Chapter 98

Sudden Unilateral Neuralgiform Pain with Conjunctival Injection and Tearing

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SHORT-LASTING UNILATERAL NEURALGIFORM HEADACHE ATTACKS WITH CONJUNCTIVAL INJECTION AND TEARING

International Headache Society (IHS) code and diagnosis: 3.3 Short-lasting unilateral neuralgiform headache attacks with conjunctival injection and tearing (SUNCT)

World Health Organization (WHO) code and diagnosis: This syndrome is not recognized in the current edition of the International Classification of Disease (ICD).

Short description: This syndrome is characterized by strictly unilateral, severe, neuralgic attacks centred on the ophthalmic trigeminal distribution that are brief in duration and very often occur in association with prominent lacrimation and redness of the ipsilateral eye.

Previously used terms: None.

EPIDEMIOLOGY

Short-lasting unilateral neuralgiform headache attacks with conjunctival injection and tearing (SUNCT) syndrome was first described relatively recently in 1978 (64). Since then 82 cases (64 primary or idiopathic, 18 secondary or symptomatic cases) have been reported in the English language literature. The prevalence and incidence of SUNCT are not known, though the extremely low number of reported cases suggests that it is a very rare syndrome. Primary SUNCT has a slight male predominance (35 males, 29 females) with a sex ratio of 1.2:1. In an earlier review of SUNCT syndrome the male predominance among primary cases was greater than that reported here, with a sex ratio of 3.75 (15 males, 4 females) (52). The trend toward decreasing male:female ratios over time probably reflects an ascertainment issue. The typical age of onset is between 35 and 65 years (69% of primary SUNCT cases), though it ranges from 10 to 77 years (mean 50 ± 15 years) (36).

PATHOPHYSIOLOGY

The trigeminal autonomic cephalgias (TACs) are a group of primary headache disorders characterized by unilateral trigeminal distribution pain that occurs in association with ipsilateral cranial autonomic features (17). The group comprises SUNCT, paroxysmal hemicrania (PH), and cluster headache (CH). Any pathophysiologic construct for TACs must account for the two major clinical features characteristic of the various conditions that comprise this group: trigeminal distribution pain and ipsilateral autonomic features. The pain-producing innervation of the cranium projects through branches of the trigeminal and upper cervical nerves to the trigeminocervical complex, from which nociceptive pathways project to higher centers. This implies an integral role for the ipsilateral trigeminal nociceptive pathways in TACs. The ipsilateral autonomic features suggest cranial parasympathetic activation and sympathetic hypofunction. Goadsby and Lipton have suggested that the pathophysiology of the TACs revolves around the disinhibition of the trigeminal–autonomic reflex (17). There is considerable experimental animal literature to document that stimulation of trigeminal afferents can result in cranial autonomic outflow, the trigeminal–autonomic reflex (44). In fact, some degree of cranial autonomic symptomatology is a normal physiologic response to cranial nociceptive input (16,43) and patients with other headache syndromes may report these symptoms (1,3,6). The distinction between the TACs and other headache syndromes is the degree of cranial autonomic activation (18).

Cranial autonomic symptoms are likely to be prominent in the TACs due to a central disinhibition of the

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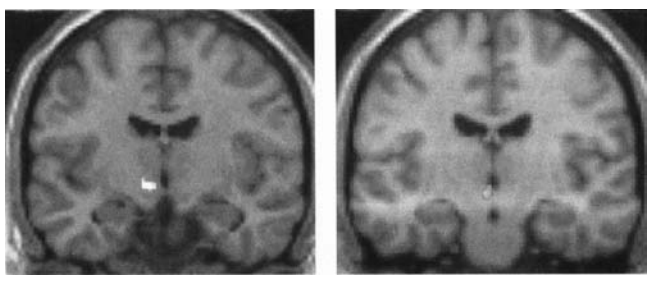

FIGURE 98-1. Blood oxygen level dependent functional magnetic resonance imaging (BOLD fMRI) study demonstrating posterior hypothalamic activation during spontaneous pain attacks in a SUNCT patient. (Reused from May A, Bahra A, Buchel C, et al. Functional magnetic resonance imaging in spontaneous attacks of SUNCT: short-lasting neuralgiform headache with conjunctival injection and tearing. *Ann Neurol* 1999;46:791–794.) (See color plate)

trigeminal–autonomic reflex (18). Supporting evidence is emerging from functional imaging studies: a functional magnetic resonance imaging (fMRI) study in SUNCT (42) (see Fig. 98-1) and a positron emission tomography (PET) study in CH (41) have both demonstrated ipsilateral hypothalamic activation. Hypothalamic activation is specific to these syndromes and is not seen in migraine (2,71) or experimental ophthalmic trigeminal distribution head pain (45). There are direct hypothalamic–trigeminal connections (34) and the hypothalamus is known to have a modulatory role on the nociceptive (10,70) and autonomic pathways (33). Hence, SUNCT is possibly due to an abnormality in the hypothalamus, with subsequent trigeminovascular and cranial autonomic activation.

Several studies have demonstrated the diverse parasympathetic manifestations of SUNCT. Forehead sweating is usually increased during bouts (28), unlike PH, in which it is normal. Pupillary studies using pupillometry and pharmacologic approaches have revealed no abnormalities (73). Since conjunctival injection occurs during SUNCT, it is not surprising that intraocular pressure and corneal temperatures are elevated during attacks (67), most likely reflecting local vasodilation consequent to the marked parasympathetic activation. Similarly, a report of bradycardia in association with attacks of SUNCT indicates increased parasympathetic outflow (27). Systolic blood pressure is sometimes elevated, although ventilatory function is normal (26).

Orbital phlebography is reported to be abnormal in SUNCT syndrome, with a narrowed superior ophthalmic vein and the cavernous sinus homolateral to the pain (29). This finding leads to the suggestion that SUNCT syndrome may be a form of orbital venous vasculitis (20), although there are similar reports in CH, Tolosa-Hunt syndrome, and PH (17). However, transcranial Doppler and SPECT (single photon emission computed tomography) studies have not demonstrated convincing change in the vasomotor activity (63) or cerebral blood flow during attacks of pain (55). Similarly, no intracranial vessel activation was apparent in an fMRI study (42). The hypothalamic activation and the diverse parasympathetic manifestations taken together with the paucity of evidence for vascular changes favor a central pathogenesis for SUNCT rather than a peripheral vasculitic cause.

CLINICAL FEATURES

Site of Pain

The pain is usually maximal in the ophthalmic distribution of the trigeminal nerve, especially the orbital or retroorbital regions, forehead, and temple. It may radiate to the other ipsilateral trigeminal divisions and, rarely, even to extratrigeminal regions such as the ear and occiput.

Laterality of Attack

The attacks are typically strictly unilateral and side-locked; however, in four patients the pain was simultaneously experienced on the opposite side (52,60), while three patients reported strictly unilateral but side-alternating attacks (11,12,39). The pain is present more frequently on the right side than the left in patients with side-locked attacks (36).

Severity of Pain

The intensity of the pain is generally severe. The usual pain intensity was described as moderate in 14% of patients, severe or intense in 63% of patients, and very severe or excruciating in 23% of patients (36).

Character of Pain

The pain had a neuralgic character in the majority of patients (85%), being usually described as stabbing, sharp, burning, pricking, piercing, shooting, lancinating, or electric shock–like. The character of pain has also been described as pulsatile or throbbing (6%), steady (4%), spasmodic (2%), staccato (2%), and pressing (2%) (36).

Duration of the Individual Attack

Three different types of pain have been described in SUNCT syndrome: relatively short-lasting attacks; long-lasting attacks; and a continuous or intermittent background ache.

The individual short-lasting attacks are very brief, lasting between 5 and 120 seconds in the majority of patients (84%). The attacks were described as lasting less than

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5 seconds in 8% of patients and more than 120 seconds in 8% of patients. The median duration of the usual attack was 40 seconds. The range of the usual attack duration was between 2 seconds and 20 minutes (7,11,36,39). In a study that objectively measured the duration of 348 attacks in 11 patients, the range of duration was 5 to 250 seconds and the mean duration was 49 seconds (50).

There are four reported cases of SUNCT syndrome in which prolonged attacks, lasting 1 to 2 hours, are described (37,42,49). In all these patients, the majority of the attacks were short-lasting and were otherwise typical for SUNCT syndrome; the relatively long-lasting attacks were phenotypically similar to the short-lasting attacks except for the duration of the attack.

Most patients are completely pain free between attacks, although there are seven case reports of a dull interictal ipsilateral discomfort over the same site; the interictal discomfort has been reported to be continuous in five cases (7,39,52,60) and intermittent in the other two (62,69). There is also one case report of a bilateral continuous interictal discomfort over the forehead (39). In addition, a burning sensation lasting 2 hours following the attacks has also been reported (57).

Temporal Profile of the Individual Attack

The short-lasting paroxysms begin abruptly, reaching the maximum intensity within 2 to 3 seconds (52). In the majority of patients, the pain is maintained at the maximum intensity before abating rapidly. However, several temporal patterns for the individual attack, besides the plateau-like pattern, have been described, including a repetitive pattern of spike-like paroxysms; a saw-tooth pattern in which repetitive spike-like paroxysms occur without reaching the pain-free baseline between the individual spikes; and plateau-like plus exacerbations pattern in which a plateau-like attack had superimposed, random, ultrashort exacerbations of 1 to 2 seconds (51) (Figure 98-2).

Frequency and Periodicity of Attacks

Both episodic and chronic forms of SUNCT exist (36). In the majority of patients, SUNCT syndrome presents in an episodic manner; the temporal pattern is quite variable, with the symptomatic periods alternating with remissions in an erratic manner. Symptomatic periods generally last from a few days to several months and occur once or twice annually, although a maximum of 22 bouts per annum have been reported (52). Remissions typically last a few months, though they can range from 1 week to 8.5 years (24). Symptomatic periods appear to increase in frequency and duration over time (52). Circannual periodicity is not typically a feature of SUNCT.

In 28% of patients the disorder is chronic, with the symptomatic period lasting more than 1 year. In these

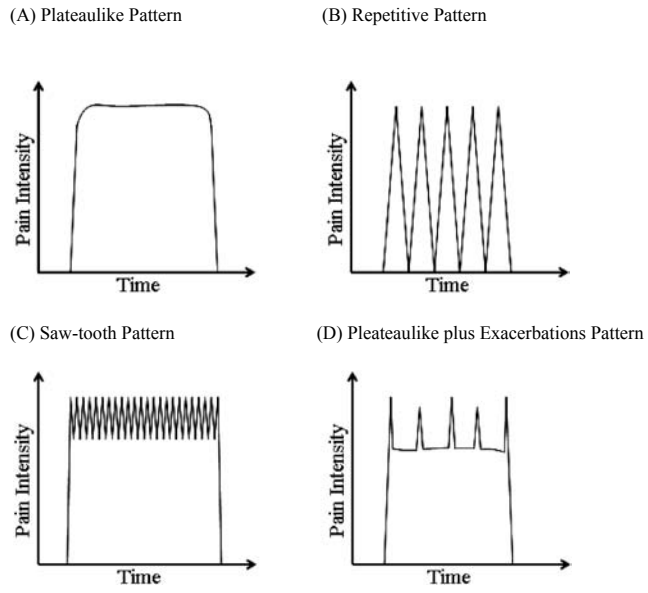

FIGURE 98-2. Temporal profile of SUNCT attacks: (A) *plateau-like* pattern: rapid onset of pain, which is maintained at maximum intensity before abating rapidly; (B) *repetitive* pattern of spike-like paroxysms; (C) *saw-tooth* pattern in which repetitive spike-like paroxysms occur without reaching the pain-free baseline between the individual spikes; and (D) *plateau-like plus exacerbations* pattern in which a plateau-like attack had superimposed, random exacerbations (adapted from ref. 41).

cases the mean duration of the chronic phase was 5.3 ± 5.0 years (range: 1 to 17 years). In the majority of these patients the disorder is chronic from onset (72%), though in some the chronic form develops from the episodic variety (28%) (36). In addition, there are several case reports of SUNCT syndrome in which the chronic phase alternates with the episodic phase (5,20,21,65). Earlier reviews noted that the chronic form of SUNCT was not sufficiently validated (53). The chronic form of SUNCT is now sufficiently validated and, therefore, the subclassification of SUNCT syndrome should include episodic and chronic forms, thereby bringing it in line with that of cluster headache and paroxysmal hemicrania.

The attack frequency during the symptomatic phase varies immensely between sufferers and within an individual sufferer. Attacks may be as infrequent as once a day or less to more than 60 per hour (46). Objective assessment of the frequency of attacks in four patients demonstrated a mean of 16 attacks daily with a range of 1 to 86 attacks daily (50). There are six case reports of a SUNCT-like status, when patients experience severe exacerbations with frequent, easily triggered, high-intensity pain attacks in a repetitive and overlapping fashion for several hours or days at a time (21,46,52).

SUNCT attacks occur either exclusively or predominantly during the daytime. Exclusively diurnal attacks were reported in five (17%) patients. Nocturnal attacks

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were reported to occur occasionally in 20 (66%) patients and frequently in 5 (17%) patients. However, these results need to be interpreted cautiously as no data were available in over half of the reported cases (34 patients; 53%). Objective assessment of the timing of 585 attacks in four patients demonstrated that a bimodal distribution occurs with morning and afternoon/evening predominance, with only 1.2% occurring at night; however, given the small number of patients studied these data need to be interpreted cautiously (50).

Associated Features

Acute headache episodes in SUNCT syndrome are accompanied by a variety of associated symptoms. The attacks are virtually always accompanied by both ipsilateral conjunctival injection (100%) and lacrimation (95%). Ipsilateral rhinorrhea (55%), nasal congestion (44%), eyelid edema (30%), ptosis (19%), meiosis (6%), and facial redness (6%) or sweating (3%) are less commonly reported. These cranial autonomic symptoms, particularly conjunctival injection and lacrimation, are typically very prominent in SUNCT syndrome. The associated conjunctival injection and tearing usually begin 1 to 2 seconds after onset of the pain and may outlast the pain by a few seconds. Rhinorrhea, when present, is delayed, occurring relatively late in the course of the headache (52). Unlike in CH, restlessness is not a prominent feature of SUNCT syndrome and has only been described in four patients (39,69).

Nausea (2%), vomiting, photophobia (5%), phonophobia (2%), osmophobia, and worsening of pain with movement (3%) are not normally associated with SUNCT syndrome. A migrainous aura in association with SUNCT has been reported in two patients. There is a report of a tingling sensation over the ipsilateral temple in association with the attacks (46) and another report of ipsilateral facial tingling lasting 5 to 10 minutes and occurring 5 to 15 minutes prior to the SUNCT attacks (39).

Triggers

Most SUNCT patients have both spontaneous and triggered attacks, while a minority seem to exhibit exclusively spontaneous attacks. No patients have been reported to have exclusively triggered attacks (36). The majority of patients can precipitate attacks by touching certain trigger zones within trigeminal innervated distribution and, occasionally, even from an extratrigeminal territory. Precipitants include touching the face or scalp, washing, shaving, eating, chewing, brushing teeth, talking, and coughing (52). Neck movements can also precipitate attacks, although some patients can lessen or abort attacks by continuously rotating their neck (52,65).

Refractory Period

Unlike in trigeminal neuralgia, most patients have no refractory period (52). There have only been two case reports of absolute refractory periods in SUNCT syndrome (66,72). In addition, there is a report of a patient with both short-lasting and long-lasting attacks who demonstrated the presence of an absolute refractory period after the prolonged attacks in the absence of even a relative refractory period after the shorter attacks (37). However, most case reports of SUNCT syndrome make no mention of a refractory period. This may be due to a lack of awareness of this feature and, consequently, refractoriness in SUNCT syndrome may be underreported.

Physical Examination

The physical examination is normal in the vast majority of patients. Slight allodynia or hyperesthesia over the ophthalmic and mandibular trigeminal divisions has been reported in seven patients (19,52,57,59,60). Trigeminal hypoesthesia has been reported in four patients (7,39,52) though two of these patients had previously had invasive surgical procedures of the trigeminal nerve (7,39). Horner syndrome was previously thought not to be a feature of SUNCT. However, there have been two recent descriptions of persistent Horner syndrome in association with SUNCT (56,61). Persistent ipsilateral ptosis (69) and eyelid edema (61,62) have been reported in one and two patients, respectively. One patient was described as having transient ipsilateral meiosis without ptosis that lasted one bout only (65).

CLASSIFICATION AND DIAGNOSTIC CRITERIA

The revised IHS classification criteria (22) recognize SUNCT syndrome as a subgroup of TACs (see Table 98-1).

The IHS classification criteria for SUNCT have some notable problems. First, the name of the syndrome and the proposed classification criteria imply that all patients must have both conjunctival injection and tearing. Our clinical experience and the data from this review suggest that this is not the case. Indeed, cases of SUNCT without either conjunctival injection or tearing may have been erroneously mislabeled as other syndromes and hence underreported in the medical literature. It has been proposed that a more appropriate term for the syndrome may be short-lasting unilateral neuralgiform headache attacks with cranial autonomic symptoms (SUNA) (22,38). Second, the attack frequency of SUNCT is rather unhelpful given its breadth. Since the attacks are usually daily, changing the frequency requirement may be helpful. Third, the pain attacks can be difficult to differentiate from ophthalmic division trigeminal neuralgia. The main differentiating features in the

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TABLE 98-1 IHS Diagnostic Criteria for Short-Lasting Unilateral Neuralgiform Headache Attacks with Conjunctival Injection and Tearing

Diagnostic criteria

A. At least 20 attacks fulfilling B through E
B. Attacks of unilateral orbital, supraorbital, or temporal, stabbing or pulsating pain lasting from 5–240 seconds
C. Attack frequency from 3 to 200 per day
D. Pain is accompanied by ipsilateral conjunctival injection and lacrimation
E. Not attributed to another disorder

Comment:

This syndrome was described after the publication of the first edition of The International Classification of Headache Disorders (57) and has become well recognized in the last decade.

Patients may be seen with only one of conjunctival injection or tearing, and other cranial autonomic symptoms, such as nasal congestion, rhinorrhea, or eyelid edema, may be seen. SUNCT may be a subset of SUNA, short-lasting unilateral neuralgiform headache attacks with cranial autonomic symptoms (see Table 98-2). A chronic (unremitting) form is not yet sufficiently described but probably exists. Both forms should be coded here. The literature suggests that the most common mimic of SUNCT would be a lesion in the posterior fossa or involving the pituitary gland (see Table 98-3).

SUNCT with coexistent trigeminal neuralgia:

Patients have been described in whom there is an overlap between SUNCT and trigeminal neuralgia. Such patients should receive both diagnoses. This differentiation is clinically difficult.

From ref. 54.

proposed criteria are duration of attack and presence of autonomic features. However, there can be a considerable overlap in the duration of the attack and autonomic features can occur with trigeminal neuralgia but are generally not prominent. One possibility is to propose the requirement of prominent cranial autonomic feature(s), but this would introduce subjectivity into the criteria, unless what constitutes "prominent" autonomic symptoms is clearly defined. Another suggestion might be to introduce the absence of a refractory period to cutaneous stimulation as a criterion, though the drawback is that this feature is not adequately validated in this syndrome. Fourth, both episodic and chronic forms are now adequately validated. These subdivisions should be incorporated into the classification system, thereby bringing it in line with that of CH and paroxysmal hemicrania. Some of these problems have been addressed in the proposed criteria for SUNA and require validation (Table 98-2) (22).

PROGNOSIS

The natural history of SUNCT syndrome is poorly understood yet. The average duration of symptoms at reporting was 7 + 8 years. In 20 of these patients the duration of SUNCT exceeded 10 years. The longest reported duration of SUNCT is 48 years (52). It appears to be a lifelong disorder once it starts, though more prospective data are needed. The syndrome itself is not fatal and does not cause any long-term neurologic sequelae.

DIAGNOSTIC WORKUP

There are several case reports of secondary SUNCT (see Table 98-3). Secondary SUNCT is associated with pituitary and posterior fossa abnormalities, thereby emphasizing the absolute need for a cranial MRI, including an adequate view of the pituitary. All the reported cases of SUNCT secondary to prolactinoma initially presented with headache, and there can be a considerable time lag before the onset of pituitary-related symptoms. Hence, SUNCT patients should have a screen for basal hormone measurements including prolactin, thyroid-stimulating hormone, free thyroxine, cortisol, adrenocorticotropic hormone,

TABLE 98-2 Proposed Classification Criteria for Short-lasting Unilateral Neuralgiform Headache Attacks with Cranial Autonomic Symptoms (SUNA)

Diagnostic criteria

A. At least 20 attacks fulfilling B through F
B. Attacks of unilateral orbital, supraorbital, or temporal stabbing or pulsating pain lasting from 2 seconds to 10 minutes
C. Pain is accompanied by one of the following:
   1. Conjunctival injection and/or lacrimation
   2. Nasal congestion and/or rhinorrhea
   3. Eyelid edema
A. Attacks occur with a frequency of $\geq$1 per day for more than half of the time
B. No refractory period follows attacks triggered from trigger areas
C. Not attributed to another disorder

Episodic SUNA:

Description:

SUNA attacks occurring in periods lasting 7 days to 1 year separated by pain-free intervals lasting one month or longer

Diagnostic criteria:

A. Attacks fulfilling criteria A through F for SUNA
B. At least 2 periods of attacks lasting (if untreated) from 7 days to 1 year and separated by pain-free remission periods of $\geq$1 month

Chronic SUNA

Description:

SUNA attacks occur for more than 1 year without remission or with remissions lasting $<$1 month

Diagnostic criteria:

A. Attacks fulfilling criteria A through F for SUNA
B. Attacks recur over $>$1 year without remission periods or with remission periods lasting $<$1 month

From ref. 54.

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TABLE 98-3 Causes of Secondary SUNCT

Posterior fossa lesions
Cerebellopontine angle arteriovenous malformations (9,47)
Brainstem cavernous hemangioma (13)
Posterior fossa lesion in an HIV/AIDS patient (17)
Severe basilar impression causing pontomedullary compression in a patient with osteogenesis imperfecta (68)
Craniosynostosis resulting in a foreshortened posterior fossa (48)
Ischemic brainstem infarction (54)
Pontocerebellar astrocytoma (8)
Meningiomas at the homolateral cerebellopontine angle and frontal lobe (58)
Pituitary tumors
Nonfunctioning pituitary macroadenoma (15)
Macroprolactinoma with cavernous sinus invasion (35,40)
Microprolactinoma (31,35)
Miscellaneous
Cavernous sinus leiomyosarcoma (25)
Orbital cyst (32)
HIV-positive patient with no opportunistic infections and normal brain imaging (4)
Trigeminal nerve compression by the superior cerebellar artery has been reported (14)

SUNCT, short-lasting unilateral neuralgiform headache attacks with conjunctival injection and tearing

luteinizing hormone, follicle-stimulating hormone, estrogen, testosterone, and growth hormone. In addition, a therapeutic trial of indomethacin should be considered to exclude an indomethacin-responsive headache.

MANAGEMENT

Drug Therapies

Until recently, SUNCT was thought to be highly refractory to treatment. Numerous pharmacologic strategies have been reported to be ineffective in open-label studies including non-steroidal anti-inflammatory drugs (indomethacin, diclofenac, ibuprofen, ketoprofen, mefenamic acid, naproxen, and piroxicam), cyclooxygenase 2 inhibitors (nimesulide and celecoxib), simple analgesics (acetaminophen, aspirin, metamizole), opiates (tramadol, buprenorphine, codeine, dihydrocodeine, hydrocodone, meperidine, morphine), oxygen, ergots, methysergide, β-blockers (propranolol, timolol), β-adrenoceptor agonists and antagonists (clonidine and doxazosin), histamine desensitization and receptor antagonists (cyproheptadine), calcium channel antagonists (verapamil, nifedipine, amlodipine, flunarizine, diltiazem), γ-aminobutyric acid (GABA) agonists (baclofen), benzodiazepines, tricyclic antidepressants (amitriptyline, nortriptyline, lofepramine, imipramine, desipramine), selective serotonin reuptake inhibitors (fluoxetine), lithium, phenytoin, valproic acid, neuroleptics (pimozide, levomepromazine), central nervous system stimulants (methylphenidate, pemoline), somatostatin, angiotensin-converting enzyme inhibitors (captopril, enalapril), aprotinin, tranexamic acid, omeprazole, vitamin B_{12}, and acyclovir (36).

Recently, several agents have been suggested to be partially or completely effective, including lamotrigine, topiramate, gabapentin, carbamazepine, intravenous lidocaine, corticosteroids, and sumatriptan. On the basis of the limited, open-label data available in the literature, lamotrigine is the treatment of choice, while topiramate and gabapentin are reasonable second-line agents. Some patients have a useful response to carbamazepine and its use can be considered if the other agents are ineffective. When patients with SUNCT syndrome experience severe exacerbations with frequent, easily triggered, high-intensity pain attacks, acute interventions are needed because the patients are severely affected. They may not be able to eat or drink because these actions trigger attacks. In that situation, intravenous lidocaine can be utilized to temporarily ameliorate the attacks while conventional therapy is being optimized. Corticosteroids can also be considered in this situation but need to be used with caution because of the potential for serious side effects, especially with prolonged courses. Sumatriptan has, at best, a short-lasting transient effect and therefore has no role in the management of SUNCT syndrome.

Table 98-4 lists all the drugs that have been applied in at least two patients and their efficacy as reported by the respective authors.

Surgery

Local blockades of the stellate ganglion, pericranial nerves (including the supraorbital, infraorbital, greater occipital, and lacrimal nerves), orbicularis oculi muscles, and the retrobulbar region have generally been reported to be ineffectual. Pterygopalatine ganglion and superior cervical ganglion blockades have been reported to be partially effective in one patient each.

There are nine case reports of either apparently successful treatment or temporary relief of SUNCT syndrome with invasive surgical procedures involving the trigeminal nerve. The procedures that have been utilized include the Janetta procedure, percutaneous trigeminal ganglion compression, trigeminal ganglion thermocoagulation, retrogasserian glycerol rhizolysis, and trigeminal nerve balloon compression. These procedures provided complete pain relief, though the duration of the benefit ranged from 3 months to 4.5 years. Furthermore, as the follow-up period in some of these patients was limited to less than 18 months, it is difficult to assess the actual effectiveness of the procedures given the variable nature of attack frequency in this syndrome. This is further underscored by the observation that the benefit was temporary in all reported cases with prolonged follow-up. Conversely, there

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TABLE 98-4 Different Drug Treatments in the Case Reports on SUNCT (Number of Patient Reports in Parentheses)

	Efficacy		
Drug Name	***None***	***Moderate***	***Good***
Promising or partial clinical effect			
Lamotrigine (45)	4	—	12
Topiramate (1)	3	—	5
Gabapentin (41)	10	—	3
Carbamazepine (21)	30	11	4*
Intravenous lidocaine (43)	2	—	4
Corticosteroids			
Prednisone (18)	7	2*	2
Prednisolone or methylprednisolone (44)	3	1*	1*
Sumatriptan (2)	10	1	4
Clinically ineffective			
Indomethacin (57)	39	1	—
Oxygen (3)	9	—	—
Ergotamine or dihydroergotamine (42)	12	—	—
Methysergide (1)	7	1	—
Propranolol (36)	3	—	—
Timolol eye drops (52)	2	—	—
Doxazosin (52)	2	—	—
Verapamil (70)	16	1*	1*
Flunarizine (52)	2	—	—
Baclofen (1)	8	—	—
Clonazepam (17)	4	—	—
Amitriptyline (41)	12	1	—
Lithium (16)	6	1*	—
Phenytoin (41)	11	2	—
Valproic acid (18)	9	1	1*
Lidocaine			
Intranasal (43)	6	—	—
Mouthwash (44)	3	2	—

Only drugs tried in at least two patients are presented. The efficacy is classified according to the statements of the respective authors.
*Drug response in some of these patients reported in combination with other treatments.

are four reports of patients who were submitted to trigeminal procedures without any benefit.

Given the uncertain efficacy of trigeminal procedures together with the potential for complications, surgery should only be considered as a last resort and only when the pharmacologic options have been exploited to the fullest.

Based on the finding of ipsilateral posterior hypothalamic activation in SUNCT, Leone and colleagues (30) have treated one patient with intractable SUNCT by electrode implantation and stimulation of this region. The patient was successfully treated and the procedure was well tolerated, with no significant adverse events. Nondestructive surgical treatment modalities demand further examination, though caution needs to be exercised since deep brain stimulation procedures are associated with a small risk of mortality.

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