

# Tears of Pain

## SUNCT and SUNA

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# IHS Classification – 1989 (updated 2004)

- **Primary Headaches** – 4 categories
  - Migraine
  - Tension-type
  - **Cluster and other trigeminal autonomic cephalalgia**
  - Other primary headaches
    - Trigeminal Neuralgia
    - Primary cough, exertional and coital headache

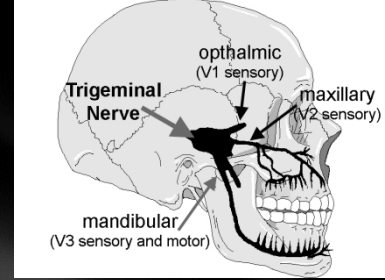
# IHS Classification – 1989 (updated 2004)

## • Secondary Headaches

- Head and or neck trauma
- Cranial or cervical vascular disorders
- Non vascular intracranial disorders
- Substance or withdrawal
- Infection
- Disorders of homeostasis
- Disorders of cranium, neck, eye, ear, nose, teeth, mouth or other facial or cranial structures
- Psychiatric disorders



# Trigeminal Neuralgia



- **Intense, paroxysms of sharp, stabbing pain**
  - Lasting few seconds to 2 minutes
  - Pain free between attacks
  - Attacks are stereotyped
- **Precipitated from trigger**
  - Light touch of the face (washing, shaving, make-up)
  - Chewing, talking, cold wind
- **No clinical neurological deficit**
  - Numbness or weakness
- **Not caused by another disorder**
- **Classical TN responds well to Tegretol**

# Trigeminal Autonomic Cephalgias

Group of Primary Headache with

- Unilateral trigeminal distribution
- Associated with ipsilateral photophobia and cranial autonomic symptoms
- Differ in duration, frequency and response to therapy

**Cluster Headache – longest duration and low frequency**

**Paroxysmal Hemicrania – immediate duration and frequency**

**SUNA** (major DD for TN)

- Short lasting unilateral neuralgiform attacks with cranial autonomic symptoms
- **Shortest duration and highest frequency**

**SUNCT**

- Short lasting unilateral neuralgiform attacks with conjunctival injection and tearing +/- other cranial autonomic symptoms
- **Shortest duration and highest frequency**

# CAS symptoms SUNCT vs SUNA vs CH

**Table 2** Associated cranial autonomic symptoms\*

	SUNCT	SUNA	CH** (%)
Conjunctival injection	43 (100%)	2 (22%)	77
Lacrimation	43 (100%)	3 ipsilateral, 1 contralateral (44%)	91
Nasal blockage	17 (40%)	2 (22%)	75
Rhinorrhoea	23 (53%)	2 (22%)	72
Eyelid oedema	21 (49%)	1 (11%)	74
Ptosis	22 (51%)	3 (33%)	74
Facial flushing	2 unilateral, 2 bilateral (9%)	1 (11%)	
Sweating	2 unilateral, 1 bilateral (7%)	1 bilateral (11%)	
Other	4 (9%)	3 (33%)	

\*By definition, 100% of SUNCT patients had both conjunctival injection and lacrimation and no patients with SUNA had both;

\*\*CH, cluster headache (after Bahra *et al.*, 2002).



# IHS criteria for episodic cluster headache

- A. At least 5 attacks fulfilling B-D
- B. Severe unilateral orbital, supra-orbital and/or temporal pain lasting 15-180 minutes untreated
- C. Headache associated with at least one of the following signs:
  - Conjunctival injection
  - Lacrimation
  - Nasal congestion
  - Rhinorrhea
  - Forehead and facial sweating
  - Miosis, ptosis
  - Eyelid oedema
- D. Frequency once every other day to 8 per day

# Cluster headache

Chronic refers to similar attacks but occurring for  $> 1$  year without remission or with remission lasting  $< 14$  days

80-90% are episodic

10-20% are chronic

85% with episodic cluster headaches are males

Females  $>$  males for chronic

Characteristically occur at night



# Cluster Headache Treatment

## Acute treatment

- High-flow (12 L/min) 100% nasal oxygen for 10-15 minutes
- Nasal lignocaine
- Dihydroergotamine via IV or rectal suppository
- Triptans – sc or nasal
- Nasal capsaicin spray

## Once controlled

- Prednisone 50 mg daily, tapered slowly over the following 3 weeks to prevent recurrence

## Chronic Cluster Headache

- Verapamil 80 mg bd up to 320 mg bd (Heart block, gum hypertrophy, constipation, SOA)
- Lithium with or without clonazepam
- Valproate and baclofen have been tried
- Methysergide

# Cluster Headache - New Treatments

## **New combinations of old drugs**

- **Treximet** - 85 mg sumatriptan and 500 mg naproxen

## **New formulations of old drugs**

- **Intranasal zolmitriptan (Zomig)**
  - Better tolerated than Imigran (unpleasant taste)
- **Inhaled DHE**

## **New acute and preventive treatments**

- **Telcagepant** - a CGRP (Calcitonin Gene-Related Peptide) receptor antagonist - phase III
- **TRPV 1** (Transient Receptor Potential Vanilloid) receptor antagonist on ion channels
  - Selective for calcium, magnesium and sodium

## **Established treatments for other neurologic conditions**

- **Memantine** - glutamate receptor antagonist (NMDA) - prevention of refractory migraine

## **Gene therapy**

- Mayo Clinic collecting genomic data on 4,500 patients migraine with and without aura and on gender- and age-matched controls

# Chronic Paroxysmal Hemicrania

Severe, unilateral, (orbital, supraorbital, temporal)

Shorter lasting - 2-45 mins and more frequent (1-40/day)

>5/day lasting 2-30min

Generally V<sub>1</sub>

Females > Males

Ipsilateral parasympathetic activation – miosis

Episodic in 20% lasting for a week per month

Chronic with no remission

Distinctively responds very well to **Indomethacin 150 mg or less**

Also COX-2 and topiramate



## **Short-lasting Unilateral Neuralgiform headaches Attacks with cranial autonomic symptoms**

Prominently pain in V1 more than V2, V3 or in teeth  
Strictly unilateral stabbing  
Very brief (5 - 240 secs) occurring at least 20-200 times a day

Conjunctival injection or Tearing only but not both  
Ipsilateral photophobia, phonophobia c.f. bilateral in migraine  
Other cranial autonomic symptoms

- Rhinorrhoea
- Eyelid oedema
- Facial sweating / flushing
- Ear flushing

Cutaneous stimulation triggers attacks in 22%  
No refractory period between attacks

High rate of pituitary tumours in both so MRI scan indicated

# SUNCT

- **Short-lasting Unilateral Neuralgiform Headaches with Conjunctival injection and Tearing**
- ? Subset of SUNA or separate
- Prominently pain in V<sub>1</sub> more than V<sub>2</sub>, V<sub>3</sub> or in teeth
- Strictly unilateral stabbing
- Very brief (5 - 240 secs) occurring at least 20-200 times a day

Ipsilateral conjunctival injection and tearing always present

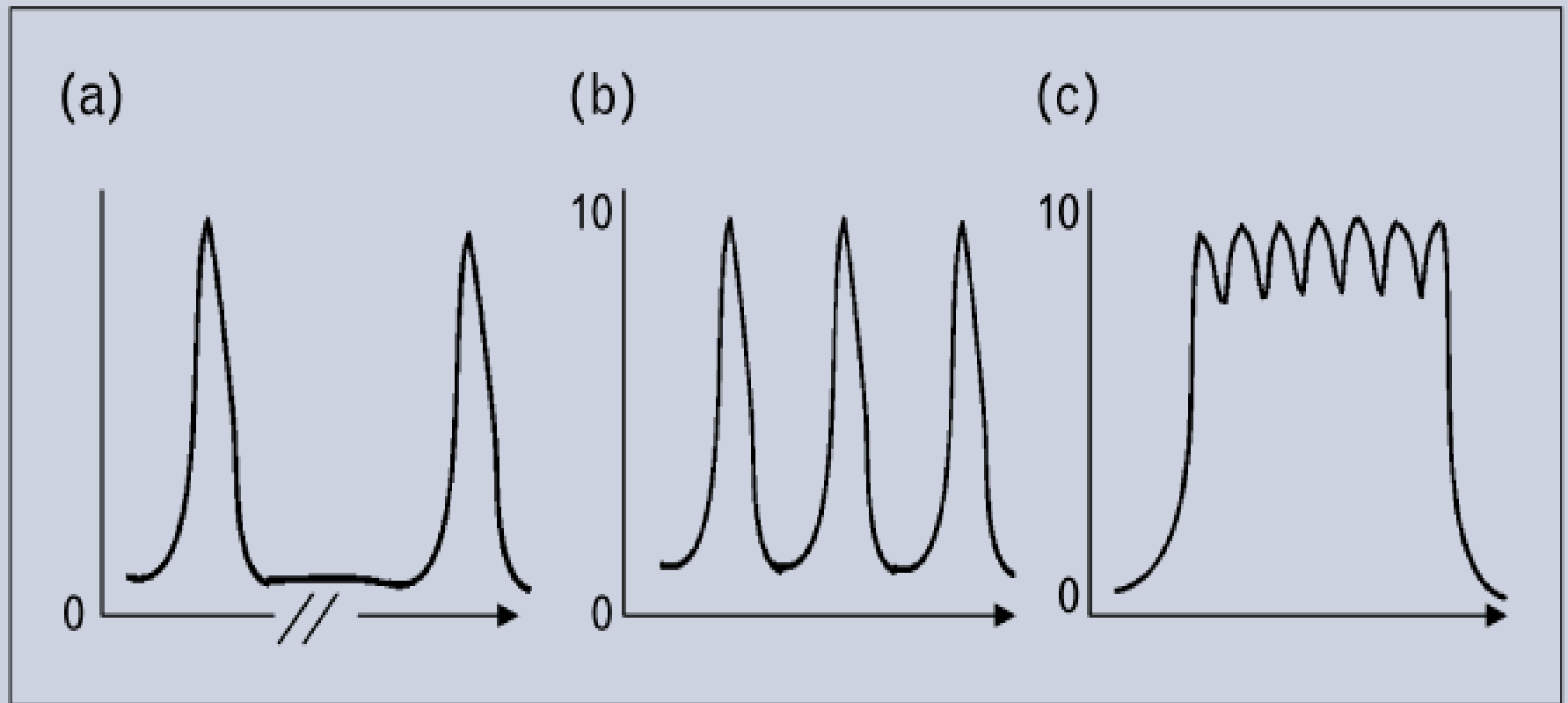
Cutaneous stimulation triggers attacks in 74%  
touching, washing, chewing, eating, talking, shaving

No refractory period between attacks  
Dull ache

Men > Female

**Figure 1 The three types of clinical picture of attacks of SUNCT/SUNA**

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Pain (verbal rating scale from 1 to 10). (a) Single stabs. (b) Each attack is a group of stabs. (c) Saw-tooth pattern. Reproduced with permission from [7].



**Table 1 Comparison on the trigeminal autonomic cephalalgias based on cohorts we have studied<sup>a</sup> and patients we have reviewed<sup>b</sup>**

	Cluster headache	Paroxysmal hemicrania	SUNCT/SUNA
Sex	3 M to 1 F	M = F	1.5 M to 1 F
Pain			
Quality	Sharp/stab/throb	Sharp/stab/throb	Sharp/stab/throb
Severity	Very severe	Very severe	Severe
Distribution	V1 > C2 > V2 > V3	V1 > C2 > V2 > V3	V1 > C2 > V2 > V3
Attacks			
Frequency (per day)	1–8	20	100
Length (min)	30–180	2–30	1–5
Triggers			
Alcohol	+++	+	–
Nitroglycerin	+++	+	–
Cutaneous	–	–	+++
Agitation/restlessness	90%	80%	65%
Episodic vs. chronic	90 : 10	35 : 65	10 : 90
Circadian/circannual periodicity	Present	Absent	Absent
Treatment effects			
Oxygen	70%	No effect	No effect
Sumatriptan, 6 mg	90%	20%	<10%
Indomethacin	No effect	100%	No effect
Migraine features with attacks			
Nausea	50%	40%	25%
Photophobia/phonophobia	65%	65%	25%

C, cervical; F, female; M, male; SUNCT/SUNA, short-lasting unilateral neuralgiform headache attacks with conjunctival injection and tearing/short-lasting unilateral neuralgiform headache attacks with cranial autonomic features; V, trigeminal.

# SITE of Attacks SUNCT vs SUNA

Table 1 Site of attacks

	SUNCT	SUNA
Eye	29 (67%)	2 (22%)
Retro-orbital region	24 (56%)	5 (56%)
Eyebrow	3 (7%)	0 (0%)
Forehead	16 (37%)	1 (11%)
Temple	14 (33%)	5 (56%)
Side of head	4 (9%)	4 (44%)
Top of head	9 (21%)	0 (0%)
Back of head	12 (28%)	2 (22%)
Nose	16 (37%)	1 (11%)
V <sub>2</sub>	14 (33%)	3 (33%)
V <sub>3</sub>	0 (0%)	3 (33%)
Teeth	9 (21%)	2 (22%)
Neck	1 (2%)	1 (11%)
Ear	2 (5%)	1 (11%)

# Attack Triggers

**Table 4** Triggered attacks and refractory period

	SUNCT	SUNA	Total
<b>Triggers to attacks</b>			
Touch	27 (63%)	0 (0%)	27 (52%)
Chew/eat	26 (60%)	2 (22%)	28 (54%)
Wind	17 (40%)	1 (11%)	18 (35%)
Wash face	17 (40%)	0 (0%)	17 (33%)
Brushing teeth	16 (37%)	0 (0%)	16 (31%)
Move	14 (33%)	2 (22%)	16 (31%)
Talk	9 (21%)	1 (11%)	10 (19%)
Wash or brush hair	5 (12%)	0 (0%)	5 (10%)
Exercise	5 (12%)	1 (11%)	6 (12%)
Light	4 (9%)	0 (0%)	4 (8%)
Shower	4 (9%)	0 (0%)	4 (8%)
Shaving	4 (9%)	0 (0%)	4 (8%)
Blow nose	3 (7%)	0 (0%)	3 (6%)
Alcohol	0 (0%)	1 (11%)	1 (2%)
Smoke	1 (2%)	0 (0%)	1 (2%)
Smells	1 (2%)	0 (0%)	1 (2%)
Warm	1 (2%)	0 (0%)	1 (2%)
Others	5 (12%)	0 (0%)	5 (10%)
<b>Factors making attacks worse</b>			
Stress	3 (7%)	1 (11%)	4 (8%)
Tired	1 (2%)	1 (11%)	2 (4%)
Travel	3 (7%)	0 (0%)	3 (6%)
Weather	1 (2%)	0 (0%)	1 (2%)
All triggered	1 (2%)	0 (0%)	1 (2%)
Mostly triggered	12 (28%)	2 (22%)	14 (27%)
Equal triggered and spontaneous	7 (16%)	0 (0%)	7 (13%)
Mostly spontaneous	12 (28%)	0 (0%)	12 (23%)
All spontaneous	6 (14%)	6 (67%)	12 (23%)
Unknown	5 (12%)	1 (11%)	6 (12%)



# SUNCT / SUNA Treatment

- Responds very well to
  - Indomethacin
  - Lamotrigine – 2/3 respond
  - Topiramate – 2/5 received 50mg bd improved over 10 days
  - Gabapentin
- Also responsive to
  - sub-occipital stimulation
  - sub-cutaneous Lignocaine infusion

# Sub-Occipital Stimulators

Electrical stimulation of primary sensory afferents

- Anti-nociceptive effects
- PET studies
  - Changes in regional cerebral blood flow
    - Dorsal rostral pons
    - Anterior cingulate cortex
- Occipital neuralgia
- Chronic migraine
- Cluster Headache and SUNCT

# SUNA / SUNCT

## SUNA/SUNCT vs TN

- Prominent V<sub>1</sub> distribution of pain

- Attacks triggered by touch

- Lack of refractory period between attacks

## SUNA/SUNCT vs CH

- Lack of response to O<sub>2</sub>, Triptans and Verapamil

Goadsby experience - overdiagnosis of TN in UK

- <1 in 100 diagnosed with TN actually have TN



# Mrs BB

- 58 yo
- 3 year hx of left facial pain
  - Sharp, shooting and knife like lasting seconds
  - Pain increased by touching nose and face
  - **Dx as TN**
- BUT
  - Conjunctival injection
  - Lacrimation left eye
  - Nasal congestion
- **SUNCT**
- Indocid 25 mg three times a day
- Complete resolution of pain

# SUNCT vs SUNA

SUNCT – most painful condition ever (worse than childbirth)

VRS rating 10/10

84% SUNCT vs 33% SUNA

SUNCT - more attacks / day

Triggered by touching face

74% SUNCT vs 22% in SUNA

Conjunctival Injection and Tearing

100% SUNCT vs 22-44% in SUNA

Mean age of onset similar - 40-44

Mean duration of symptoms similar – 8 - 10 years

Mean duration for diagnosis similar - 6.7 - 7.1 years

# Conclusion

Trigeminal Autonomic Cephalgias are a group of Primary Headache with

- Unilateral trigeminal distribution pain
- Associated with ipsilateral cranial autonomic symptoms
  - Lacrimation, conjunctival injection or nasal symptoms
- Important to differentiate given selective response to treatment

**Cluster Headache – longest duration and low frequency**

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