Background
Cluster Headaches (CH) are classified as a primary headache with a prevalence 0.1-0.9%. They present with repeated attacks of excruciating severe unilateral headache pain usually occurring several times a day. Most sufferers with chronic cluster headache (CCH) have an unremitting illness that requires daily preventive therapy and in many cases abortive therapy for years. Increasing evidence shows that neurostimulation of peripheral afferents of C1 – C3, as in occipital nerve stimulation, may benefit cluster headache sufferers by affecting the trigeminocervical complex, which is thought to be closely involved in the pathophysiology of the cluster headache (Jasper and Hayek, 2008).

Chronic cluster headaches, and intractable cluster headaches, represent a very significant personal health, quality of life, and economic issue. One treatment modality for chronic refractory headache involves the use of subcutaneous implanted neurostimulator leads in the occipital region. Cluster headache sufferers have been studied with peripheral nerve field stimulation and found responsive to stimulation of the sub-occipital region, known commonly as occipital nerve stimulation (ONS).

Objective
Evaluate the effectiveness of ONS as treatment, Preventive / Abortive, for the chronic cluster headache, and the intractable chronic cluster headache.

Case Report / Study Design
Conduct a systematic review of the collected data and results obtained from the occipital nerve stimulation as treatment for intractable chronic cluster headache.

Method
I was the study subject, a 49-year-old (year 1960) male with no other co-morbid medical conditions. I’ve suffered from intractable chronic cluster headaches averaging 8 Episodes daily for over three years. These Episodes failed to respond to the entire formulary of the most frequently prescribed preventative medications.

I achieve some relief from the Ganglionic Local Opioid Application (GLOA with Buprenorphine / Bupivacaine) block but they lasted on average only a week.

I had a more favorable response to the occipital nerve blockade, (ONB) but an adverse reaction to continued use of the ONB (betamethasone dipropionate and rapid acting salt of Betamethasone disodium phosphate mixed with 2% xylocaine and lidocaine) made that impossible.

We discontinued this method of intervention when the reaction became apparent. It was also accompanied by a reduction of endocrine levels, (Listyo, A. Ott, Kantonsspital St. Gallen, Department of Pain therapy, Switzerland).
Standard abortive drug treatments as well as other interventional procedures proved only marginally effective. Only oxygen therapy at a flow rate of 15 liters/minute proved effective most of the time. Although Swiss law currently prevents the prescription of higher oxygen flow rates that support hyperventilation, I’ve recently tried the demand valve method of oxygen therapy as it easily provides the minute volume of lung ventilation needed to support hyperventilation. I have found that hyperventilating with this method of oxygen therapy until I reach respiratory alkalosis with mild symptoms of paresthesia, provides significantly greater efficacy with faster abort times in aborting my acute cluster headaches than oxygen therapy at flow rates of 15 liters/minute.

After a close look at the narrow choice of alternatives with my neurologist, (D. Flügel Kantonsspital St. Gallen, Department of Neurology, Switzerland), we selected occipital nerve stimulation as the best option for a comprehensive alternative treatment.

During the first surgery, my doctors positioned the two 8-contact percutaneous leads shown below on the left, bilaterally over the two greater occipital nerve branches as shown in the X-Ray above. This was accomplished under general anesthesia (J. Y. Fournier, M. Land, Kantonsspital St. Gallen, Department of Neurosurgery, Switzerland). This surgery was originally scheduled to be conducted under local anesthesia so I could be awake and respond during the lead implantation. Unfortunately as luck would have it, I suffered a violent cluster headache attack that left the neurosurgeons and operating room staff white faced and shaking. On top of that they were unable to locate a sterile non-rebreathing mask for the oxygen therapy that was planned for just such an event.

After a week of trials with an external pulse generator, my doctors surgically implanted a rechargeable pulse generator in my abdomen. I used a remote control unit, shown below on the left, to activate the implanted pulse generator by holding it over my abdomen and pressing the appropriate function button.

During the study period we made several adjustments to the preset ONS program parameters in order to achieve a higher efficacy (A. Wenk Lang SR Neuromodulation, Switzerland, Boston Scientific). As the ONS system in its present instantiation is essentially reactive in nature, it must be activated when the user senses a cluster headache attack is starting. Accordingly, I also implemented a timing routine to activated one of the preset programs to see if we could improve efficacy during sleep. This proved ineffective by triggering when no attack were present missing the actual episodes.

During the review period of the occipital nerve stimulation, I was also treated at Kantonsspital St. Gallen, Department of Endocrinology (M. Brändle, H-U Mellinghoff, Kantonsspital St. Gallen, Department of Endocrinology, Switzerland) for an adrenal gland insufficiency that appeared concomitant with the adverse effects of the occipital nerve blockade. This treatment did not appear to have any effect (positive or negative) on the operation or effectiveness of the ONS, or on the frequency and intensity of my cluster headaches.
Results

A review of data I collected over the year since the implant was activated revealed an initial moderate efficacy of 63% (5 out of every 8 episodes responded to the ONS, while the remaining episodes required bailout abortives). After we made the adjustments to the preset stimulation program, the efficacy of the ONS gradually improved to 85%, but only during daytime when I was awake. Efficacy of the ONS during sleep was marginal at best.

CH attacks that occur during sleep frequently escalate to high pain levels by the time they wake me up. At that point the ONS was rarely effective. This forced me to use oxygen therapy and when that wasn’t effective after 20 minutes, I resorted to the triptan-based bailout abortives.

ONS has only marginal effectiveness during sleep. Although the continuous signal required for an effective prophylactic effect can be modulated to relatively low levels, even then, it still represented a source of irritation significant enough to make restful sleep impossible. The effectiveness of ONS during meetings is also marginal due to the distracting effects of the stimulating signals.

There are also times when ONS signals cannot block my cluster headache pain. When this happens, my first abortive of choice is oxygen therapy with a portable unit at a flow rate of 15lt/min. When oxygen fails to abort the CH attack, I’m compelled to use triptan based bailout abortives. I’ve also found that head position can be a factor in the effectiveness of the ONS. Sometimes even the slightest head movement creates tension on the stimulation leads causing them to be repositioned with respect to the underlying occipital nerve. This reduced the effectiveness of the ONS at times, and on rare occasions, this appeared to actually help precipitate cluster headache attacks.

I refrained from using any of the preventive medications during the yearlong test period and only used abortive medications in extreme situations, or when oxygen therapy was unavailable as an abortive. The bailout abortive medications I used included Zomig™ (zolmitriptan) nasal spray and Sumatriptan-Mepha® subcutaneous injection pen.

Conclusion

ONS in its present instantiation can be effective while the user is awake, but not while sleeping. It is a very expensive and invasive method of intervention that requires constant user attention to be effective. ONS is clearly not for every intractable chronic cluster headache sufferer unless they are technically inclined and willing to endure constant and ongoing adjustments to achieve optimum ONS functions capable of reliably preventing and aborting their cluster headache attacks.

The ONS system shows great promise as a treatment of last resort for intractable cluster headaches. However, having lived with it for a year, it is clear to me that it needs additional research to investigate the feasibility of implementing a sensor function similar to a cardiac pacemaker that senses electrical impulses of an irregular heartbeat and reacts accordingly. If such a function could be implemented using the stimulation leads to sense impending cluster headaches, and a program developed to process that signal in order to trigger selected ONS functions, this system could provide efficacy while sleeping and lessen the level of user attention required while awake.
**Recommendations**

Although the ONS system holds great promise as a method of intervention for intractable cluster headaches and it is clearly worthy of continued investigation through controlled studies with patients willing and capable of enduring lengthy periods of adjustment, I cannot recommend it for use as a treatment of last resort until it can provide meaningful efficacy during sleep, with a reduced level of user attention during waking hours. It is not ready for prime time use.

In addition, although my experience with the demand valve method of oxygen therapy is anecdotal at best, this method of oxygen therapy has proven to me, and many others who use it here in Europe and the US, to be a very effective method of aborting cluster headache attacks.

Given this method of oxygen therapy continues to be superior to lower oxygen flow rates that do not support hyperventilation in aborting my cluster headaches, the fact that it minimizes total exposure time to 100% oxygen, it's non-invasive with no real side effects, and is clearly more cost effective, I recommend this method of oxygen therapy be given the highest priority for further clinical studies.

In short, the demand valve method of oxygen therapy appears to be a very effective method of aborting cluster headache attacks that is ready for prime time and only needs minimal medical proof to become an evidence-based alternative to the far more invasive and expensive triptan-based abortives prescribed today.

*Know! This world is a very narrow bridge. The important thing is not to be afraid.*

--Reb Nachman of Breslov (1772-1810)