Surgical Treatment of Chronic Headaches and Migraines

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To my patients, residents, colleagues, and staff at the University of Wisconsin, this book is possible because you all contributed to it.
To my mentors in Madison, Cleveland, Pittsburgh, and Cairo, for believing in me and being the role models that you are.
To my wife, Iman, for your unconditional dedication, love, and support.
And to our kids, Mariam, Sara, and Omar, you are the greatest source of joy and motivation, and I am looking forward to many more miles traveled, giggles shared, and tournaments conquered.

– Ahmed M. Afifi

To my mentors and colleagues for their wisdom, patience, teaching, and experience.
To my parents who taught me the value of hard work, perseverance, and drive.
To my children, Simon, Charlotte, and Eveline, who are a daily source of light and joy.
To my wife Anne, the love of my life, who fills more buckets than anyone and shares my hopes and dreams as her own.
I am grateful for and love you all more than you know. This book doesn't exist without you.

– Ziv M. Peled

To my wonderful wife, Emily, for her unwavering love, support, patience, and understanding.
To our phenomenal children, Jackson, Brinkley, and Holden, who are the source of my inspiration, and whose smiles and joy make the sun rise every day.
To my patients, whom I serve gladly and to the best of my ability in the never-ending quest to improve their quality of life.
To my friends, colleagues, teachers, and mentors, who have guided and supported me every step of the way. I am eternally grateful.

– Jeffrey E. Janis

Foreword

Listening to patients and learning from their observations led me to research and describe the contemporary surgical techniques for the treatment of chronic headaches going back to late 1999. The investigations to prove the concept and improve the results have continued over the past 20 years. The evolution of this field has been the most exhilarating aspect of my professional career. Sharing the information I have gleaned with others throughout the world and developing a whole new subspecialty of plastic surgery where surgical talents can be applied to eliminate dreadfully painful condition have become the most fulfilling part of my life.

For those who do not suffer from migraine headaches, understanding the magnitude of what these patients endure is difficult. Even minor headaches can ruin an entire day. Those who writhe at the insignificant, yet annoying, ache can appreciate the enormity of what these patients who have chronic, daily migraine headaches tolerate. Those of us who care for these people repeatedly hear from them that we are their last resort, and should we fail to help them, they prefer not to continue living. The role of the plastic surgeon in eliminating deformities to render affected individuals presentable to society, help them enjoy their lives, and be accepted by their peers has been recognized to some degree. However, prevailing opinions about plastic surgeons are that they deal mostly with vain and frivolous concerns. Elimination of pain through peripheral nerve surgery has not been acknowledged by the lay public to the degree that it should. I am hopeful that by propagating information about migraine/headache surgery, the public will become more aware of how great an impact plastic surgeons can make in altering the quality of life of so many patients.

It is remarkably gratifying to see so many younger colleagues enthusiastically embrace this surgical field. They are not only eager to learn and implement these headache operations but seem to be emboldened with a sincere desire to improve the lives of so many. The growing number of surgeons keen to distribute their personal knowledge and experience through this book and previously published books is a resolute testimony to the efficacy of the operations described herein, despite initial disbelief among some of our other medical colleagues. Over the years, collectively, we have offered enough evidence for those skeptics to become convinced that our success neither is a placebo effect nor is it transient. We now have patients who have been followed for close to 18 years with no migraine symptoms and no chronic headaches. There are very few surgical procedures that have been scrutinized

through rigorous studies including a retrospective analysis, prospective pilot study, prospective randomized trial with comprehensive inclusion of different trigger sites, and prospective, randomized study with sham surgery and 5-year follow-up. Through basic science studies involving plastic surgery, neurobiology, and proteomics departments at Case Western Reserve University, we have also demonstrated structural pathology in the peripheral nerves of migraine patients compared with asymptomatic patients. Similarly, recent discoveries from the neurology literature strengthen these findings. Collectively, these results form the foundation for the peripheral concept of chronic headaches, which is now being revisited by some open-minded neurologists.

While initial skepticism could have dissuaded us from our efforts, a firm belief in the validity of the peripheral concept, steadfast determination, and unwavering commitment, fueled by continual positive outcomes, had led to the development of multiple procedures that reduce or eliminate migraine headache pain. We will continue this work and hope that by publishing this text, we can entice more surgeons to incorporate this surgical technique into their practice, with the aim of helping more and more patients, both nationally and internationally.

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Introduction: The Surgical Treatment of Chronic Headaches and Migraines

The contributors to this book consider the surgical treatment of chronic headache and migraine patients a true labor of love. We have seen first-hand the devastation that these types of chronic headaches have on patients, their loved ones, as well as their relationships with the outside world. Moreover, when studying this condition in detail, the magnitude of the problem both in the United States and worldwide becomes truly staggering. It is for these reasons that the improvement in those same peoples' lives following these operative procedures is both so dramatic and rewarding. At the same time, it has allowed us to be at the forefront of a new field within plastic and peripheral nerve surgery, one that continues to evolve, and as such, is ripe with innovation.

We have several goals with this text. The first is to instill in the reader a true understanding of the problem being treated. Second, we hope that future headache surgeons understand the clinical workup of these patients, the relevant anatomy, and the surgical approaches to the various nerves and structures that are the targets for intervention. Third, and perhaps just as importantly, we hope to impart to others the true, substantial, positive impact these concepts and techniques can have on a patient's life. What is described herein is not "experimental or investigational," but rather is the summary of over 20 years of clinical experience and published, peer-reviewed scientific evidence that supports the surgical treatment of headaches. The patients' outcomes and the improvement in their quality of life speak for themselves. You can help make a difference in patients whose hope may be waning as they live lives of chronic pain. We hope you derive as much from the following pages as we have gotten from writing and editing them. We would also like to acknowledge the people at Springer, especially Asja Rehse, for sharing our vision and helping to bring it to life.

Madison, WI, USA — Ahmed M. Afifi, MD
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1 Pathophysiology and Diagnosis of Nerve Compression Headache

Pamela Blake

Background

Successful reduction of chronic headaches by decompression of the occipital nerves was described in the 1990s [1, 2], and studies in the early 2000s similarly reported significant clinical outcomes with decompression of the trigeminal branches of the forehead and temple [3, 4]. Despite these promising results, widespread adoption of nerve decompression surgery was slow to gain acceptance by practitioners of Headache Medicine due to numerous factors, including the lack of evidence of the mechanism of extracranial processes in chronic headache, the presence of only a single placebo-controlled study demonstrating efficacy [5], and, most importantly, the contradictory nature of the concept of extracranial factors as causative in chronic headache with the prevailing theory of the intracranial etiology of migraine [6]. The grip of the intracranial theory as a cause of all headache is loosening, however, as evidence emerges regarding the role of the periphery in chronic headaches, in particular. These advances are due to important new research findings as well as to the development of effective treatments including botulinum toxin [7], peripheral neuromodulation [8], and calcitonin gene-related peptide (CGRP) antagonists [9, 10], all of which are directed at peripheral structures and mechanisms. In this more favorable environment, the concept of extracranial nerve compression as a factor in chronic headache will likely gain a greater foothold, and surgical decompression of involved nerves will find greater application. The most critical step toward a good surgical outcome remains correct patient selection, and thus the ability to correctly detect nerve compression in the patient with chronic headache is paramount.

Since the diagnosis of nerve compression headache follows directly from an understanding of the mechanism of the condition and the distinction of the resulting pain from other causes of headache, a useful point at which to begin consideration of this matter is to review the current concepts of chronic headache, the pathophysiology of nerve compression, and the clinical presentation of nerve compression headaches.

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Current Concepts of Chronic Headache and the Pathophysiology of Nerve Compression Headache

The criteria for each of the primary headaches (i.e., headache arising from the structures of the head and neck) can be found in the *International Classification of Headache Disorders*, third edition (ICHD-3) [11], the widely employed headache classification system generated by the International Headache Society. The ICHD has organized the types of headaches into categories based on common presenting symptoms; it does not, generally, speak to the etiology of the cause of the headache disorder. *Episodic migraine (EM), in* which there are truly pain-free days in between infrequent attacks, is the most common cause of recurrent headaches. Episodic migraine, particularly when associated with aura, is generally considered to be a disorder of the brain and attributable to dysfunction at the neuronal level [12]. Migraine attacks are comprised of distinct clinical stages including (1) the prodrome, a period of up to 24 hours prior to a migraine attack, characterized by fatigue, mood changes, and gastrointestinal symptoms; (2) the aura in some attacks, considered to be most likely due to a cortical phenomenon called spreading depression, associated with a gradually moving wave of depolarization that begins at the occipital cortex and commonly results in visual, sensory, and language disturbance; (3) the headache, which is often frontal and throbbing in nature and must be associated with either nausea (with or without vomiting) or photophobia and phonophobia; and (4) the postdrome, a period of fatigue, cognitive changes, and malaise which may last for 24–48 hours following a migraine and which can result in significant disability. Episodic migraine is defined as fewer than 15 days of headache per month; migraines most commonly occur four or fewer times a month [13]. The interictal period is thus a pain-free period with regard to the head and the neck, although the interictal migraine brain is characterized by hypersensitivity to stimuli and an increased risk of mood disorders and often a sense of fear of the next headache attack.

Chronic migraine (CM) is described as headache occurring on 15 or more days/month for more than 3 months, which, on at least 8 days/month, has the features of migraine, including sensitivity to lights and sounds or nausea and/or vomiting, and also at least two of the following: unilateral location, pulsating pain, moderate-severe intensity, and aggravated by routine physical activity. The migraine may also be accompanied by fully reversible aura symptoms which may include disruptions in visual, sensory, speech/language, motor, brainstem, or retinal function. The aura symptoms should spread gradually, usually resolve within 60 minutes, and are usually followed by headache. The characteristic pain of the headache of migraine, while not specified in the ICHD, is typically frontal and throbbing, reflecting neurogenic inflammation of the trigeminovascular tree. CM is usually considered to develop from EM; the phenomenon of progressively increasing frequency in EM to the point of being daily or near-daily has been described and was previously termed "transformation of EM to CM," and rates of such transformation of 2.5% of EM patients per year have been reported [14]. It has been noted that a large percentage – about 70% – of individuals who meet the criteria for CH will experience remission of CM and revert back to EM when followed longitudinally [15]; the remaining approximately 30% will have persistent CM.

A more common chronic headache is *chronic tension-type headache (CTTH)*. CTTH, as opposed to migraine, is characterized as a bilateral headache of pressing or tightening quality, usually of mild-moderate intensity that is accompanied by neither nausea and/or vomiting nor sensitivity to lights and sounds. CTTH has not been as well studied and even less is known about the pathophysiology of CTTH than is known about migraine. The first population survey of chronic headaches in the United States [16], published in 1998, reported the prevalence of daily or near-daily headaches to be 4.1%. CTTH was the most common diagnosis of chronic headaches, present in 53% of the population with chronic headaches and representing a total of 2.2% of the general population. CM, on the other hand, represented about one third of the patients with chronic headaches and about 1.3% of the overall

population in this study. This figure, incidentally, is close to the 0.9% prevalence of chronic migraine reported in a later large population study [17]. The primary feature of chronic headache is the refractory nature of the pain; unlike episodic migraine, which generally responds well to acute interventions such as triptans and antiemetics, chronic headache presents significant disability to those afflicted [18] and a significant challenge to healthcare providers.

Another relevant headache condition included in the ICHD is the new daily persistent headache (NDPH). This condition is characterized solely by the presence of unremitting pain for more than 3 months, the onset of which is distinct and clearly remembered. As with the ICHD categories for CM and CTTH, no characterization of the pain of NDPH is stated with regard to location or character of pain.

The pathophysiology of primary headache disorders has been the subject of substantive and ongoing research. Clear pathophysiologic data, however, is available for only one type of headache, namely, migraine with aura. The ability to trigger aura in the laboratory setting and observe with sophisticated functional imaging the neuronal changes occurring at the cortical level has added greatly to our understanding of this most disruptive of headache disorders [19]. The migraine aura is now reliably attributed to cortical spreading depression, a depolarizing response of the cortex to noxious stimuli. Similar pathophysiologic information is not available, however, for other types of headache such as CM or CTTH, although recent anatomic studies have led to fascinating insights regarding the role of extracranial tissues in chronic headache.

An important study published in 2009 reported the presence of a network of intracranial trigeminal afferents in pup and adult mice crossing calvarial bone and suture lines to terminate in the extracranial space [20]; in the adult mice, the nerve fibers were only visualized in the suture lines, suggesting that the nerves in the bone degenerated as the pup mouse aged. The staining patterns of these fibers indicated that they are important for both sensory and pain functions. Corroborating findings were later reported in the rat and human skull in 2013 [21] and 2014 [22]; in these works, the extracranial nerves were found to be branches of the mandibular nerve, the third branch of the trigeminal ganglion [21, 22]. Finally, it was recently reported that the dura overlying the cerebellum is innervated by neurons in C2–C3 ganglia that contain CGRP and which enter the intracranial space through bony canals and foramina of the occipital bone [23]. This network of nerves demonstrated a pathway by which extracranial forces or stimuli may reach the intracranial compartment and the spinal trigeminal nucleus of the brainstem and ultimately give rise to migraine-like headaches. An inciting stimulus for the activation of pain pathways, however, remained elusive, as did evidence of extracranial pathology.

In 2016, the first evidence of extracranial pathophysiology in patients with chronic occipital headache was reported, when biopsies of the occipital periosteum from this population revealed a significant upregulation of genes that code for inflammatory proteins [24]. This important finding represented the first evidence of extracranial pathophysiology in chronic headache and indicated a robust proinflammatory environment in the occipital periosteum in these patients. It has previously been demonstrated that compression of peripheral nerves leads to an inflammatory response around the involved nerves in animal studies [25, 26], and a similar brisk inflammatory response around compressed nerves has been described in human subjects [27]. A theory is thereby emerging in which anatomic compression of extracranial nerves results in an inflammatory state leading not only to local pain and tenderness but also to radiation of pain to more distal, frontal locations on the head via activation of the trigeminovascular tree through trigeminal afferents crossing suture lines. Consistent with this theory, decompression of the occipital nerves was again reported to be helpful [28, 29] for patients presenting with a variety of headache presentations and with enduring benefit. Our knowledge of the pathophysiology of extracranial nerve compression and the types of associated pain has thus advanced significantly, although much remains to be learned.

The diagnosis and management of headache disorders is ideally conducted by a specialist in Headache Medicine. Such specialists are usually, but not always, neurologists; subspecialty board certification by the United Council for Neurologic Subspecialties (UCNS) is the highest level of certification in the discipline. The benefit of close collaboration of a Headache Medicine specialist with the surgeon performing nerve decompression surgery lies not only in the appropriate diagnosis and management of nerve compression headache but also in the diagnosis and management of other comorbid headache conditions as well as other medical and psychological conditions that commonly coexist with headache.

The identification of nerve compression headache is the first and perhaps most important step toward a good outcome with nerve decompression surgery. In the author's practice, the key feature of nerve compression headache has been the presence of daily, unremitting pain. In patients with occipital nerve compression, the pain may be predominantly or even solely in the upper neck (also referred to as the suboccipital space) and will often also radiate to the forehead and temples. In trigeminal branch compression, it will follow the distribution of the affected nerve. In our experience, nerve compression more often affects the occipital nerves than the trigeminal nerves, although both are possible and both may be present at the same time. The unremitting nature of pain reflects, and aligns with, the pathophysiology of nerve compression; constant pressure on a nerve would not be expected to result in intermittent pain. Many patients will describe a prolonged period of time, often years, during which their head pain will become progressively more frequent, likely reflecting progressive morphologic changes in the nerve in response to the compression [30]. It is possible that, prior to the development of unremitting pain, the patient may present experiencing episodic pain that might be triggered by certain positions or maneuvers. This could represent an intermittent compression of the nerves due to particular muscle engagement. It is important to determine the frequency of head and neck pain, even mild, in such patients. Migraine-like features are often present, and the response to migraine-specific acute medications such as triptans may be adequate or even good; however, the resistance of the headaches to treatments to prevent the headache and the generally progressive nature of the condition suggest the presence of nerve compression.

A question that surgeons commonly ask is "Which headache diagnosis should I operate on?". This is an important question, considering that the diagnosis is usually made by the neurologist(s) before the surgical consult. Patients with headaches due to nerve compression may meet the diagnostic criteria for chronic migraine, chronic tension-type headaches, or new daily persistent headaches [29]. In fact, most patients presenting for a surgical consultation will have had many diagnoses over the years. A formal diagnostic category for nerve compression headaches may be appropriate following further careful study of the entity.

Clinical Presentation of Nerve Compression Headache

The information presented herein is drawn from the experience of the author and based on treatment of over 1000 patients and 15 years of experience in the diagnosis and management of patients with headaches due to nerve compression, as well as close collaboration with surgeons performing nerve decompression surgery. Many patients have been followed for years following the surgery. Clinical studies confirming these observations are, however, necessary.

The clinical presentation of nerve compression headaches conforms to the pathophysiology of nerve compression. Many nerve compressions are persistent, and therefore the associated pain is generally an unremitting pain [28]. Such unremitting pain is considered to be a strong indicator of nerve compression. However, as with other nerve compressions in the extremities, the nerve compression could also be intermittent, and good surgical results have been achieved in individuals with episodic pain. Many

of the nerve compression headaches in this author's practice involve the occipital nerves, and this discussion will thus involve that entity to a greater extent. The *frequency* of the pain is thus usually daily, and the *duration* is usually constant and unremitting. It is very important to note that while the majority of patients will report unremitting pain, a significant minority will be unaware of the daily nature of the pain. This may occur particularly if the daily pain is mild and long-standing; additionally, patients often do not report neck pain in discussion about headaches. In such circumstances, education of the patient, as discussed below, and the maintaining of a careful log are needed to determine the true frequency of head and neck pain. If there are truly pain-free days, it is possible that episodic migraine may be present and treatment should be directed at that entity.

It is common for the pain of occipital nerve compression, in particular, to begin as episodic pain that gradually increases in frequency to the point of becoming daily; this gradual increase in pain from episodic to unremitting appears, in our experience, to occur in a majority of patients with nerve compression pain (and aligns with the "transformation" of EM to CM as discussed above). The remainder of patients with nerve compression headache present with an abrupt onset of headache, the date of which is often clearly recalled by the patient; such headaches may fulfill the ICHD-3 criteria for new daily persistent headache, the designation for a primary headache disorder which is abrupt in onset, becomes unremitting within 24 hours, and lasts for more than 3 months. NDPH may incorporate tension-type or migraine-like features, and the characterization of headache as NDPH, as noted above, has not designated any particular pathophysiology.

The *location* of the pain conforms to the anatomic distribution of the nerve and is the most important factor in deciding if the patient is a surgical candidate and determining the exact surgical site and sequence. This is more straightforward for trigeminal branch compression. The pain of supratrochlear and supraorbital nerve compression is generally at or above the level of the eyebrow and radiating into the forehead. The pain of zygomaticotemporal nerve compression is in the anterior temple and that of auriculotemporal nerve compression in the lateral temple and parietal area, above the ear or just behind the temporal hairline. The pain of occipital nerve compression can be much more variable and conforms not only to the distribution of the occipital nerves but also to trigeminal locations more anteriorly on the head, likely through the anatomic associations of the nerves in the extracranial compartment. Occipital nerve compression is usually associated with some degree of daily discomfort in the occiput or suboccipital space, representing the nerve within the nuchal musculature. This location of pain is often described by patients as "neck pain," and the examiner obtaining the history must be careful to include questions as to the presence of neck pain. From the subocciput and occipital space, the pain will often radiate directly along the pathway of the nerves: for the greater occipital nerve, ascending lateral to the midline on the posterior aspect of the head, often to the vertex, and even to the forehead, and for the lesser occipital nerve, superiorly in a retro-auricular location radiating to the temple. Occasionally, compression of the great auricular nerve is also present, causing pain of the posterior ear and immediate retro-auricular location. Greater occipital nerve pain may also radiate directly anteriorly and be felt as a retro-orbital pressure (generally perceived as pushing the eye forward). Clinical experience indicates that most patients with occipital nerve compression have some degree of pain in the anterior head, either the forehead, the temple, the anterior vertex, or the retro-orbital space; sometimes, the pain may be more intense anteriorly, and infrequently the head pain is perceived only anteriorly. In such patients, the presence of associated symptoms and aggravating factors of pain (see below) is indicative of occipital nerve compression. In the experience of the author, it is often common for individuals with chronic headache to have a low level of awareness of where precisely their chronic pain is located. Patients are often surprised by the attention we pay to determining the exact location and pattern of radiation of the pain, which is often different than previous consults they had over the years. It is helpful in such instances to conduct with the patient a review of the relevant anatomy and to ask the patient to maintain a careful log for 1 to 2 weeks, including in

particular the location of pain on the head and neck area. A follow-up visit to review such information, and also for the presence of previously unrecognized associated symptoms and aggravating factors, may add significantly to the history obtained at the original evaluation. It is also important to recognize that, on occasion, certain conditions such as attentional deficiencies or anxiety may interfere with a patient's ability to record or report such pain, and repeated questioning at follow-up visits will be helpful to elucidate a history that is consistent with nerve compression.

The *severity* of pain of nerve compression similarly varies. It is helpful to review initially the pain scale with patients so that consistent measures of pain are being recorded (in our practice, we place the pain scale on the reverse of the log). It is common for occipital nerve compression to be associated with daily mild pain in the occiput or suboccipital space; there may be anterior radiation of the pain a few times a week to the eye or forehead, and when such occurs, the pain intensity will usually increase. As with determination of location, it is not always immediately apparent that there is daily mild pain, and the careful recording of pain by the patient on a daily log after the initial visit is often the best way to elicit this history. Trigeminal branch compression pain may also range from mild to severe.

The *character* of pain of occipital nerve compression is often that of pressure, most commonly an imploding (pushing in) sensation in the occiput that may take on more of a throbbing quality as it radiates anteriorly. There will occasionally be neuropathic qualities to the pain at the occiput, with stinging, burning, or stabbing sensations, and when such occurs, it is usually superimposed on the baseline of constant pressure. Occipital nerve compression is often associated with the presence of allodynia, which is defined as the presence of pain generated by non-painful stimuli. The presence of occipital allodynia is often a prominent feature of occipital nerve compression. Allodynia is most often present with the placement of the head on the pillow, although the specific presentation may vary. Sometimes it is present immediately or early in the night; in others, it occurs later during the period of sleep. Questions may be posed to the patient to elicit this history in a number of ways, reflecting the different manners in which allodynia is experienced. These may include: Does it hurt to put your head on the pillow? Do you have to arrange your head in a certain way on the pillow to be comfortable? Do you awaken during the night with your head hurting and have to rearrange your head on the pillow to be comfortable? Do you ever wake up at night and find that your head hurts so much, with the pain aggravated by the pressure of the pillow, that it is necessary to sit up in bed or get out of bed altogether to be comfortable? This last phenomenon in particular may be pathognomonic of occipital nerve compression. It is important to eliminate the possibility that the patient is exiting the bed simply for pain medications or to use the restroom. The patient who exits the bed and then spends the rest of the night sleeping elsewhere such as a recliner or on a couch that does not exert pressure at the tender area of the occiput, or who remains in bed but so propped up by pillows that he or she is essentially upright, suggests a feature uniquely suggestive of marked occipital allodynia that is likely due to occipital nerve compression. Allodynia may also be experienced by resting the head against the back of a chair or a headrest, placing the head in a shampoo bowl, or with pressure on the head from a cap, helmet, or hard hat. Finally, manipulation of the hair such as brushing the hair or pulling the hair into a ponytail may be uncomfortable and will be avoided by a patient with occipital allodynia, although this milder form of allodynia may also be experienced with episodic migraine. Given the importance of this clinical feature, and the extended nature of the process to obtain the history, it is helpful to list questions addressing the varied manifestations on a questionnaire.

Associated symptoms of trigeminal branch compression often include typical migraine features such as photophobia and phonophobia, but these symptoms are not always present. The associated symptoms of occipital nerve compression generally can be considered in two categories, the first relating to involvement of cervical muscles and the trapezius muscle and the second relating to the common occurrence of symptoms also encountered in migraine headaches. Involvement of cervical

musculature is more indicative of an occipital nerve involvement, particularly if engagement of the muscles triggers or aggravates headaches, as will be discussed below. It is helpful to distinguish cervical muscle spasm or tightness, which usually occurs in the lower cervical muscles or the middle or inferior portion of the trapezius muscle, from the presence of pain in the suboccipital space, which is likely due directly to compression of the occipital nerves. It should be noted, however, that compression of the occipital nerves in the suboccipital space is often associated with muscle spasms that may radiate to the middle and inferior fibers of the trapezius. It may be difficult to distinguish what is likely the pain of nerve compression in the upper neck from spasm of the remainder of the trapezius. The symptoms of muscular involvement present as a constant tightness or spasm in the lower neck or upper back, and the onset of these symptoms has been observed by the author to occasionally predate by years the onset of the head pain; this may be indicated by a history of long-standing treatments with chiropractic manipulation or massage, either professionally or by family members. Patients with occipital nerve compression often report that their headaches "always begin in my neck" or even in the middle of the upper back, in the inferior fibers of the trapezius, medial to the scapula. It is often reported that tightness originates in the middle or inferior portion of the trapezius, with the pain then radiating up the posterior neck to the occiput and then often, but not always, onto the top of the head and the forehead. Patients are often able to draw this line of pain progression very specifically. Often the degree of tightness and discomfort of cervical and trapezius muscle spasm will parallel the intensity of the headache. Physical activities that engage the trapezius or other cervical muscles will often aggravate pain, and these activities are often of a daily nature, commonly including housework, vacuuming, lifting grocery bags, and prolonged use of a computer, particularly a laptop, which demands prolonged cervical flexion which will also engage the posterior cervical muscles. Involvement of the cervical muscles is a very common feature of occipital nerve compression, observed to be present in almost all patients, and while the absence of this symptom does not exclude the diagnosis of occipital nerve compression, it should cause one to consider the diagnosis carefully. The cervical muscles are generally not involved in trigeminal branch compression.

The cause of symptoms such as photophobia and phonophobia in the presence of occipital nerve or trigeminal nerve compression is unclear; however it seems likely that these symptoms of heightened central sensitivity are mediated via the same pathways involved in migraine and can occur due to the anatomic and functional connections between the extracranial occipital nerve and intracranial trigeminal nerve pathways as outlined above for individuals with occipital nerve compression and more directly from trigeminal pathways in individuals with trigeminal branch compression. Individuals with occipital nerve compression will often report that photophobia and phonophobia will typically occur as their pain travels into more frontal ("trigeminal") locations such as the forehead and the temple. Interestingly, pain that is perceived in the retro-orbital space seems less often to be associated with migraine-like features. Other very common symptoms of migraine include nausea and vomiting and also dizziness. These symptoms are very commonly encountered in occipital nerve compression, perhaps more often with lesser occipital nerve involvement, and often are as bothersome to the patient as the pain is. Finally, aural symptoms have been reported often in individuals with ON compression, particularly the lesser occipital nerves. These symptoms may include fullness, pressure, tinnitus, and auditory distortions, and they are often ipsilateral to the side of pain, or bilateral if the pain is bilateral, and they often subside with nerve decompression. The etiology of such symptoms is unclear; it is interesting to note that the mandibular nerve, the third branch of the trigeminal nerve, and the primary branch crossing the calvarial sutures to innervate the periosteum and extracranial space, gives off a branch to the tensor tympani of the middle ear While much research is required to understand the etiology of these symptoms, the co-occurrence of aural symptoms with lesser occipital nerve involvement, in the experience of the author, is specific enough to lesser occipital nerve compression that it should be a strongly suggestive feature of nerve compression if it is present.

Triggers of headaches are understood to be factors that incite the headache and thus are encountered before the onset of pain and lead an individual from a state of no pain to a state of having pain. These are distinct from aggravating factors (described below), which worsen underlying pain; it is possible and even common for a condition to be both a trigger for headache and an aggravating factor. As the pain of nerve compression is as often described as noted above as a constant, daily pain, aggravating factors have more clinical relevance. For the patients with episodic pain, triggers similar to those seen in migraine may cause headaches. These include menstrual period or other hormonal fluctuations, stress, weather changes, and strong sensory stimuli of smell, sound, or light.

Most of the patients who present with nerve compression headache do not report a clear event for the initial onset of their head and neck pain. When there does appear to be a triggering event for the onset of pain, it is most often trauma. Trauma may be defined broadly and may incorporate a number of events including a direct blow to the back of the head such as may be incurred during a fall, athletic play, or automobile collision or sustained firm pressure on the back of the head such as may occur during prolonged dental work or a surgical procedure. Indirect injuries that involve more of a whiplash-like forced flexion-extension injury can occur during an automobile collision or exposure to a blast injury in a military setting. In the setting of a traumatic injury with resulting head pain, it is important to note that pain in the distribution of the occipital nerves may be due to injury and inflammation of the nerves and may be amenable to treatment with steroids, whether oral or injected at the occiput, and, thus if seen acutely, does not always indicate the presence of a nerve compression process or that surgery is needed. This is similar to postcraniotomy headaches, which are discussed later in this book. If pain proves to be refractory to treatment after a period of several months, then nerve compression is likely present. It should be noted additionally that the surgical pathology of patients with post-traumatic headaches appears to be identical to those of patients who do not have a history of trauma. Other triggering factors that may be present at the onset of nerve compression headache involve strong immunogenic stimuli, such as significant stress or systemic illness. Emerging evidence of the presence of permanently altered neuroimmune development following early life adversity [31] may underlie the finding that headaches are much more common in individuals who have had adverse childhood experiences [32] such as emotional, physical, and sexual abuse, neglect, etc.

Aggravating factors for occipital nerve compression headaches commonly include pressure to the back of the head (as noted above, termed allodynia) and engagement of the cervical muscles or the trapezius. A history of these aggravating factors should be carefully sought as this is an important diagnostic feature. The movements that most commonly appear to cause pain include prolonged cervical flexion (i.e., looking down at a cell phone or a book), cervical extension (looking up to change a lightbulb, or extending the neck to allow vision through a bifocal section of eyeglasses), and turning the head to the side for a prolonged period of time (most often encountered in a meeting or a religious service). Trapezius muscle engagement, with either vigorous engagement, such as lifting heavy items (including groceries, small children and lifting weights as part of exercise program), or less vigorous but more prolonged actions (working at a desktop computer for several hours or driving with the hands at the '10 and 2' positions on the steering wheel) may also be a common trigger for worsening pain. Avoidance of such activities may be so long-standing and common for the individual with nerve compression that it is a normal part of life, and such behaviors may not be immediately reported as problematic. *Alleviating factors* of nerve compression headache include moving the neck and head out of an offending position and the application of ice to the occiput or the neck. Many patients will describe having a large collection of cool packs in the freezer which are routinely changed, as well as creative methods for the application of the packs and also for keeping a supply of cool or ice packs close by, such as in a cooler at the side of the bed. Additionally, afflicted individuals will often apply heat to the muscles of the neck and shoulders. In our experience, the practice of frequent application of ice or a cool source to the occiput or neck is highly suggestive of occipital nerve compression.

Trigeminal branch headaches are less likely to have aggravating or alleviating factors, and when such are present, they tend to be typical migraine-like factors such as strong sensory stimuli of bright light, loud noises, etc.

The *response to medications* is also an important aspect of the history. *Preventive medications* in general do not provide significant, sustained reduction in head pain frequency and intensity. Some medications, such as gabapentin and pregabalin, may provide a moderate reduction in pain (unpublished observation) for some patients. The recent entry of preventive medications targeting calcitonin gene-related peptide (CGRP), a pro-inflammatory molecule, to the pharmacopeia may provide more useful options, however, as these medications do address the inflammatory system and one which may be more relevant to the pathophysiology of nerve compression than is the stabilization of neuronal membranes accomplished by the centrally acting agents. Botulinum toxin injections administered in the chronic migraine protocol [7], in our experience, do provide a substantial benefit for approximately 40–50% of patients with nerve compression. Adjustment of dosage, especially an increase to the posterior neck and trapezius muscles for patients with significant cervical muscle spasm, may provide additional benefit. Furthermore, targeted injections of botulinum toxin to the areas of pain specific to the individual patient have also been reported to significantly reduce headache frequency and intensity at long-term follow-up [33]. The lack of central side effects and the reduction of compliance issues add to the appeal of botulinum toxin; however, the need for regular office visits and the usual brisk return of pain as the effect of botulinum toxin declines are relative disadvantages; additionally the presence of an alternative, more enduring solution in the form of nerve decompression surgery usually causes the treatment strategy of never-ending botulinum injections eventually to diminish in appeal. However botulinum toxin has a role in patients who are not medically fit or interested in surgery or while other medical and psychological conditions are being addressed. Moreover, as the pathophysiology of nerve compression and subsequent inflammation advances, the headaches of nerve compression may also continue to progress even as botulinum injections are being administered, and many patients who responded well initially to botulinum injections may find the response declining. In our experience, therefore, while botulinum toxin can provide for some patients a useful and well-tolerated treatment for nerve compression headache, it is generally not a satisfying long-term treatment strategy in patients for whom surgery is a viable option.

Acute medications for headache include analgesics and triptans. Most patients will have tried a variety of over-the-counter analgesics and will often find these to be helpful however impractical due to the constant daily nature of pain. It is not, however, uncommon to encounter a patient taking large quantities of OTC analgesics (e.g., 10–12 tablets a day) to achieve some degree of pain control. Gastric irritation and ulcers are often encountered, and it is not uncommon for patients to present for initial treatment of chronic headaches when NSAIDs are stopped by a gastroenterologist following a GI bleed. It is also not uncommon for patients to attain pain control only from the use of daily opiate medications, and the ongoing role of this treatment option requires very careful consideration, as will be addressed below. Triptan medications often do provide benefit. This factor – the presence of a triptan-responsive headache – will often suggest and support the diagnosis of migraine as the underlying headache pathophysiology and may lead to some confusion as to diagnosis. The responsiveness to triptans is, however, in line with the shared role of trigeminal nerve pathophysiology in even occipital nerve compression, however, and does not rule out the presence of occipital nerve pathophysiology. It is common for patients to report that triptans are more helpful for a frontal (i.e., trigeminal distribution) aspect of head pain and less so for occipital pain. Muscle relaxants may provide a useful treatment for occipital nerve compression and for some patients highly useful, presumably by addressing the muscular issues described above.

The *disability* of nerve compression headaches is an important aspect of the history that should be established early in the course of treatment, not only to determine the intensity of management that