Surgical treatment of cluster headache

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Cluster headache is ordinarily managed medically, but may become refractory to such medical management. In this setting, surgical treatment has occasionally been performed, based on evidence that pertinent pain pathways and parasympathetic pathways may be interrupted at the main sensory root of the trigeminal nerve and at the nervus intermedius. Between 1976 and 1987, 13 patients underwent surgery for treatment of cluster headache that was refractory to medical therapy (15 procedures). Partial sectioning of the main sensory root and sectioning of the nervus intermedius were performed in nine patients; only partial sectioning of the main sensory root in one; only sectioning of the nervus intermedius in one; and nervus intermedius sectioning plus microvascular decompression of the trigeminal nerve in two. The average postoperative period for the 13 patients was 37 months (range 2 to 135 months). All patients had return of their headaches postoperatively except for one patient who obtained relief after a repeat procedure. Headache began to return between 2 days and 2 years postoperatively. Three patients are currently free of headache, including both patients who had nervus intermedius sectioning plus microvascular decompression of the trigeminal nerve. Together with recurrence of headache, cluster-associated autonomic disturbances recurred after 14 of the 15 operations but are currently absent in the three headache-free patients. Partial sectioning of the main sensory root and sectioning of the nervus intermedius, as performed in these patients, seem to have limited value in the treatment of cluster headache.

Key Words - cluster headache - greater superficial petrosal nerve - nervus intermedius - trigeminal nerve

Cluster headache (also called migrainous neuralgia, Horton’s syndrome, and histamine cephalalgia among other terms) is a typical form of cranial pain syndrome that can occur episodically or chronically.7,11,13,16,28,38 It is predominately a disorder of adult men, with reported male to female ratios ranging between 4.5:1 and 6.7:1. The pain is typically unilateral and retro-orbital, oculotemporal or oculofrontal in location, excruciating in severity, and boring and non-throbbing in character. In association with the pain, the patient may exhibit ipsilateral signs of autonomic dysfunction such as rhinorrhea or nasal congestion, increased lacrimation, ptosis, and miosis.

Methysergide, corticosteroid preparations, and ergotamine tartrate are among the medications given prophylactically to prevent episodic cluster headache. Lithium carbonate, calcium channel blockers such as verapamil, and indomethacin are some of the drugs that are administered to prevent chronic cluster headache. Histamine desensitization is also used prophylactically with some success. Oxygen inhalation, ergotamine tartrate administration, and local anesthesia of the sphenopalatine fossa region with cocaine or lidocaine are employed to ameliorate an attack of migrainous neuralgia once it has begun. However, the headaches of some patients are or become refractory to such medical management. This has led to the development of surgical strategies designed to interrupt the transmission of nociceptive and autonomic impulses.

Partial destruction of the trigeminal nerve and surgical division of the nervus intermedius are two of the surgical approaches that have been taken when treating cluster headache. Because some degree of success has been reported for these operations, one of the authors (R.H.W.) performed 15 such procedures on 13 patients with intractable cluster headache.

Summary of Cases

Patient Population

Data were obtained from chart review and patient interviews. The patient population consisted of 12 men and one woman (Table 1). All patients had classic symptoms of cluster headache, including unilateral orbital, frontotemporal, or maxillary pain with associated autonomic symptoms. Seven patients were in the epi-
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**TABLE 1**  
Results in 13 patients treated surgically for cluster headache (CH)*

<table>
<thead>
<tr>
<th>Case No.</th>
<th>Sex, Age at Onset (yrs)</th>
<th>Age at Treatment (yrs)</th>
<th>Symptoms</th>
<th>Medications of Benefit†</th>
<th>Operation</th>
<th>Results</th>
<th>Return of Autonomic Symptoms</th>
<th>Postop Follow-Up (Mos)</th>
</tr>
</thead>
<tbody>
<tr>
<td>1</td>
<td>M, 29</td>
<td>36</td>
<td>rt orbit, chronic CH weekly + autonomic sx</td>
<td>corticosteroid prep (16)</td>
<td>PSV, SNI</td>
<td>rec. 5 wks</td>
<td>yes</td>
<td>3</td>
</tr>
<tr>
<td>2</td>
<td>M, 17</td>
<td>23</td>
<td>rt temple, episodic CH 3–4/day + autonomic sx</td>
<td>corticosteroid prep (11)</td>
<td>PSV, SNI</td>
<td>rec. 2 mos</td>
<td>yes</td>
<td>2</td>
</tr>
<tr>
<td>3</td>
<td>M, 27</td>
<td>32</td>
<td>lt maxilla, episodic CH several/day + autonomic sx</td>
<td>hydrocodone bitartrate + acetaminophen (16)</td>
<td>PSV</td>
<td>rec. &lt; 1 wk</td>
<td>less severe</td>
<td>10</td>
</tr>
<tr>
<td>4</td>
<td>M, 16</td>
<td>24</td>
<td>rt orbit, chronic CH several/day + autonomic sx</td>
<td>lithium carbonate, corticosteroid prep (11)</td>
<td>PSV, SNI</td>
<td>rec. &lt; 1 mo</td>
<td>yes</td>
<td>12</td>
</tr>
<tr>
<td>5</td>
<td>M, 16</td>
<td>30</td>
<td>lt orbit, chronic CH 4–5/mo + autonomic sx, transient hemiparesis with a few headaches</td>
<td>none (17)</td>
<td>PSV, SNI</td>
<td>rec. &gt; 2 wks</td>
<td>less severe</td>
<td>3</td>
</tr>
<tr>
<td>6</td>
<td>F, 29</td>
<td>46</td>
<td>lt orbit, chronic CH 1–9/day + autonomic sx</td>
<td>lithium carbonate (14)</td>
<td>SNI</td>
<td>rec. &gt; 2 yrs</td>
<td>yes</td>
<td>51</td>
</tr>
<tr>
<td>7</td>
<td>M, 54</td>
<td>67</td>
<td>rt orbit, chronic CH 1–3/day + autonomic sx, trauma to forehead</td>
<td>lithium carbonate (9)</td>
<td>MVD, SNI</td>
<td>rec. &gt; 1 wk; less frequent; CH-free for 8 yrs</td>
<td>rec with CH, now none</td>
<td>120</td>
</tr>
<tr>
<td>8</td>
<td>M, 35</td>
<td>41</td>
<td>lt orbit, chronic CH 4–5/day + autonomic sx, V neuralgia</td>
<td>none (7)</td>
<td>PSV, SNI</td>
<td>rec. at 2 days</td>
<td>yes</td>
<td>30</td>
</tr>
<tr>
<td>9</td>
<td>M, 26</td>
<td>39</td>
<td>lt orbit, episodic CH 2/day + autonomic sx, trauma to face</td>
<td>verapamil, corticosteroid prep (2)</td>
<td>1: PSV, SNI 2: PSV, SNI</td>
<td>1: rec &lt; 1 mo 2: no CH</td>
<td>yes</td>
<td>1</td>
</tr>
<tr>
<td>10</td>
<td>M, 16</td>
<td>29</td>
<td>lt orbit, episodic CH 3/day + autonomic sx</td>
<td>lithium carbonate (11)</td>
<td>PSV, SNI</td>
<td>initial relief; rare mild headache &gt; 1 mo; facial trauma at 18 mos, pain rec</td>
<td>yes</td>
<td>25</td>
</tr>
<tr>
<td>11</td>
<td>M, 31</td>
<td>43</td>
<td>lt orbit/frontal/occipital, episodic CH several/day + autonomic sx</td>
<td>lithium carbonate, corticosteroid prep, oxygen (11)</td>
<td>PSV, SNI</td>
<td>rec. &lt; 2 wks</td>
<td>yes</td>
<td>48</td>
</tr>
<tr>
<td>12</td>
<td>M, 33</td>
<td>35</td>
<td>rt orbit, episodic CH several/day + autonomic sx</td>
<td>lithium carbonate, oxygen (11)</td>
<td>1: PSV, SNI 2: PSV, SNI</td>
<td>1: rec &gt; 5 mos 2: rec at 5 days</td>
<td>yes</td>
<td>7</td>
</tr>
<tr>
<td>13</td>
<td>M, 32</td>
<td>50</td>
<td>rt maxilla, episodic CH 3/day + autonomic sx</td>
<td>lithium carbonate, corticosteroid prep (12)</td>
<td>MVD, SNI</td>
<td>rec &gt; 1 yr, remission, rec years later; CH-free for 3 yrs</td>
<td>yes</td>
<td>135</td>
</tr>
</tbody>
</table>

* Abbreviations: SNI = sectioning of the nervus intermedius; MVD = microvascular decompression of the trigeminal nerve; PSV = partial sectioning of the main sensory root of the trigeminal nerve; rec = recurrence; sx = symptoms; V = trigeminal nerve.

† Total number of medications tried given in parentheses.

Sodic phase and six patients were in the chronic phase of headache. One patient (Case 8) had concurrent trigeminal neuralgia (a cluster-tic syndrome). The patients' age at onset of symptoms ranged from 16 to 54 years (mean 28 years). The duration of symptoms before operation ranged from 1 to 13 years (mean 7 years).

The patients had been treated with a wide range of medical therapies, an average of 11 therapies per patient. Most frequently used were ergot preparations, carbamazepine, beta receptor blockers, calcium channel blockers, lithium carbonate, corticosteroid preparations, oxygen, methysergide maleate, antidepressant drugs, and narcotic agents. The best results had been obtained with corticosteroid preparations (in six of nine patients so treated), lithium carbonate (in seven of 12 patients), and oxygen (in two of four patients).

All patients had become refractory to medical treatment before operative treatment was undertaken. Between 1976 and 1987, 15 retromastoid craniectomies were performed on these 13 patients. Nine patients underwent partial sectioning of the main sensory root of the trigeminal nerve; four patients had partial sectioning of the nervus intermedius; two patients had both operations; and in one patient, only partial sectioning of the main sensory root was performed. One patient had only partial sectioning of the main sensory root; one patient had only sectioning of the nervus intermedius; and two patients (the first two in the series) had nervus intermedius sectioning plus microvascular decompression.
of the ipsilateral trigeminal nerve. The postoperative follow-up period ranged from 1 to 135 months (average 37 months).

Results of Surgery

Partial sectioning of the main sensory root of the trigeminal nerve plus sectioning of the nervus intermedius was performed 11 times in nine patients (Table 1). Two patients (Cases 8 and 11) noted the return of headache within 2 weeks after the first operation; the remainder experienced relief for 2 weeks to 5 months postoperatively. A second similar procedure in two of these patients resulted in relief in one for the 2-year follow-up period, but the other patient experienced the return of pain on the 5th postoperative day. All patients in this group had return of their autonomic symptoms with recurrence of their headaches. The sole patient who had headache relief through the 2-year follow-up period also remained free of autonomic symptoms.

The two patients who underwent either partial sectioning of the main sensory root or nervus intermedius sectioning experienced recurrence of headache and autonomic symptoms. This occurred within one week in Case 3 (partial main sensory root sectioning alone), but did not occur until after 2 years in Case 6 (nervus intermedius sectioning alone).

Although the two patients (Cases 7 and 13) who had microvascular decompression of the trigeminal nerve plus nervus intermedius sectioning noted recurrence of headache and autonomic symptoms after 1 week and after 1 year, respectively, each had a subsequent remission. At the time of the last follow-up assessment 120 and 135 months postoperatively, these two patients had been pain-free and without related autonomic dysfunction 8 and 3 years, respectively. Overall, when symptoms returned postoperatively they were often less severe initially than the preoperative symptoms had been. But with time they tended to return to their preoperative level.

All patients who had partial main sensory root sectioning had some degree of trigeminal sensory loss postoperatively; however, dense hypalgiesia in the upper face was not produced. Two patients had cerebrospinal fluid (CSF) rhinorrhea postoperatively which was treated successfully by a short course of external lumbar CSF drainage. The one patient who was treated by nervus intermedius sectioning alone had ipsilateral postoperative deafness.

Discussion

The nervus intermedius ordinarily consists of a series of filaments that lie between, and communicate with, the facial and vestibular nerves.25,27 The greater superficial petrosal nerve leaves the nervus intermedius within the temporal bone at the geniculate ganglion and enters the cranial cavity through the hiatus of the facial canal. It runs forward on the floor of the middle fossa, passing under the gasserian ganglion and lateral to the internal carotid artery. It is joined by the deep petrosal nerve from the sympathetic plexus around the internal carotid artery to form the vidian nerve, which passes forward through the pterygoid canal to the sphenopalatine ganglion.

Preganglionic parasympathetic fibers from the lacrimal nucleus travel through the nervus intermedius and greater superficial petrosal nerve to reach the sphenopalatine ganglion.22 Postganglionic fibers mediate secretory impulses to the lacrimal gland via the zygomatic nerve and its anastomosis with the lacrimal nerve. Other postganglionic fibers carry secretory and vasodilatory impulses to the mucous glands and vessels of the nasal epithelium.22,29 Additional preganglionic parasympathetic fibers, leaving the medulla with the fibers mentioned above, pass through the nervus intermedius but leave the greater superficial petrosal nerve to join the internal carotid artery plexus. Within this plexus are neurons that give rise to vasodilatory postganglionic fibers that pass to the cerebral arteries.4,22,29

The nervus intermedius also contains afferent fibers,4,29,34,37 and there is some evidence that nociceptive impulses from deep facial structures may pass to the brain through the greater superficial petrosal nerve and nervus intermedius.32 According to Solomon,29 “many neuroanatomists believe that deep sensation from the face is carried by neurons whose cell bodies are in the geniculate ganglion. . . . Afferent pathways from the internal carotid artery and from the dura are carried via the [greater superficial petrosal nerve] to the geniculate ganglion. . . . The [nervus intermedius] carries all of the afferent impulses from the geniculate ganglion to the brain stem, where some fibers synapse in the nucleus of the tractus solitarius and somatic sensory pathways end in the nucleus of the descending tract of the trigeminal nerve.”

Beginning in 1940, Gardner and his associates9 treated cluster headache by resecting the greater superficial petrosal nerve in an attempt to interrupt abnormal parasympathetic discharges responsible for dilation of the cerebral, meningeal, and nasal mucosal blood vessels, the pain resulting from which was being transmitted over the trigeminal nerve. However, they also thought it possible that the operation interrupted painful impulses coming over the geniculate somatic afferent fibers from the dura mater, internal carotid artery, and vidian nerve. By the time of their report in 1947, Gardner, et al., had performed this procedure on 13 patients. The results were excellent in 25% and fair to good in 50%; 25% of the patients experienced no relief.

Other surgeons subsequently performed this same procedure for cluster headache.1,2,23,24,33-35,38 Beginning in 1948, White and Sweet38 treated periodic migraine neuralgia by greater superficial petrosal nerve sectioning, either alone or in association with sectioning of the lesser superficial petrosal nerve, the external nerve, or the middle and accessory middle meningeal arteries. The results were quite varied. One of their nine patients noted no improvement; all of the others experienced
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recurrence of symptoms, but there were often significant periods of relief. In 1953, Trowbridge, et al., reported four patients with cluster headache treated by dividing the greater superficial petrosal nerve. One patient had complete relief and two had marked improvement, but the longest follow-up period was only 8 months. Aubin, et al., in 1953 and Ponti and Valerio in 1960 reported surgical sectioning of the greater superficial petrosal nerve as treatment of post-otic cephalgia. Ponti and Valerio described seven patients who received this type of treatment for what were probably cluster headaches. In 1970, Stowell reported 36 patients so treated for cluster headache over a 20-year period. Of these, 32 had complete relief, but 15 subsequently experienced recurrence of headache (eight by 1 year, three more by 2 years, and four more by 3 years). Alvarez Garijo, et al., published in 1975 an account of nine patients whom they had treated for cluster headache by greater superficial petrosal neurectomy plus division of the superficial temporal and middle meningeal arteries. The results were excellent in four cases, fair to good in two, and a failure in three. In 1977, Denecke presented three patients who had had cluster headache for a period of 10 to 20 years and who were relieved by resection of the greater superficial petrosal nerve; however, the follow-up period was less than 1 year in all three cases. Among the patients reported in 1983 by Watson, et al., four were treated for chronic cluster headache by greater superficial petrosal neurectomy. One of the patients had no improvement, one experienced pain relief for 12 months before recurrence, and two were still pain-free at 4 and 5 years after operation, respectively.

From the foregoing, it seems that the main drawback to the treatment of cluster headache by greater superficial petrosal neurectomy is the incidence of headache recurrence, which in some cases may result from nerve regeneration. Because of the relatively high failure and recurrence rates associated with this type of surgical treatment, other approaches have been tried.

Two of these approaches involve interruption of the same neural system, either peripheral to the greater superficial petrosal nerve (the sphenopalatine ganglion) or central to it (the nervus intermedius). In 1962, Brown reported that the injection of alcohol into the sphenopalatine ganglion of 28 patients with cluster headache did not reliably stop the headache. Meyer, et al., presented 13 patients with intractable cluster headache who were treated by sphenopalatine ganglionectomy; seven obtained little relief, four were improved, and only two had complete relief (for more than 1 year).

Beginning in 1952, Sachs divided the nervus intermedius for intractable face and head pain, and in 1968 reported four patients who had been treated successfully in this way; at least one of these patients had cluster headache. By 1969, he had performed this procedure on two more patients with cluster headache. As of that date, all six patients had been relieved of pain.

A variation on the surgical approach to the nervus intermedius was tried by Apfelbaum and his associates. In 1986, they reported their experience with decompressing the facial nerve at the brain stem in five patients with chronic cluster headache. Two of these patients also had decompression of the trigeminal nerve at its entry zone into the pons; one of them experienced improvement in the severity of the cluster headache, but the other did not. Of the remaining three patients, two noted improvement. However, none of the five patients achieved complete relief.

Kunkel and Dohn reported in 1974 that since 1970 they had treated 12 patients with chronic migrainous neuralgia by sectioning the greater superficial petrosal nerve and neurolysis of the sensory root of the trigeminal nerve. Three of the patients continued to have similar headaches, but without parasympathetic manifestations. Of the nine patients with immediate relief, three later experienced recurrence of headache, again without parasympathetic dysfunction. The remaining six patients (50%) were free of headache at their last follow-up examination. Four of the 12 patients experienced postoperative facial weakness that resolved completely.

Procedures designed to interrupt pain transmission in the trigeminal system have been employed for some 80 years to treat cluster headache. White and Sweet summarized the various approaches taken prior to 1969 and this information was updated by Maxwell in 1982. In 1936, Harris noted that "since 1909 I have treated a considerable number of migrainous neuralgias with alcohol injection, of either the supra-orbital or the infra-orbital nerves, being guided by the principal location of the pain. Of late years I have, instead, in several cases injected the inner portion of the Gasserian ganglion so as to obtain a permanent anaesthesia and a lasting cure." In 1970, Stowell stated that 16 of his 21 patients were relieved by a block or avulsion of the supraorbital nerve and that all of five patients obtained relief after sectioning the first trigeminal division.

In more recent years, the trigeminal sensory root has been divided surgically or injured by a percutaneous radiofrequency trigeminal gangliorhizolysis. Onofrio and Campbell noted excellent pain relief in 54% of 26 patients so treated and fair to good relief in another 15%. These authors stressed "the absolute need to achieve either corneal anesthesia or dense analgesia to accomplish relief of cluster headaches ...." O'Brien and MacCabe performed radiofrequency lesioning on 27 occasions in 13 patients; only 12 of these 27 procedures provided significant pain relief for at least 5 months. In another eight patients, they performed nine intracranial trigeminal sensory root sections; good results were obtained following five of the nine operations, lasting a median of 44 months (range 24 months to 8 years).

Maxwell achieved better results than these with percutaneous radiofrequency trigeminal gangliorhizo-
lysis, probably because of more refined patient selection for the procedure. In addition to the usual selection criteria based on an accurate diagnosis and the failure of adequate medical therapy, Maxwell also tested the patient’s response to a lidocaine injection of the ipsilateral gasserian ganglion. Of 23 patients so tested, eight experienced immediate but transient relief. These eight patients were then treated by gangliolysis and all received prolonged relief.

With this background information about the interruption of aspects of the nervus intermedius and trigeminal systems for the treatment of cluster headache, we elected to treat the patients in the present series by sectioning the nervus intermedius, either alone (one patient) or in combination with partial sectioning of the trigeminal sensory root (nine patients) or decompression of the trigeminal sensory root (two patients). One additional patient was treated by partial trigeminal sensory root sectioning alone. The initial results were gratifying, but the recurrence rate has been disappointing.

It is interesting that the two patients who were treated by microvascular decompression of the trigeminal nerve and sectioning of the nervus intermedius had a distinctly better outcome than the other 11 patients. If this represents a true difference rather than a chance occurrence, the reason is not obvious to us.

At least two factors may explain the recurrence of headache and autonomic symptoms in our patients. First, the caudal portion of the main trigeminal sensory root was cut. Perhaps a better result would follow sectioning of the cephalad portion, which should contain more of the sensory fibers from the upper face. Second, because of the variable and extensive branching of the nervus intermedius, it is difficult to identify and section all of its components. Perhaps sectioning of the greater superficial petrosal nerve in the treatment of unilateral headache would give a more complete effect. Finally, because of the relatively short time to recurrence of symptoms in most of our patients, it does not seem likely that such recurrence resulted from regeneration of the nervus intermedius.

Addendum

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Manuscript received October 3, 1989.
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