Curr Pain Headache Rep (2013) 17:332
DOI 10.1007/s11916-013-0332-0

Role of Trigeminal Microvascular Decompression in the Treatment of SUNCT and SUNA

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Published online: 6 April 2013

Abstract Short-lasting unilateral neuralgiform headache with conjunctival injection and tearing (SUNCT) and short-lasting unilateral neuralgiform headache attacks with cranial autonomic symptoms (SUNA) are primary headache disorders. Evidence suggests that SUNCT/SUNA have similar pathophysiology to the trigeminal autonomic cephalalgias and involves the trigeminal autonomic reflex. This review provides an overview of microvascular decompression of the trigeminal nerve and other surgical therapeutic options for SUNCT/SUNA. We have undertaken a mini-meta-analysis of available case reports and case series with the aim of providing recommendations for the use of such therapies in SUNCT/SUNA. There is some evidence supporting microvascular decompression of the trigeminal nerve in selected patients who have medically refractory SUNCT/SUNA and a demonstrable ipsilateral aberrant vessel on magnetic resonance imaging (MRI). We also consider what further investigations could be undertaken to assess the role of surgical interventions in the treatment of these often debilitating conditions.

Keywords SUNCT Syndrome · SUNA Syndrome · Surgery · Trigeminal Nerve · Treatment · Microvascular Decompression

This article is part of the Topical Collection on *Trigeminal Autonomic Cephalalgias*

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Introduction

Short-lasting unilateral neuralgiform headache with conjunctival injection and tearing (SUNCT) [1•] and short-lasting unilateral neuralgiform headache attacks with cranial autonomic symptoms (SUNA) [2] are primary headache disorders characterised by brief, severe, ipsilateral, orbitotemporal pain with autonomic features [3]. Patients with SUNCT describe short episodes of severe stabbing pains predominantly in the distribution of the first division of the trigeminal nerve [4••]. Attacks generally last between 5 and 240 seconds, with an attack frequency of 3 to 200 episodes in 24 hours and are associated with significant ipsilateral autonomic features such as conjunctival injection and tearing [4••]. In SUNA the range of attack duration is wider (2 seconds to 10 minutes) and the range of associated ipsilateral autonomic symptoms is also more wide ranging, but generally less severe and includes rhinorrhoea, ptosis and facial flushing in addition to lacrimation and conjunctival injection [5]. SUNCT is frequently debilitating and medical management is often unsatisfactory [6]. The pain associated with this condition can be so severe as to result in suicidal ideation in some cases [7, 8••]. Recently, there has been a renewed interest in the role of surgical interventions and neurostimulation therapy as possible therapeutic modalities in the treatment of SUNCT [8••, 9].

In this article, we review the small number of studies that have examined the role of surgical interventions, including microvascular decompression of the trigeminal nerve in SUNCT/SUNA. The use of surgical interventions has coincided with the development of increasingly more sophisticated neuroimaging, which has helped identify vascular compression of the trigeminal nerve by aberrant arterial and venous loops. The Jannetta procedure involves microvascular decompression of the trigeminal nerve and has been an accepted treatment for intractable trigeminal neuralgia for many years [10]. More recently, there have been encouraging reports

of successful decompression of the trigeminal nerve in cases with SUNCT/SUNA [8••]. However, one concern about neurosurgical interventions has been the potential for both intraoperative and postoperative complications [11].

The use of functional imaging has transformed our understanding of the various mechanisms involved in the pathogenesis of trigeminal autonomic cephalalgia (TAC) [12], which includes SUNCT/SUNA. Functional imaging has shown regions of hypothalamic activation in patients with TAC, which supports the important role of the hypothalamus in the generation of these headaches [13]. Central neuromodulation techniques, including posterior hypothalamic deep brain stimulation (DBS) and occipital nerve stimulation (ONS), have been used for a range of primary headache disorders including migraine and TAC with encouraging results [14, 15]. This further raises the question as to whether neurostimulation could be a therapeutic option for SUNCT/SUNA.

Pathophysiology

The pathophysiology of SUNCT/SUNA is complex and incompletely understood with both central and peripheral models having been proposed. There is increasing support for the view that posterior hypothalamic abnormalities are involved in TAC [7]. The current model for the generation of pain in TAC is summarised in Fig. 1.

Anatomical Considerations

The trigeminocervical complex can be characterised as a functional brainstem unit consisting of an overlap of trigeminal and upper cervical painful receptive fields [16]. The functional connection between the trigeminal nucleus and the superior salivatory nucleus leads to activation of parasympathetic efferents via the greater superficial petrosal nerve. The hypothalamus is thought to regulate the parasympathetic outflow from the superior salivatory nucleus [17]. Anatomical studies in rats have identified the trigeminohypothalamic tract connecting the posterior hypothalamus with the trigeminal caudal nucleus. This tract forms the afferent pathway for transmission of sensory information from the trigeminal cervical complex to the posterior hypothalamus [18]. Stimulation of the posterior hypothalamus in this manner modulates the activity of the trigeminal nucleus via the same tract resulting in heightened sensitivity to various noxious stimuli. Studies have now established that the posterior hypothalamus is a physiological modulator of activity in the trigeminal nucleus caudalis [19].

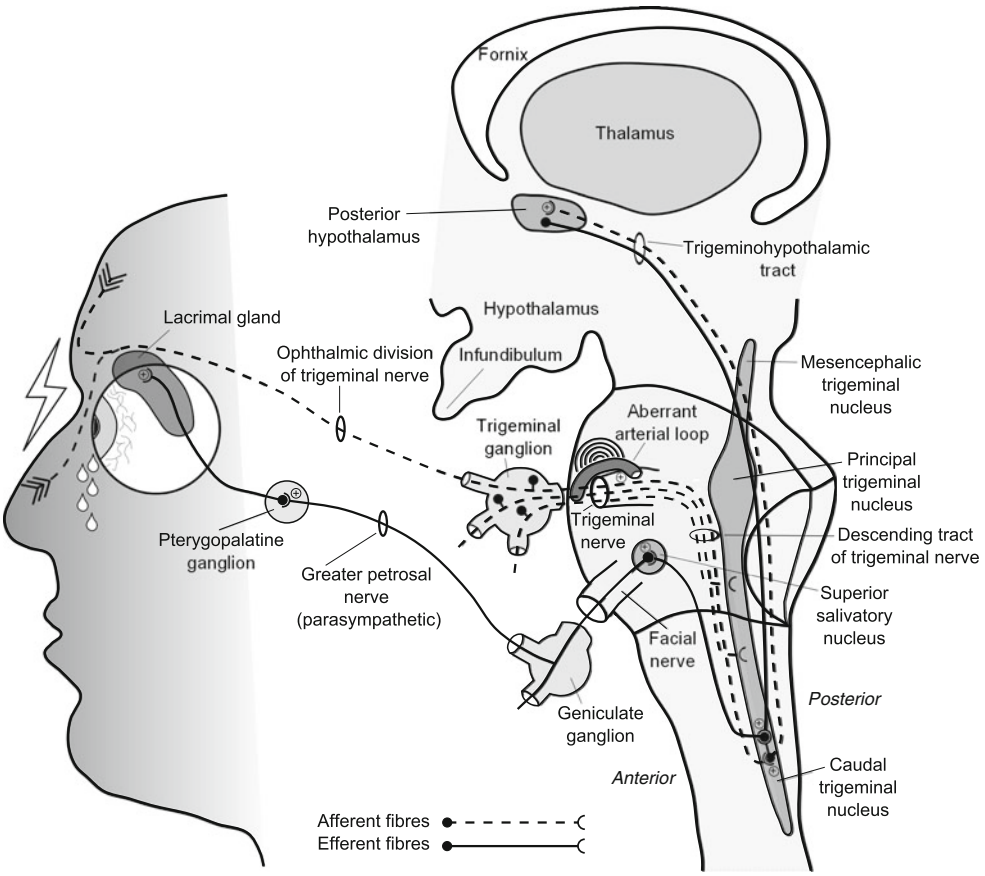

Fig. 1 Schematic representation of the brainstem and diencephalon showing the principal pathways and nuclei involved in a proposed model of pain and autonomic features seen in SUNCT syndrome and other trigeminal autonomic cephalgias. A left lateral view of the brainstem is shown with lateral nuclei together with a paramedian section of the diencephalon

Neuroimaging

Advances in functional and structural neuroimaging have provided new insights into our understanding of TAC. Cluster headaches, paroxysmal hemicrania and SUNCT/SUNA have strikingly similar phenotypic manifestations suggesting a shared pathogenesis [13]. It is generally accepted that theories of vascular changes and neurogenic inflammation alone cannot explain causation of these headaches [13]. Positron emission tomography (PET) studies of experimental head and facial pain models have delineated the widespread brain network involved in nociceptive processing. This network has been referred to as the 'pain neuromatrix' and includes the anterior cingulate cortex, frontal and prefrontal cortices, primary and secondary somatosensory cortices, thalamus, basal ganglia and regions within the temporal and parietal cortices [20].

It has been demonstrated that hypothalamic activation is only seen in patients with cluster headache during an active bout [12, 21]. In a study of experimentally induced cluster headache in 17 patients, activation of the pain neuromatrix structures on PET scans was seen in all. However, only the nine patients in an active bout showed activation of the hypothalamus, reinforcing the theory that the hypothalamus has a permissive or triggering role in the generation of these headaches [12, 21]. A similar phenomenon has been demonstrated in patients with SUNCT using blood oxygen level-dependant functional magnetic resonance imaging (BOLD-fMRI) [22]. It has also been shown that there are posterior hypothalamic changes on BOLD fMRI in nine patients with SUNCT [4••]. Five had bilateral posterior hypothalamic activation: two had contralateral activation and two patients showed ipsilateral deactivation in the same area. Other studies have confirmed the occurrence of the same phenomena in SUNCT [7, 22]. Furthermore, structural neuroimaging studies using voxel-based morphometry have confirmed the involvement of the posterior hypothalamic grey matter in patients with cluster headache. An increase in volume of the posterior hypothalamic grey matter was demonstrated in cluster headache patients as compared to healthy volunteers.

Evidence for a central aetiology has also emerged from a limited number of studies that have investigated the role of neurostimulation in the form of DBS of the posterior hypothalamus in TAC which has resulted in improvement in the number of attacks and severity of pain [23, 24]. This use of DBS in SUNCT/SUNA is discussed further below.

With regards to a peripheral origin of pain in TAC, most of the evidence is drawn from studies on patients with cluster headache. Increased concentrations of calcitonin gene-related peptide (a surrogate marker for trigeminal activation) in the ipsilateral jugular vein have been documented during attacks of cluster headache (CH) and paroxysmal hemicrania (PH) [25]. Observational studies in patients with secondary forms of TAC, where improvement in symptoms after sectioning of the trigeminal nerve or resection of causative lesions has been seen, support the role of a peripheral origin for pain in these cases [7, 26]. Interestingly, SUNCT has been reported in association with a variety of intracranial lesions (so called 'symptomatic SUNCT'), including vascular malformations, basilar impression, leiomyosarcoma of the cavernous sinus, craniosynostosis, prolactinoma, acromegaly and brain stem infarction [27–35]. It is interesting to note that the clinical phenotype of secondary TAC is indistinguishable from the primary forms [36]. Neuroimaging studies with BOLD-fMRI failed to show any changes in the hypothalamus in symptomatic cases of SUNCT. It has been concluded that while the hypothalamus may be the central generator of pain in primary TAC, this may not be true for symptomatic cases [4••].

Aberrant vascular loops in contact with the trigeminal nerve are seen in about 47–90 % of cases of trigeminal neuralgia [37]. Aberrant vessels and anatomical variants have also been described in SUNCT [38, 39]. In one case series in which dedicated fine cut constructive interference steady state (CISS) images of the trigeminal nerves were obtained (Fig. 2a), 15/17 (88 %) patients with SUNCT showed an aberrant arterial loop [40]. Blinded analysis of the symptomatic versus asymptomatic side indicated that 90 % of these aberrant vessels were symptomatic, while only 7 % were asymptomatic [40]. This supports the notion of peripheral compression or disruption of the trigeminal nerve being a potential cause of SUNCT syndrome. However, other series have found a much lower incidence of aberrant vessels (2–8 %) [41, 42], but in one of these series, dedicated views of the trigeminal nerve were not obtained (Matharu, MS; personal communication) and the other was a literature review of cases in whom for the vast majority such images were similarly not obtained. The fact that both viral meningitis [43] and idiopathic pachymeningitis [44] can be associated with SUNCT also points to peripheral stimulation of the trigeminal nerve being a mechanism for pain generation in this syndrome.

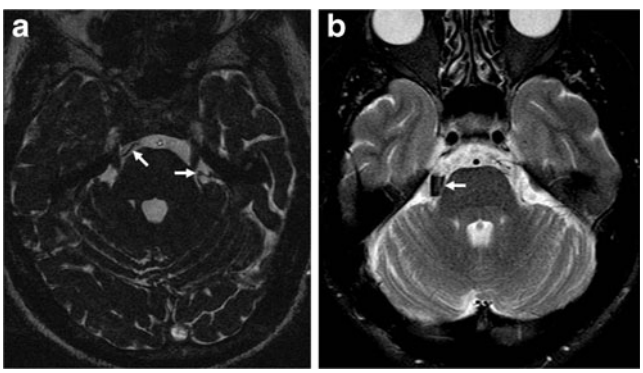

Fig. 2 Axial MRI of the brain stem at the level of the trigeminal nerve. (**a**) CISS image showing bilateral aberrant arterial loops (arrows) preoperatively and (**b**) T2 image showing placement of a silicone sleeve around the right trigeminal nerve (arrow)

It is likely that both central and peripheral mechanisms play a role in the pathophysiology of SUNCT/SUNA, but one may predominate over the other, resulting in differential responses to various surgical treatment modalities.

Surgical Management of SUNCT/SUNA

We undertook a systematic search of PubMed, Medline and Cochrane databases using the search terms: SUNCT; SUNA; trigeminal autonomic cephalalgia and surgery, microvascular decompression; Jannetta procedure; decompression; block; stimulation and secondary. Articles were limited to those with abstracts written in English. Careful review of titles and abstracts of these 481 articles identified 41 potentially relevant articles. Where there was ambiguity, the full article was reviewed. A further ten articles were identified from the bibliographies of all selected articles. As a result of this process, a total of 20 articles reporting either single cases or case series of surgical interventions for SUNCT/SUNA were identified. There were no randomised clinical trials. Other relevant articles have been referenced as necessary.

In the case of microvascular decompression, there were sufficient cases to perform a mini-meta-analysis of case reports and case series. We have also reviewed available case reports for ONS, gamma knife radiosurgery, greater occipital nerve block, superior cervical ganglion block and Gasserian ganglion decompression. We have not reviewed the management of secondary cases of SUNCT (excluding aberrant vascular loops), which has generally been specific to the underlying pathology involved.

Microvascular Decompression of the Trigeminal Nerve

Microvascular decompression of the trigeminal nerve involves releasing arachnoid adhesions from any aberrant vessels in contact with the trigeminal nerve and dorsal root entry zone. The most commonly used technique is the Jannetta procedure [10] with further adaptations utilising a silicone cuff (Fig. 2b) [8••]. In the lateral position with the neck flexed, stereotactic navigation and a retro-sigmoid craniotomy behind the mastoid are used to approach the trigeminal nerve superolateral to the cerebellum.

We identified ten case reports [7, 11, 23, 45–50] and one case series of nine patients [8••] with SUNCT/SUNA who underwent trigeminal microvascular decompression between 1997 and 2011. One further case which has been briefly reported as an addendum only has not been included as no details were provided [51]. Median age was 54 years (range, 28–73 years), which is similar to previous cross sectional series of SUNCT/SUNA [4••, 42]. Fourteen cases (74 %) were male, which is slightly higher than the 1.3:1 male:female ratio seen in other series of SUNCT/SUNA [4••, 40, 42]. Clinical details of the 19 cases are summarised in Table 1. Fourteen cases were SUNCT, three were SUNA and two were a combination of SUNCT and SUNA. There was evidence of an aberrant vessel abutting the ipsilateral trigeminal nerve on neuroimaging in 15/17 (88 %) cases, with details of imaging findings not being reported for two cases. In 13/17 (76 %) cases the aberrant vessel was an ectatic loop of the superior cerebellar artery. In 10/12 (83 %) of cases where the pattern of attacks was noted, the pattern was chronic rather than episodic. It has been noted previously that episodic SUNCT/SUNA is more likely to respond to medical therapies such as lamotrigine [40], and would therefore be less likely to result in consideration for surgical intervention. Patients had trialled a median of 6 (range, 3–16) different medical or surgical therapies prior to microvascular decompression and the median disease duration was 4 years (range, 1 month to 26 years), indicating that these were intractable cases resistant to multiple therapies.

After a median follow-up of 14 months (range 0.5–32 months) 12/19 (63 %) of cases had complete resolution of attacks. In the remainder, surgery appeared to have little effect upon the frequency or severity of attacks although a transient improvement was noted in some. Transient complications were seen in five cases (wound infection, chest infection, vertigo, jaw pain and dural sinus bleed). More persistent symptoms of hearing loss and ataxia were seen in 2/19 (11 %) of cases. A longer period of follow-up would be ideal, as SUNCT is known to have periods of remission lasting for up to 7 years [4••]. What is of some reassurance is that the reported outcomes for the individual case reports and the one series of nine cases were identical, both showing improvement in two thirds of cases. This suggests that there has not been any significant bias in the reporting of single cases.

As summarised in Table 2, there were no significant differences in the majority of baseline characteristics between those who did and did not have a favourable outcome. The only significant difference was that patients who did not respond to microvascular decompression were exposed (on average) to a greater number of prior medical and surgical therapies ($p=0.03$, χ^2 test). Of the non-responders, 3/7 (43 %) had undergone prior ablative interventions (including occipital nerve block, glycerol rhizotomy and gamma knife radioablation of the trigeminal nerve) compared with only 1/12 (8 %) amongst the responders (this difference was not statistically significant). These findings may reflect more severe or refractory SUNCT/SUNA, but may also indicate a window of opportunity to treat these conditions when caused by vascular compression. The meta-analysis also hints at better outcome for cases with identifiable trigger factors.

Table 1 Review of case reports and case series of trigeminal nerve decompression for SUNCT/SUNA

Ref	Age (Yrs)	Sex (M/F)	Disease Duration (yrs)	Attack Freq (/day)	Attack Duration (s)	Trig	Course	Type	MRI	No. Prior R_x	Follow-Up (mths)	Response	Complications
[23]	73	M	6	<480	1–60	No	Chronic	SUNA	SCA	16	0.5	–	None
[47]	70	M	16	3–10	5–10	Yes	ns	SUNCT	SCA	6	14	+++	None
[8••]	71	M	6	30–200	60–120	Yes	Chronic	SUNCT	SCA	6	32	+++	None
[8••]	54	M	0.1	15–20	1–300	Yes	Episodic	SUNCT/ SUNA	SCA	4	32	+++	Wound infection
[8••]	46	M	3	20–30	30–120	No	Chronic	SUNCT	AICA Vein Adhesions	4	30	+++	Chest infection
[8••]	61	M	3	5–10	60–300	No	Chronic	SUNCT/ SUNA	SCA Vein	3	22	–	Transient vertigo
[8••]	56	M	1	20–30	120–300	Yes	Chronic	SUNA	SCA	3	20	+++	Right jaw pain
[8••]	48	F	2	30–50	300	Yes	Chronic	SUNA	AICA Vein	3	20	–	Dural sinus bleed
[8••]	51	F	5	90–120	3–10	Yes	Chronic	SUNCT	SCA	5	10	+++	Hearing loss/ataxia
[8••]	49	F	5	100–300	30–120	Yes	Chronic	SUNCT	SCA	6	10	–	None
[8••]	49	M	26	8–10	20–180	Yes	Episodic	SUNCT	SCA	3	9	+++	None
[46]	57	M	0.25	100–200	30–120	Yes	ns	SUNCT	SCA	ns	ns	+++	None
[48]	56	F	2	ns	5–120	Yes	ns	SUNCT	SCA	7	24	+++	None
[7]	49	M	2	30–40	5–60	Yes	ns	SUNCT	SCA	3	7	+++	None
[50]	60	M	12	200–300	5–60	No	Chronic	SUNCT	ns	12	ns	– –	None
[11]	39	M	2	60–400	3–30	No	Chronic	SUNCT	NA	9	5	–	None
[11]	28	M	10	100–200	20–30	ns	ns	SUNCT	NA	15	2	–	Unilateral deafness, vertigo, chronic disequilibrium
[45]	48	F	5	6–7	30–45	Yes		SUNCT	SCA	6	17	+++	None
[49]	54	M	3	ns	ns	ns	ns	SUNCT	ns	6	3	+++	None

Ref=reference; Freq=frequency; Trig=triggers; R_x=treatment; ns=not stated; NA=no abnormality; M=male; F=female; SCA=superior cerebellar artery; AICA=anterior inferior cerebellar artery; yrs=years; mths=months; s=seconds

Response key; +++=complete resolution of symptoms; ++ =>50 % improvement; +=<50 % improvement; –=no change in symptoms; – –=worsening of symptoms

Table 2 Comparison of clinical features in responders and non-responders (complete resolution of symptoms) for microvascular decompression

Clinical feature	Clinical Response		*p* value*
	Yes	No	
n	12	7	
Age (Yrs) – mean (SD)	55.1 (8.0)	51.1 (15.0)	ns
Gender (male) – *n* (%)	9 (75)	5 (83)	ns
Duration (Yrs) – mean (SD)	5.9 (7.7)	5.7 (3.9)	ns
Trigger – *n* (%)	10 (91)	2 (40)	ns
Phenotype			
SUNCT	10 (83)	4 (67)	ns
SUNA	1 (8)	2 (33)	
SUNCT/SUNA	1 (8)	1 (17)	
Aberrant vessel on MRI – *n* (%)	11 (92)	4 (80)	ns
Prior R_x – mean (SD)	4.8 (1.5)	9.1 (5.4)	0.03

ns=non-significant; R_x=treatment

* χ^2 test for categorical variables and Wilcoxon *T* test for continuous variables

In a recent, large series of posterior fossa surgery, 110 patients underwent microvascular decompression for trigeminal neuralgia and hemifacial spasm [52]. The most common complications were postoperative cerebrospinal fluid (CSF) leak (14.5 %), meningitis (10.9 %) and post-op wound infection (8.1 %). Temporary or permanent cranial nerve palsies were seen in four cases (3.6 %). The overall mortality rate was 1.8 % [52]. Another large study (with 1,185 patients) looking at the long-term outcome of microvascular decompression in trigeminal neuralgia found a 1 % incidence of ipsilateral hearing loss. One patient was found to have a brainstem infarction, and the overall mortality from the procedure in this series was 0.2 % [53]. These studies indicate that microvascular decompression is not without risk and the risk versus benefit needs to be weighed carefully before embarking on surgery [54].

A recent study of autonomic symptoms in trigeminal neuralgia has indicated that autonomic symptoms may be present in up to a third of cases [55]. This study also found that patients who had autonomic symptoms associated with trigeminal neuralgia had a poorer prognosis with regards to pain relief after microvascular decompression [55]. It is unclear as to whether the severity of autonomic symptoms affects the surgical outcome in patients with SUNCT/SUNA and further studies are needed in this area.

Neurostimulation

In recent years, neurostimulation as a treatment modality for TAC has gained popularity. This includes DBS and ONS. A better understanding of the role of the posterior hypothalamus in the generation of TAC and the recognition of the trigeminocervical complex has led to the increasing use of these treatment modalities.

Deep Brain Stimulation

Activation of the posterior hypothalamus in patients with cluster headache on PET has been demonstrated [12]. Subsequently, DBS of the posterior hypothalamus was used to successfully treat a patient with chronic cluster headache [56]. The first case of successful DBS in a patient with chronic SUNCT was reported by the same group in 2005, using the same stereotactic correlates [57]. There have been three further case reports of SUNCT treated with posterior hypothalamic DBS [23, 58]. In all three cases, the beneficial effects of DBS were not immediate, but were noticed after variable periods from 1 to several months. There was a definite and sustained decrease in attack frequency. Improved responsiveness to lamotrigine at a lower dose post-operatively was also reported in one patient, thereby inferring that long term DBS modifies the pain circuits that involve the hypothalamus [56].

Most of the experience in the use of posterior hypothalamic DBS comes from case series of patients with intractable cluster headache. A recent review summarised the results of posterior inferior hypothalamic stimulation in patients with refractory cluster headache [59]. DBS was found to be effective with 36/58 (62 %) of patients achieving a pain free/almost pain-free state [59]. The follow-up period varied from 1 to 4 years. Hypothalamic stimulation was found be ineffective in acute attacks [60]. The most frequently reported adverse effect with DBS was diplopia [59]. This was directly related to the amplitude of stimulation and may be the limiting adverse effect. Vertigo has also been reported [9]. There is a small risk of intracerebral haemorrhage related to the implantation procedure, which can result in death [61]. Long-term stimulation has been found to be safe, with no effects on the baroreflex, cardiorespiratory function, sympathetic function or vagal efferent function. Asymptomatic impairment of orthostatic adaptation was the only effect noted on the autonomic nervous system [62]. The limited experience in patients with SUNCT and other TAC suggests that DBS could be an option in refractory cases. However, this is an invasive procedure and patient selection needs to be done carefully with due consideration of the risks.

Occipital and Trigeminal Nerve Stimulation

The effects of peripheral nerve stimulation on pain perception were described in 1978 [63]. It is now known that such stimulation can modify the perception of pain and lead to analgesic effects [64]. The practical application from our understanding of the trigeminocervical complex is that

stimulation of the afferent fibres of the occipital nerves (C2 and C3) results in activation of the trigeminal nucleus caudalis and the posterior hypothalamus [65]. ONS has been shown to be effective in chronic migraine with a responder rate (50 % drop in headache days or 3 point drop on headache scale) of 39 % with adjustable stimulation, 8 % with preset stimulation and no response in medically managed patients [66]. The patients enrolled into this study had responded to occipital nerve blocks as a prerequisite prior to ONS [66]. With regards to TAC, ONS has been found to be effective in cluster headache and hemicrania continua [67, 68]. In SUNCT, experience with ONS is limited to two patients, one of whom responded [69]. Further details of these patients and duration of follow-up have not been published. ONS is considered to be a safe procedure. However, electrode migration both traumatic and spontaneous, electrode fracture and malfunction are known to occur necessitating surgical revision. Local discomfort and paraesthesias are the other potential problems [69]. Stimulation of the supraorbital and supratrochlear nerves has been used as a treatment for cluster headache and has also been successfully used to treat one patient with SUNCT syndrome [70].

Ablative Procedures

A summary of reported cases and series of other ablative procedures for SUNCT/SUNA is given in Table 3 and these are discussed further below. Three patients who had poor outcomes with ablative procedures (glycerol rhizotomy, trigeminal radiofrequency thermocoagulation, gamma knife radiosurgery) have been reported [11, 50]. These procedures may have irreversible complications, such as residual hypoaesthesia, anaesthesia dolorosa and keratitis [11, 16, 50]. The most feared of these complications is anaesthesia dolorosa, but in studies of rhizotomy for trigeminal neuralgia, this outcome is extremely rare even with prolonged follow-up [71]. It must be born in mind that the following data on ablative procedures are limited to isolated cases reports, and there is considerable potential for bias due to under-reporting of unsuccessful cases or those with adverse outcomes.

Balloon Compression of the Gasserian Ganglion

Ablative compression of the Gasserian ganglion can be achieved using a trochar and Fogarty catheter with an approach through the maxilla and foramen ovale under fluoroscopic guidance [72]. The balloon is inflated for 1.5–2 minutes. There are two case reports of patients with SUNCT who benefitted from percutaneous balloon compression of the Gasserian ganglion, with one remaining pain free for ten years [73] and the other having a partial response [74].

Glycerol Rhizotomy

The use of retro-Gasserian glycerol rhizolysis has been described in a case series of three patients [51] and in one other single case report [11]. In all four cases treated with this therapy, further interventions were required.

Other Procedures

The outcomes of these procedures are also summarised in Table 3.

Gamma Knife Radiosurgery

Gamma knife radiosurgery of the trigeminal nerve and sphenopalatine ganglion has been used to treat SUNCT with mixed results [11, 75].

Occipital Nerve Block

Outcomes from two detailed case reports of this therapy have shown mixed results, with one showing an excellent response [76] and the other having only a partial response requiring ongoing medical therapy [77]. A further nine cases of SUNCT/SUNA treated in this way have been reported with minimal clinical data [41]. These cases suggest an overall response rate for this treatment modality of 6/11 (55 %). The duration of effect for this procedure is generally short (1 week to 6 months) [41], and its role in predicting response to ONS is unclear.

Superior Cervical Ganglion Block

Opioid blockade of the superior cervical ganglion was effective in one case who required ongoing medical therapy, but had resolution of symptoms over a 16 month period following a series of five procedures [78].

Discussion

SUNCT and SUNA are rare but disabling primary headache disorders that can often be refractory to medical management [3]. Advances in structural and functional imaging have demonstrated a potential role for the posterior hypothalamus in the generation of SUNCT [22]. However, whether these changes are pathogenic or an epiphenomena is a matter of considerable debate [79•]. Our understanding of the anatomical pathways involving the trigeminocervical complex and the trigeminal autonomic reflex, and how these pathways are influenced by the hypothalamus, is growing. The posterior inferior

Table 3 Review of case reports of other surgical treatments for SUNCT/SUNA

Ref	Age (yrs)	Sex (M/F)	Disease Duration (yrs)	Attack Freq (/day)	Attack duration (s)	Trig	Course	Type	MRI	No Prior Rx	Follow-Up (mths)	Resp	Complications
Deep brain stimulation													
[23]	73	M	6	ns	1–60	No	ns	SUNCT	SCA	16	15	++	None
[58]	44	M	36	30–200	60–120	ns	ns	SUNCT	NA	26	12	++	Erectile dysfunction
[57]	66	F	14	70–300	2–20	Yes	Chronic	SUNCT	NA	9	15	++	Diplopia at high stimulation levels
Occipital nerve stimulation													
[69]	ns	ns	ns	ns	ns	ns	ns	SUNCT	ns	ns	ns	++	ns
[69]	ns	ns	ns	ns	ns	ns	ns	SUNCT	ns	ns	ns	–	ns
Trigeminal nerve stimulation													
[70]	ns	ns	ns	ns	ns	ns	ns	SUNCT	ns	5	36	++	Skin erosion, superficial infection
Gasserian ganglion balloon compression													
[73]	66	F	0.1	3–5	15–30	No	ns	SUNCT	NA	7	120	+++	None
[74]	68	F	17	30–100	5–60	Yes	Chronic	SUNCT	NA	9	18	++	None
Glycerol rhizotomy													
[51]	80	M	25	ns	30–120	Yes	Episodic	SUNCT	NA*	7	87	++	Facial sensory loss
[51]	72	F	10	ns	20–120	Yes	Chronic	SUNCT	NA	6	90	++	Facial sensory loss
[51]	52	M	8	ns	ns	Yes	Chronic	SUNCT	NA*	9	7	+++	
[11]	38	M	2	60–400	3–30	No	Chronic	SUNCT	NA	9	5	–	Residual hypoaesthesia
Gamma knife radiosurgery													
[75]	82	M	6	3–10	30–120	Yes	Episodic	SUNCT	NA	6	39	+++	None
[11]	39	M	2	60–400	3–30	No	Chronic	SUNCT	NA	9	5	+	Anesthesia dolorosa
[11]	28	M	10	100–200	20–30	ns	ns	SUNCT	NA	15	2	–	
Greater occipital nerve block													
[76]	62	F	0.5	40–50	120–480	Yes	ns	SUNCT	NA	6	0	+++	None
[77]	82	F	0.1	10–20	120	Yes	ns	SUNCT	ns	5	0	+	None
[41]	ns	ns	ns	ns	ns	ns	ns	SUNCT	ns	ns	ns	+++	ns
[41]	ns	ns	ns	ns	ns	ns	ns	SUNCT	ns	ns	ns	+++	ns
[41]	ns	ns	ns	ns	ns	ns	ns	SUNCT	ns	ns	ns	+++	ns
[41]	ns	ns	ns	ns	ns	ns	ns	SUNCT	ns	ns	ns	+++	ns
[41]	ns	ns	ns	ns	ns	ns	ns	SUNCT	ns	ns	ns	+++	ns
[41]	ns	ns	ns	ns	ns	ns	ns	SUNCT	ns	ns	ns	–	ns
[41]	ns	ns	ns	ns	ns	ns	ns	SUNCT	ns	ns	ns	–	ns
[41]	ns	ns	ns	ns	ns	ns	ns	SUNCT	ns	ns	ns	–	ns
[41]	ns	ns	ns	ns	ns	ns	ns	SUNA	ns	ns	ns	–	ns

Ref=reference; Freq=frequency; Trig=triggers; R_x=treatment; Resp=response; ns=not stated; NA=no abnormality: M=male; F=female; SCA=superior cerebellar artery; yrs=years; mths=months; s=seconds

*=CT only performed

Response key; +++=complete resolution of symptoms; ++ =>50 % improvement; +=<50 % improvement; –=no change in symptoms; – –=worsening of symptoms

part of the hypothalamus has an important role in the generation and modulation of these headache disorders. A better understanding of the pathophysiology of SUNCT has given impetus to the development of new surgical interventions in this area. High resolution neuroimaging (particularly CISS images [80]) has helped to identify aberrant vascular loops compressing the trigeminal nerve resulting in secondary SUNCT. This does raise questions regarding the differentiation between SUNCT/SUNA and first division trigeminal neuralgia. However, lower severity of pain, longer duration of pain (greater than 5 seconds), prominence of autonomic

features, absence of a refractory period and incomplete response to carbamazepine have been noted as features indicative of SUNCT syndrome [81]. Autonomic symptoms are common in trigeminal neuralgia, although are generally milder. In first division trigeminal neuralgia, tearing was reported in 45 % of cases and conjunctival injection in 30 % [55]. Thus, there is certainly some potential for confusion.

There is some evidence to suggest that chronic cases of SUNCT/SUNA (daily symptoms for at least 12 months with no remission greater than 1 month) are more likely to be resistant to medical therapy [40]. In chronic cases of SUNCT/SUNA refractory to multiple therapies, the present review would suggest that surgical treatments are certainly worth considering. Other factors that should always be born in mind include: existence of comorbidities and risk factors that might impact upon the risks of a surgical intervention, the perception and personal relevance of specific potential complications of the procedure, psychological consequences of SUNCT/SUNA and of course patient preference.

A critical evaluation of the small number of patients who have had microvascular decompression for SUNCT/SUNA suggests that this approach is an option for patients with ipsilateral trigeminal nerve compression due to aberrant vascular loops and who have previously not responded to maximal medical management. However, caution needs to be exercised in patients who have undergone previous ablative surgical procedures of the trigeminal nerve, as there is a suggestion that this subgroup may do less well. Overall a positive response should be expected in two thirds of cases, but this benefit needs to be carefully weighed against the risks. Reviews of larger studies of patients who underwent microvascular decompression for trigeminal neuralgia suggest that risk of complications is low, but the consequences can be significant. Apart from the immediate perioperative risks, permanent vertigo, deafness, ataxia and facial pain have been reported.

DBS of the posterior hypothalamus has yielded some promising results in chronic migraine and cluster headache. The results in SUNCT have been encouraging, but experience is limited to three cases. DBS, being an invasive procedure, carries distinct risks and this needs to be weighed carefully against any potential benefit.

Preliminary experience of ONS in SUNCT shows some promise and warrants further study. This procedure lacks the more significant risks associated with microvascular decompression and DBS. The experience of this therapy in chronic cluster headache suggests that this could be an option in refractory cases of SUNCT/SUNA. Although local complications and device malfunction are common, there is a low incidence of permanent and disabling complications, and this could render ONS as an attractive option.

Ablative surgical procedures have yielded variable results, and can be associated with extremely troublesome adverse outcomes, such as facial sensory loss and anaesthesia dolorosa. The potential for this latter condition alone means that enthusiasm for these therapies must be guarded. However, in extreme cases where all else has failed they remain an option.

Little is known about whether certain subgroups may benefit from one particular type of procedure. Defining groups of patients who stand to benefit from one particular intervention will be important to guide management for medically refractory cases of SUNCT. The studies discussed above are limited by small numbers, non-randomised design and unblinded assessment. A randomised, assessor blinded, clinical trial should be possible in SUNCT/SUNA. Assuming a population prevalence of 6.6/100,000 [40], a frequency of chronic disease of 60 % and a frequency of aberrant vascular loops of 30 %, there would be approximately 4,000 prevalent cases of chronic SUNCT/SUNA in the USA. For a clinical trial comparing microvascular decompression (60 % response rate) against lamotrigine (less than 50 % response rate for chronic cases—for purpose of power calculation, response rate of 30 % used) a total sample size of 30 would have 50 % power to detect a difference with $\alpha=0.05$. It should be noted that the spontaneous remission rate for SUNCT/SUNA is also approximately 30 % and probably much lower in chronic cases. Thus, a placebo-controlled or "best medical treatment" comparison trial would also be feasible with a similar number of participants. Given that spontaneous remission can frequently last for 2–3 years [40] a minimum follow-up period of 2 years would be essential.

Conclusion

In the absence of randomised clinical trials, we conclude that microvascular decompression is a therapeutic option in patients with medically refractory SUNCT/SUNA, in whom trigeminal nerve compression is demonstrated on MRI (Level II-2 evidence). In the absence of structural pathology, central or peripheral neurostimulation with ONS/DBS may be considered and from a purely pragmatic perspective, it would seem sensible to consider the less invasive peripheral approach first. In cases that remain resistant, ablative interventions are an option, but the potential for decreasing efficacy and greater complications (facial numbness in particular) must be considered carefully. It must be emphasised that the evidence for these further surgical interventions is limited and results have been, at best, rather mixed with some potentially significant adverse outcomes.

Conflict of interest Swapna Sebastian declares that she has no conflict of interest.

Daniel Schweitzer declares that he has no conflict of interest.

Leong Tan declares that he has no conflict of interest.

Simon A. Broadley declares that he has no conflict of interest.

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Papers of particular interest, published recently, have been highlighted as:

- • Of importance
- •• Of major importance

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