

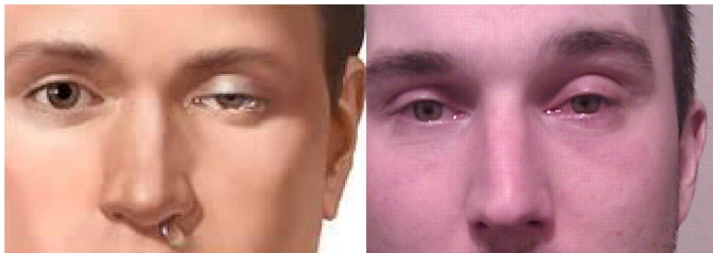
# Trigeminal Autonomic Cephalalgias



문 희 수

성균관대학교 의과대학 신경과학교실

## Trigeminal Autonomic Cephalalgias



↑ Parasympathetic discharge (conjunctival tearing, rhinorrhea)

↓ Sympathetic paresis (Miosis, Horner's)

## Short lasting headaches (<4 hours)

### With autonomic Symptoms

- 3.1 Cluster headache
- 3.2 Paroxysmal hemicrania
- 3.3 Short-lasting unilateral neuralgiform headache attacks
- 3.4 Hemicrania continua
- 3.5 Probable trigeminal autonomic cephalalgia

**Cluster-like headache (2<sup>nd</sup>) :**  
**Pituitary region pathology**

### Without autonomic symptoms

- 4.1 Primary cough headache
- 4.2 Primary exercise headache
- 4.3 Primary headache associated with sexual activity
- 4.7 Primary stabbing headache
- 4.9 Hypnic headache
- 13.1 Trigeminal neuralgia

## Classification: ICHD

### 3. Trigeminal Autonomic Cephalalgias

#### ICHD-2

- 3.1 Cluster headache
- 3.2 Paroxysmal hemicrania
- 3.3 SUNCT
- 3.4 Probable trigeminal autonomic cephalalgia

4.7 Hemicrania continua

A 3.3 SUNA

#### ICHD-3 beta

- 3.1 Cluster headache
- 3.2 Paroxysmal hemicrania
- 3.3 Short-lasting unilateral neuralgiform headache attacks
  - 3.3.1 SUNCT
  - 3.3.2 SUNA
- 3.4 Hemicrania continua
- 3.5 Probable trigeminal autonomic cephalalgia

## Criteria of Autonomic Symptoms in TAC

C. Either or both of the following:

1. at least one of the following symptoms or signs, ipsilateral to the headache
  - a) conjunctival injection and/or lacrimation
  - b) nasal congestion and/or rhinorrhoea
  - c) eyelid oedema
  - d) forehead and facial sweating
  - e) forehead and facial flushing
  - f) sensation of fullness in the ear
  - g) miosis and/or ptosis
2. a sense of restlessness or agitation

ICHD3

1/8

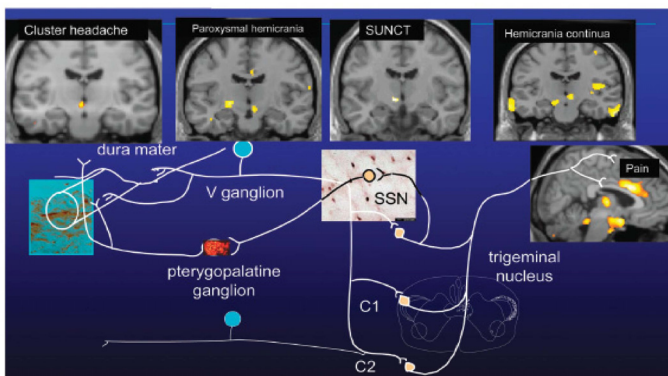
C. Headache is accompanied by at least one of the following:

1. ipsilateral conjunctival injection and/or lacrimation
2. ipsilateral nasal congestion and/or rhinorrhoea
3. ipsilateral eyelid oedema
4. ipsilateral forehead and facial sweating
5. ipsilateral miosis and/or ptosis
6. a sense of restlessness or agitation

ICHD2

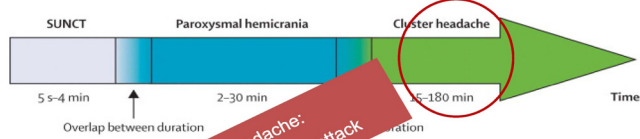
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## Pathophysiology in TACs

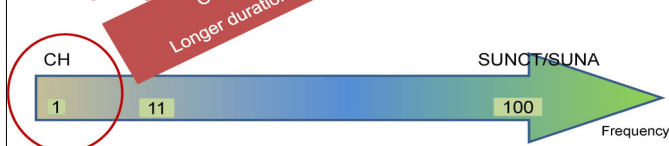


## Distinguished from each other..

## 1. Attack duration



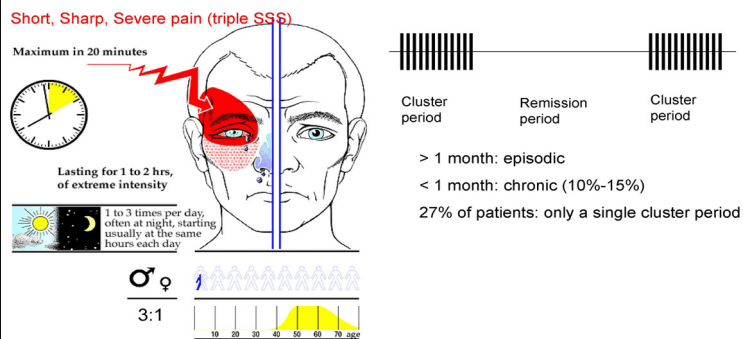
## 2. Frequency of



## Case 1

32세 남자 환자가 2주 전부터 시작된 우측 눈 부위에 반복되는 통증을 주소로 응급실에 내원하였다. 두통은 하루 1-2차례 발생하여 1시간 정도 지속되었다. 두통은 칼로 찌르는 듯 심한 통증이었으며 통증이 시작되면 우측 눈이 충혈되고 콧물이 동반되었다. 두통이 시작되면 도저히 가만히 있을 수 없어서 안절부절 못하고 때로는 벽에 머리를 찌기도 하였다. 낮에도 발생하지만 주로 새벽 1시경에 통증이 왔다. 약간의 구역이 동반되기도 하고 술을 먹으면 두통이 유발되었다. 과거력상 특이사항이 없었으며 하루에 20PY의 흡연을 하고 있다. 응급실에서 시행한 brain CT 및 혈액검사상 정상이었다.

## Clinical Pearls on Diagnosing CH



## How To Make The Diagnosis: If no autonomic features..

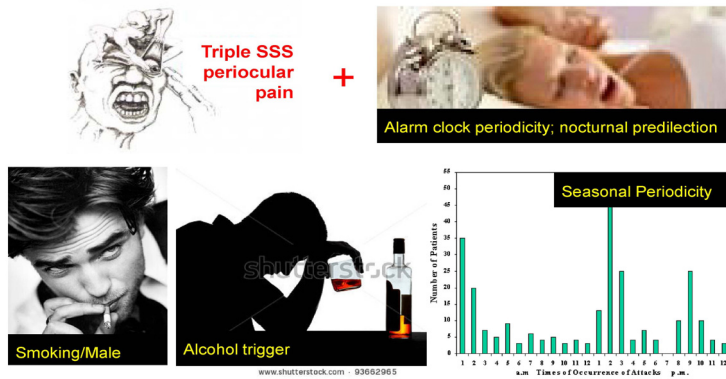
### Cluster Headache Without Autonomic Symptoms: Why Is It different?

Isabel P. Martins, MD, PhD; Raquel G. Gouveia, MD; Elsa Parreira, MD

No autonomic features, cluster suspected (**5-7% in CH**)

- Ask about agitation
- Rocking, pacing, groaning, holding his involved eye

## How To Make The Diagnosis: If no autonomic features..



## How To Make The Diagnosis: CH vs. Migraine

Unilateral Photophobia or Phonophobia Can Distinguish Migraine from Cluster Headache

December 15, 2010

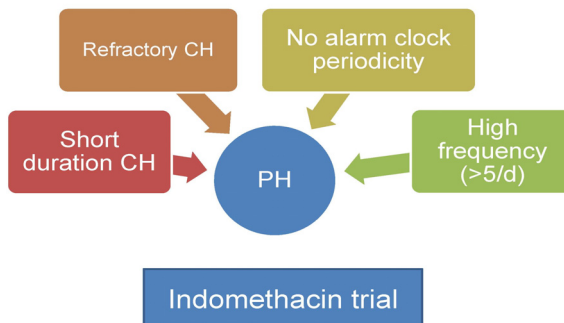
- Photo/phonophobia in 50 % TACs
- Cranial Autonomic symptoms in Migraine
- less 5% unilateral migraine with ipsilateral photo/phonophobia

CH	Migraine
<ul style="list-style-type: none"> <li>• Unilateral</li> <li>• Prominent</li> <li>• Stereotypical</li> </ul>	<ul style="list-style-type: none"> <li>• Bilateral</li> <li>• Less prominent</li> <li>• Variable</li> </ul>

**55세 여자** 환자가 8개월 전부터 시작된 **좌측** 안와주위와 측두부에 생긴 간헐적인 두통을 주소로 내원하였다. 두통은 찌르는 듯한 심한 통증으로 최근 빈도와 강도가 증가하여 **하루 10회 이상** 발생하였으며 1회 지속시간은 **10분 정도** 되었다. 동반증상으로는 두통이 있을 때 **좌측 눈물 흘림과 눈부심**이 동반되었다. 신경학적 진찰 및 MRI는 정상이었다.

**Indomethacin 50mg**을 복용한 첫날부터 두통이 호전되었다

### Clinical Pearls On Diagnosing Paroxysmal Hemicrania



### Treatment of Paroxysmal Hemicrania

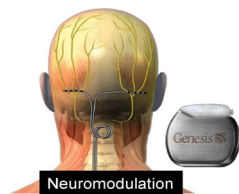
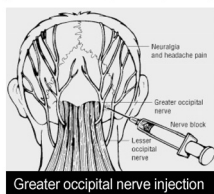
#### Indomethacin Trial

start with 25 mg tid

→ 50mg tid in partial response after 1 week

→ 75 mg tid in partial response after 2 week

Other drugs: NSAIDs, verapamil, COX-2 inhibitors, Sumatriptan



*Almost Always relieved with indomethacin*

5-240s : average 50s  
Tics like pain

Short-lasting **Unilateral Neuralgiform** Headache Attacks  
With **Conjunctival Injection And Tearing** (SUNCT)

Short-lasting **Unilateral Neuralgiform** Headache Attacks  
With **Cranial Autonomic Symptoms** (SUNA)

Conjunctival Injection OR Tearing

C. Either or both of the following:

- at least one of the following symptoms or signs, ipsilateral to the headache
  - conjunctival injection and/or lacrimation
  - nasal congestion and/or rhinorrhoea
  - eyelid oedema
  - forehead and facial sweating
  - forehead and facial flushing
  - sensation of fullness in the ear
  - miosis and/or ptosis
- a sense of restlessness or agitation

SUNA SUNCT

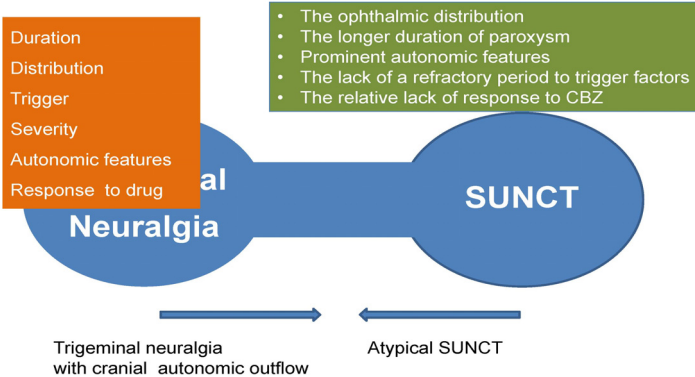
Short-lasting **Unilateral Neuralgiform** Headache Attacks  
With **Conjunctival Injection And Tearing** (SUNCT)

Short-lasting **Unilateral Neuralgiform** Headache Attacks  
With **Cranial Autonomic Symptoms** (SUNA)

63세 남자가 5개월 전 시작된 우측 안와부 주위의 반복되는 심한 통증으로 방문하였다. 두통은 **항상 우측**에만 발생하였고 빈도는 **하루 중 수 차례에서 수십 차례 정도** 발생하였다. 칼로 찌르는 듯한 날카로운 심한 통증과 작열감이 우측 안와부와 상악부 및 비익에 약 **30-40초 동안** 지속되었다. 구역, 구토, 빛 공포증이나 소리 공포증은 동반되지 않았고 거의 매번 **심한 결막출혈, 눈물, 비루와 비폐색이 동반**되었으며 이런 증상은 두통의 지속기간동안 계속된 뒤 소실되는 양상이었다. 또한 두통이 없을 때는 자율신경증상은 발생하지 않았다.



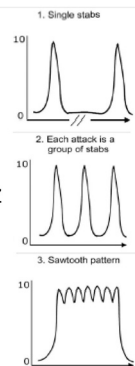
## Trigeminal neuralgia vs SUNCT/SUNA



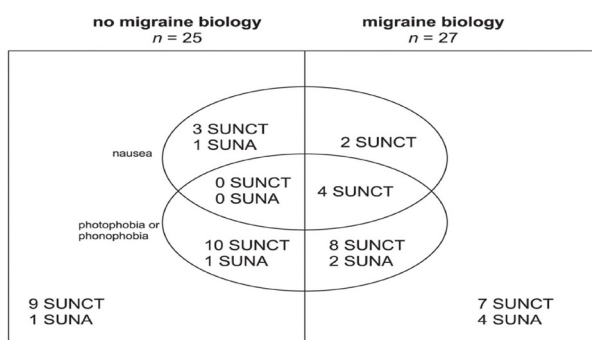
## Clinical Pearls On Diagnosing SUNCT/SUNA

- Pain is maximal in V1 distribution
- Short-lasting attacks of unilateral pain & lacrimation and redness of the ipsilateral eye
- Triggers are common
- No latency
- Refractory to all treatments including indomethacin/ CBZ
- SUNCT is rare which is why the workup is crucial

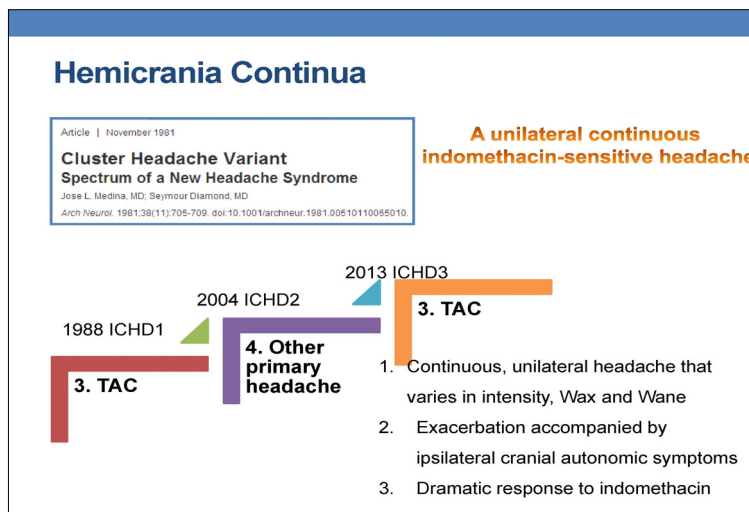
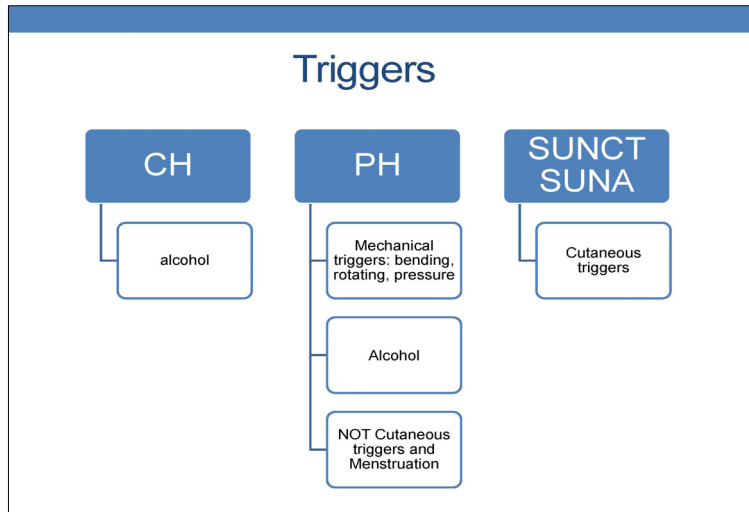
**Note that SUNCT may be secondary/ symptomatic** (posterior fossa, pituitary lesion, cavernous sinus)



## Photo/Phonophobia in SUNCT/SUNA



Brain (2006), 129, 2746–2760



### Caution: TAC-like headache !!

A case of 'cryptogenic' type D carotid cavernous fistula presenting initially with Cluster-like headache  
 J.S. George<sup>a</sup>, W. Adams<sup>b</sup>, M. Sadler<sup>c</sup>, S.J. Weatherby<sup>c,\*</sup>, P. Ellis<sup>d</sup>  
 Clinical Neurology and Neurosurgery

Cluster-like headache and a cystic hypothalamic tumour as first presentation of sarcoidosis Cephalalgia April 2013 33: 421-424, first	Headache in patients with pituitary adenoma: Clinical and paraclinical findings Cephalalgia December 2012 32: 1198-1207,
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- Secondary headache in **10% of TACs**  
Hypothalamus, pituitary, posterior fossa regions
- Make sure that diagnosis of a TAC should provoke a workup.

PJ Goadsby Continuum: 2012;18(4) 883-895



**Table 2.12** Differential points among the TACs

Features	Cluster headache	Paroxysmal hemicrania	SUNCT/SUNA
Gender (M/F)	3–6/1	1/1	1.5/1
Pain quality	Stab/sharp/throb/poker	Stab/sharp/throb/poker	Stab/sharp/throb/poker
Severity	Very severe	Severe – very severe	severe
Distribution	$V_1 > C2 > V_2 > V_3$	$V_1 > C2 > V_2 > V_3$	$V_1 > C2 > V_2 > V_3$
Attack frequency	Every other day–8/day	Mean 11; up to 30/day	Mean 100; >100/day
Length	15–180 min	2–30 min	4–240 s
Migraine features			
Nausea	50%	40%	25%
Photo-/phonophobia	65%	65%	25%
Triggers			
Alcohol	Yes	Yes	No
Nitroglycerin	Yes	Yes	No
Cutaneous triggers	No	No	Yes
Agitation/restlessness	90%	80%	65%
Episodic/chronic	9/1	1/2	1/9
Circadian/circannual periodicity	Yes	No	No
Treatment efficacy			
Oxygen	70%	None	None
Sumatriptan	90%	20%	10% or less
subcutaneously			
Indomethacin	None	100%	None

**Table 11.4** Treatment recommendations for cluster headache, paroxysmal hemicrania, and SUNCT syndrome. For exact doses see text. (A denotes effective, B denotes probably effective, C denotes possibly effective)

	Cluster headache	paroxysmal hemicrania	SUNCT syndrome
Attack treatment	oxygen inhalation (A) sumatriptan 6 mg s.c. (A) zolmitriptan 5 mg nasal (A) sumatriptan 20 mg nasal (A) zolmitriptan 10 mg oral (B) lidocaine nasal (B) octreotide (B)	none	none
Prophylactic treatment	verapamil (A) steroids (A) lithium (B) methysergide (B) topiramate (B) ergotamine tartrate (B) valproic acid (C) melatonin (C) baclofen (C)	indomethacin (A) verapamil (C) NSAIDs (C) topiramate (C)	lamotrigine (C)