, the tumor size does not appreciably diminish or change with irradiation and histologically viable tumor cells (chief cells) have been shown to be present after radiation therapy.26,27 In addition, radiation therapy has the potential for irradiation-induced tumors as well as temporal bone complications (osteoradionecrosis).28

From the Department of Otolaryngology—Head and Neck Surgery, Vanderbilt University Medical Center (Drs Netterville, Miller, and Wanamaker), and The Otology Group PC (Drs Jackson and Glasscock), Nashville, Tenn.
MATERIALS AND METHODS

The medical records of all patients with VP who presented to Vanderbilt University Medical Center and The Otology Group PC, Nashville, Tenn, between 1978 and 1998 were retrospectively reviewed. Eighteen of these patients have previously been described. The medical records were reviewed for the following information: demographics (age, sex, race, laterality); clinical manifestations; radiographic findings; methods of treatment, including the surgical approach used; and the incidence of complications. The occurrence of multicentric, familial, malignant, and functional tumors was also recorded. Information on the methods of rehabilitation used in patients with lower cranial nerve deficits and the long-term outcome of these lower cranial nerve deficits on speech and swallowing was accumulated. Follow-up information was obtained from patient charts, referring physicians, and telephone contact as needed to update status.

The neoplasms were classified as being VP on the basis of radiographic demonstration of anterior displacement of the carotid artery and by intraoperative findings. Because of the nature of this group's referral pattern, the series was heavily weighted toward VP tumors with lateral skull base involvement.

The operative approach selected depended on the location and the extent of the tumor. The transcervical approach was used for neoplasms located in the high cervical/lower parapharyngeal space, while a combined transcervical and transtemporal approach was used for tumors that are located high in the parapharyngeal space and for tumors that involve the skull base. This combined approach is also used to obtain distal internal carotid artery control. This approach has been detailed in previous publications.

RESULTS

The study group consisted of 46 patients (30 women and 16 men). The female-to-male ratio was 1.87 to 1. The mean age of the patients at the time of diagnosis was 43 years (range, 16-77 years). Thirty-five (76%) of the VPs occurred on the right side and 11 (24%) occurred on the left side.

Overall, multiple paragangliomas of the head and neck were seen in 17 patients (37%). The incidence of multicentricity was 78% (7/9) in patients with familial paraganglioma vs 22% (8/37) in patients with nonfamilial paragangliomas. The majority of multicentric paragangliomas were carotid body tumors seen in conjunction with a VP.

Two patients were diagnosed as having malignant VPs on the basis of regional nodal metastases and both demonstrated carotid artery invasion. Two patients were diagnosed as having catecholamine-secreting VP on the basis of elevated serum and urinary catecholamine levels. Preoperative cranial nerve deficits were present in 13 (36%) of 36 previously untreated patients with VP. The 10th cranial nerve was the most commonly affected and was partially paralyzed or paralyzed in 10 (28%) of the 36 patients. Other cranial nerve deficits included hypoglossal (13%), spinal accessory (13%), glossopharyngeal (11%), and facial (6%). Two patients presented with Horner syndrome. In the remaining 10 patients, preoperative cranial nerve palsies could not be assessed due to previous attempts at surgical excision.

The presenting signs and symptoms in this group of patients are shown in the Table. In 6 (14%) of the patients, there were no presenting signs and symptoms.

<table>
<thead>
<tr>
<th>Presenting Signs and Symptoms (N = 46)</th>
</tr>
</thead>
<tbody>
<tr>
<td><strong>Sign/Symptom</strong></td>
</tr>
<tr>
<td>Neck mass</td>
</tr>
<tr>
<td>Pulsatile tinnitus</td>
</tr>
<tr>
<td>Pharyngeal mass</td>
</tr>
<tr>
<td>Hoarseness</td>
</tr>
<tr>
<td>Hearing loss</td>
</tr>
<tr>
<td>Aural mass</td>
</tr>
<tr>
<td>Dysphagia</td>
</tr>
<tr>
<td>Cervical bruit/pulsatile mass</td>
</tr>
<tr>
<td>Aural pressure/fullness</td>
</tr>
<tr>
<td>Facial paresthesias</td>
</tr>
<tr>
<td>Facial weakness</td>
</tr>
<tr>
<td>Headache</td>
</tr>
<tr>
<td>Cough with manipulation</td>
</tr>
</tbody>
</table>

Figure 1. Cranial nerves IX, X, XI, and XII, and the sympathetic trunk are endangered by either the growth of a vagal paraganglioma or its removal.
and the neoplasms were diagnosed when an imaging study was performed as a screening examination. The mean duration of the symptoms was 30 months (range, 2 months to 11 years).

The radiographic evaluation included bilateral carotid angiography, computed tomography (CT) (with contrast), and magnetic resonance (MR) imaging (with gadolinium). Angiography was performed in all cases. Early on in the series, CT was the imaging study of choice, while later MR became the imaging modality of choice. In cases of suspected skull base involvement, both CT (high resolution with bony windows) and MR were used. Arteriography demonstrated both a tumor “blush” and the anterior and medial displacement of the internal carotid artery without widening of the carotid bifurcation (Figure 2). In several cases, the specific diagnosis of a VP was either made or confirmed at the time of surgery. The neoplasms were enhanced on contrast CT.

On MR imaging, the tumors demonstrated an intermediate signal on T1- and T2-weighted scans, and enhancement with gadolinium. In some cases, there was a “salt-and-pepper” appearance due to the areas of low signal intensity produced by flow voids of rapidly flowing blood (Figure 3). Arteriography with selective embolization of tumor vasculature was performed in 28 patients.

The extent of the tumor in these patients can roughly be divided into 3 groups based on tumor encroachment of the skull base (Figure 4). These groups are outlined in the tabulation below.

<table>
<thead>
<tr>
<th>Location</th>
<th>No. (%) of Patients</th>
</tr>
</thead>
<tbody>
<tr>
<td>High cervical</td>
<td>15 (33)</td>
</tr>
<tr>
<td>Cervical and cranial base</td>
<td>21 (45)</td>
</tr>
<tr>
<td>Intracranial extension</td>
<td>10 (22)</td>
</tr>
<tr>
<td>Gross</td>
<td>6 (13)</td>
</tr>
<tr>
<td>Occult</td>
<td>4 (9)</td>
</tr>
<tr>
<td>Encased internal carotid artery</td>
<td>7 (15)</td>
</tr>
</tbody>
</table>

There was variable involvement of the upper neck, parapharyngeal space, and skull base. Ten patients had intracranial extension. This involvement varied from 0.5 cm to greater than 5 cm. The most common location was the high parapharyngeal space, with tumors “abutting” the cranial base.

The treatment and operative approach of this series of patients included surgery in 40 patients (transcervical approach in 12, combined transcervical and transtemporal approach in 28 [canal preservation in 10 and canal obliteration in 18]), radiation therapy in 4 (external beam radiation in 3 and stereotactic radiation in 1), and observation in 2. Ten patients had undergone previous operations on the ipsilateral side where the VP was found. Seven of these patients had prior resections of a VP and presented with recurrent disease, whereas 3 patients had procedures for removal of ipsilateral paragangliomas (most commonly a carotid body tumor). In addition to the procedures listed above, 3 patients underwent a suboccipital craniotomy for removal of the intracranial disease. Two of these procedures were performed at the time of the primary resection, while 1 was performed as a 2-staged procedure. In 2 patients, a translabyrinthine-transcochlear dissection was also necessary.

At the time of surgery, the relationship of the tumor to the carotid artery was quite variable. In some cases, a plane could be easily developed between the tumor and carotid artery, while in other cases the tumor capsule was densely adherent to the vessel, making dissection and
tumor removal extremely difficult. There were 5 patients in whom the carotid artery was encased by the VP. Two had clinical evidence of carotid artery invasion. One of these patients underwent carotid artery sacrifice, while the other patient underwent biopsy and was treated with radiation therapy. One of the 5 patients underwent carotid artery replacement, while 2 of the patients experienced carotid artery lacerations during tumor excision.

Six patients did not undergo surgical treatment. One elderly patient was observed because he had a contralateral vocal cord paralysis. A second elderly patient with multiple medical problems, including colon cancer, was observed. Four patients were treated with radiation therapy, including 1 patient with stereotactic radiation therapy. With a mean follow-up of 4½ years, these patients remain stable with tumor.

There were 40 surgical resections in this series. In 3 cases, the vagus nerve was preserved and in 37 cases it was sacrificed, with all 40 demonstrating a permanent vocal fold paralysis. The vagus was the only nerve sacrificed in 20 cases, while various combinations of the lower cranial nerves (IX-XII) were removed in the remaining 20 cases. The sympathetic chain was resected in 10 patients.

The large number of patients with lateral skull base involvement necessitated transtemporal approaches in combination with the transcervical approach. These approaches require some form of facial nerve management, either in the exposure of the skull base and/or tumor removal. In 15 of the 40 surgical cases, the facial nerve was not exposed. In 18 cases, the facial nerve was mobilized; in 6 cases, it was transected and either grafted or reanastomosed; and in 1 case, it was sacrificed.

One patient underwent subtotal excision. This was a patient with extensive carotid artery involvement who was not a candidate for carotid sacrifice or replacement based on a preoperative balloon occlusion test.

Dysphasia and/or aspiration was common in the immediate postoperative period in patients undergoing surgical resection. Twenty-seven of the patients were taking adequate nutrition by mouth at the time of discharge, while 10 patients were still dependent on tube feedings (nasogastric or gastrostomy). The status of the remaining 3 patients at the time of discharge could not be determined from the medical records. Long-term follow-up revealed that no patients had intractable aspiration or severe dysphasia preventing oral alimentation. All tracheostomy, nasogastric, and gastrostomy tubes were eventually removed. In most patients, the initial oral diet was a required considerable effort, resulting in an early diet that was limited by bolus size and texture. Many commented that they had to avoid dry, "crumbly" foods and remarked that meat was especially difficult to swallow. Other compensatory swallowing techniques were used by most patients months and years after their surgery. Most patients contacted reported that the length of time it took to finish a meal was often 1 hour or twice as long as other family members.

A variety of procedures aimed at the rehabilitation of lower cranial nerve deficits were performed either at the time of surgery or in the postoperative period. Early on in the series it was routine to perform tracheotomies.

Figure 4. The 3 extensions of the vagal paraganglioma with reference to its extension to involve the skull base. A, Tumor confined to the cervical region. B, Tumor abutting against the jugular foramen and the skull base with either anterior displacement and/or encasement of the internal carotid artery. C, Tumor extending into the jugular foramen, often with intracranial extension.
at the time of surgery, followed by Teflon injections some-
time in the postoperative period. More recently, 12 pa-
tients have undergone primary medialization laryn-
goplasty with Silastic (MLS) at the time of the cranial nerve loss and an additional 16 patients have undergone sec-
ondary MLS. Later in the series, arytenoid adduction was combined with MLS to improve phonation and de-
crease aspiration. In addition, 6 patients have under-
gone a unilateral palatal adhesion for velopharyngeal in-
sufficiency. Patients with preexisting cranial nerve palsies or paralyses without dysphasia did not undergo trache-
ostomy or primary MLS. Facial nerve deficits were man-
aged with eye precautions (artificial tears, ophthalmic lu-
ostomy or primary MLS). Facial nerve deficits were man-
aged with eye precautions (artificial tears, ophthalmic lu-
bricating ointment, and moisture chambers) in all patients,
as well as gold weight eyelid implants or canthoplasty when indicated.

The complications that occurred in this series of pa-
ients are listed in the following tabulation.

<table>
<thead>
<tr>
<th>Complication</th>
<th>No. of Patients</th>
</tr>
</thead>
<tbody>
<tr>
<td>Death</td>
<td>1</td>
</tr>
<tr>
<td>Cerebrovascular accident</td>
<td>2</td>
</tr>
<tr>
<td>Cerebrospinal fluid leak</td>
<td>3</td>
</tr>
<tr>
<td>Wound</td>
<td>6</td>
</tr>
<tr>
<td>Hematoma</td>
<td>3</td>
</tr>
<tr>
<td>Necrosis</td>
<td>2</td>
</tr>
<tr>
<td>Infection</td>
<td>1</td>
</tr>
<tr>
<td>Pneumothorax</td>
<td>2</td>
</tr>
<tr>
<td>Endocarditis</td>
<td>1</td>
</tr>
<tr>
<td>Airway obstruction</td>
<td>3</td>
</tr>
<tr>
<td>Major depression syndrome</td>
<td>2</td>
</tr>
</tbody>
</table>

A single operative death occurred and was secondary to a cerebrovascular accident. Sequelae related to cranial nerve loss other than dysphonia, dysphasia, and aspiration developed in a number of patients and included per-
sistent shoulder pain and weakness, gastrointestinal dys-
function (eg, persistent nausea and vomiting, gastroesophageal reflux), facial pain, and baroreceptor failure (eg, orthostatic hypotension, episodic hyperten-
sion, and paroxysmal tachycardia).

The mean follow-up in this series of 46 patients was
68 months (range, 8-146 months). During the fol-
low-up period, there have been no tumor recurrences in the 39 cases with total tumor removal. The 1 patient with subtotal tumor resection has had no tumor progression.

### COMMENT

The diagnosis and treatment of these neoplasms is par-
ticularly challenging for a number of reasons. The clinical behavior of these tumors is variable. They generally are benign slow-growing neoplasms associated with low morbidity and mortality; however, there is the potential to surround and invade neurovascular structures, in-
vade the skull base, and extend intracranially. Because of the tumor's origin from the vagus nerve, and the in-
timate relationship to the other cranial nerves at the skull base, surgical excision may be associated with signifi-
cant morbidity. In addition, the propensity of these tu-
mors to be multicentric and bilateral puts this popula-
tion at risk not only for multiple cranial neuropathies but also for bilateral neuropathies, and certainly adds to the complexity of medical decision making. Future studies need to critically assess the growth pattern or rate, de-
termine the natural history of untreated and partially re-
sected paragangliomas, and to better define the role of radiation therapy.

Advances in imaging have improved the way in which paragangliomas are managed. Magnetic reso-
nance imaging is helpful in defining the location and the extent of the neoplasm. In the case of cranial base in-
volveinent, the complementary use of CT and MR achieves the best assessment of bony and soft tissue detail. This facilitates treatment planning and helps to determine the best surgical approach. Since paragangliomas common-
ly involve the parapharyngeal space, the use of MR imaging helps distinguish between the various neo-
plasms occurring in this area such as neurogenic (ie, schwannoma) or salivary gland (ie, deep lobe parotid, mi-
nor salivary gland) neoplasms. Many patients in this se-
ries were referred to us either because the superior extent of the tumor was not accurately determined preopera-
tively or because a parapharyngeal space mass was mis-
interpreted as a deep lobe parotid tumor. Magnetic reso-
nance imaging is also an effective noninvasive screening tool when looking for paragangliomas in other family
members.

Because the excision of these tumors is a formi-
dable undertaking, the surgeon should have at his or her disposal surgical approaches that provide cranial base ex-
posure and allow for carotid artery control. In addition, a “skull base operative team” consisting of an otologist, head and neck surgeon, vascular surgeon, and neuro-
surgeon should be assembled, and be available should it become necessary. The favored approach to tumors ex-
tending to or involving the skull base in this series was via a lateral transtemporal approach. Biller et al preferred cranial base exposure through a combined trans-
cervical and transmandibular approach. These authors cited facial nerve paralysis and “limited” exposure as disadvantages of the transmastoid exposure. They repor-
ted that the direct exposure of the medially dis-
placed carotid artery was an advantage of the transman-
dibular route.

We believe that the lateral approach allows for a con-
tinuous progression of surgical exposure, starting in the neck and extending superiorly, as needed to accom-
plish a complete tumor resection. With tumors con-
fined to the high cervical region, resection of the stylo-
oid process along with its associated muscles will provide the necessary exposure for complete tumor removal. With further cephalic extension of the tumor, the mastoid is removed down to the jugular foramen. By operating deep to the stylomastoid foramen, the extension of the tumor to the jugular foramen can be resected, clearing it away from the internal carotid artery. If the tumor extends into the foramen, the facial nerve is mobilized and the sig-
moid ligated, which allows the jugular bulb to be re-
oved. This allows the necessary exposure to further re-
sect minimal intracranial extension. Occult intracranial extension, as we saw in several patients, can be well iden-
tified and removed by this approach. With more exten-
tive tumors eroding the skull base with gross intracra-
nial extension, the procedure is further extended to isolate the internal carotid artery deep to the glenoid fossa to
gain superior vascular control. The artery is followed through the carotid canal, in the middle fossa floor, as far as necessary to clear all disease. The lateral approach allows good exposure of the glossopharyngeal and the hypoglossal nerves as they pass over the tumor surface. With cephalic extension of the tumor to contact the skull base, the removal of the mastoid allows exposure to mobilize these nerves off the surface of the tumor. With extension into the jugular foramen, the 12th nerve can be followed posterior to the tumor, but often the nerve is encased by tumor at this point. The hypoglossal and vagal nerves form an intimate association, with shared nerve fibers, just caudal to the jugular foramen. Thus, with tumor extension up to the foramen, the 12th nerve is often involved in the tumor as evidenced from our patient group.

Second, patients with bilateral vagal tumors, or those who present with a previous palsy of the contralateral vagus or hypoglossal nerve, are not good candidates for surgical resection. We recommend observation until growth of the tumor is appreciated. At that point, either external beam or stereotactic irradiation can be used for treatment. Radiation therapy can slow or stop the growth rate of these tumors. Resection may still be needed in the future when further growth of the tumor occurs. The addition of stereotactic radiation as a treatment modality is unproven as yet, but it appears to have promise for tumors less than 3 cm in patients who are not good candidates for surgical resection.

The major challenge facing the surgeon after excision of VP's with lower cranial nerve injury or sacrifice is speech and swallowing rehabilitation. These deficits may result in varying degrees of lingual, palatal, pharyngeal, and laryngeal dysfunction. Many or most patients compensate for loss or damage to any one of these cranial nerves; however, damage to more than 1 nerve compounds the problem and prolongs the rehabilitation period for speech and swallowing. An approach to the treatment of patients with deficits of cranial nerves IX, X, XII is outlined in Figure 5. While tracheotomies, tube feedings, and Teflon injections were the mainstay of phonatory and deglutitional rehabilitation in the past, recent advances in laryngeal framework surgery have changed our management approach.

Primary medialization laryngoplasty by placement of carved Silastic implant is performed under general anesthesia at the time of lower cranial nerve injury or sacrifice. Primary medialization laryngoplasty provides immediate glottal competence in the postoperative period, thus avoiding the need for tracheotomy and its associated morbidity. Our preliminary results with this technique revealed a trend toward earlier intake by mouth (both initial and adequate), and earlier hospital discharge. Silastic medialization is an effective alternative to Teflon in the management of glottal incompetence secondary to vocal cord paralysis. It is well tolerated and consistently produces good-quality voices with few complications. The details of the surgical procedure, including the size of the implants used, has been previously described. Aggressive postoperative swallowing therapy is also crucial to the successful rehabilitation of these patients. When possible, patients underwent a preoperative consultation with a speech therapist. All patients were seen in the first few postoperative days, and followed up closely until swallowing without aspiration was attained.

The motor branch to the palate takes its origin from the vagus adjacent to the nodose ganglion. Only rarely can it be saved during resection of VP's that extend superior to the glossopharyngeal nerve. This results in unilateral paralysis of palate, with resulting velopharyngeal insufficiency. Although nasal regurgitation of liquids is frequently seen with paralysis, it does not usually limit swallowing rehabilitation. Some patients compensate well over a 4- to 6-month period and need no further treatment. Others continue to have significant nasal leak of fluids with swallowing or have decrease in voice quality that impairs communication. This group is offered a unilateral palatal adhesion. In this procedure, the naso-
pharyngeal surface of the soft palate is sutured to the posterior pharyngeal wall on the side of the paralysis via a transoral approach. This obturates the paralyzed heminasopharynx, preventing the velopharyngeal insufficiency. If the palatal branch was only injured and not resected at the time of tumor removal, we advocate waiting 12 months before performing the procedure, which we consider irreversible. This should allow for adequate time for the palate to return to function.

Several sequelae to cranial nerve loss that are not well documented in the literature were observed in this series and include facial pain, gastrointestinal dysfunction, and baroreceptor dysfunction.

In this series, there were some patients who developed severe pain in the parotid region associated with eating. This facial pain is characterized by a severe cramping or spasm in the parotid region with the first bite of each meal that diminishes over the next several bites. The intensity of the pain is increased with strong sialagogues. In the early postoperative period, the pain may be so severe that it deters intake by mouth. Gradually, the symptoms improved. We have selected the term first bite syndrome to describe this symptom complex.

It is probably secondary to either damage to, or removal of, the cervical sympathetics with loss of sympathetic innervation to the parotid gland. This results in a denervation supersensitivity of the sympathetic receptors that control the myoepithelial cells. With oral intake and release of the parasympathetic neurotransmitter, there is cross-stimulation of these receptors resulting in a supramaximal response of the myoepithelial cells. The symptom complex that results from this includes severe pain from spasm of the myoepithelial cells with initial oral intake after a period of salivary rest. It is worse with the first meal of the day owing to the longer period of time between the evening meal and the next day’s breakfast. Nine of the patients in this series demonstrated first bite syndrome. The sympathetic trunk was resected in 8 of these patients and the ninth patient had loss of sympathetic function postoperatively as manifested by Horner syndrome. Two other patients underwent resection of the sympathetic trunk and did not complain of parotid pain.

Early treatment consists of dietary modifications with “bland” food. Other surgical treatments that might improve the patient’s condition would be the resection of the parasympathetic innervation to the parotid gland either by removal of the Jacobson plexus or by removal of the auriculotemporal nerve up to the foramen ovale. Although we have performed the latter procedure in 1 patient with excellent initial results, we believe that eventually reinnervation will occur after either of these procedures. We are monitoring a large group of patients with this symptom complex and hope that more concrete answers will be forthcoming. This pain can be a significant cause of postoperative facial pain. It should be anticipated and not confused with temporomandibular joint pain, which could lead to inappropriate surgical treatment.

Gastrointestinal dysfunction also occurred when the vagus nerve was sacrificed and visceral sensory, motor, and parasympathetic innervation was impaired. Symptoms are much more severe with bilateral vagal injury but can also occur with unilateral injury. This derangement is manifested in persistent nausea, vomiting, and regurgitation and was the result of a decrease in gastrointestinal motility. Treatment includes periods of bowel rest, gastric decompression, metoclopramide, and, in severe cases, fundoplication. Prolonged periods of ileus also have been reported and have been attributed to changing levels of gastrointestinal neuropeptides secreted by the parangangliomas.10

Both immediate and sustained alterations in the blood pressure and the pulse were observed in some patients who underwent excision of VPs. These hemodynamic changes were attributed to damage to the carotid sinus reflex mediated through the glossopharyngeal nerve, and has been termed baroreceptor dysfunction. This is more

---

**Figure 5. Algorithm for the management of lower cranial nerve deficits following the excision of a vagal paraganglioma.** 1° medialization indicates Silastic medialization laryngoplasty done at the time of surgery; SLN, superior laryngeal nerve; PEG, percutaneous endoscopic gastrostomy; IX, glossopharyngeal nerve; X, vagus nerve; and XII, hypoglossal nerve.
commonly observed in cases of bilateral injury; however, it is also observed in unilateral cases, and it is hypothesized that in the case of unilateral damage, there may be impaired compensation via the contralateral normal carotid sinus.11

With unilateral injury to the glossopharyngeal nerve, or its branch the carotid sinus nerve, transient hypertension is seen in the first 24 hours after which the contralateral normal side usually compensates. The patients who are at risk for severe baroreceptor dysfunction are those who have previously undergone any surgical procedure to the contralateral side of the neck that could have injured the carotid sinus reflex. The 2 patients in this group who developed prolonged symptoms of baroreceptor failure had previously undergone the resection of a contralateral carotid body tumor. Although some degree of compensation occurs, treatment is often necessary and is aimed at controlling excess sympathetic tone. Therapy includes vasoactive medications such as clonidine and β-blockers.

In summary, the management of VP remains difficult. Treatment must be individualized, taking into account patient age, tumor site and size, multicentricity, preexisting cranial nerve deficits, and patient treatment preference. Despite numerous cases in the literature, there is no consensus regarding the application of surgery vs radiation therapy in the management of VP. In selected cases, clinical observation confined with serial radiographic studies may be a good alternative. In the hands of an experienced skull base team, surgical removal of VP can be achieved with low morbidity and excellent prospects for rehabilitation of any associated cranial nerve deficits.

Accepted for publication July 8, 1998.

Presented at the Meeting of the American Society for Head and Neck Surgery, Palm Beach, Fla, May 15, 1998.

Reprints: James L. Netterville, MD, Department of Otolaryngology—Head and Neck Surgery, S-210 Medical Center North, Vanderbilt University Medical Center, Nashville, TN 37232-2559 (e-mail: james.netterville@mcmail.vanderbilt.edu).

REFERENCES