

- Becser, N. and M. Berky (1995). "SUNCT syndrome: a Hungarian case." *Headache* 35(3): 158-60.
 A Hungarian patient with short-lasting, unilateral, neuralgiform headache attacks with conjunctival injection and tearing (SUNCT) is presented in this paper. This male patient was first diagnosed as having first division trigeminal neuralgia. The location and duration of the attacks and the prominent accompanying autonomic features on the symptomatic side, such as conjunctival injection, lacrimation, nasal stuffiness, and the inefficacy of drugs, led to a reconsideration of the diagnosis. The pain paroxysms occurred frequently during a 3- to 4-month period, followed by a longer remission phase. Mechanical precipitating maneuvers were observed during bouts of pain. The clinical picture is reminiscent of the SUNCT syndrome, first described by Sjaastad et al in 1978. SUNCT and trigeminal neuralgia are in many ways similar, although, some decisive differences have also been noted. Further observations are needed to distinguish the two disorders and to clarify this syndrome as a new headache type or as a trigeminal neuralgia variant.
- Benoliel, R. and Y. Sharav (1998). "Trigeminal neuralgia with lacrimation or SUNCT syndrome?" *Cephalalgia* 18(2): 85-90.
 An intimate relationship between trigeminal neuralgia (TN) and short-lasting unilateral neuralgiform headache attacks with conjunctival injection and tearing (SUNCT) syndrome, based on similar clinical signs and symptoms and on cases demonstrating possible "transformation" from one entity to the other, has been widely accepted. We evaluated the presence of lacrimation in 22 consecutive cases that had been diagnosed as TN. Ipsilateral lacrimation was reported by 6 such cases (5M, 1F). These cases responded to antineuralgic therapy with concomitant resolution of lacrimation and were clinically very similar to TN. The differential diagnosis and the possibility of lacrimation in TN are discussed.
- Benoliel, R. and Y. Sharav (1998). "SUNCT syndrome: case report and literature review." *Oral Surg Oral Med Oral Pathol Oral Radiol Endod* 85(2): 158-61.
 The case of a woman with short neuralgiform paroxysmal pain of 2 years duration is described. Pain attacks were always accompanied by ipsilateral lacrimation and conjunctival injection. Standard anti-neuralgic therapy, amitriptyline and indomethacin, failed to eliminate or reduce pain. At the end of a 30-month active period the patient seemed to have gone into remission. We believe this to be a case of short-lasting, unilateral, neuralgiform headache attacks with conjunctival injection and tearing (SUNCT), the first reported in the dental literature and the 24th in the general medical literature. The differential diagnosis of the case and relevant literature are discussed.
- Black, D. F. and D. W. Dodick (2002). "Two cases of medically and surgically intractable SUNCT: a reason for caution and an argument for a central mechanism." *Cephalalgia* 22(3): 201-4.
 We report two cases of SUNCT that demonstrate the medically and surgically refractory nature of this disorder and support the hypothesis that the causative 'lesion' lies within the central nervous system. After both patients had failed medical therapies, the first underwent a glycerol rhizotomy, gammaknife radiosurgery and microvascular decompression of the trigeminal nerve. The second patient underwent gammaknife radiosurgery of the trigeminal root exit zone and two microvascular decompression surgeries. Neither patient benefited from these procedures. Currently, the first patient suffers from anaesthesia dolorosa and the second patient from unilateral deafness, chronic vertigo and dysequilibrium as a result of surgical trauma. These cases of SUNCT highlight the uncertainty regarding the role of surgery given the potential for significant morbidity. These cases also suggest that SUNCT originates and may be maintained from within the CNS and this central locus explains why SUNCT is not typically amenable to interventions aimed at the peripheral portion of the trigeminal nerve.
- Bouhassira, D., N. Attal, et al. (1994). "'SUNCT' syndrome. A case of transformation from trigeminal neuralgia?" *Cephalalgia* 14(2): 168-70.
 A patient with typical trigeminal neuralgia involving the first branch of the nerve developed short-lasting unilateral attacks in the same area which were associated with severe vasomotor phenomena consistent with the recently described SUNCT syndrome. This evolution suggests that SUNCT might correspond, at least in this case, to a "transformed" trigeminal neuralgia and emphasizes the close relationship between these unilateral facial pain syndromes.
- Calvo, J. F., O. C. Bruera, et al. (2004). "SUNCT syndrome: clinical and 12-year follow-up case report." *Cephalalgia* 24(10): 900-2.
- Chakravarty, A. and A. Mukherjee (2003). "SUNCT syndrome responsive to lamotrigine: documentation of the first Indian case." *Cephalalgia* 23(6): 474-5.

- Chakravarty, A., A. Mukherjee, et al. (2004). "Trigeminal autonomic cephalgias and variants: clinical profile in Indian patients." *Cephalalgia* 24(10): 859-66.
The present study summarizes the authors' experience of the clinical profile of short-lasting trigeminal autonomic cephalgias (TAC) in Indian patients. Over a period of 17 years a total of 41 cases of episodic cluster headache, seven cases of chronic cluster headache, six cases of variant cluster headache, three cases of paroxysmal hemicrania, and a single case of SUNCT syndrome were encountered. TACs appear to be rare in Indian patients and cluster headache seems to be exclusively a disease of men. The present report is to the best of our knowledge the first of its kind to be reported from India.
- Cohen, A. S., M. S. Matharu, et al. (2004). "SUNCT syndrome in the elderly." *Cephalalgia* 24(6): 508-9.
- D'Andrea, G., F. Granella, et al. (1999). "Possible usefulness of lamotrigine in the treatment of SUNCT syndrome." *Neurology* 53(7): 1609.
- D'Andrea, G., F. Granella, et al. (2001). "Lamotrigine in the treatment of SUNCT syndrome." *Neurology* 57(9): 1723-5.
Short-lasting, unilateral, neuralgiform headache attacks with conjunctival injection and tearing (SUNCT) is considered an intractable condition. The authors treated five patients with lamotrigine (125 to 200 mg daily), obtaining a complete remission in three patients and a substantial reduction (about 80%) of attack frequency in the other two. No adverse effects were noted.
- D'Andrea, G. and F. Granella (2001). "SUNCT syndrome: the first case in childhood. Short-lasting unilateral neuralgiform headache attacks with conjunctival injection and tearing." *Cephalalgia* 21(6): 701-2.
- De Benedittis, G. (1996). "SUNCT syndrome associated with cavernous angioma of the brain stem." *Cephalalgia* 16(7): 503-6.
A 62-year-old male patient with 2 years of SUNCT syndrome is described. The patient presented with long-lasting periods of frequent attacks of intense orbital pain with a duration of about 1 min, associated with ipsilateral conjunctival injection, lacrimation and rhinorrhea. Cranial MRI and cerebral angiography demonstrated an ipsilateral cavernous angioma of the pons, involving the trigeminal roots. As the pain was refractory to most treatments, including carbamazepine, the patient asked for, and eventually underwent, direct surgical excision of the malformation. Following the operation, his neurological conditions rapidly deteriorated and he died of postoperative complications (haemorrhage).
- Di Monda, V. (1999). "Chronic paroxysmal hemicrania and SUNCT syndrome." *Ital J Neurol Sci* 20(2 Suppl): S49-52.
- el Amrani, M., H. Massiou, et al. (2001). "[Idiopathic SUNCT (short lasting unilateral neuralgiform headache attacks with conjunctival injection, tearing, sweating and rhinorrhea) syndrome: 2 new cases]." *Rev Neurol (Paris)* 157(12): 1519-24.
The syndrome of short-lasting unilateral, neuralgiform attacks of pain in the peri orbital area associated with conjunctival injection and tearing (SUNCT) is a rare disorder affecting mainly males. We report two French patients (1 male and 1 female) with SUNCT syndrome, 27 and 28 years of age respectively. Both patients had short (30 sec), frequent (30-100/day) excruciating pain located at the peri orbital area, associated with conjunctival injection, tearing, rhinorrhea, ptosis and others vasomotor symptoms. Clinical examination and imaging were normal. Most drugs used in the treatment of migraine, cluster headache, trigeminal neuralgia, and other short-lasting headaches were not successful.
- Forderreuther, S. and A. Straube (1999). "[A rare headache syndrome. SUNCT syndrome, hemicrania continua and red ear syndrome]." *Nervenarzt* 70(8): 754-8.
Rare headache syndromes that are so far not yet admitted to the classification system of the International Headache Society are the SUNCT syndrome (short-lasting, unilateral, neuralgiform headache attacks with conjunctival injection and tearing), the hemicrania continua and the red ear syndrome. The clinical characteristics, differential diagnoses and therapeutic strategies of these pathophysiologically unclarified diseases are presented with three casuistic reports.
- Gardella, L., A. Viruega, et al. (2001). "A case of a patient with SUNCT syndrome treated with Jannetta procedure." *Cephalalgia* 21(10): 996-9.

Ghose, R. R. (1995). "SUNCT syndrome." *Med J Aust* 162(12): 667-8.

Gladstone, J., E. Eross, et al. (2003). "Chronic daily headache: a rational approach to a challenging problem." *Semin Neurol* 23(3): 265-76.

Chronic daily headache (CDH) is a significant public health problem with 3 to 5% of the population worldwide experiencing daily or near-daily headaches. Patients with CDH can be particularly challenging, and clinicians require a systematic approach to help guide investigations and management. The revised 2004 International Headache Society Classification Criteria introduces formalized criteria for several CDH disorders including chronic migraine and medication overuse headache as well as new daily persistent headache, hemicrania continua, hypnic headache, and SUNCT syndrome. Medication overuse is common in patients with CDH who present to physicians. Familiarity and comfort with drug-withdrawal and detoxification strategies is therefore essential. Patients with chronic migraine and chronic cluster experience significant disability and diminished quality of life. The ability to manage these patients effectively is a rewarding clinical experience.

Goadsby, P. J. and R. B. Lipton (1997). "A review of paroxysmal hemicranias, SUNCT syndrome and other short-lasting headaches with autonomic feature, including new cases." *Brain* 120 (Pt 1): 193-209. The short-lasting primary headache syndromes may be conveniently divided into those exhibiting marked autonomic activation and those without autonomic activation. The former group comprise chronic and episodic paroxysmal hemicrania, short-lasting unilateral neuralgiform headache with conjunctival injection and tearing (SUNCT syndrome) and cluster headache. These headache syndromes are compared with other short-lasting headache disorders, such as hypnic headache, and persistent headache with milder autonomic features such as hemicrania continua. Cluster headache is included with the shorter-lasting headaches to attempt a nosological analysis of these syndromes. The paroxysmal hemicranias are characterized by frequent short-lasting attacks of unilateral pain usually in the orbital, supraorbital or temporal region that typically last minutes. The attack frequency usually ranges from 5 to 40 attacks per day. The pain is severe and associated with autonomic symptoms such as conjunctival injection, lacrimation, nasal congestion, rhinorrhoea, ptosis or eyelid oedema. Almost all reported cases respond to treatment with indomethacin, but respond poorly to other treatments including other nonsteroidal anti-inflammatory drugs. A recent case study demonstrated the release of both trigeminal and parasympathetic neuropeptides during a bout of pain in the same pattern previously described in cluster headache. The SUNCT syndrome is a distinctive rare condition characterized by less severe pain but marked autonomic activation during attacks. Consistent with previous reports, the present case of SUNCT syndrome was intractable to therapy. The similarities of these syndromes suggests a considerable shared pathophysiology. It is suggested that the syndromes are sufficiently well established for inclusion in the International Headache Society Classification system and that trigeminal-autonomic cephalalgias should be classified as a group together. A proposed re-classification is presented.

Goadsby, P. J., M. S. Matharu, et al. (2001). "SUNCT syndrome or trigeminal neuralgia with lacrimation." *Cephalalgia* 21(2): 82-3.

Graff-Radford, S. B. (2000). "SUNCT syndrome responsive to gabapentin (Neurontin)." *Cephalalgia* 20(5): 515-7.

A 48-year-old male suffering with SUNCT (severe unilateral neuralgiform headache with conjunctival injection and tearing, rhinorrhea and sub-clinical sweating) presented in 1996 after a 10-year history of multiple failed therapies. The symptoms included strictly left-sided ocular, as well as facial and temple pain. The pain attacks were burning, sharp, shooting and occurred 25 times daily, lasting 2 to 3 minutes with tearing and conjunctival injection. There was no associated nausea or vomiting, but there was photophobia. No other autonomic changes were reported and the pain was not triggerable. Initially Indocin (indomethacin) was tried without significant benefit. Gabapentin (Neurontin) was then started with improvement at 1800 mg per day. The patient was then lost to follow-up for 3 years, as he moved from the Los Angeles area. He returned in 1999 having stopped the gabapentin after his prescription ran out in 1996, reporting the pain returned immediately. Again gabapentin was prescribed and at 900 mg three times daily he has been pain free for 12 months.

Grechko, V. E. and A. M. Dobrovitskaia (2000). "[A specific form of headache with neuralgia-like manifestations (SUNCT-syndrome)]." *Zh Nevrol Psikhiatr Im S S Korsakova* 100(12): 28-31.

A recently described SUNCT is a rare head ache syndrome characterized by shortlasting, unilateral, neuralgia-like headache attack with conjunctival injection and tearing; 26 cases of SUNCT-syndrome have been reported in medical literature. The authors observed the described symptoms in 3 cases

(among 86 patients with trigeminal paroxysmal manifestations). Clinical features of SUNCT-syndrome are presented. In addition, the authors discuss possible etiologic factors and treatment as well as a differential diagnosis of SUNCT-syndrome, from trigeminal neuralgia, cluster headache, chronic paroxysmal hemicrania and combined syndromes.

- Gutierrez-Garcia, J. M. (2002). "SUNCT syndrome responsive to lamotrigine." *Headache* 42(8): 823-5.
BACKGROUND: Short-lasting, unilateral, neuralgiform headache attacks with conjunctival injection, tearing, rhinorrhea (SUNCT syndrome) is a headache form generally refractory to drug therapy. Occasional patients with SUNCT have been reported with a successful response to lamotrigine. OBJECTIVE: To report two patients with SUNCT treated with lamotrigine. METHODS: Clinical history, neurologic examination, and brain magnetic resonance imaging. RESULTS: Both patients with SUNCT syndrome were successfully treated with lamotrigine. In both cases, when lamotrigine was tapered off, the attacks reappeared, only to disappear when the dose was again increased. In addition, lamotrigine was well tolerated and no undesired side-effects were reported. CONCLUSION: If the positive effect of lamotrigine in patients with SUNCT is confirmed in other cases, lamotrigine could become the first specific treatment for SUNCT syndrome.
- Hannerz, J., D. Greitz, et al. (1992). "SUNCT may be another manifestation of orbital venous vasculitis." *Headache* 32(8): 384-9.
A patient with more than 20 years of SUNCT, i.e., long lasting periods with frequent attacks of intense orbital pain with a duration of about one minute, associated with ipsilateral conjunctival injection, lacrimation, rhinorrhea and facial sweating is described. Some attacks were possibly related to increased cerebral blood flow but could also be triggered from the oral area. Orbital phlebography showed pathologic changes on the side of the pain, changes which were normalized when these attacks ceased to appear. Due to these findings in conjunction with serum evidence of inflammation, associated systemic symptoms and susceptibility to steroids and azathioprine, venous vasculitis is suggested to be the cause of SUNCT in this patient. Carbamazepine and sumatriptan decreased the frequency, intensity and duration of attacks, although not completely.
- Hannerz, J. and B. Linderöth (2002). "Neurosurgical treatment of short-lasting, unilateral, neuralgiform hemicrania with conjunctival injection and tearing." *Br J Neurosurg* 16(1): 55-8.
Trigeminal neuralgia solely involving the upper trigeminal nerve branch is rare. The SUNCT syndrome (short-lasting, unilateral, neuralgiform hemicrania with conjunctival injection and tearing) in which the periorbital pain lasts for 60-120 s, and is accompanied by conjunctival injection and tearing is even less common. Unlike trigeminal neuralgia, SUNCT is usually not relieved by medication. Three patients with SUNCT were treated with retrogasserian glycerol rhizolysis, two of them twice. All five treatments provided complete pain relief and the duration of the effects was 2 to more than 4 years. One of these three patients also had a third treatment with compression of retroganglionic fibres with a Fogarty balloon, according to Mullan, of the upper trigeminal nerve with excellent results.
- Hunt, C. H., D. W. Dodick, et al. (2002). "SUNCT responsive to gabapentin." *Headache* 42(6): 525-6.
- Jimenez-Huete, A., O. Franch, et al. (2002). "SUNCT syndrome: priming of symptomatic periods and worsening of symptoms by treatment with calcium channel blockers." *Cephalalgia* 22(10): 812-4.
We report a SUNCT patient who showed both a precipitation and worsening of symptomatic periods after treatment with L-type calcium channel blockers. This pharmacological response may provide us with important clues for understanding the pathophysiology of SUNCT, and hopefully to find a remedy for the victims of this syndrome. This observation could also support a verapamil trial in SUNCT patients as a precipitating of attacks.
- Kaphan, E., A. Eusebio, et al. (2003). "Shortlasting, unilateral, neuralgiform headache attacks with conjunctival injection and tearing (SUNCT syndrome) and tumour of the cavernous sinus." *Cephalalgia* 23(5): 395-7.
- Klimek, A. and P. Sklodowski (2001). "[SUNCT syndrome--case report]." *Neurol Neurochir Pol* 35(1): 153-8.
Short-lasting unilateral neuralgiform headache attacks with conjunctival injection and tears (SUNCT) syndrome is a very rare disorder, first described by Sjaastad et al. in 1989. The authors report a case of that type of headache in an 80 years old woman. They also discuss the pathophysiology and treatment.

Koseoglu, E., Y. Karaman, et al. (2005). "SUNCT syndrome associated with compression of trigeminal nerve." *Cephalalgia* 25(6): 473-5.

Kruszewski, P., M. L. Fasano, et al. (1991). "Shortlasting, unilateral, neuralgiform headache attacks with conjunctival injection, tearing, and subclinical forehead sweating ("Sunct" syndrome): II. Changes in heart rate and arterial blood pressure during pain paroxysms." *Headache* 31(6): 399-405.
The recently described "Sunct" syndrome is a rare picture of unilateral, shortlasting headache attacks accompanied by autonomic phenomena (conjunctival injection, tearing, etc.) on the symptomatic side. Heart rate and blood pressure were monitored in two elderly "Sunct" patients during and outside headache attacks. An ultrasound Doppler servo method was used for the non-invasive, continuous, beat-to-beat determination of instantaneous arterial blood pressure. In a third patient, systolic and diastolic blood pressure, both outside and during pain paroxysms, were assessed using the standard Korotkoff method. Heart rate was found to be significantly decreased during pain paroxysms. Systolic blood pressure was observed to be significantly increased during attacks, when compared with the inter-attack period, while a less consistent pattern was observed for diastolic blood pressure. Some of the changes in the cardiovascular system seemed to start prior to pain onset. Therefore, it seems unlikely that these changes were caused by pain activation of the sympathetic nervous system or the oculocardiac reflex.

Kruszewski, P. (1992). "Shortlasting, unilateral, neuralgiform headache attacks with conjunctival injection and tearing (SUNCT syndrome): V. Orbital phlebography." *Cephalalgia* 12(6): 387-9.
We studied six patients with SUNCT, a unilateral headache syndrome with shortlasting attacks and ipsilateral autonomic phenomena with orbital phlebography and MRI. All but one orbital phlebogram showed abnormalities on the headache side (in one patient bilaterally), involving the superior ophthalmic vein or/and the cavernous sinus. No systematic changes were demonstrated on MRI. The phlebography findings were similar to those observed in the Tolosa-Hunt syndrome and in cluster headache.

Kruszewski, P., T. Sand, et al. (1992). "Short-lasting, unilateral, neuralgiform headache attacks with conjunctival injection and tearing (SUNCT syndrome): IV. Respiratory sinus arrhythmia during and outside paroxysms." *Headache* 32(8): 377-83.
SUNCT is a headache syndrome characterized by short-lasting (usually 15-120 sec), unilateral head pain paroxysms localized in the peri-ocular area, accompanied by conjunctival injection, lacrimation, nasal stuffiness, rhinorrhea, and subclinical forehead sweating, all on the symptomatic side. A relative bradycardia seems to be an integral part of the paroxysm; a parasympathetic stimulation could theoretically be the causative factor for the bradycardia. In 3 SUNCT patients, vagal nerve function (E:I ratio) has been monitored outside and during pain paroxysms, while 3 other patients could be studied in the attack-free period only. E:I ratio is obtainable in the course of a maximally deep breath and represents the ratio of the longest R-R interval during a 5 sec long expiration to the shortest R-R interval during a 5 sec long expiration. The mean E:I ratio of SUNCT patients outside paroxysms was significantly higher than the mean E:I ratio in an aged-matched control group. The E:I ratio was, however, significantly decreased during paroxysms in comparison with ratios obtained outside the pain paroxysms. After 0.6 mg atropine administration s.c. to one of the patients in the symptomatic phase, the heart rate increased, and the relative bradycardia during headache paroxysm was diminished (but not completely abolished). The E:I ratio was lowered but it was still slightly larger outside than during attacks. The reason for the abrupt and seemingly clear attack-related decrement in E:I ratio together with the previously described relative bradycardia remains enigmatic, however the possibility of increased parasympathetic tone cannot be excluded.

Kruszewski, P., J. M. Zhao, et al. (1993). "SUNCT syndrome: forehead sweating pattern." *Cephalalgia* 13(2): 108-13.
The forehead sweating function has been assessed in SUNCT syndrome--a short-lasting, unilateral, neuralgiform headache syndrome with autonomic phenomena on the symptomatic side (conjunctival injection, lacrimation, etc.). In the three patients (of a total of six) who could be studied during paroxysms, increased evaporation was present on the symptomatic side of the forehead compared to the non-symptomatic side during attacks or to the symptomatic side between attacks. Basal sweating was generally within control limits, so long as the attack frequency was not so high as to influence the interictal level. During attacks precipitated by eating chocolate or sour apple (in the case of one of the patients), forehead sweating was also increased on the symptomatic side. The forehead sweating responses to heating and pilocarpine were without any notable or systematic asymmetries. The forehead sweating pattern in SUNCT syndrome may differ from the patterns in unilateral headaches like cluster headache, on the one hand (in which there is generally an asymmetry during

heating and pilocarpine tests), and chronic paroxysmal hemicrania (CPH) and cervicogenic headache, on the other (where there is no systematic increase during attacks).

- Kruszewski, P., L. R. White, et al. (1995). "Respiratory studies in SUNCT syndrome." *Headache* 35(6): 344-8. Seven SUNCT patients (six men, one woman) took part in this study. In four patients, respiratory variables were compared during and outside attacks. In five patients, peripheral chemosensitivity was tested and compared with a control group matched with respect to age, sex, and smoking habits. The results indicate that SUNCT patients hyperventilate during attacks. Moreover, they appear to hyperventilate slightly under basal conditions. The tests for peripheral chemoreceptor activity indicated no differences between the SUNCT and the control groups except for one variable, namely the mean ventilatory response to a single breath of 13% CO₂. It is possible that this indicates a blunted response of the peripheral chemoreceptors. On the other hand, it may also represent a chance finding, since none of the other results presented suggested such a conclusion, and the size of the test group was very small. The results do not indicate that a reduction in oxygen saturation can trigger SUNCT since low levels of oxygen saturation were only rarely accompanied by SUNCT, whereas many attacks were not associated with any appreciable lowering in arterial oxygen saturation.
- Kuhn, J., M. Vosskaemper, et al. (2005). "SUNCT syndrome: a possible bilateral case responding to topiramate." *Neurology* 64(12): 2159.
- Kuhn, J. and H. Bewermeyer (2005). "[Trigeminal autonomic headache, hemicrania continua and hypnic headache. A review of rare primary headache forms]." *Dtsch Med Wochenschr* 130(19): 1221-6. Tension-type headache and migraine are the most common types of primary headaches. Apart from these well known diseases, the group of primary headaches includes other relatively rare headache disorders. Some of these seldom syndromes have been described for the first time within the last twenty years and have been newly included in the revised IHS classification from 2004. Their typical symptomatic is less known, but offers an excellent opportunity to diagnose these syndromes. The importance of recognising these disorders is underlined by the fact, that rare primary headaches response often complete and rapid to a specific treatment. This review summarizes the current knowledge on the clinical presentation and treatment of cluster headache, paroxysmal hemicrania, SUNCT syndrome, hemicrania continua and hypnic headache.
- Lain, A. H., A. B. Caminero, et al. (2000). "SUNCT syndrome; absence of refractory periods and modulation of attack duration by lengthening of the trigger stimuli." *Cephalalgia* 20(7): 671-3.
- Lanusse, S., O. Senechal, et al. (1999). "[Sunct syndrome: case report and literature review]." *Rev Neurol (Paris)* 155(12): 1071-3. The case of a woman with short neuralgiform paroxysmal attacks located in orbital-periorbital area and associated with autonomic features of ten years duration is reported. This headache syndrome is compared with trigeminal neuralgia involving the first branch of the nerve. Duration, intensity, spreading of the pain and presence of accompanying ipsilateral vasomotor phenomena may be of help in the differential diagnosis. According to the latest reports, sex distribution which passed from 17 men/2 women to 18/6 and effect of the carbamazepine on pain would not appear to have an effect. Nevertheless other reports are needed to distinguish these two clinical syndromes and to develop an etiological and pathogenesis hypothesis.
- Leone, M., A. Rigamonti, et al. (2000). "Two new SUNCT cases responsive to lamotrigine." *Cephalalgia* 20(9): 845-7. Short-lasting unilateral neuralgiform headache attacks with conjunctival injection and tearing (SUNCT) is a rare and debilitating headache form generally unresponsive to treatment. Following a recent report of a SUNCT patient who responded to lamotrigine, we tried this drug in two new SUNCT patients, reported here. In both cases prophylaxis was successful, suggesting lamotrigine might be the first effective treatment for this rare and debilitating headache syndrome.
- Lim, E. C. and H. L. Teoh (2003). "Headache - it's more than meets the eye: orbital lesion masquerading as SUNCT." *Cephalalgia* 23(7): 558-60.
- Malik, K., S. Rizvi, et al. (2002). "The SUNCT syndrome: successfully treated with lamotrigine." *Pain Med* 3(2): 167-8. Short-lasting unilateral neuralgiform headache attacks with conjunctival injection and tearing (SUNCT) syndrome is an uncommon headache syndrome. It is usually resistant to treatment. We

describe a patient who responded successfully to the anticonvulsant lamotrigine, but not other anticonvulsants commonly used in the treatment of migraine and other cephalgias.

- Massiou, H., J. M. Launay, et al. (2002). "SUNCT syndrome in two patients with prolactinomas and bromocriptine-induced attacks." *Neurology* 58(11): 1698-9.
- Matharu, M. S., C. J. Boes, et al. (2002). "SUNCT syndrome: prolonged attacks, refractoriness and response to topiramate." *Neurology* 58(8): 1307.
- Matharu, M. S., M. J. Levy, et al. (2003). "SUNCT syndrome secondary to prolactinoma." *J Neurol Neurosurg Psychiatry* 74(11): 1590-2.
Short-lasting unilateral neuralgiform headache attacks with conjunctival injection and tearing (SUNCT) syndrome is a rare form of primary headache disorder, although secondary causes, particularly posterior fossa abnormalities, are well known. We report a case of SUNCT syndrome secondary to a prolactinoma. Administration of dopamine agonists led to complete resolution of the SUNCT attacks. This case, together with other similar case reports in the literature, highlight the importance of excluding a diagnosis of pituitary adenoma in all suspected cases of SUNCT syndrome, especially as the headache can precede more classical pituitary symptoms by a considerable period of time. Clinicians managing patients with suspected SUNCT syndrome should elicit a history of symptoms associated with pituitary neoplasms, perform a magnetic resonance imaging scan of the brain and pituitary, and screen for serum hormonal abnormalities.
- Matharu, M. S., C. J. Boes, et al. (2003). "Management of trigeminal autonomic cephalgias and hemicrania continua." *Drugs* 63(16): 1637-77.
The trigeminal autonomic cephalgias (TACs) are a group of primary headache disorders characterised by unilateral trigeminal distribution pain that occurs in association with ipsilateral cranial autonomic features. This group of headache disorders includes cluster headache, paroxysmal hemicrania and short-lasting unilateral neuralgiform headache attacks with conjunctival injection and tearing (SUNCT syndrome). Although hemicrania continua has previously been classified amongst the TACs, its nosological status remains unclear. Despite their similarities, these disorders differ in their clinical manifestations and response to therapy, thus underpinning the importance of recognising them. We have outlined the clinical manifestations, differential diagnoses, diagnostic workup and the treatment options for each of these syndromes.
- Matharu, M. S., A. S. Cohen, et al. (2003). "Short-lasting unilateral neuralgiform headache with conjunctival injection and tearing syndrome: a review." *Curr Pain Headache Rep* 7(4): 308-18.
The clinical features of short-lasting unilateral neuralgiform headache attacks with conjunctival injection and tearing (SUNCT) syndrome have been reviewed in 50 patients reported in the English language literature. SUNCT syndrome is a rare condition that predominates slightly in men. The mean age at onset is 50 years. It is characterized by strictly unilateral attacks centered on the orbital or periorbital regions, forehead, and temple. Generally, the pain is severe and neuralgic in character. The usual duration ranges from 5 to 250 seconds, although the reported range of duration is 2 seconds to 20 minutes. Ipsilateral conjunctival injection and lacrimation are present in most, but not all patients. Most patients are thought to have no refractory periods and this has probably been unreported in the past. Episodic and chronic forms of SUNCT exist. The attack frequency varies from less than one attack daily to more than 60 attacks per hour. The attacks are predominantly diurnal, although frequent nocturnal attacks can occur in some patients. A functional magnetic resonance imaging study in SUNCT syndrome has demonstrated ipsilateral hypothalamic activation. SUNCT was thought to be highly refractory to treatment. However, recent open-label trials of lamotrigine, gabapentin, topiramate, and intravenous lidocaine have produced beneficial therapeutic responses. These results offer the promise of better treatments for this syndrome, but require validation in controlled trials.
- Matharu, M. S., A. S. Cohen, et al. (2004). "SUNCT syndrome responsive to intravenous lidocaine." *Cephalalgia* 24(11): 985-92.
Short-lasting unilateral neuralgiform headache with conjunctival injection and tearing (SUNCT) is a primary headache syndrome that has been reported to be resistant to treatment with intravenous lidocaine. We report four cases of SUNCT in whom intravenous lidocaine (1.3-3.3 mg kg⁻¹ h⁻¹) completely suppressed the headaches for the duration of the infusion. The headache returned after cessation of treatment. Two patients went on to have their symptoms controlled on topiramate (50-300 mg daily). One patient had typical migrainous aura in association with some of the attacks of pain but never migrainous headaches. These cases suggest that treatment with lidocaine can be

considered when acute intervention is required to suppress a severe exacerbation of SUNCT, and further broaden the therapeutic and clinical background of this syndrome.

- May, A., A. Bahra, et al. (1999). "Functional magnetic resonance imaging in spontaneous attacks of SUNCT: short-lasting neuralgiform headache with conjunctival injection and tearing." *Ann Neurol* 46(5): 791-4.
A 71-year-old woman presented with a short history of episodes of severe left-sided orbital and temporal pain in paroxysms lasting 60 to 90 seconds, and accompanied by ipsilateral lacrimation of the eye, rhinorrhea, and conjunctival injection. Results of clinical examination and structural imaging were normal and a clinical diagnosis of SUNCT (short-lasting unilateral neuralgiform pains with conjunctival injection and tearing) was made. The patient had a BOLD contrast-magnetic resonance imaging study in which significant activation was seen in the region of the ipsilateral hypothalamic gray, comparing the pain to pain-free state. The region of activation was the same in this patient as has been reported in acute attacks of cluster headache.
- May, A. (2004). "[Headache attacks with ipsilateral autonomic symptoms]." *Schmerz* 18(5): 370-7.
Following the new IHS-classification, cluster headache, paroxysmal hemicrania and SUNCT syndrome are included into the classification as trigemino-autonomic cephalgias (TAC's). Clinically, they share strictly half-sided head pain with autonomic symptoms. The headaches often occur during particular sleep stages and are associated with other chronobiologic factors. Broadly the management of TAC's comprises acute and prophylactic treatment. Paroxysmal hemicrania and hemicrania continua have a very robust response to indomethacin. Acute cluster headache attacks can be treated with inhalation of oxygen or serotonin agonists, whereas verapamil is the drug of choice in the prophylactic treatment. This review covers the clinical picture and therapeutic options. Although studies following the criteria of evidence based medicine (EBM) are rare, most patients can be treated sufficiently.
- May, A., S. Evers, et al. (2005). "[Treatment and prophylaxis for cluster headaches and other trigeminal autonomic headaches. Revised recommendations of the German Migraine and Headache Society]." *Schmerz* 19(3): 225-41.
Following the new IHS classification, cluster headache, paroxysmal hemicrania, and short-lasting unilateral neuralgiform headache with conjunctival injection and tearing (SUNCT syndrome) are included in the classification as trigeminal autonomic cephalgias (TAC). The similarities of these syndromes suggest a considerable shared pathophysiology. These syndromes have in common that they involve activation of trigeminovascular nociceptive pathways with reflex cranial autonomic activation. Clinically, this physiology predicts pain with some combination of lacrimation, conjunctival injection, nasal congestion, or eyelid edema. Broadly the management of TAC comprises acute and prophylactic treatment. Some types of trigeminal autonomic headaches such as paroxysmal hemicrania and hemicrania continua have, unlike cluster headaches, a very robust response to indomethacin, leading to a consideration of indomethacin-sensitive headaches. This review covers the clinical picture and therapeutic options. Although studies following the criteria of evidence-based medicine (EBM) are rare, most patients can be treated sufficiently.
- Montes, E., R. Alberca, et al. (2001). "Statuslike SUNCT in two young women." *Headache* 41(8): 826-9.
OBJECTIVE: To describe the statuslike pattern of SUNCT (short-lasting unilateral neuralgiform pain with conjunctival injection and tearing) in two young women. BACKGROUND: SUNCT syndrome is a rare condition characterized by a short-lasting periocular pain associated with marked autonomic symptoms. Twenty-five cases have been reported in the literature with a high preponderance of males and a mean age of 51 years. The frequency of episodes shows a wide variability, not just among individuals but also in the same patient, and a statuslike pattern of almost continuous attacks has been described. METHODS: We report the cases of two young women (aged 26 and 23 years) with typical SUNCT features who suffered bouts of up to 60 paroxysms of pain per hour. Paraclinical investigations showed no abnormalities. CONCLUSIONS: Although unusual, paroxysms in SUNCT may overlap into a clinical status. A strong relationship with hormonal changes was noted in one patient. In both cases, the pain was refractory to treatment with indomethacin, carbamazepine, and hypnotics, and only intravenous methylprednisolone with oral carbamazepine may have been partially effective in one case.
- Morales, F., E. Mostacero, et al. (1994). "Vascular malformation of the cerebellopontine angle associated with "SUNCT" syndrome." *Cephalgia* 14(4): 301-2.
A 70-year-old man complained of two distinct types of unilateral headache during the past fifteen years. When the illness began, the pain was intermittent and short-lasting. In successive years, the

crises appeared in clusters and lasted weeks to months. At present, the pain occurs daily, and is located on the right side, from the forehead supraorbitally to the temporal region. Some attacks last 30 sec and are accompanied by tearing, conjunctival injection, rhinorrhea and a subjective need to micturate. Other headaches last 1/2-1 h and are occasionally accompanied by local ipsilateral dysautonomic symptoms. Attacks of pain are provoked by movements of the trunk and neck. A vascular malformation in the right cerebellopontine angle was demonstrated on cranial CT and MRI, and by angiography.

Morales Asin, F. (1997). "[Chronic paroxysmal hemicrania]." *Neurologia* 12 Suppl 5: 44-9.

In order to provide an update of clinical, pathogenic, diagnostic and therapeutic aspects of chronic paroxysmal hemicrania (CPH), also known as Sjaastad syndrome, we review the relevant literature. The term CPH was proposed by Sjaastad in 1976. Although continuous and non continuous cases have been described, Sjaastad prefers to use the term "prechronic" for the non continuous form, given that the adjective "chronic" denotes an important defining feature of CPH. CPH, which has been included in International Headache Society classification system since 1988, is much less common than cluster headache. CPH can be defined as pain that mainly affects women, is unilateral, always on the same side, and generally oculo-fronto-temporal. It can appear at any hour of the day or night, can be triggered by various phenomena and is accompanied by dysautonomic phenomena, generally on the same side as the pain, such as red eyes, tearing, nasal congestion and sometimes rhinorrhea. This headache is distinguished by its response to indomethacin therapy. The pathogenesis of CPH is unknown, although it is believed to resemble cluster headache, at least in its final stages (involving the trigeminal vascular system). Differential diagnosis should include cluster headache, SUNCT syndrome, continuous hemicrania and cervical headache, as well as facial neuralgia.

Moris, G., R. Ribacoba, et al. (2001). "SUNCT syndrome and seborrheic dermatitis associated with craniostenosis." *Cephalgia* 21(2): 157-9.

Narbone, M. C., S. Gangemi, et al. (2005). "A case of SUNCT syndrome responsive to verapamil." *Cephalgia* 25(6): 476-8.

Neumeier, S., T. Brinkschmidt, et al. (2003). "[Case report on a patient with SUNCT-syndrome]." *Schmerz* 17(2): 142-6.

Case report on a patient with SUNCT-syndrome (short lasting, unilateral neuralgiform headache attacks with conjunctival injection, sweating, and rhinorrhoea) who was successfully treated with gabapentin. SUNCT, a still relatively unknown headache syndrome, is characterized by attacks of periorbital pain with accompanying ipsilateral autonomic symptoms. Along with this case report the differences of SUNCT to similar headaches are emphasized. Due to clear diagnostic criteria the inclusion of SUNCT in the IHS classification (International Headache Society) as a separate clinical entity should be favoured.

Newman, L. C. (2001). "Effective management of ice pick pains, SUNCT, and episodic and chronic paroxysmal hemicrania." *Curr Pain Headache Rep* 5(3): 292-9.

Idiopathic stabbing headaches, the SUNCT syndrome, and the paroxysmal hemicranias are a group of primary headache disorders that are characterized by brief, short-lived attacks of head pain, which recur multiple times throughout the day. These syndromes are much less prevalent than other types of primary headaches such as migraine and tension-type headaches but are significantly more disabling. Recognition of these uncommon disorders is important because their management differs from standard headache therapies.

Pareja, J. A. and O. Sjaastad (1994). "SUNCT syndrome in the female." *Headache* 34(4): 217-20.

Two female patients suffering from SUNCT syndrome are presented. They are the first female SUNCT cases reported (of a total of 16 cases hitherto seen by us, reported to us, or published). Also some minor modifications of the hitherto known clinical symptomatology of SUNCT syndrome have been observed. Attacks could be triggered in a variety of ways. A short latency between the application of a precipitating stimulus and the onset of pain was noted. An occasional inability of a given precipitating maneuver to activate the pain was also noted. The temporal pattern of pain was partly characterized by the typical, "plateau-like" pattern, but with ultrashort exacerbations. In part, the pain attacks were characterized by steep or spike-like pain waves, that did not quite subside to the baseline ("repetitive pattern"). Autonomic signs such as lacrimation and conjunctival injection were rather marked, as is generally the case in SUNCT syndrome. Rhinorrhea was present in both patients, and in one patient the rhinorrhea was unusually marked. Carbamazepine treatment seemingly brought about a slight decrease in the frequency of attacks.

Pareja, J. A., J. Pareja, et al. (1994). "SUNCT syndrome: repetitive and overlapping attacks." *Headache* 34(2): 114-6.

A 79-year-old patient had been suffering from right orbital pain attacks since he was 77. The pain, of an "electrical" or burning character, was moderate to severe in intensity. Single attacks lasted for 30-60 seconds each and were accompanied by prominent ipsilateral conjunctival injection and tearing as well as slight rhinorrhoea. The temporal pattern of attacks was irregular with spontaneous remissions alternating with unpredictable symptomatic phases. During active periods, attacks could be triggered by several precipitants acting on trigeminal and extra-trigeminal innervated areas. On most occasions, the patient had 2-3 attacks daily but, at worst, he might have almost continuous short-lasting attacks in periods of 1 to 3 hours length. When attacks had such frequency they presented either as repetitive or overlapping single episodes. According to the clinical features it is concluded that the case reported is a new case of SUNCT syndrome.

Pareja, J. A., P. Kruszewski, et al. (1995). "SUNCT syndrome: trials of drugs and anesthetic blockades." *Headache* 35(3): 138-42.

Nine patients with the SUNCT syndrome (Spanish and Norwegian patients) have, over many years, been given several drugs effective in the cluster headache syndrome, trigeminal neuralgia, and other headaches, as well as drugs not previously used in headache. Various cranial nerves were also anesthetized in an endeavor to ameliorate the suffering of those patients. Although a partial effect was obtained with carbamazepine and corticosteroids in some patients, none of the drugs or anesthetic blockades had consistent, lasting, complete effect on headache paroxysms in SUNCT. The essentially negative outcome of this study aids in further characterizing SUNCT as a separate disorder, and, above all, in distinguishing it from trigeminal neuralgia and the cluster headache syndrome.

Pareja, J. A., V. Caballero, et al. (1996). "SUNCT syndrome. Statuslike pattern." *Headache* 36(10): 622-4.

Four SUNCT patients with a clinical "SUNCT status" witnessed by the investigators are reported. Such an extreme aggregation of typical attacks lasting for the better part of the day, 1 to 3 days in a row has not previously been observed. Appropriate supplementary examinations ruled out an intracranial lesion in all four patients. Indeed, an observation period of more than 5 years speaks against a symptomatic etiology of SUNCT in these cases. Although rarely seen, clinical status may be a feature of the SUNCT syndrome.

Pareja, J. A., J. M. Shen, et al. (1996). "SUNCT syndrome: duration, frequency, and temporal distribution of attacks." *Headache* 36(3): 161-5.

Duration, frequency, and temporal distribution of attacks have been objectively estimated in 11 SUNCT patients (3 women and 8 men). The mean age at the time of the study was 69 years (range 52 to 81). The duration of a total of 348 attacks was measured from videotape records, polygraphic tracings, or by stopwatch. The duration of attacks ranged from 5 to 250 seconds, with an unweighted mean of 61 seconds. Both frequency and exact timing of attacks were assessed in four patients who filled in a time chart with the exact onset of 585 consecutive attacks. The majority of attacks occurred during daytime, with a bimodal distribution; ie, morning and afternoon/evening peaks, and only a few attacks were noted at night (ie, 1.2% of the attacks). The unweighted mean frequency of attacks was 28 per day (range 6 to 77). Duration and timing of attacks in SUNCT syndrome may be of help in the differential diagnosis versus other disorders with the same localization, especially first division trigeminal neuralgia.

Pareja, J. A., J. Joubert, et al. (1996). "SUNCT syndrome. Atypical temporal patterns." *Headache* 36(2): 108-110.

New clinical features of the SUNCT syndrome are described in a series of 3 men (mean age of 65 years, range 56 to 80). The mean age at the onset of symptoms was 55 years (range 39 to 77). Although in all patients the great majority of attacks were typical, on a few occasions unusual features of the painful attacks were either reported or witnessed by the investigators. We have classified these clinical phenomena as (1) low-grade background pain or discomfort, and (2) relatively long-lasting attacks. Neither neurological examination nor neuroimaging studies revealed structural lesions as responsible for the atypical features. The fact that these atypical attacks occurred in the usual symptomatic area and were accompanied by the usual ipsilateral autonomic signs, suggests that they are an integral part of the clinical picture of SUNCT. The possibility that another, concurrent headache was responsible for the unusual attacks is considered unlikely. These clinical phenomena should, accordingly, provisionally be considered as additional but rare clinical features of the SUNCT syndrome.

Pareja, J. A., P. Kruszewski, et al. (1997). "SUNCT syndrome. Diagnosis morbi. Shortlasting Unilateral Neuralgiform headache attacks, with Conjunctival injection, Tearing and rhinorrhoea." *Neurologia* 12 Suppl 5: 66-72.

The present communication represents an updating of the clinical features of SUNCT. The characteristics of SUNCT have been weighed against other orbital/periorbital headache syndromes towards which differential diagnosis is mandatory. In this group, there are various headaches: cluster headache syndrome, first branch trigeminal neuralgia, and idiopathic stabbing headache (jabs and jolts syndrome). SUNCT emerges as a fairly clear-cut, clinical syndrome. A set of diagnostic criteria of SUNCT syndrome is also given.

Pareja, J. A. and O. Sjaastad (1997). "SUNCT syndrome. A clinical review." *Headache* 37(4): 195-202.

The clinical features of SUNCT syndrome have been reviewed in 21 patients. There were 17 men and 4 women, rendering a clear male preponderance (ratio of 4.25). The mean age at onset was around 51 years. Attacks were experienced mostly in the orbital/periorbital area and always recurred on the same side, with an erratic temporal pattern and remissions of varying lengths. Most attacks were moderate to severe in intensity and burning, electrical, or stabbing in character. The attacks were regularly accompanied by prominent, ipsilateral, conjunctival injection; tearing; and rhinorrhea or nasal obstruction. There were many precipitating mechanisms. Exclusively spontaneous attacks were described in 3 patients. The usual duration of paroxysms ranged from 10 to 60 seconds, whereas the longest duration varied from 60 to 300 seconds. The frequency of attacks during the symptomatic periods varied from less than 1 attack daily to more than 30 per hour. In the majority of patients, supplementary examinations failed to show any notable abnormality. However, 2 patients were documented to have a symptomatic form of SUNCT, with a vascular malformation in the ipsilateral cerebellopontine angle. A variety of drugs and local anesthetic blockades, inclusive of tic douloureux drugs, were tried, but a persistent, convincingly beneficial effect was generally lacking. SUNCT syndrome is in the differential diagnosis when encountering unilateral, orbital/periorbital headache syndromes.

Pareja, J. A., P. Kruszewski, et al. (1999). "SUNCT syndrome versus idiopathic stabbing headache (jabs and jolts syndrome)." *Cephalalgia* 19 Suppl 25: 46-8.

Both SUNCT syndrome and idiopathic stabbing headache (ISH) (jabs and jolts syndrome) have to be considered when encountering shortlasting headaches. Since there are no specific tests for these headaches, the differential diagnosis depends entirely upon assessment of the clinical features. These headaches are generally easily distinguishable clinically. There seem to be symptomatic forms of SUNCT.

Pareja, J. A., A. B. Caminero, et al. (2002). "SUNCT Syndrome: diagnosis and treatment." *CNS Drugs* 16(6): 373-83.

Short-lasting Unilateral Neuralgiform headache attacks with Conjunctival injection and Tearing (SUNCT) is a syndrome predominant in males, with a mean age of onset around 50 years. The attacks are strictly unilateral, generally with the pain persistently confined to the ocular/periorbital area. Most attacks are moderate to severe in intensity and burning, stabbing or electrical in character. The mean duration of paroxysms is 1 minute, with a usual range of 10 to 120 seconds (total range 5 to 250 seconds). Prominent, ipsilateral conjunctival injection and lacrimation regularly accompany the attacks. Nasal stuffiness/rhinorrhoea are frequently noted. In addition, there is subclinical forehead sweating. During attacks, there is increased intraocular pressure on the symptomatic side and swelling of the eyelids. No changes in pupil diameter have been observed. Attacks can be triggered mostly from trigeminally innervated areas, but also from the extratrigeminal territory. There are also spontaneous attacks. An irregular temporal pattern is the rule, with symptomatic periods alternating with remissions in an unpredictable fashion. During active periods, the frequency of attacks may vary from <1 attack/day to >30 attacks/hour. The attacks predominate during the daytime, nocturnal attacks being seldom reported. A SUNCT-like picture has been described in some patients with either intra-axial or extra-axial posterior fossa lesions, mostly vascular disturbances/malformations. In the vast majority of patients, however, aetiology and pathogenesis are unknown. In SUNCT syndrome, there is a lack of persistent, convincingly beneficial effect of drugs or anaesthetic blockades that are generally effective in cluster headache, chronic paroxysmal hemicrania, trigeminal neuralgia, idiopathic stabbing headache ('jabs and jolts syndrome'), and other headaches more faintly resembling SUNCT syndrome. Single reports have claimed that carbamazepine, lamotrigine, gabapentin, corticosteroids or surgical procedures may be of help. However, caution is recommended when assessing any therapy in a disorder such as SUNCT syndrome, in which the rather chaotic and unpredictable temporal pattern makes the assessment of any drug/therapeutic effect

per se a particularly difficult matter.

Pareja, J. A. and M. L. Cuadrado (2005). "SUNCT syndrome: an update." *Expert Opin Pharmacother* 6(4): 591-9.

SUNCT (Shortlasting Unilateral Neuralgiform Headache attacks with Conjunctival injection and Tearing) is a syndrome characterised by shortlived (5-240 s), strictly unilateral, orbital/periorbital, moderate-to-severe pain attacks, accompanied by rapidly developing conjunctival injection and lacrimation. Most attacks are triggered by mechanical stimuli, but there are also spontaneous attacks. Symptomatic periods alternate with remissions in an unpredictable fashion. In active periods, the attacks predominate during daytime, with a frequency that ranges from < 1 attack/day to > 30 attacks/h SUNCT is mainly a primary disorder, but is sometimes associated with intracranial structural lesions (symptomatic SUNCT). SUNCT has been included in the group of trigeminal autonomic cephalalgias, which are thought to depend on the activation of the trigeminal system together with the disinhibition of a trigeminofacial autonomic reflex. According to a few reports, SUNCT patients may benefit from carbamazepine, lamotrigine, gabapentin, topiramate or various surgical procedures. However, well-designed clinical trials are required before these therapeutic options can be sufficiently validated.

Penart, A., M. Firth, et al. (2001). "Short-lasting unilateral neuralgiform headache with conjunctival injection and tearing (SUNCT) following presumed dorsolateral brainstem infarction." *Cephalalgia* 21(3): 236-9.

Peraire, M. (1997). "[Diagnosis and treatment of the patient with trigeminal neuralgia]." *Neurologia* 12(1): 12-22.

Neuralgia of the trigeminus (NT) is the most common of cranial nerve neuralgias. Its diagnosis is entirely clinical and its most common form of presentation is well understood. Questions of differential diagnosis can emerge with certain entities such as atypical trigeminal neuralgia, short-duration unilateral neuralgiform cephalgia of the trigeminus (SUNCT) arising from injection to the conjunctival, lacrimal or other glands, cluster headache, chronic paroxysmal hemicrania, pain arising in the teeth and myofascial pain syndrome. The three main causative factors of NT are compression of the nerve root by an artery in the prepontine space, thereby creating an area of demyelination, compression of the nerve by a tumor, and multiple sclerosis. The first is the most common of the three. NT can be classified as essential in 10 to 30% of patients. Recent advances in magnetic resonance (MR), and its advantages over other imaging systems, have made MR the diagnostic method of choice. The first treatment is medical and the basic drugs involved can be considered classic. Other therapies have been suggested in recent years, however, and should probably be studied further. Two substances stand out among those proposed: tocainide, an antiarrhythmic drug, and pimozide, an antipsychotic. Surgical treatment of NT can address either the cause (tumor or vascular compression) or symptoms, the latter being indicated when medical treatment fails. Surgery can be performed on peripheral nerves, on the gasserian ganglion and on the posterior fossa. The indications, outcomes and possible complications are quite different for each approach, making choice controversial.

Piovesan, E. J., C. Siow, et al. (2003). "Influence of lamotrigine over the SUNCT syndrome: one patient follow-up for two years." *Arq Neuropsiquiatr* 61(3A): 691-4.

The SUNCT syndrome is characterized by a short-lasting headache in the first division of the trigeminal nerve, associated with ipsilateral autonomic symptoms. It is highly refractory to prophylactic medication. We describe a case where lamotrigine reduced the intensity, duration, and frequency of attacks and increased the remission period of this disorder. Over a two-year period, the attacks came back immediately whenever the patient reduced the dose or neglected treatment. We concluded that lamotrigine is effective in treating SUNCT syndrome when used in high doses for a prolonged period of time.

Porta-Etessam, J., J. Benito-Leon, et al. (2002). "Gabapentin in the treatment of SUNCT syndrome." *Headache* 42(6): 523-4.

Porta-Etessam, J., A. Martinez-Salio, et al. (2002). "Gabapentin (neuronetin) in the treatment of SUNCT syndrome." *Cephalalgia* 22(3): 249; author reply 249-50.

Poughias, L. and J. Aasly (1995). "SUNCT syndrome: cerebral SPECT images during attacks." *Headache* 35(3): 143-5.

Two patients with SUNCT syndrome (short-lasting, unilateral, neuralgiform headache attacks with

conjunctival injection and tearing) were investigated. Blood flow velocity in the middle cerebral artery was monitored before, during, and outside four spontaneous attacks. An interhemispheric asymmetry was observed. In the second case, velocity decreased significantly on both sides during attacks in comparison with preattack values. Cerebral SPECT (single photon emission computed tomography) images were obtained during a bout and between attacks in one patient. The radiocompound was injected 5 to 10 seconds after the start of an attack. In both patients, normal tracer uptake and symmetric perfusion was observed during headache periods.

Prakash, K. M. and Y. L. Lo (2004). "SUNCT syndrome in association with persistent Horner syndrome in a Chinese patient." *Headache* 44(3): 256-8.

This is the first case report of a Chinese patient with SUNCT (shortlasting, unilateral, neuralgiform headache attacks with conjunctival injection and tearing) presenting with persistent Horner's syndrome. She had episodic, brief, right periorbital pain in association with ipsilateral eye injection, lacrimation and rhinorrhea as well as persistent ipsilateral miosis and ptosis. She had partial response to a combination of indomethacin and carbamazepine therapy.

Putzki, N., A. Nirkko, et al. (2005). "Trigeminal autonomic cephalalgias: a case of post-traumatic SUNCT syndrome?" *Cephalalgia* 25(5): 395-7.

Raimondi, E. and L. Gardella (1998). "SUNCT syndrome. Two cases in Argentina." *Headache* 38(5): 369-71. Two patients suffering from SUNCT syndrome are presented. Some features are remarkable. The first patient was a 69-year-old man whose first crisis was located in the right supraorbital region. After a 4-month spontaneous remission, the pain returned to the upper part of the cheek, radiating to the supraciliary region on the same side, with lacrimation and conjunctival injection. Rhinorrhea was absent. The painful attacks were triggered by head movements. Clinical improvement occurred with carbamazepine treatment. The second patient was a 48-year-old woman whose painful attacks lasted from 30 to 45 seconds followed by a burning sensation lasting 2 hours. Autonomic signs such as conjunctival injection, lacrimation, and edema and ipsilateral ptosis of the upper lid were rather marked. There was never any rhinorrhea. Her attacks were triggered by head and eye movements. She responded to the administration of corticosteroids and carbamazepine. According to these features, the two patients had SUNCT syndrome, and the positive carbamazepine response suggests a relationship with trigeminal neuralgia.

Ramirez-Moreno, J. M., I. Fernandez-Portales, et al. (2004). "[SUNCT syndrome and neoplasms in the central nervous system. A new association]." *Neurologia* 19(6): 326-30. Short-lasting unilateral neuralgiform headache attacks with conjunctival injection and tearing (SUNCT) syndrome is a rare form of primary headache disorder, although secondary causes are well known. A growing number of cases have been described since its description in 1978 up to now. We report a new case in a 72 year old woman suffering SUNCT syndrome secondary to two ipsilateral intracranial lesions in the basal portion of the left frontal lobe of the sphenoid wing and the left pontocerebellar angle, suggestive of meningiomas. The patient was successfully treated with carbamazepine. We discuss the differential diagnosis with other very brief headaches associated with cranial autonomic features and the therapeutics possibilities. We review the secondary cases previously reported. The possible pathophysiological mechanism in this case is discussed.

Rossi, P., F. Cesarino, et al. (2003). "SUNCT syndrome successfully treated with topiramate: case reports." *Cephalalgia* 23(10): 998-1000.

Rozen, T. D. (2004). "Short-lasting headache syndromes and treatment options." *Curr Pain Headache Rep* 8(4): 268-73.

A number of primary headache syndromes are marked by their short duration of pain. Many of these syndromes have their own unique treatment, so they must be recognized by practicing physicians. In this article, a number of the short-lasting headache disorders are reviewed, including chronic paroxysmal hemicrania, SUNCT syndrome, hypnic headache, exploding head syndrome, primary stabbing headache, and cough headache.

Rozen, T. D., G. V. Haynes, et al. (2005). "Abrupt onset and termination of cutaneous allodynia (central sensitization) during attacks of SUNCT." *Headache* 45(2): 153-5.

The presence of central sensitization and cutaneous allodynia has not been readily studied in other primary headache syndromes outside of migraine. If central sensitization does occur, is the temporal profile any different in the short-lasting more aggressive syndromes such as SUNCT than in migraine? A patient with SUNCT was examined during and in between attacks looking for the

presence and duration of cutaneous allodynia.

Sabatowski, R., M. Huber, et al. (2001). "SUNCT syndrome: a treatment option with local opioid blockade of the superior cervical ganglion? A case report." *Cephalalgia* 21(2): 154-6.

Santos-Garcia, D., J. M. Prieto, et al. (2005). "[SUNCT syndrome secondary to megadolichobasilar anomaly]." *Rev Neurol* 41(10): 638-9.

Schwaag, S., A. Frese, et al. (2003). "SUNCT syndrome: the first German case series." *Cephalalgia* 23(5): 398-400.

Sekhara, T., K. Pelc, et al. (2005). "Pediatric SUNCT Syndrome." *Pediatr Neurol* 33(3): 206-7.
This report describes a 5-year-old male with sudden unilateral headache attacks (2-50 seconds) accompanied by conjunctival injection, lacrimation, and nasal congestion. The episodes occurred without a precipitating factor, never during sleep. Brain imaging was normal. The attacks resolved spontaneously within 5 months. This headache syndrome (short-lasting unilateral neuralgiform headache attacks with conjunctival injection and tearing) was previously described in two other children aged 10 and 11.

Selekler, M. and M. Alemdar (2004). "[Trigeminal autonomic cephalgias: diagnosis, therapy, atypical forms and pathophysiology]." *Agri* 16(4): 17-27.
Trigeminal autonomic cephalgias; cluster headache, paroxysmal hemicrania, hemicrania continua and SUNCT (short-lasting unilateral neuralgiform headache attacks with conjunctival injection and tearing) syndrome are characterized by unilateral trigeminal distribution of pain and accompanying ipsilateral autonomic symptoms. Other than cluster headache, all of these syndromes have been described within last twenty years, and are found relatively less frequent and less known forms. Diagnosis of paroxysmal hemicrania and hemicrania continua, especially atypical forms, essentially depends on indomethacin responsiveness. For SUNCT syndrome, there is not such a drug which provides a practical approach to both diagnosis and therapy and its diagnosis depends on clinical features. So far, case reports from various countries helped us realize the existence of secondary forms of these syndromes and the necessity of imaging techniques, especially for recently described autonomic cephalgias.

Sesso, R. M. (2001). "SUNCT syndrome or trigeminal neuralgia with lacrimation and conjunctival injection?" *Cephalalgia* 21(2): 151-3.

Shen, J. M. and H. J. Johnsen (1994). "SUNCT syndrome: estimation of cerebral blood flow velocity with transcranial Doppler ultrasonography." *Headache* 34(1): 25-31.
Four patients with SUNCT syndrome (Short-lasting, Unilateral, Neuralgiform headache attacks with Conjunctival injection and Tearing) were investigated with Doppler ultrasonography. Blood flow velocity (V) was measured in all intracranial arteries during both normocapnia and voluntary hyperventilation in 4 patients outside attacks (2 during remission; 2 during bout, but outside attacks) and in 8 healthy individuals. Vasomotor reactivity (VMR) was calculated on the basis of the formula of percentage change in V divided by the reduction in end-tidal PCO₂ (PETCO₂). Under the basal condition, the patients had a slightly, but non-significantly higher V in the middle cerebral artery (MCA) ($P > 0.1$) and lower V in the basilar artery ($P > 0.05$) than controls. During hyperventilation, a significant reduction in V was observed in the anterior and posterior cerebral arteries, at a level 1.5-2 SD above that in controls ($P < 0.05$), but a non-significant difference in VMR in comparison with controls. VMCA was continuously insonated during spontaneous (n = 8) and precipitated (n = 4) attacks in one particular patient on different days. Prior to attack, VMCA was significantly lower on the symptomatic side than on the non-symptomatic side ($P < 0.014$). VMCA decreased significantly during spontaneous attacks on both sides ($P < 0.01$) in comparison with the pre-attack stage, and returned to baseline before the cessation of attack. Similar findings were made during precipitated attacks. PETCO₂ was rather constant throughout the entire attack study. Our data suggest that abnormal cerebral circulation may be part of the SUNCT syndrome. The vascular changes may have underlying mechanisms differing from those of the pain.

Sjaastad, O., J. M. Zhao, et al. (1991). "Short-lasting unilateral neuralgiform headache attacks with conjunctival injection, tearing, etc. (SUNCT): III. Another Norwegian case." *Headache* 31(3): 175-7.
"SUNCT" is a recently reported head pain syndrome characterized by shortlasting, unilateral neuralgiform paroxysms with conjunctival injection and tearing, and to a lesser extent nasal secretion, and (subclinical) sweating. Another case--that of a 56 year old male--is reported herein.

The attacks lasted 1/2-1 min. and occurred only infrequently, i.e. once or twice in 1-4 weeks, except for a week recently when there were up to 20 or more typical attacks per day, with the usual ipsilateral, autonomic accompaniments. Due to the benign nature and low frequency of attacks, the diagnosis would have been most difficult to establish prior to the occurrence of this shortlasting period with more marked symptoms. A variety of precipitation mechanisms were present, partly concerning the V 2-3 areas, partly concerning the neck. Precipitation mechanisms in "SUNCT" to some extent seem to differ from those in trigeminal neuralgia. It is remarkable that all four hitherto reported cases are males.

Sjaastad, O., P. Kruszewski, et al. (1992). "SUNCT syndrome: VII. Ocular and related variables." *Headache* 32(10): 489-95.

SUNCT is a unilateral headache syndrome with shortlasting attacks, accompanied by e.g. conjunctival injection and lacrimation on the painful side. Intraocular pressure (IOP), corneal indentation pulse (CIP) amplitudes, episcleral venous pressure, and corneal, tympanic, and facial temperature have been studied in 6 SUNCT patients. IOP and CIP amplitudes increased on the painful side during headache paroxysms, while episcleral venous pressure remained unchanged. Corneal temperature seemed to increase during attack on both sides. However, the number of observations during attacks is scanty. Outside of attacks, the corneal temperature on the symptomatic side seemed to be higher when compared with the non-symptomatic side (generally $> \text{ or } = 0.5$ degrees C), provided that the attack frequency was high. The facial temperature seemed to be even on both sides or slightly higher on the symptomatic than on the non-symptomatic side in the periocular area. This pattern seems to be different from the one in trigeminal neuralgia, in which the temperature has been reported to be lowest on the painful side of the face. During attacks, there seemed to be a tendency for the temperature to increase in the periocular area, but not over the mandible or in the neck. The results obtained could be caused by increased blood supply to the eye (and the surrounding skin) on the symptomatic side because of vasodilatation during repeated pain attacks. As far as the ocular changes are concerned, probably the arteriolar side of the vascular bed is involved.

Sjaastad, O. and P. Kruszewski (1992). "Trigeminal neuralgia and "SUNCT" syndrome: similarities and differences in the clinical pictures. An overview." *Funct Neurol* 7(2): 103-7.

SUNCT is a recently described unilateral headache with frequently occurring, shortlasting pain attacks in the ocular area accompanied by ipsilateral conjunctival injection, lacrimation, and (subclinical) forehead sweating. In some patients, attacks may be triggered by cutaneous stimuli. In this communication, SUNCT patients (n = 5) are compared with the considerable clinical series of trigeminal neuralgia in the literature (e.g. Harris, 1940, 1433 cases). In several respects (unilaterality, triggering, brevity and frequency of paroxysms), SUNCT shows similarity to trigeminal neuralgia. SUNCT seems to differ clearly from trigeminal neuralgia in other respects: sex distribution (SUNCT patients are often males), pain localization (SUNCT patients have the pain in the ocular area), the carbamazepine effect, presence of conjunctival injection, lacrimation, etc. SUNCT may accordingly altogether seem to be distinct from trigeminal neuralgia.

Sjaastad, O., J. A. Pareja, et al. (1997). "Trigeminal neuralgia. Clinical manifestations of first division involvement." *Headache* 37(6): 346-57.

A series of 19 patients with what originally had been diagnosed as a first division (V1) trigeminal neuralgia was collected. The inclusion criteria were severe, rather short-lasting pain attacks within the V1 area, combined with trigger mechanisms. There were 10 women and 9 men, and the mean age of onset was 57.8 years. Fifteen of 16 with adequate information on attack duration had paroxysms of a "few seconds" duration or less, whereas 10 patients had paroxysms lasting $< \text{ or } = 2$ seconds. In an exceptional case, only "more long-lasting" attacks (greater than 30 seconds' duration) were experienced. In regard to autonomic phenomena, lacrimation was most frequently present (in a total of 8 patients; 3 rather regularly, 5 more irregularly). The combination of lacrimation, conjunctival injection, and rhinorrhea was present in only 2 (of 19), and in neither of them in a major way. Typically, autonomic phenomena occurred during the later stages of disease and during particularly severe and long-lasting attacks. Seven of 14 with adequate information also had nocturnal attacks. Initially, a more or less complete carbamazepine effect was reported by 10 of 13 patients. Precipitation mechanisms were the same as with second and third division tic, but were mainly located within the V1 area, particularly initially. A comparison with SUNCT syndrome has been made. SUNCT is a predominantly male disorder, with only exceptional attacks of $< \text{ or } = 10$ seconds' duration, and generally with attacks of 15 seconds or longer. Autonomic symptoms and signs are more pronounced than in V1 tic. Carbamazepine generally provides minor, if any, benefit in SUNCT. The present work strongly indicates that the two disorders are essentially different.

- Sprenger, T., M. Valet, et al. (2005). "SUNCT: bilateral hypothalamic activation during headache attacks and resolving of symptoms after trigeminal decompression." *Pain* 113(3): 422-6.
 Short-lasting unilateral neuralgiform headache with conjunctival injection and tearing (SUNCT) is a primary head-pain syndrome, which is often refractory to any medical treatment. Concerning the pathophysiology of SUNCT, hypothalamic involvement ipsilaterally to the pain has been suggested based on the clinical features and one functional imaging case report. Here we now report a new case with SUNCT and the concomitant cerebral activation pattern (fMRI) during the pain attacks. In addition to an activation of several brain structures known to be generally involved in pain processing, bilateral hypothalamic activation occurred during the pain attacks, arguing for a central origin of the headache. Interestingly, this patient became completely pain free after surgical decompression of the ipsilateral trigeminal nerve. We hypothesize that in this case with a central predisposition for trigeminal autonomic cephalgias, a peripheral trigger with ectopic excitation might have contributed to the clinical picture of SUNCT.
- ter Berg, J. W. and P. J. Goadsby (2001). "Significance of atypical presentation of symptomatic SUNCT: a case report." *J Neurol Neurosurg Psychiatry* 70(2): 244-6.
 A patient with a SUNCT-like syndrome caused by severe basilar impression in association with osteogenesis imperfecta is described. Initially symptoms of both the first and second branch of the trigeminal nerve were prominent, on which carbamazepine had only a temporary and mild effect. Progressive symptoms with prominent ipsilateral autonomic features, unexplained triggering by photostimulation, and increasing duration of pain attacks occurred with relentless progressive basilar impression associated with pontomedullary compression. Pathophysiologically a dysfunction in ephaptic transmission is hypothesised.
- Tilignac, C. and G. Dordain (2001). "[SUNCT syndrome sensitive to lamotrigine]." *Presse Med* 30(22): 1106.
- Trucco, M., F. Mainardi, et al. (2004). "Chronic paroxysmal hemicrania, hemicrania continua and SUNCT syndrome in association with other pathologies: a review." *Cephalalgia* 24(3): 173-84.
 We present a review of 22 cases of headache mimicking chronic paroxysmal hemicrania (CPH) (17 female and five male; F : M ratio 3.4), nine cases mimicking hemicrania continua (HC) (seven female and two male) and seven cases mimicking SUNCT syndrome (five male and two female) found in association with other pathologies published from 1980 up to the present. All case reports were discussed with respect to diagnostic criteria proposed by International Headache Society (IHS) for CPH, by Goadsby and Lipton for HC and SUNCT, and evaluated to identify a possible causal relationship between the pathology and the onset of headache. The aim of the present review was to evaluate if the presence of associated lesions and their location could help elucidate the pathogenesis of trigeminal autonomic cephalgias (TACs).
- Valade, D. and M. El Amrani (2000). "[Idiopathic facial pain other than vascular pain]." *Rev Neurol (Paris)* 156 Suppl 4: 4S57-61.
 Cluster Headache associates a severe pain generally unilateral and autonomic symptoms with a remarkable periodicity. In the first part we tried to explain the conception of physiopathology of these short lasting headache syndromes and in the second part we described the clinical features. The short lasting primary headaches are divided into two groups: those with marked autonomic activation which comprise chronic and episodic paroxysmal hemicrania, short lasting unilateral neuralgiform headache with conjunctival injection and tearing (SUNCT syndrome) and cluster headache. The second group includes two entities, one without autonomic activation: Hypnic Headache and one with mild autonomic features: Hemicrania Continua. The paroxysmal hemicranias are characterized by attack frequency ranges from 15 to 20 per day of short lasting attacks of unilateral pain that typically last 2 to 10 minutes, the severe pain is associated with autonomic symptoms and responds to treatment with indomethacin. The SUNCT syndrome has a less severe pain but marked autonomic activation during attacks, this syndrome is actually resistant to proposed therapy. The Hypnic Headache and the Hemicrania Continua have yet less severe pain with very mild or without autonomic features.
- van Vliet, J. A., M. D. Ferrari, et al. (2003). "SUNCT syndrome resolving after contralateral hemispheric ischaemic stroke." *Cephalalgia* 23(3): 235-7.
- Vikelis, M., M. Xifaras, et al. (2005). "SUNCT syndrome in the elderly." *Cephalalgia* 25(11): 1091-2.
- Vincent, M. (1998). "SUNCT, lacrimation, and trigeminal neuralgia." *Cephalalgia* 18(2): 71.

Wober, C., C. Wober-Bingol, et al. (1993). "[SUNCT syndrome--case report and review of the literature]." *Fortschr Neurol Psychiatr* 61(11): 378-82.

The SUNCT syndrome is a recently described headache disorder characterised by shortlasting, unilateral headache attacks with conjunctival injection and tearing. Our report presents a further case and compares the findings in our patient with those in the cases described earlier. In addition, we review the literature and discuss possible aetiological and pathogenetic factors as well as the differential diagnosis of SUNCT syndrome, trigeminal neuralgia and cluster headache syndromes.

Zhao, J. M. and O. Sjaastad (1993). "SUNCT syndrome: VIII. Pupillary reaction and corneal sensitivity." *Funct Neurol* 8(6): 409-14.

SUNCT is a newly described headache characterized by shortlasting, unilateral attacks of a neuralgiform character, accompanied by conjunctival injection and tearing on the symptomatic side. At present, 8 patients have been observed, all males. In the present work, vertical pupillary diameters have been estimated by the binocular, Whittaker infrared pupillometer (Gulf & Western). Measurements have been carried out both during the basal state and after topical, pharmacologic stimulation (by sympathicomimetic agents, i.e. tyramine (2%), OH-amphetamine (1%) and phenylephrine (1%). In the basal state, there was no clear tendency to anisocoria. After sympathicomimetic drugs, there was a tendency to underreaction on the symptomatic side, mainly as regards the indirectly acting ones (OH-amphetamine and tyramine). These two agents should supposedly give similar results, but, nevertheless, the results partly deviated. Phenylephrine did not invariably normalize the symptomatic/non-symptomatic side ratio. Corneal sensitivity was investigated with the Cochet-Bonnet esthesiometer. Two of 6 patients studied outside attack showed moderately lowered values on the symptomatic side. During attack, there seemed to be reduced corneal sensitivity in both patients investigated, in one bilaterally, in the other, on the symptomatic side only.

Zidverc-Trajkovic, J., M. Mijajlovic, et al. (2005). "Vertebral artery vascular loop in SUNCT and concomitant trigeminal neuralgia. Case report." *Cephalgia* 25(7): 554-7.