Chapter 1.B. Periodic or Chronic Migrainous Neuralgia — Cluster Headache

An early dramatic description, Opperman’s [1747] doctoral thesis, is of a 35-year-old woman suffering from 15-min. attacks of hemicrania of agonizing intensity recurring every hour day and night for 5 years. At the onset there had been 2 periods of 4 weeks of such attacks with a 6-month remission between them. The remissions and the intensity and duration of the attacks, but not their location, were typical of this syndrome. A general physician, Muller [1813], first described the full-blown picture with typical frightful episodes in himself continuing for 34 years with onset at age 20. The spontaneous attacks lasting a few to 30 min began in the eyebrow and included photophobia, ‘inflaming’ of the conjunctiva and tearing, and a pounding temporal pulse [Isler, 1987]. Harris [1926] gave an excellent comprehensive description of this disorder under the rubrics of migrainous neuralgia, periodic migrainous neuralgia and ciliary neuralgia. He emphasized the differential features between this and essential trigeminal neuralgia. His last publication on the subject [Harris, 1940] describes briefly complete relief after 19 of 29 alcohol injections of the Gasserian ganglion with 5 other patients much improved and ‘less violent recurrences’ in the remaining 5. ‘Cluster headache’, the term introduced by Kunkle et al. [1952], has become the more popular designation for the disorder.

Clinical Features

These include as differing from trigeminal neuralgia: (1) spontaneous onset of the pains rather than provocation by local stimuli to specific facial areas; (2) a constant rather than paroxysmal character; (3) a duration of minutes to many hours; (4) a tendency to occur at a specific time of the day or night for a period with spontaneous changes in this schedule; (5) a greater tendency to waken the patient from sound sleep; (6) usually origin around orbit, forehead or temple, but often with radiation not only elsewhere on the face but also to ear, back of head, side or back of neck and shoulder; (7) tendency to remain unilateral in any one attack, but more frequent involvement of the second side in other attacks; (8) ipsilateral autonomic features are much commoner, mainly of parasympathetic hyperactivity appearing as ipsilateral redness of eyeball or cheek; dilation, pounding and tenderness of the superficial temporal artery, ipsilateral profuse tearing; watery rhinorrhea.
or nasal congestion and mucus; less often swelling of eyelids, cheek or nasal mucosa and rarely excessive salivation.

Sympathetic paresis is common in the form of miosis and ptosis. These signs may occur associated, oddly enough, with hyperhidrosis of the ipsilateral forehead when the pain is referred mainly to cheek and jaw, ‘the lower syndrome’ of Ekbom [1970].

Of 416 cases reported in 10 publications the pain spread to the other side during an attack in only 7, and three quarters of the patients were males. Roughly half of them [White and Sweet, 1969, pp. 348-354] provoke their attacks by drinking an alcoholic beverage. A small amount may suffice to do so promptly. The clinical features may change to those of classical migraine and vice versa.

Although these attacks tend to occur in ‘clusters’ for weeks or months and then stop for a time, the onset in a few patients is primarily chronic without remissions from the beginning. A larger group of patients stop having remissions, becoming secondarily chronic as the disease continues. On the basis of at least two attacks per week for a year Ekbom and Olivarius [1971] placed 10% of their 167 patients in this chronic category. It is mainly from this group that the surgical candidates are drawn, although in some patients each attack is so devastating that they are unwilling to await a remission and seek surgery.

The panoply of drugs now available for this disorder results in medical control in the great majority, e.g. Graham has needed to refer for surgery only 4% of his circa 550 patients. Having worked with him for the last 20 years, W.H.S. has a surgical series confined to the medical truly intractable cases. Indomethacin, which so promptly controls the variant described by Sjaastad and Dale [1976] as chronic paroxysmal hemicrania, should not be forgotten. However, as we shall be commenting frequently in this book, medical management is all too often being carried to undesirable lengths and the opportunity lost to return the patient to productivity by timely surgery. Thus, one of our patients persisted in taking the lone agent which relieved him, cafergot, until the coronary arterial disease it provoked required 2 open heart operations. He is now a cardiac invalid despite neurosurgical complete control of his pain.

The distinguished neurologist Sir Charles Symonds [1956] has stated that in these patients there is ‘no evidence... that psychological factors were operative’. Although this may well be true of the dramatic attacks themselves there are patients whose personality traits and social situations make the chance of surgical success too small to justify it. One of us cites details of 4
such examples in a forthcoming publication [Sweet et al., 1989].

Neural Pathways Activated during These Pains

Present data point to at least two afferent pathways in the cranial roots which may be taken by the impulses related to this pain: (1) trigeminal, and (2) nervus intermedius and conceivably glossopharyngeal and upper vagal. No one has as yet proposed any clinical feature which characterizes one route more than the other. Mayberg et al. [1984] have depicted afferent fibers from durai and cerebral arteries and venous sinuses in the cat which traverse the trigeminal first division; these could account for vascular pains related to intracranial vessels being referred to this area of the face as so commonly occurs in this disorder.

Temporary Trigeminal Denervation

Retrogasserian Lidocaine Block

The trigeminal pathway seems to be involved oftener than the intermedius-greater superficial petrosal alternative; hence we begin by trying to determine if the former is implicated by a selective block of the pain fibers in the trigeminal rootlets. The severity and relatively short duration of many attacks usually preclude placement of the needle to try to abort the attack once the pain has started. However, we think that the state of partial sensory loss of analgesia without anesthesia is not as well tolerated by patients with cluster headache as by those with trigeminal neuralgia. Moreover, first-division analgesia is more likely to lead to major dysesthesias in both groups of patients than is second- and third-division loss. Hence we think it advisable to let the patient sample the sensations both of analgesia only and as well that combined with loss of appreciation of touch. We seek also to provoke an attack once the needle is placed and then try with lidocaine to stop it, the crucial test of the value of differential or total rhizotomy. We have done such blocks on 36 patients, twice in 4 of them with a less equivocal result the second time in 2 of the 4. In 4 others, whose attacks were readily provoked by drinking alcoholic beverages, we have given oral whiskey or i.v. alcohol without ever initiating an attack. Ekbom’s 1968 suggestion of using sublingual nitroglycerin has worked better, with 2 of 8 patients developing typical clinical attacks. Recently the second of two doses of i.v. naloxone totaling 0.11 mg in one and a combination of increasing doses to a total of 0.33 mg i.v. naloxone and 1.7 mg nitroglycerin sublingually in another
patient provoked an attack. In 16 other patients a major attack began during the needle placement or during electrical stimulation to correct the placement. If feasible, the whole procedure should be timed just to precede an expected attack. In the 19 who developed an attack it was clearly arrested in 13 of them by the lidocaine-produced analgesia. Only 24 of the 36 patients found the new sensation of analgesia acceptable. The remaining 12 were not submitted to any form of trigeminal rhizotomy. We think this diagnostic procedure helps select the type of surgery; in those who have found the sensation of analgesia acceptable no patient has developed major facial dysesthesias after a subsequent trigeminal rhizotomy. A longer temporary trigeminal denervation advocated by Harris [1940] consisted of peripheral injections of alcohol into supra- or infraorbital or temporal areas. Neurectomy of the appropriate peripheral branches may be tried when an attack cannot be provoked at the lidocaine block.

Radiofrequency Rhizotomy

As we have already commented these patients have a greater tendency to develop anesthesia or analgesia dolorosa after trigeminal lesions than those with trigeminal neuralgia. Hence these patients especially benefit from the fact that the control of the locus and degree of sensory loss is better with percutaneous RF heating than with open operations in the middle or posterior fossa. Usually the reference of pain is to the first two divisions to which consequently we confine lesions, extending them to the third division of course for pain in this zone. Because of so many recurrences when only hypalgesia was present we have been more aggressive about producing full analgesia in the areas of pain than we have in trigeminal neuralgia, using a 5mm bare 20-gauge electrode heated to 100 °C for 4 min if needed. Much lower temperatures have usually sufficed, and we start about 5 °C above the threshold temperature for pain. In one patient in whom at 46 °C there was a ‘burning’ in the eye, the first lesion at 50 °C for 1 min with a 5-mm bare electrode yielded the desired analgesia of the first and upper half of the second division.

We have now carried out 28 RF procedures on 20 patients. Watson et al. [1983] with the next largest series have done 27 RF procedures on 13 patients. Both groups have had the disappointing result of inadequate or no relief from a presumably appropriate trigeminal sensory loss. Of our 6 such patients 2 had analgesia throughout all three trigeminal divisions and 4 had analgesia in much or all of the first two divisions in the zone of the worst pains; yet they
had pain referred to these areas. Only one of these had had an attack aborted by a lidocaine block. Another, after an open rhizotomy elsewhere, had had 4 years of relief until recovery of pinprick sensation. We have found it advisable to secure analgesia throughout all areas of pain before classifying the result as a failure. Thus, in one patient it took us three procedures before we achieved sustained useful relief by rendering analgesic and anesthetic all of the second division’s painful oral mucosa. Another patient’s pain predominantly in cheek and temple was stopped by a V2, V3 lesion. Attacks then gradually built up over 18 months in the typical orbital area and were eliminated by a VI,2 lesion. In 3 other of our patients two or three lesions were required to deal with initial failures to secure adequate analgesia or later recovery of algesia. Relief continues in 11 of our 20 patients followed from 10 months to 20 years for an average of 5.3 years; 4 of these require supplemental medication. Two others were relieved for 3 and 10 years before recurrence and one other with early relief was lost to follow-up. Of these 14 patients with complete VI analgesia some decreased corneal sensation to touch was retained in 5, another advantage of RF lesion over open rhizotomy.

The experience with RF lesions of Watson et al. [1983] can be summarized as follows: 14 without complete analgesia in VI,2 were unrelieved; 3 with such ‘inadequate’ sensory loss were relieved. However, one with full analgesia of VI,2 and another with analgesia in VI continued with pain in the analgesic zone. Of 8 with analgesia throughout V1,2, relief continued in 4 and was maintained until recovery of sensation in the other 4. Relief continues in 7 from 10 to 36 months with an average of 24 months. Anesthesia dolorosa developed in one patient who was nevertheless glad he had the operation.

The series of Onofrio and Campbell [1986] includes 24 RF procedures on 22 patients. The analgesia they produced was incomplete in 12 procedures with 11 early failures and a recurrence at 6 months in the 12th. In 6 of these patients they proceeded directly to open rhizotomy in the posterior fossa. Of the 12 RF procedures followed by good to excellent relief 11 had complete VI,2 analgesia and the 12th severe hypalgesia in VI with analgesia in V2 confirming the thought that full analgesia in the painful zone is usually necessary. Follow-up ranged from 12 to 32 months with an average of 21 months. Some corneal sensation to touch was retained in 3 of the 12 patients with relief. These authors also describe a successful outcome in one patient following bilateral RF lesions for attacks on both sides. Tew et al. [1982] comment briefly that their VI,2 lesions gave ‘satisfactory relief to 6 of 8 cluster headache patients.'
Maxwell [1982], with the preliminary criterion of stopping an attack by retrogasserian lidocaine, has compiled the perfect record of 8 initial successes after RF lesions. His objective of producing only hypalgesia is, judging from the results of the three groups just described, likely to be followed by more severe late recurrences than the 3 mild ones he has described.

Glycerol Rhizotomy

The results of 13 injections of an average of 0.4 ml of glycerol in 6 severely afflicted patients followed 5-47 months (mean 16 months) were described by Ekbom et al. [1984]. In 3 patients a total of 7 injections yielded slight VI sensory loss and poor relief. In 3 others immediate complete relief was followed after 2, 6, and 11 months, respectively, by recurrence and a repeat injection. Complete relief was again achieved in 2 followed for 9 and 18 months and 90% relief in the third was maintained at 4 years. In these 3, analgesia around the eye and loss of corneal reflex also ensued. Waltz et al. [1985] treated 5 patients by glycerol injection with no benefit in one and with enough improvement in the other 4 to render them 'more easily managed medically'. The attacks were not stopped in any of them. We think the lidocaine-RF lesion sequence is more logical and effective.

Open Trigeminal Rhizotomy

Dott [1951] and White and Sweet [1969] have described relief following this operation. In the 2 cases of the latter authors this result persisted at 4 and 15 years. Mazars and collaborators [1976] in an extensive experience from Neurosurgical Treatment of Persistent Pain 76

1948 to 1975 sought to limit their rhizotomy to first-division rootlets. They used electrical stimulation at operation to identify ‘those rootlets responsible for meningeal sensation’ and cut only this ‘meningeal contingent’ in 11 of their 31 patients. They describe only 3 failures — one apparently after relief for one year.

The Toronto group [Watson et al., 1983] did 8 open rhizotomies on 8 patients with initial cessation of the pain attacks in 5. There was a recurrence at 30, 36, and 44 months in 3. Relief continued at 48 months and 8 years in the other 2. They achieved anesthesia at the site of the main pain in these 5 — only one of whom had the result marred by anesthesia dolorosa. Noteworthy are 3 failures despite adequate sensory loss. One of them with pain only in the upper gum was not relieved by complete V2,3 anesthesia. These emphasize again the importance of a temporary procedure to assess the value of
trigeminal analgesia during an attack. The brief accounts of O’Brien and MacCabe [1984] and of Campbell [1984] tell us of 6 more encouraging results; there is no indication that either group preceded the rhizotomy by a temporary block.

Onofrio and Campbell’s [1986] 11 posterior fossa rhizotomies included 6 carried out only after prior failures of an RF lesion, 4 more as the primary procedure and the 11th operation when the first open operation produced only minor sensory loss. There were 5 excellent results and 2 failures to control the pain correlated in one with incomplete VI,2 analgesia and in a second despite complete anesthesia including the cornea. There were 2 other results classified as poor even though relief of pain was achieved associated with complete anesthesia. One patient went into a severe depression and deserted his family; the other with anesthesia dolorosa became totally disabled and vindictive.

Of Hitchcock’s [Wake and Hitchcock, 1987] 4 patients treated by open rhizotomy with extensive anesthesia, 3 have maintained complete or good relief at 12, 15 and 25 years. One of the 3 was not relieved by a prior lesion producing only complete hypalgesia. The 4th patient was unrelieved despite production of complete trigeminal anesthesia, again pointing to the utility of temporary trigeminal loss as a preliminary procedure.

We conclude that production of any necessary analgesia without anesthesia in order to decrease the incidence of major dysesthesia is an objective best achieved by differential thermal rhizotomy or by bulbar tractotomy of the descending cephalic pain pathway. Our first patient with this disorder treated by a dense extensive RF lesion found the areas of anesthesia dolorosa far worse than her neuralgic pains (which never recurred). When sensory recovery set in she was grateful only to the physician whose medication made her hypalgesia dolorosa more tolerable. The course in 3 of our patients treated by other neurosurgeons by open rhizotomy reemphasizes its disadvantages. One, although free of his attacks of migrainous neuralgia for 29 months, is infuriated and states he is totally disabled by anesthesia dolorosa associated with the total anesthesia; he is suing and threatening to murder the neurosurgeon. (These patients seem prone to more violent responses to an adverse result than those with trigeminal neuralgia.) The other 2 patients’ operations were followed by recovery of sensation and recurrence.

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Superficial Petrosal Neurectomy

Chorobski and Penfield [1932] found in the monkey efferent vasodilator
fibers to the arteries of the supratentorial pia mater only in the greater superficial petrosal nerve. This nerve also carried secretory and vasodilator fibers to the mucosa of nose, mouth and upper pharynx, as well as secretory fibers to the lacrimal gland. They also identified a distinct bundle of small myelinated and unmyelinated fibers in the nervus intermedius just proximal to the geniculate ganglion passing directly into the greater superficial petrosal nerve without interruption in the ganglion (fig. 1/12). Further on, a distinct bundle of nerve fibers emerged from the greater superficial petrosal nerve passing to the nerve plexus on the internal carotid. These appear to be efferent pathways. Coupling these observations to the evidence that vasodilation causes this type of headache, Gardner et al. [1947] proposed that superficial petrosal neurectomy might be effective treatment, suggesting the term petrosal neuralgia for the disorder. Although at first encouraged by his results Gardner [1951] later lost his enthusiasm for the procedure. Table 1/VII summarizes the results in 13 communications reporting on this operation. It is noteworthy that Gardner’s successors at the Cleveland Clinic, Kunkel and Dohn [1974], have persisted with the operation in carefully selected patients, adding ‘neurolysis’ of the trigeminal sensory root to the petrosal neurectomy. By neurolysis Gardner [1962] meant: ‘freeing the sensory rootlets from their durai sleeve, gently brushing them with a cotton pledget and spraying them forcefully with a Ringer’s solution using a 21gauge needle.’ Since section of the greater superficial petrosal nerve is distal to the geniculate ganglion from which its afferent fibers arise, one would expect regeneration with a recurrence of the pain in most of the cases if these afférents are responsible for it, just as we see after peripheral trigeminal neural avulsions in trigeminal neuralgia. Hence we now consider this operation to be stage 1 in assessing the likelihood that the more permanent rhizotomy of nervus intermedius, IX and possibly the upper fibers of X or a high bulbar tractotomy may be in order.

In 1955 and 1969, White and Sweet reported on the results of nearly
always dividing both the greater and lesser superficial petrosal nerve, and
rarely the usually unidentified external superficial petrosal nerve as well, plus
the middle meningeal artery in 10 patients. In 3 patients there was no relief;

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Table 1/VII. Chronic migrainous neuralgia

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Table 1/VIII. Petrosal neurectomy

in them stimulation of neither superficial petrosal nerve at operation reproduced
the patient’s pain. In the remainder the results were gratifying as
described in table 1/VTI. The subsequent experience of W.H.S. in 13 patients
has been much less favorable with no relief in 7 patients, temporary ’50% relief in one, and full or good relief varying from 6 to 27 months with an
average of 18 months in the remaining 5. Complete recurrence has taken
place in all 5. Hitchcock [Wake and Hitchcock, 1987] also cuts both lesser
and greater petrosal nerves. Other authors describe cutting only of the greater
superficial petrosal nerve.

W.H.S. has always awakened the patient after the nerves were exposed and
stimulated them electrically. In the cases since 1968, the use of atiny concentric
needle electrode has permitted greater precision than with earlier larger
electrodes. Both the two intact nerves and then their central and peripheral cut
ends were stimulated. At times gentle suction to the nerve or cutting it gave a
more decisive reproduction of the patient’s pain than the electrical stimulus.
Table 1/VIII summarizes the results, including those in 2 patients whose
absence of a bony roof over the carotid permitted stimulation of the neural

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plexus on the intrapetrous internal carotid artery as well. Hitchcock also
stimulated the exposed petrosal nerves in the awakened patient. In one
patient he cut the trigeminal root when petrosal nerve stimulation failed to
reproduce the pain — securing relief supplemented by medication for 15
years. The possibility that the mechanism of evoking the pain induced an
autonomic effect or an effect in the periphery which then excited nociceptive
afferents was assessed by stimulation of the cut medial (peripheral) end of the
nerves before they passed beneath the Gasserian ganglion. In only 2 patients
and only in the greater superficial nerves did low voltage elicit pain like the
clinical pain. In one of them stimulation of the lateral cut end of both nerves
also provoked the same response. All the rest, upon stimulus to the medial cut end of the nerves, experienced no facial contraction and either no pain or pain referred to the medial part of the third trigeminal division’s zone, hence clearly related to spread to these nearby trigeminal nerve fibers.

We conclude, as did White and Sweet [1969, p. 363] from earlier studies on other patients, that these two nerves contain pain fibers and that when their section relieves pain this is a consequence of cutting an afferent nociceptive pathway.

Division of the Nervus intermedius

In their first monograph on pain, White and Sweet [1955, p. 501] proposed that patients deriving clear-cut relief from petrosal neurectomy should, upon recurrence of severe chronic pain, have division of the n. intermedius (the sensory rootlets of the geniculate ganglion). Dr. E. Sachs, Jr., is the pioneer who first actually carried out this operation in patients with chronic cluster headache, doing his first operation in 1952 and reporting his first 4 patients in 1968 (fig. 1/12). In his first case stimulation of the n. intermedius with a forceps reproduced the patient’s clinical pain. However, when this nerve was cut similar stimulation of the VUIth cranial nerve also reproduced her pain, so with her assent this nerve was also cut. The second patient also had a complete hearing loss on the side of the cut n. intermedius after operation. Two previous operations for section of the greater superficial petrosal nerve had relieved him for 3 and 2 years, respectively. Both patients, after complete lesions of n. intermedius and VIII, had remained pain-free at the last follow-ups 16 and 11 years later [Sachs, 1969]. By 1971 [Sachs, 1976], he had done rhizotomy of n. intermedius in 8 patients with this disorder with excellent very long-term results in 4. One recurred promptly on the 3rd postoperative day; in another relief persisted on the operated side but the pain recurred on the opposite side a year later. In 2 the pain spread during the attacks out of the usual fronto-facio-aural area to the back of the head and side of the neck. In these 2 ‘who also had unstable personalities’ and ‘were poor candidates for the operation’, it failed. Sachs [1976] also reports a patient of C. Drake in whom initial good result was followed by recurrence. W.H.S. has cut the n. intermedius in 3 patients (2 women and one man); in all 3 we first did greater and lesser superficial petrosal neurectomy, at which operation, in all 3, stimulation of the intact petrosal nerves and of their cut central ends reproduced the clinical pain. This report was especially striking in the man in whom cutting the greater nerve provoked a grade 3/4
attack of his clinical pain. The petrosal neurectomy gave only 50% relief to Janice D., but had yielded complete relief for 19 and 17 months, respectively, to the other 2. In Janice D. n. intermedius section bilaterally at a 4-month interval between the two sides was soon followed by a major recurrence equally severe on the two sides with up to 12 attacks per day by the following year. A lidocaine block with V1,2 analgesia had also failed to stop her pain. In the man it was especially disconcerting at the posterior fossa operation to have him report that stimulation of n. intermedius caused only aural pain, not at all like his clinical pain. His n. intermedius consisted of three unusually large fibers, but cutting them gave him no relief despite the good result for 19 months after petrosal neurectomy. In Maureen L. we cut the glossopharyngeal as well as the two well-developed rootlets of n. intermedius. Pain in her ear had been particularly prominent. She was completely relieved of the original attacks for over 4 years until December 1980. Since then infrequent less severe groups of attacks have been well controlled by nontoxic doses of medication. In another of our female patients with poor relief from retrogasserian lidocaine block we concluded that her personality and environment made her a poor candidate for surgery. Subsequently she had three sections of n. intermedius elsewhere, first on the left side, then on the right and finally on the left again with only a few months of benefit following each.

A problem with the anatomy of the n. intermedius is its propensity to have some of its fibers incorporated into either the VIIth or VUth cranial nerves as beautifully depicted in Bischoff's [1865] dissections and republished in small part by White and Sweet [1969, p. 626]. The clinical importance of such disposition was clearly demonstrated in Sachs' first 2 patients in whom the VUth nerve function was sacrificed but with excellent sustained relief. Although cranial n. IX and the upper fibers of X also supply the ear and pharynx we are not certain that dividing those at the time of an operation on n. intermedius would help in this disorder, having divided IX in only one of our 3 patients. However, our failure in Janice D. suggests that the operation about to be described would have been preferable.

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Fig. 1/13. Cross-section of the bulb just below the obex. Site of incision which surrounds descending trigeminal tract indicated by faint continuous lines. From Krieg [1942].

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Tractotomy of Bulbar Descending Cephalic Pain Tract
The initial observations of one of us (W.H.S.) in 1942 and 1943 [White and Sweet, 1955, pp. 463-466] revealed that all cephalic pain fibers from V, VII, n. intermedius, VIII, IX and X travel in what was originally denoted the spinal or descending trigeminal tract. Extensive incisions into this tract in the medulla oblongata at about the obex were likely to give the full analgesia of inner head and face indicated above. Our first operation which deliberately achieved such analgesia was carried out on June 8, 1945 [White and Sweet, 1955, pp. 476-478]. However, analgesia of the first 2 trigeminal divisions requires a less extensive incision placed more caudally. Figure 1/13 shows that if one deliberately divides a dorsal bit of the spinothalamic fibers ventral to the descending tract and produces some V3 as well as VI,2 analgesia at operation, one can be reasonably certain of having divided all of the upper two thirds of the trigeminal fibers, which are the most important for the management of chronic migraineous neuralgia in the usual picture. We have done the more rostral operation with an incision extending dorsally into the fasciculus cuneatus in 2 patients with migraineous neuralgia, both of whom had extensive reference of their pain, and in whom we wished to divide as much as feasible of the descending cephalic pain pathway.

Patient Eric M. Born 8/57, had his first attack at age 12. The pain involved left periorbital area, temple, ear and the whole lateral and posterior neck. He had had only 7 attacks of right-sided pain. He was totally disabled from work or social activities by the severity of his pains uncontrolled after 4 years of a thorough trial of medications by a cluster headache specialist (J. Graham). A lidocaine block 10/16/80 yielded VI,2 analgesia and stopped an attack; this hypersensitive man thought the numbness was probably acceptable. On 10/17/80 an RF lesion produced an initially excellent analgesia which soon faded to hypalgesia with recurrence of attacks. On 11/26/80 a bulbar descending cephalic pain tractotomy was carried out to a depth 3 mm in the transverse plane 1 mm caudal to the obex. Extensive intraoperative testing revealed and postoperative testing confirmed in more detail analgesia of the opposite distal lower limb from mid calf on down as well as throughout trigeminal skin and mucosa, both walls of the external auditory canal, tympanic membrane and tonsillar fossa but not oral pharynx. Temporary ipsilateral latéropulsion confirmed an adequate dorsal extent of the incision. In the 86-month period since operation he has had many mild attacks controlled by modest doses of medication, but he works full time having only an occasional bout of pain and has contracted a successful marriage, in contrast to preoperative total disability.

Patient John B. Bom 6/43. Pain attacks in left temple radiating to left jaws and lateral neck since 1970. Cafergot, the only useful drug, controlled the attacks but led to 3 episodes of congestive heart failure and in 1977 an aortic and mitral valve replacement. On 3/2/82
left greater and lesser superficial petrosal neurectomy was done with no relief. On 5/7/82 a bulbar lesion in the plane 2 mm rostral to the obex was made deeper than in the previous patient, in the hope of destroying ascending fibers within the nucleus of the descending cephalic pain pathway. Analgesia of the ipsilateral trigeminal zone and opposite limbs and torso has controlled the facio-cephalic reference in the attacks of pain, but they resumed within 3 months in a zone about 36 cm2 in an area in the left side of the neck with the same time course and disabling intensity as the original attacks. They remained under control only with massive doses of cafergot, which he refused to give up despite its leading to another open heart operation in November 1983. On 7/11/84 right C2-3 posterior rhizotomy was done (C. Poletti) yielding anesthesia of posterior scalp and upper two thirds of neck. Complete relief of pain persists at 3.5 years follow-up.

Patient Eric M.’s original extensive cervical reference of pain has not required rhizotomy of this region.

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Patient Stephen H. Born 2/51. First attack left cluster headache 7/83 superimposed on bilateral migraine attacks since age 8 years. Pain in left posterior scalp as well as face. Two retrogasserian glycerol injections, the 1st with VI,2 numbness and inadequate relief; the 2nd with V2,3 denser numbness and relief 6 months — the numbness ‘extremely unpleasant’ — same for 1 h numbness after a lidocaine block 5/86. At Massachusetts General Hospital 6/11/86 analgesic, then anesthetic lidocaine block during attack of pain — no benefit. 6/24/86 — bulbar descending cephalic pain tractotomy and Cl,2,3 posterior rhizotomy (Poletti) producing analgesia all V, VII, IX, X zones but no cervical analgesia; tested 6th postoperative day. Although the cluster attacks have been greatly relieved, the patient is disabled by analgesia dolorosa — a constant severe burning throughout left V2,3 and behind eye. Not realized was the hope that the more complete preservation of touch associated with bulbar tractotomy than with RF rhizotomy would avert dysesthesias. These have been helped only temporarily by Dr. R. Tasker’s implantation of a stimulating electrode in the area of the trigeminal ganglion and root. In retrospect since the glycerol injections had also provoked dysesthesias we should have cut VII, IX and upper X posterior roots.

Autonomic Activity vis-à-vis Attacks of Pain

Although the frequent linkage between the manifestations of autonomic activity and the episodes of pain led to Gardner’s performance of petrosal neurectomy, numerous observers have noted that after surgical treatment, episodes of pain may be stopped while the autonomic signs continue and vice versa. Thus, Dott [1951] states: ‘After trigeminal rhizotomy in some cases periodic watering and suffusion of the eye and throbbing in the temple
(continue) with no pain or even discomfort.’ Mazars [1976] noted after his rhizotomy confined to the trigeminal first division that although the attacks of pain disappeared, ‘vasomotor and secretory episodes may persist — decreasing over several months.’ One of Maxwell’s [1982] thermally rhizotomized patients, though relieved of pain continued with attacks of mild tearing, hemicranial flush, and nasal fullness. Campbell [1984] too had a pain-free patient in whose first year after open trigeminal rhizotomy there was a painless episode of unilateral tearing and ptosis. We have had 2 patients with persistent pain relief but dramatic attacks of autonomic manifestations. In one patient trigeminal VI,2,3 analgesia and VI nearly complete anesthesia with elimination of all symptoms was followed 32 months later by the sudden recurrence with clock-like regularity of these features of the original attacks; right-sided profuse tearing, watery rhinorrhea, flaming red face and eyeball, miosis, and ptosis with nausea, generalized weakness and sweating on both sides of the face and all limbs. These attacks occurred at 3-4 p.m. or awakened him at 3-4 a.m. — all now ‘with absolutely no pain whatsoever’. The second patient’s total trigeminal anesthesia and complete cessation of all symptoms was followed 5 years later by episodes beginning with drooping of the right comer of the mouth, then miosis, ptosis and profuse ipsilateral watery rhinorrhea without tearing, but generalized weakness so striking that he must lie down for the 25- to 35-min duration of the episodes. These, persisting for 8 years, include only minor initial stabs of pain ipsilaterally behind the right eyeball and in the right ear canal.

Conversely, Kunkel and Dohn [1974] describe 3 patients whose pain recurred after greater superficial petrosal neurectomy and trigeminal sensory ‘rhizolysis’ but without lacrimation, conjunctival injection or other such symptoms. In 3 more of their similarly treated patients the attacks of parasympathetic discharge were stopped but the pain persisted without a period of relief. Wake and Hitchcock’s [1987] case 8 showed the same behavior. We have a patient whose tearing and rhinorrhea stopped but the pains continued despite excellent VI,2 analgesia.

These observations all make it clear that the impulses regarding the pain may travel by grossly disparate routes from those for the autonomic discharge which probably travel via the fiber bundle described by Chorobski and Penfield as bypassing the geniculate ganglion.

Summary
Pain in this disorder may be transmitted over trigeminal, n. intermedius, glossopharyngeal and possibly upper vagal afferent paths. We recommend as initial evaluation for possible invasive therapy a retrogasserian needle placement followed by a sustained effort to provoke an attack and to ascertain if analgesic block of rootlets will stop the attack at the price of an acceptable denervation sensation. The great majority of the successful surgical cases have had appropriately sited trigeminal analgesia. If the attack is not stopped or the numbness is not tolerated, petrosal stimulation and neurectomy is the next step. Should this peripheral neurectomy yield relief but be followed by recurrence, section of the roots of nervus intermedius, IX and perhaps upper X may yield longer relief. Section of the bulbar descending cephalic pain pathway may be advisable in severe cases with widespread neural involvement or poor tolerance of sensory deprivation. The rare case may require supplemental cervical posterior rhizotomy.

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Conclusions

Although far fewer patients with this disorder require surgery than do those with trigeminal neuralgia and the management is more complex, the desperate medically intractable patients who are benefited are truly grateful even for a degree of relief which requires medical supplementation. Since we have not been able to identify features indicative of the specific neural pathways involved we favor the sequence of diagnostic and therapeutic procedures we have described.