Occipital Nerve Stimulation for Refractory Headache in the Chiari Malformation Population

**BACKGROUND:** Chronic occipital and suboccipital headache is a common symptom in patients with Chiari I malformation. These headaches may persist despite appropriate surgical treatment of the underlying pathology via suboccipital decompression, duroplasty, and cerebrospinal fluid diversion. Occipital nerve stimulation has been shown to be effective in the treatment of a variety of occipital headache/pain syndromes.

**OBJECTIVE:** To review retrospectively our experience with occipital nerve stimulation in patients with a primary diagnosis of Chiari malformation and a history of chronic occipital pain intractable to medical and surgical therapies.

**METHODS:** We present a retrospective analysis of our series of 22 patients with Chiari malformation and persistent occipital headaches who underwent occipital neurostimulator trials and, after successful trials, permanent stimulator placement. A trial was considered successful with ≥ 50% pain relief as assessed with a standard Visual Analog Scale score. Patients with a successful trial underwent permanent placement approximately 1 to 2 weeks later. Patients were assessed postoperatively for pain relief via the Visual Analog Scale.

**RESULTS:** Sixty-eight percent of patients (15 of 22) had a successful stimulator trial and proceeded to permanent implantation. Of those implanted, 87% (13 of 15) reported continued pain relief at a mean follow-up of 18.9 months (range, 6-51 months). Device-related complications requiring additional surgeries occurred in 40% of patients.

**CONCLUSION:** Occipital stimulation may provide significant long-term pain relief in selected Chiari I malformation patients with persistent occipital pain. Larger and longer-term studies are needed to further define appropriate patient selection criteria and to refine the surgical technique to minimize device-related complications.

**KEY WORDS:** Cerebellar ectopia, Migraine, Occipital neuralgia, Peripheral neuromodulation, Tonsillar herniation

**ABBREVIATIONS:** CM, Chiari malformation; ONS, occipital nerve stimulation; TCC, trigeminocervical complex; VAS, Visual Analog Scale
subset of patients remains in whom occipital headaches may persist or even progress despite resolution of other CM-associated symptoms. It is thought that in some patients, postsurgical phenomena such as postoperative dural adhesions and inadvertent damage to the greater or lesser occipital nerves may also contribute to the persistence or augmentation of headache. In this subset of patients, treatment of these symptoms remains elusive.

Occipital nerve stimulation (ONS) is an alternative surgical technique that has proven efficacious in treating refractory occipital neuralgia, chronic migraine, and trigeminal autonomic cephalgias such as cluster headache. Slavin and colleagues were the first to report the use of ONS in 3 patients with persistent chronic occipital headache after Chiari decompression; these 3 patients were a subset of 14 patients with differing causes of pain undergoing ONS. A single case report noted success of ONS in a case of refractory pain after occipital cervical fusion with a dramatic reduction in Visual Analog Scale (VAS) scores (from 9 of 10 to 1 of 10) at the 1-year follow-up. Here, we reviewed our experience with this cohort of patients with CM presenting with persistent headaches refractory to previous surgical correction.

PATIENTS AND METHODS

Patient Selection

General referral criteria included patients with the clinical and radiographic diagnosis of CM with persistent and disabling occipital headaches despite appropriately performed corrective surgery and comprehensive multidisciplinary pain management. Before the neurostimulator trial, patients had failed trials with a variety of medications, including narcotic analgesics, antiepileptics, antidepressants, sleep agents, antipsychotics, and muscle relaxants. Other therapies used included cutaneous nerve stimulation, physical therapy, nerve blocks with local anesthetics and/or steroids, and botulinum toxin injections. Patients were selected for an ONS trial if their primary pain was in the occipital/suboccipital region; those with the primary complaint of frontal, temporal, or vertex headache were not selected for ONS. It should be noted, however, that 15 of the 22 patients also reported significant secondary frontal headaches. Success of occipital nerve block was not a requirement for patient selection. The VAS was used to identify a quantitative assessment of current pain before and after procedures and at each clinical visit in the office or hospital setting. This study was approved as a retrospective chart review under the auspices of the Hofstra North Shore–LIJ School of Medicine Institutional Review Board.

Surgical Technique

Percutaneous Trial

All patients underwent a percutaneous trial, lasting from 3 to 7 days, to assess successful pain relief. Patients with unilateral pain underwent a single percutaneous lead placement, whereas dual leads were used for cases of bilateral pain. The trials were performed under local anesthesia with intravenous sedation with a variety of agents, including midazolam, diprivan, and dexmedetomidine hydrochloride. Prophylactic antibiotics were given at the start of the case and throughout the duration of the outpatient trial. We preferred in all of our cases to use a lateral-to-medial approach to cannulation and electrode placement. Patients were positioned semilateral or prone, depending on the nature of the pain (unilateral or bilateral) and the presence of an occipital-cervical fusion, in which case the prone position was used for a bilateral placement. Because a portion of our CM cohort had undergone a posterior fossa decompression, cranioplasty, occipital cervical fusion, and/or ventriculoperitoneal shunt, great care was taken to keep the needles and implants away from the hardware. Given our experience with a single case of wound erosion resulting from a horizontally placed lead over the occipital bar of an occipital-cervical fusion construct, leads were placed vertically in some post–occipital-cervical fusion patients (Figure 1).

Landmarks for initial insertion included identifying the mastoid process of the respective side and entering the skin percutaneously into the subcutaneous epicraniac layer, slightly above the mastoid tip, which usually corresponds to the level of the arch of C1. After sterile prepping and draping and infiltration of the needle entry point with local anesthesia, the spinal needle was bent and then used to cannulate the subcutaneous layer of the scalp. Fluoroscopy was used continuously to identify our cannula and lead placements in respect to the odontoid process and the arch of C1. Eight-pole leads manufactured by Medtronic, Boston Scientific, or St. Jude Neuromodulation were used. The trial leads were connected to an external programmer and assessed for adequate initial paresthesia of the painful region. When lead placement was deemed satisfactory, the needles and stylets were removed, and 3-0 nylon sutures in a purse-string fashion were sewn around the external electrode wire to secure it in place. The leads were again connected to an external programmer, and the stimulation settings were then optimized in the recovery room once all sedation had worn off.

Permanent Implantation

Those patients who reported a minimum of 50% pain improvement as assessed with a VAS during the trial period underwent permanent placement. After being intubated and receiving general anesthesia, patients were placed either in the supine position with the head rotated away from side of interest or in the lateral or prone position, depending on the location of generator placement. Most patients returned for the permanent implantation with their trial leads in place. An initial x-ray was taken to identify the placement of the trial leads just before removal. Once the trial leads were removed, the operative site was prepped and draped. For a bilateral placement, a 5-mm midline incision was made down to the subcutaneous layer, and a lead was placed through the needle from the midline incision toward the distal mastoid process. A 4-cm retromastoid incision was then made vertically, and dissection was performed into a suprafascial plane. The spinal needle was then used to pass the initial lead to the retromastoid incision, and the second lead was then placed from lateral to medial, again under fluoroscopic guidance with the trial image used as comparison. The leads were then anchored to the retromastoid fascia with anchors provided by the equipment manufacturers. For generator placement in the chest region, a standard 4-cm infraclavicular incision was made, followed by a subcutaneous pocket. The leads were tunneled down subcutaneously to this infraclavicular region and plugged into the implantable pulse generator. For an abdominal or buttocck placement, depending on the size of the patient, extension leads were used when necessary. The generator at this point was checked for impedance and programmed. All incisions were irrigated, closed primarily, and covered with sterile dressings.

Data Analysis

A univariate analysis of patient age, sex, pain location, surgery location, outcomes, and complications was performed with nonparametric
(Kruskal-Wallis or Mann-Whitney) tests. A value of \( P < .05 \) was considered significant. Months to implant removal was assessed with the LIFETEST procedure. All data were evaluated with SAS 9.2 statistical software (SAS Institute, Inc, Cary, North Carolina).

RESULTS

Twenty-two patients, 19 female and 3 male, ranging in age from 14 to 54 years (mean, 33.6 years) underwent percutaneous trials for refractory occipital headaches after surgical intervention for CM. Demographics and associated diagnoses are presented in Table 1. A total of 62 procedures were performed, including unilateral and bilateral trials and, when indicated, permanent implantations (Figure 2). Few patients presented with unilateral (2 of 22, 9%) instead of bilateral (20 of 22, 91%) occipital headaches. Nineteen patients (86%) received bilateral percutaneous trials (stage 1 neurostimulation) with 1 patient wanting the trial on the right side only (right-side pain was greater than
left-side pain) despite bilateral occipital headache presentation. A trial was considered successful with 50% pain relief assessed with a standard VAS. Patients with a successful trial underwent permanent placement approximately 1 week later. Fifteen patients (68%) reported a successful trial within 1 week and progressed to the second operative stage, implantation of permanent neurostimulators. All 7 patients without reported successful trial stimulation were patients with bilateral occipital pain presentations, included both female patients and 1 male patient, and had a mean age (28.4 years; range, 16-46 years) approximately 7 years younger than the mean age of successful trials (35.6 years; range, 15-54 years). No statistically significant correlation between patient age, sex, pain location, and implant location (unilateral vs bilateral) and successful outcome was found.

Long-term follow-up was considered in CM patients who underwent successful implantation of permanent leads and were followed up for a minimum of 6 months. An implantation was considered successful with > 50% pain relief assessed with a VAS. The average long-term follow-up in this series ranged from 6 to 51 months (average, 18.9 months; Table 1). Thirteen of the 15 implanted patients (87%) reported wanting to maintain the implant in place, reported successful reduction in VAS scores at a minimum of 6 months, and were considered to have achieved a successful outcome. Of all referrals selected to undergo trial and implantation, 59% reached a successful outcome in our series. In estimations of implant life-survival, all patients who received stage 2 implantations were very likely to keep it in place by the 6-month follow-up (87%), and all patients who achieved successful outcome at 6 months went on not to require device explantation (13 of 13; 100%; Figure 3).

The surgical complications in our case series were similar in nature and in frequency to those reported in the literature for ONS. There were no complications from the trial procedures. Complications requiring 1 or more surgical revisions occurred in 6 of the 15 patients (40%) implanted (Table 2). Lead migration, a frequent occurrence in the ONS literature, occurred in 3 of the 15 patients implanted. At the time of revision, 1 patient with a lead migration was found to have a defect in the lead anchor (Titan, Medtronic) involving the inner metal tubing slipping out from the Silastic surrounding anchor. This was confirmed later by the manufacturer to be a defect in the anchor, which was recalled from clinical use and subsequently rereleased in a modified version. An example of this is depicted in an intraoperative radiograph demonstrating a single lead that migrated with the contralateral lead remaining in place (Figure 4). Use of the modified anchor has not been associated with any subsequent lead migrations in our experience or in other cases of peripheral stimulation or epidural spinal cord stimulation performed at our center, including patients without the diagnosis of CM and receiving ONS (unpublished observations).

Unique to our patient population is the high frequency of patients with prior surgeries in the occipital region, including cranietomies, cranioplastics, occipital-cervical fusion with hardware, and ventriculoperitoneal shunt placement. Initial experience with standard horizontal placement of the electrodes (parallel to the arch of C1) resulted in 1 case of lead tip erosion in which the tip was superficial to the occipital fusion plate. Given that event, subsequent leads were rarely placed over the fusion hardware but more lateral and, in 1 patient with laterally placed plates, placed in a vertical rather than a horizontal fashion (Figure 1). In 1 patient, an infection occurred at the site of the implantable pulse generator, necessitating removal and reimplantation 3 months later.

### TABLE 1. Demographic Characteristics of Chiari Malformation Patients Selected for Occipital Nerve Stimulation Trial

<table>
<thead>
<tr>
<th>Characteristic</th>
<th>Value</th>
</tr>
</thead>
<tbody>
<tr>
<td>Sex, n</td>
<td></td>
</tr>
<tr>
<td>Female</td>
<td>19</td>
</tr>
<tr>
<td>Male</td>
<td>3</td>
</tr>
<tr>
<td>Age (range), y</td>
<td>33.6 (14-54)</td>
</tr>
<tr>
<td>Location of pain, n patients</td>
<td></td>
</tr>
<tr>
<td>R suboccipital</td>
<td>2</td>
</tr>
<tr>
<td>L suboccipital</td>
<td>1</td>
</tr>
<tr>
<td>Bl suboccipital</td>
<td>19</td>
</tr>
<tr>
<td>Etiological factors</td>
<td></td>
</tr>
<tr>
<td>CM</td>
<td>22</td>
</tr>
<tr>
<td>Ehlers-Danlos syndrome</td>
<td>14</td>
</tr>
<tr>
<td>Tethered cord syndrome</td>
<td>4</td>
</tr>
<tr>
<td>Epilepsy</td>
<td>2</td>
</tr>
<tr>
<td>Fibromyalgia</td>
<td>2</td>
</tr>
<tr>
<td>Arachnoid cyst</td>
<td>1</td>
</tr>
<tr>
<td>Pseudotumor cerebri</td>
<td>1</td>
</tr>
<tr>
<td>Duration of trial (range), d</td>
<td>6 (3-7)</td>
</tr>
<tr>
<td>Long term follow-up (range), mo</td>
<td>18.9 (6-51)</td>
</tr>
</tbody>
</table>

*CM, Chiari malformation.

*a One or more diagnoses are associated with the primary diagnosis of CM in all patients.*

![Operative experience in CM population](image)

**FIGURE 2.** Operative experience with a 2-stage approach for patient selection. CM, Chiari malformation; F/U, follow-up.
DISCUSSION

Given the success of ONS in treating a variety of clinical syndromes that present with occipital and suboccipital headache, it is reasonable to apply this technique to the CM population in whom such headaches are a frequent complaint. Our patient population is entirely a postsurgical one in that all of these patients have had 1 or more surgeries aimed at treating the underlying malformation but yet remain disabled by intractable, severe headache.

The presence of prior surgery in all of these patients posed a number of technical problems not usually present in other ONS populations. Multiple prior incisions and the presence of hardware, including fusion constructs and shunt tubing, required great attention to placement of the incisions, lead tips, extension leads, and generators. Nonetheless, given that ONS is a low-risk, entirely subcutaneous procedure, there was no permanent morbidity in any patient from these procedures.

Whereas we attempted to select patients in whom occipital headaches were the primary symptom, the frequent coexistence of constant headaches in the frontal region suggests that some of these failures may be due to suboptimal pain relief in regions not covered by the ONS. Indeed, 1 patient in this series who failed the ONS trial ultimately underwent a bilateral supraorbital stimulator trial and permanent placement with good results at the 2-year follow-up.

The use of occipital stimulation to treat headache syndromes was first described by Weiner and Reed in 1999, and subsequent reports have demonstrated its use and efficacy in a variety of cranial neuralgias and headache syndromes. Although the mechanism of efficacy of ONS in the treatment is still unclear, modulation of nociceptive processing at the level of the trigeminocervical complex (TCC) of the caudal medulla/upper cervical spinal cord is thought to play a significant role. Dorsal horn neurons in the medullary TCC have been shown to respond to input from both trigeminal nerve branch stimulation (ie, supratentorial dura mater) and stimulation of greater occipital nerve afferents (ie, cervical skin overlying the C2/3 dermatomes). Chronic headache may thus represent a process of central sensitization of second-order afferent neurons as a result of chronic trigeminal afferent hyperactivity, with the common clinical observation of neck pain in association with headache a manifestation of the physiology of the TCC.

A process of central sensitization may also explain the persistence of these headaches despite appropriate treatment of the underlying condition via suboccipital decompression.
cranioplasty, shunting, or fusion. Stimulation of the occipital nerve afferents may thus result in pain relief by modulating the nociceptive processing at the level of the TCC, with ONS ultimately effecting central changes in supratentorial structures, as demonstrated in functional imaging studies of patients undergoing therapeutic ONS for chronic migraine headache.

### CONCLUSION

Occipital nerve stimulation appears to be successful in treating a subset of patients with CM with refractory occipital headaches. The unique nature of these patients has necessitated occasional modifications to the surgical technique to minimize device-related complications, which remain frequent but not serious. Optimizing the results of this technique in this series of patients clearly requires a deeper understanding of the varying causes of headache in this population, which would presumably allow further optimization of patient selection criteria and surgical technique.

### Disclosures

The operative techniques described in this article were initially displayed in part at the Pain Section of the 2008 Annual Meeting for the Congress of Neurological Surgeons.

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### TABLE 2. Complications and Revisions of Occipital Nerve Stimulation in Chiari Malformation Patients

<table>
<thead>
<tr>
<th>Patient</th>
<th>Age, y/Sex</th>
<th>Previous Medical History</th>
<th>Pain Location</th>
<th>Complication Description</th>
</tr>
</thead>
<tbody>
<tr>
<td>1</td>
<td>30/F</td>
<td>CM, Ehlers-Danlos syndrome</td>
<td>B/L</td>
<td>Lead migration</td>
</tr>
<tr>
<td>2</td>
<td>42/F</td>
<td>CM, Ehlers-Danlos syndrome</td>
<td>B/L</td>
<td>Repositioned generator from chest to abdomen because of uncomfortable position</td>
</tr>
<tr>
<td>3</td>
<td>27/F</td>
<td>CM</td>
<td>B/L</td>
<td>Lead migration</td>
</tr>
<tr>
<td>4</td>
<td>38/F</td>
<td>CM, epilepsy</td>
<td>B/L</td>
<td>B, Revision for uncomfortable position</td>
</tr>
<tr>
<td>5</td>
<td>19/F</td>
<td>CM</td>
<td>B/L</td>
<td>Wound revision for stitch abscess</td>
</tr>
<tr>
<td>6</td>
<td>22/F</td>
<td>CM</td>
<td>B/L</td>
<td>Wound infection</td>
</tr>
</tbody>
</table>

*B, bilateral; CM, Chiari malformation.*

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### FIGURE 4. Lead migration of a single lead.
Surgeons, and the presentation was a recipient of the 2010 Robert C. Erwin literary award. Preliminary results were presented as a platform presentation at the 2010 American Society for Stereotactic and Functional Neurosurgery meeting. Dr Mogilner has received grant support, consulting fees, and honoraria from Medtronic Neurological and has received grant support from St. Jude Neuro-modulation. The other authors have no personal financial or institutional interest in any of the drugs, materials, or devices described in this article.

REFERENCES


COMMENTS

The authors report a retrospective series of 22 patients with previous surgical treatment of Chiari malformation undergoing bilateral or unilateral occipital nerve stimulation (ONS) for refractory headaches. This work is interesting and addresses a common problem in neurosurgery. Management of chronic pain in patients with Chiari malformation can be challenging, and new modalities are needed to manage these patients. Occipital nerve stimulation is not new, and its use for the management of Chiari patients has previously been described. However, the authors’ contribution is important because of the significant size of the case series with mid- to long-term follow-up and a well-documented rate of complications.

Occipital nerve stimulation has been attempted in a number of headache syndromes, including cluster headaches, migraine, and post-surgical or posttraumatic pain. A large clinical trial has been reported on ONS for migraine headaches, with approximately 30% reduction in days with headaches and a 1.5-point mean reduction in pain measured on a scale of 0 to 10 in the group receiving adjustable ONS (Saper et al). Approximately 40% of patients were responders to ONS. A significant incidence in hardware complications with lead migration was reported, consistent with the findings of the present study.

Occipital nerve stimulation is a promising modality for the management of refractory headache disorders. The results of this study point to the potential value of the technique for patients with persistent pain after surgery for Chiari. Occipital nerve stimulation may become a treatment option to be discussed with these patients in addition to conventional pain management. The significant rate of implant failure and migration is still a limitation to the routine use of this modality. Implantation of ONS with paddle leads, with or without mesh, allows direct anchoring to the surgical site and should be considered an option to reduce migration rate. This option is particularly interesting in patients undergoing permanent implantation under general anesthesia.

André Machado
Cleveland, Ohio

Occipital pain in patients with Chiari malformation (CM) is frequently considered an indication for surgical decompression, and it is always very frustrating (for the patient and surgeon alike) if the pain persists after adequate decompression. Unfortunately, there is no good explanation why this occurs. If one postulates that headaches are caused by increased pressure at the level of cranio-cervical junction, then the posterior fossa decompression should relieve them in all cases. Other explanations for post-CM decompression headaches, including compression or injury of the occipital nerves caused by the surgery itself, may be the case in some patients, but would not explain those instances when preoperative pain does not improve at all, its pattern remains unchanged, and there are no sensory deficits. Same with the theory of dural scarring; The pain occurs before any scar is formed and happens even in those patients who did not have any dural closure or whose dural defect was patched with allogenic or xenogenic substitutes.

The introduction of peripheral nerve stimulation (PNS) as a non-destructive approach to the management of neuropathic pain > 40 years ago and the more recent development of a percutaneous PNS technique have changed our surgical decision-making algorithm. With the wider acceptance of the PNS modality, it is now being used to treat pain in many chronic pain conditions, including post-surgical and post-surgical neuropathy, post-heretic neuralgia, and complex regional pain syndromes. One of the most common PNS applications, ONS, has been used successfully to treat migraines, occipital neuralgia, cluster headaches, and even fibromyalgia. It is therefore not surprising that PNS has been considered for the treatment of pain after CM decompression, and the experience of these authors conclusively shows that it works in the majority (59%) of patients in whom it is tried, with an 87% success rate among those who passed a week-long trial. From these results, I believe that ONS should be strongly considered for patients with persistent postocipital decompression headaches, and like many others, I remain enthusiastic that at some point this modality will be officially recognized and approved.

In the meantime, these authors and the rest of us who use this modality are forced to use devices that are not intended for this particular
application. Therefore, the high rate of complications with 40% of patients requiring reoperations is not surprising. Once devices specifically designed, approved, and marketed for ONS become available, the rate of complications should decrease. However, one has to keep in mind that, similar to the authors’ experience, both our group and others were able to resolve all these complications with minimal morbidity and no long-term consequences.

The authors’ point about the need for larger and longer-term studies is probably the most important because this is the only way for this modality to become safer and more available to the patients who need it. As a first step in this direction, I would encourage the authors to continue similar thorough follow-up of their implanted cohort. Perhaps the next step will be to use this sizeable clinical experience to research the mechanism of PNS action and then use this knowledge to further refine the indications and selection of best responders.

Konstantin V. Slavin
Chicago, Illinois


This series of 22 patients receiving ONS for refractory head pain/headache after decompressive craniectomy and potential fusions for Chiari malformations is a welcome addition to the current literature because it represents a reasonably large number of patients followed up for an average of > 1.5 years and addresses a fairly common problem in this neurosurgical therapy. These patients are often quite disabled by their pain and have done well from their Chiari malformations symptoms otherwise, making this difficult-to-treat result frustrating for all involved. Results here were evaluated by a Visual Analog Scale and whether the device is still used or explanted, culminating in almost 90% having significant improvement if they passed an initial trial. Keep in mind that such trials allow an interventional therapy, like ONS, to be evaluated without full commitment of cost and resources. This is very unusual but exceedingly helpful in this current climate of cost containment and value in medicine.

Others have tried this approach in smaller series, and it would be helpful of course to have blinded randomized patients in all studies, but like most things we do in medicine, this is impossible to obtain with stimulation unless the authors consider subclinical stimulation in a study, perhaps at a later date. Ultimately, however, this work and others using ONS, and peripheral nerve stimulation in general, will garner enough understanding of underlying mechanisms of benefit and the appropriate parameter space required so that specific device designs and reimbursement approval can come about.

Jeff Arle
Burlington, Massachusetts