|  |
| --- |
|  |
| **REVIEW ARTICLE** |
|  |

Jose A, Prasad R S, Pai A. Trigeminal autonomic cephalalgias: The impersonators. Indian J Pain [serial online] 2019 [cited 2021 Apr 1];33:62-6. Available from: <https://www.indianjpain.org/text.asp?2019/33/2/62/264071>

|  |  |  |
| --- | --- | --- |
| **Abstract** |  |  |

Orofacial pain disorders are highly prevalent and debilitating conditions involving the head, face, and neck. Headache is the most common complaint reported to dental and medical practitioners, which manifests as a myriad of neuro-ophthalmologic symptoms, including orbital pain, disturbances of vision, aura, photophobia, lacrimation, conjunctival injection, ptosis, and other manifestations. The differential diagnosis is extensive and includes both primary and secondary headache disorders. The similarity in clinical presentation and diagnostic features is a challenge to the clinicians because of two facts: (a) the orofacial region is complex and (b) pain can arise from many sources. The term “trigeminal autonomic cephalalgias (TACs)” includes a group of headache disorders characterized by moderate-to-severe, short-lived head pain in the trigeminal distribution, with accompanying unilateral cranial parasympathetic autonomic features, such as lacrimation, rhinorrhea, conjunctival injection, eyelid edema, and ptosis. TACs include cluster headache (CH), paroxysmal hemicrania (PH), short-lasting unilateral neuralgiform headache attacks, and hemicrania continua. They are grouped on the basis of their shared clinical features of unilateral headache of varying durations and ipsilateral cranial autonomic symptoms. TACs are rare with the prevalence rate of <1%. The peculiarity of these conditions is their similar clinical presentation and overlapping features. A sound knowledge of TACs is essential for proper diagnosis. Moreover, multidisciplinary approaches for the management of these conditions are strongly recommended. Hence, the aim of this article is to provide an overview of trigeminal autonomic cephalalgias.

**Keywords: Cluster headache, orofacial pain, paroxysmal hemicrania, short-lasting unilateral neuralgiform headache attacks with conjunctival injection and tearing syndrome, trigeminal autonomic cephalalgias**

|  |  |  |
| --- | --- | --- |
| **Introduction** |  | [Top](https://www.indianjpain.org/article.asp?issn=0970-5333;year=2019;volume=33;issue=2;spage=62;epage=66;aulast=Jose#top) |

The word “pain” is most distressing to every individual. The correlation of pain by an individual is always dependent on his/her past experiences and varies from person to person.  
  
Pain is defined as an unpleasant sensory and emotional experience associated with actual or potential tissue damage or described in terms of such damage.[[1]](https://www.indianjpain.org/article.asp?issn=0970-5333;year=2019;volume=33;issue=2;spage=62;epage=66;aulast=Jose" \l "ref1)  
  
Orofacial pain is one of the highly prevalent and debilitating conditions in this modern era. They pose challenges to the treating clinicians because the origin of pain is multifactorial. Headache is the most common complaint reported to dental and medical practitioners. A wide variety of headache disorders manifest as a myriad of neuro-ophthalmologic symptoms, including orbital pain, disturbances of vision, aura, photophobia, lacrimation, conjunctival injection, ptosis, and other manifestations. The differential diagnosis in these patients is extensive and embraces both secondary or symptomatic and primary headache disorders. Awareness of various headaches and their accompanying signs and symptoms is essential to attain the correct diagnosis.[[2]](https://www.indianjpain.org/article.asp?issn=0970-5333;year=2019;volume=33;issue=2;spage=62;epage=66;aulast=Jose" \l "ref2)  
  
The unique feature of vascular pain is its primary pulsatile or throbbing quality.[[3]](https://www.indianjpain.org/article.asp?issn=0970-5333;year=2019;volume=33;issue=2;spage=62;epage=66;aulast=Jose" \l "ref3) The pulsations match the heartbeat and are usually present during exacerbations. The greater the amplitude of vascular dilation, the more pronounced is the throbbing quality of the pain.[[3]](https://www.indianjpain.org/article.asp?issn=0970-5333;year=2019;volume=33;issue=2;spage=62;epage=66;aulast=Jose#ref3),[[4]](https://www.indianjpain.org/article.asp?issn=0970-5333;year=2019;volume=33;issue=2;spage=62;epage=66;aulast=Jose" \l "ref4)  
  
Inflammatory pain, especially those which are confined in such a way that the pulsating vessels in the inflamed area cause noxious stimulation, can have a throbbing quality (pulpitis, osteitis, periostitis, and confined cellulitis). Hence, the throbbing quality of true vascular pain must be distinguished from other throbbing pains.[[3]](https://www.indianjpain.org/article.asp?issn=0970-5333;year=2019;volume=33;issue=2;spage=62;epage=66;aulast=Jose#ref3)  
  
The term “trigeminal autonomic cephalalgias (TACs)” was coined by Goadsby and Lipton. According to the International Classification of Headache Disorders-3 beta (ICHD-3 beta), this group includes cluster headache (CH), paroxysmal hemicrania (PH), and short-lasting unilateral neuralgiform headache attacks (SUNHAs) (including short-lasting unilateral neuralgiform headache attacks with conjunctival injection and tearing [SUNCT] and Shortlasting Unilateral Neuralgiform headache with cranial Autonomic symptoms [SUNAs]) with latest addition of hemicrania continua (HC) to the group (All of others except HC are characterized by short-lasting headaches while HC is characterized by a continuous unilateral headache that varies in its intensity without complete resolution. HC overlaps in the activation of the posterior hypothalamic gray and has similar indomethacin responsiveness with others).[[5]](https://www.indianjpain.org/article.asp?issn=0970-5333;year=2019;volume=33;issue=2;spage=62;epage=66;aulast=Jose" \l "ref5) These conditions seem similar, but they differ in attack duration, frequency, and their response to different treatments. For example, the attack duration of CH is the longest compared to other types. They have some overlapping features, but they are not difficult to recognize and subclassify.[[6]](https://www.indianjpain.org/article.asp?issn=0970-5333;year=2019;volume=33;issue=2;spage=62;epage=66;aulast=Jose" \l "ref6),[[7]](https://www.indianjpain.org/article.asp?issn=0970-5333;year=2019;volume=33;issue=2;spage=62;epage=66;aulast=Jose" \l "ref7),[[8]](https://www.indianjpain.org/article.asp?issn=0970-5333;year=2019;volume=33;issue=2;spage=62;epage=66;aulast=Jose" \l "ref8),[[9]](https://www.indianjpain.org/article.asp?issn=0970-5333;year=2019;volume=33;issue=2;spage=62;epage=66;aulast=Jose" \l "ref9)  
  
TACs are rare with prevalence rate of <1%. CH has a prevalence of 0.01%,[[10]](https://www.indianjpain.org/article.asp?issn=0970-5333;year=2019;volume=33;issue=2;spage=62;epage=66;aulast=Jose" \l "ref10) while PH and SUNHAs have prevalence of 0.009% and 0.006% world wide.[[11]](https://www.indianjpain.org/article.asp?issn=0970-5333;year=2019;volume=33;issue=2;spage=62;epage=66;aulast=Jose" \l "ref11) The exact prevalence rate of HC is still unknown.[[12]](https://www.indianjpain.org/article.asp?issn=0970-5333;year=2019;volume=33;issue=2;spage=62;epage=66;aulast=Jose" \l "ref12)  
  
TACs have common pathophysiology involving trigeminovascular system, trigeminoparasympathetic reflex, and centers controlling circadian rhythms.[[13]](https://www.indianjpain.org/article.asp?issn=0970-5333;year=2019;volume=33;issue=2;spage=62;epage=66;aulast=Jose" \l "ref13),[[14]](https://www.indianjpain.org/article.asp?issn=0970-5333;year=2019;volume=33;issue=2;spage=62;epage=66;aulast=Jose" \l "ref14)  
  
The majority of patients with TACs respond to standard or first-line treatments, as outlined in the international guidelines, with some exceptions that have led to the search for alternative treatments. Standard treatment recommendations for TACs are based on moderate-to-low-quality evidence, with little in the way of randomized controlled trials.[[15]](https://www.indianjpain.org/article.asp?issn=0970-5333;year=2019;volume=33;issue=2;spage=62;epage=66;aulast=Jose" \l "ref15)

|  |  |  |
| --- | --- | --- |
| **Discussion** |  | [Top](https://www.indianjpain.org/article.asp?issn=0970-5333;year=2019;volume=33;issue=2;spage=62;epage=66;aulast=Jose#top) |

The term “trigeminal autonomic cephalalgias (TACs)” includes a group of headache disorders characterized by moderate-to-severe, short-lived head pain in the trigeminal distribution, with accompanying unilateral cranial parasympathetic autonomic features, such as lacrimation, rhinorrhea, conjunctival injection, eyelid edema, and ptosis. The peculiarity of these conditions is their similar clinical presentation and overlapping features. TACs are rare with the prevalence rate of <1%. The TACs include CH, PH, SUNHAs, and HC.  
  
**CH**  
  
CH is a primary headache characterized by recurrent attacks of severe unilateral orbital or supraorbital and/or temporal pain, lasting for 15–180 min.[[16]](https://www.indianjpain.org/article.asp?issn=0970-5333;year=2019;volume=33;issue=2;spage=62;epage=66;aulast=Jose" \l "ref16),[[17]](https://www.indianjpain.org/article.asp?issn=0970-5333;year=2019;volume=33;issue=2;spage=62;epage=66;aulast=Jose" \l "ref17),[[18]](https://www.indianjpain.org/article.asp?issn=0970-5333;year=2019;volume=33;issue=2;spage=62;epage=66;aulast=Jose" \l "ref18) It is divided into episodic and chronic forms with clear male predominance, commonly affecting young adults, with a diminution in symptom severity after 70 years of age and has a mean prevalence rate of 0.1%.[[17]](https://www.indianjpain.org/article.asp?issn=0970-5333;year=2019;volume=33;issue=2;spage=62;epage=66;aulast=Jose#ref17)  
  
People with CH may overindulge in habits such as smoking, alcoholism, and consumption of coffee.[[19]](https://www.indianjpain.org/article.asp?issn=0970-5333;year=2019;volume=33;issue=2;spage=62;epage=66;aulast=Jose" \l "ref19) CH has a circannual and circadian periodicity, in which attacks are clustered (hence the name) in bouts that can occur during specific months of the year. Triggering factors include alcohol, strong odors, and napping.[[16]](https://www.indianjpain.org/article.asp?issn=0970-5333;year=2019;volume=33;issue=2;spage=62;epage=66;aulast=Jose#ref16) The attacks are primarily localized retro-orbitally or periorbitally and are described as stabbing type with severe intensity.[[18]](https://www.indianjpain.org/article.asp?issn=0970-5333;year=2019;volume=33;issue=2;spage=62;epage=66;aulast=Jose#ref18) CH presents as brief, one-sided headache attacks with ipsilateral autonomic symptoms.[[20]](https://www.indianjpain.org/article.asp?issn=0970-5333;year=2019;volume=33;issue=2;spage=62;epage=66;aulast=Jose" \l "ref20) Typical comorbid symptoms include lacrimation from the eye on the affected side, conjunctival reddening, rhinorrhea and/or nasal congestion, miosis, and ptosis.[[19]](https://www.indianjpain.org/article.asp?issn=0970-5333;year=2019;volume=33;issue=2;spage=62;epage=66;aulast=Jose#ref19),[[21]](https://www.indianjpain.org/article.asp?issn=0970-5333;year=2019;volume=33;issue=2;spage=62;epage=66;aulast=Jose" \l "ref21)  
  
The diagnostic criteria for CH given by The ICHD-II are given in [[Table 1]](https://www.indianjpain.org/viewimage.asp?img=IndianJPain_2019_33_2_62_264071_t1.jpg).[[18]](https://www.indianjpain.org/article.asp?issn=0970-5333;year=2019;volume=33;issue=2;spage=62;epage=66;aulast=Jose#ref18)

|  |  |
| --- | --- |
|  | Table 1: Diagnostic criteria for cluster headache given by the International Classification of Headache Disorder-II  [**Click here to view**](https://www.indianjpain.org/viewimage.asp?img=IndianJPain_2019_33_2_62_264071_t1.jpg) |

Various treatment modalities got from various studies,[[22]](https://www.indianjpain.org/article.asp?issn=0970-5333;year=2019;volume=33;issue=2;spage=62;epage=66;aulast=Jose" \l "ref22),[[23]](https://www.indianjpain.org/article.asp?issn=0970-5333;year=2019;volume=33;issue=2;spage=62;epage=66;aulast=Jose" \l "ref23),[[24]](https://www.indianjpain.org/article.asp?issn=0970-5333;year=2019;volume=33;issue=2;spage=62;epage=66;aulast=Jose" \l "ref24),[[25]](https://www.indianjpain.org/article.asp?issn=0970-5333;year=2019;volume=33;issue=2;spage=62;epage=66;aulast=Jose" \l "ref25) are enlisted in [[Table 2]](https://www.indianjpain.org/viewimage.asp?img=IndianJPain_2019_33_2_62_264071_t2.jpg). Apart from these treatment modalities enlisted a short course of Prednisolone (60 mg daily for 7 days followed by rapid taper) is beneficial as preventive treatment in patients with short bouts of CH.

|  |  |
| --- | --- |
|  | Table 2: Treatment modalities for cluster headache  [**Click here to view**](https://www.indianjpain.org/viewimage.asp?img=IndianJPain_2019_33_2_62_264071_t2.jpg) |

**PH**  
  
PH is a rare variety of TACs, characterized by unilateral, severe orbital, or periorbital pain (Which commonly refers to the shoulder, neck, and arm), which lasts for 2–30 min.[[26]](https://www.indianjpain.org/article.asp?issn=0970-5333;year=2019;volume=33;issue=2;spage=62;epage=66;aulast=Jose" \l "ref26) Pain can also be of throbbing, stabbing, sharp, or boring types, accompanied by at least one of the following ipsilateral autonomic features such as conjunctival injection/lacrimation, nasal congestion/rhinorrhea, eyelid edema, forehead/facial sweating, miosis, and ptosis.[[26]](https://www.indianjpain.org/article.asp?issn=0970-5333;year=2019;volume=33;issue=2;spage=62;epage=66;aulast=Jose#ref26),[[27]](https://www.indianjpain.org/article.asp?issn=0970-5333;year=2019;volume=33;issue=2;spage=62;epage=66;aulast=Jose" \l "ref27),[[28]](https://www.indianjpain.org/article.asp?issn=0970-5333;year=2019;volume=33;issue=2;spage=62;epage=66;aulast=Jose" \l "ref28) About 20% of PH behave episodically and eventually develop into a chronic form.[[26]](https://www.indianjpain.org/article.asp?issn=0970-5333;year=2019;volume=33;issue=2;spage=62;epage=66;aulast=Jose#ref26)  
  
The frequency of pain attacks is 8–30 per day.[[26]](https://www.indianjpain.org/article.asp?issn=0970-5333;year=2019;volume=33;issue=2;spage=62;epage=66;aulast=Jose#ref26) Malignancy, central nervous system disease, and benign tumors can cause secondary PH.  
  
Indomethacin 75–225 mg/day (75 mg/day for first 3 days; followed by 150 mg/day for a further 3 days is recommended) is the drug of choice.[[26]](https://www.indianjpain.org/article.asp?issn=0970-5333;year=2019;volume=33;issue=2;spage=62;epage=66;aulast=Jose#ref26) The mechanism of action of indomethacin is poorly understood, and it seems that it is not entirely dependent on inhibition of cyclooxygenase activity. Indomethacin-resistant PH may respond to topiramate. Other alternatives include calcium channel blockers, naproxen, carbamazepine, acetazolamide. Nerve block such as greater occipital nerve block (GONB) is also found to be effective.[[24]](https://www.indianjpain.org/article.asp?issn=0970-5333;year=2019;volume=33;issue=2;spage=62;epage=66;aulast=Jose#ref24),[[27]](https://www.indianjpain.org/article.asp?issn=0970-5333;year=2019;volume=33;issue=2;spage=62;epage=66;aulast=Jose#ref27),[[29]](https://www.indianjpain.org/article.asp?issn=0970-5333;year=2019;volume=33;issue=2;spage=62;epage=66;aulast=Jose" \l "ref29),[[30]](https://www.indianjpain.org/article.asp?issn=0970-5333;year=2019;volume=33;issue=2;spage=62;epage=66;aulast=Jose" \l "ref30)  
  
**SUNHAs**  
  
It clinically presents as unilateral ocular and periocular pain, accompanied by ipsilateral conjunctival injection and lacrimation. Pain is usually stabbing or pulsating and lasts from 5 to 240 s (with frequency of attacks is from 3 to 200 daily). SUNHAs have three patterns of attacks: (a) classical single attacks, (b) groups of a number of stabs/attacks, and (c) “saw tooth” pattern with numerous stabs/attacks lasting minutes.[[26]](https://www.indianjpain.org/article.asp?issn=0970-5333;year=2019;volume=33;issue=2;spage=62;epage=66;aulast=Jose#ref26),[[27]](https://www.indianjpain.org/article.asp?issn=0970-5333;year=2019;volume=33;issue=2;spage=62;epage=66;aulast=Jose#ref27),[[28]](https://www.indianjpain.org/article.asp?issn=0970-5333;year=2019;volume=33;issue=2;spage=62;epage=66;aulast=Jose#ref28)  
  
SUNHAs are categorized into two subtypes: SUNCT and SUNA. Patients with SUNCT exhibit both conjunctival injection and tearing, whereas patients with SUNA have only one or neither of them.[[27]](https://www.indianjpain.org/article.asp?issn=0970-5333;year=2019;volume=33;issue=2;spage=62;epage=66;aulast=Jose#ref27)  
  
SUNCT is a relatively rare TAC marked by short-lasting attacks of lateralized severe head pain associated with prominent cranial autonomic features and often triggered by cutaneous stimulus.[[28]](https://www.indianjpain.org/article.asp?issn=0970-5333;year=2019;volume=33;issue=2;spage=62;epage=66;aulast=Jose#ref28) SUNCT syndrome is a unilateral headache/facial pain characterized by brief paroxysmal attacks accompanied by ipsilateral local autonomic signs, usually conjunctival injection and lacrimation. SUNCT shows similarities to trigeminal neuralgia, particularly in the triggering mechanism.[[26]](https://www.indianjpain.org/article.asp?issn=0970-5333;year=2019;volume=33;issue=2;spage=62;epage=66;aulast=Jose#ref26),[[31]](https://www.indianjpain.org/article.asp?issn=0970-5333;year=2019;volume=33;issue=2;spage=62;epage=66;aulast=Jose" \l "ref31)  
  
Since the episodes of SUNCT/SUNA are of very short duration, thus the concept of acute treatment is not very useful. For long term pharmacological prevention lamotrigine, topiramate, gabapentin or carbamazapine may be advised. Nerve blocks such as GONB; multiple cranial nerve block; and surgical procedures such as microvascular decompression (MVD), occipital nerve stimulation (ONS), and deep brain stimulation are found to be effective.[[24]](https://www.indianjpain.org/article.asp?issn=0970-5333;year=2019;volume=33;issue=2;spage=62;epage=66;aulast=Jose#ref24),[[27]](https://www.indianjpain.org/article.asp?issn=0970-5333;year=2019;volume=33;issue=2;spage=62;epage=66;aulast=Jose#ref27),[[30]](https://www.indianjpain.org/article.asp?issn=0970-5333;year=2019;volume=33;issue=2;spage=62;epage=66;aulast=Jose#ref30)  
  
**HC**  
  
HC is an indomethacin-responsive primary headache disorder. Several studies suggest that HC is common and is probably an underdiagnosed condition. It commonly affects middle-aged people, with female preponderance.[[32]](https://www.indianjpain.org/article.asp?issn=0970-5333;year=2019;volume=33;issue=2;spage=62;epage=66;aulast=Jose" \l "ref32),[[33]](https://www.indianjpain.org/article.asp?issn=0970-5333;year=2019;volume=33;issue=2;spage=62;epage=66;aulast=Jose" \l "ref33)  
  
HC is characterized by a strictly unilateral, continuous headache of moderate intensity, with superimposed exacerbations of severe intensity (associated with cranial autonomic features, restlessness, and migrainous features) [[Table 3]](https://www.indianjpain.org/viewimage.asp?img=IndianJPain_2019_33_2_62_264071_t3.jpg) and [[Table 4]](https://www.indianjpain.org/viewimage.asp?img=IndianJPain_2019_33_2_62_264071_t4.jpg).[[26]](https://www.indianjpain.org/article.asp?issn=0970-5333;year=2019;volume=33;issue=2;spage=62;epage=66;aulast=Jose#ref26),[[27]](https://www.indianjpain.org/article.asp?issn=0970-5333;year=2019;volume=33;issue=2;spage=62;epage=66;aulast=Jose#ref27),[[32]](https://www.indianjpain.org/article.asp?issn=0970-5333;year=2019;volume=33;issue=2;spage=62;epage=66;aulast=Jose#ref32) The pain is dull, but during exacerbation period, it will be either throbbing or stabbing type with severe intensity. Patients may also have nocturnal exacerbations. A sense of agitation or restlessness is another important feature.[[26]](https://www.indianjpain.org/article.asp?issn=0970-5333;year=2019;volume=33;issue=2;spage=62;epage=66;aulast=Jose#ref26),[[27]](https://www.indianjpain.org/article.asp?issn=0970-5333;year=2019;volume=33;issue=2;spage=62;epage=66;aulast=Jose#ref27) HC is also associated with symptoms such as nausea, vomiting, photophobia, and phonophobia. The triggering factors of HC are stress, alcohol, irregular sleep, and menstruation.[[32]](https://www.indianjpain.org/article.asp?issn=0970-5333;year=2019;volume=33;issue=2;spage=62;epage=66;aulast=Jose#ref32),[[34]](https://www.indianjpain.org/article.asp?issn=0970-5333;year=2019;volume=33;issue=2;spage=62;epage=66;aulast=Jose" \l "ref34)

|  |  |
| --- | --- |
|  | Table 3: International Headache Society Diagnostic Criteria for hemicrania continua  [**Click here to view**](https://www.indianjpain.org/viewimage.asp?img=IndianJPain_2019_33_2_62_264071_t3.jpg) |

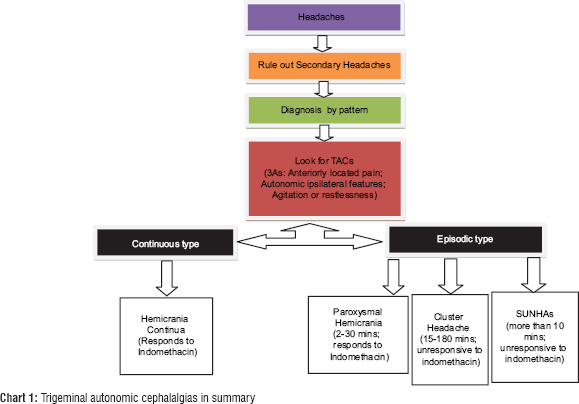
|  |  |
| --- | --- |
|  | Table 4: Diagnostic criteria of different subtypes of hemicrania continua (International Classification of Headache Disorder-3 beta)  [**Click here to view**](https://www.indianjpain.org/viewimage.asp?img=IndianJPain_2019_33_2_62_264071_t4.jpg) |

Indomethacin, supraorbital nerve blockade (SONB), GONB, celecoxib, piroxicam, minor occipital nerve blockade, oxygen, sumatriptan, methylprednisolone, ibuprofen, dorsal root ganglion blockade, sphenopalatine ganglion blockade (SPGB), and ergotamine are the treatment options available for HC.[[35]](https://www.indianjpain.org/article.asp?issn=0970-5333;year=2019;volume=33;issue=2;spage=62;epage=66;aulast=Jose" \l "ref35),[[36]](https://www.indianjpain.org/article.asp?issn=0970-5333;year=2019;volume=33;issue=2;spage=62;epage=66;aulast=Jose" \l "ref36)  
  
Indomethacin is given with an initial dose of 25 mg three times a day and gradually titrated (25 mg tid every 3–5 days) up to 100 mg tid or until the patient gets complete relief. The dose required ranges from 25 to 500 mg/day. The mean indomethacin dose varies between 94 and 176 mg/day.[[32]](https://www.indianjpain.org/article.asp?issn=0970-5333;year=2019;volume=33;issue=2;spage=62;epage=66;aulast=Jose#ref32),[[35]](https://www.indianjpain.org/article.asp?issn=0970-5333;year=2019;volume=33;issue=2;spage=62;epage=66;aulast=Jose#ref35),[[36]](https://www.indianjpain.org/article.asp?issn=0970-5333;year=2019;volume=33;issue=2;spage=62;epage=66;aulast=Jose#ref36) Other alternative drugs are topiramate, rofecoxib, celecoxib, ibuprofen, acetylsalicylic acid, gabapentin, melatonin, piroxicam derivative, amitriptyline, acemetacin, and verapamil. Surgical interventions include peripheral nerve blocks (SONB, GONB, and SPGB), radiofrequency ablation ONS, and vagus nerve stimulation.[[24]](https://www.indianjpain.org/article.asp?issn=0970-5333;year=2019;volume=33;issue=2;spage=62;epage=66;aulast=Jose#ref24),[[28]](https://www.indianjpain.org/article.asp?issn=0970-5333;year=2019;volume=33;issue=2;spage=62;epage=66;aulast=Jose#ref28),[[32]](https://www.indianjpain.org/article.asp?issn=0970-5333;year=2019;volume=33;issue=2;spage=62;epage=66;aulast=Jose#ref32),[[37]](https://www.indianjpain.org/article.asp?issn=0970-5333;year=2019;volume=33;issue=2;spage=62;epage=66;aulast=Jose" \l "ref37),[[38]](https://www.indianjpain.org/article.asp?issn=0970-5333;year=2019;volume=33;issue=2;spage=62;epage=66;aulast=Jose" \l "ref38),[[39]](https://www.indianjpain.org/article.asp?issn=0970-5333;year=2019;volume=33;issue=2;spage=62;epage=66;aulast=Jose" \l "ref39)  
  
The summary of clinical features of various TACs and their various treatment modalities are given in [[Table 5]](https://www.indianjpain.org/viewimage.asp?img=IndianJPain_2019_33_2_62_264071_t5.jpg) and [[Table 6]](https://www.indianjpain.org/viewimage.asp?img=IndianJPain_2019_33_2_62_264071_t6.jpg).

|  |  |
| --- | --- |
|  | Table 5: Summary of clinical features of trigeminal autonomic cephalalgias  [**Click here to view**](https://www.indianjpain.org/viewimage.asp?img=IndianJPain_2019_33_2_62_264071_t5.jpg) |

|  |  |
| --- | --- |
|  | Table 6: Summary of suggested treatment for trigeminal autonomic cephalalgias  [**Click here to view**](https://www.indianjpain.org/viewimage.asp?img=IndianJPain_2019_33_2_62_264071_t6.jpg) |

The Diagnosis flow chart for TACs is given in [Chart 1].



In short, TACs clinically resemble each other which not only throw a challenge to the treating clinician but also cause havoc in attaining the right diagnosis. The unique diagnostic features and treatment response are different for each entities, which will help differentiate and reach a precise diagnosis which forms the basis for the treatment protocol. For example, the duration of pain attacks (starting from continuous nature of HC to CH (up to 180 min), to PH (up to 30 min), to SUNHAs (up to 10 mins) and frequency of pain attacks (starting from less frequent CH (1–8/day), to more frequent SUNHAs (1–200/day), to continuous nature of HC) and the indomethacin response (PH and HC shows complete resolution to indomethacin) will help us to solve the puzzle of diagnosis.  
  
**Financial support and sponsorship**  
  
Nil.  
  
**Conflicts of interest**  
  
There are no conflicts of interest.

|  |  |  |
| --- | --- | --- |
| **References** |  | [Top](https://www.indianjpain.org/article.asp?issn=0970-5333;year=2019;volume=33;issue=2;spage=62;epage=66;aulast=Jose#top) |

|  |  |
| --- | --- |
| [1.](https://www.indianjpain.org/article.asp?issn=0970-5333;year=2019;volume=33;issue=2;spage=62;epage=66;aulast=Jose" \l "ft1) | Radu BM, Bramanti P, Osculati F, Flonta ML, Radu M, Bertini G, *et al.* Neurovascular unit in chronic pain. Mediators Inflamm 2013;2013:648268.  [Back to cited text no. 1](https://www.indianjpain.org/article.asp?issn=0970-5333;year=2019;volume=33;issue=2;spage=62;epage=66;aulast=Jose#ft1) |
| [2.](https://www.indianjpain.org/article.asp?issn=0970-5333;year=2019;volume=33;issue=2;spage=62;epage=66;aulast=Jose" \l "ft2) | Sharav Y, Benoliel R, editors. Orofacial Pain and Headache. Philadelphia, USA: Elsevier Health Sciences; 2008.  [Back to cited text no. 2](https://www.indianjpain.org/article.asp?issn=0970-5333;year=2019;volume=33;issue=2;spage=62;epage=66;aulast=Jose#ft2) |
| [3.](https://www.indianjpain.org/article.asp?issn=0970-5333;year=2019;volume=33;issue=2;spage=62;epage=66;aulast=Jose" \l "ft3) | Okeson JP. Bell's orofacial pains: the clinical management of orofacial pain. Chicago, Ill, USA: Quintessence Publishing Company; 2005.  [Back to cited text no. 3](https://www.indianjpain.org/article.asp?issn=0970-5333;year=2019;volume=33;issue=2;spage=62;epage=66;aulast=Jose#ft3) |
| [4.](https://www.indianjpain.org/article.asp?issn=0970-5333;year=2019;volume=33;issue=2;spage=62;epage=66;aulast=Jose" \l "ft4) | Okeson JP. Bell's orofacial pains: the clinical management of orofacial pain. Chicago, Ill, USA: Quintessence Publishing Company; 2014.  [Back to cited text no. 4](https://www.indianjpain.org/article.asp?issn=0970-5333;year=2019;volume=33;issue=2;spage=62;epage=66;aulast=Jose#ft4) |
| [5.](https://www.indianjpain.org/article.asp?issn=0970-5333;year=2019;volume=33;issue=2;spage=62;epage=66;aulast=Jose" \l "ft5) | Wei DY, Yuan Ong JJ, Goadsby PJ. Overview of trigeminal autonomic cephalalgias: Nosologic evolution, diagnosis, and management. Ann Indian Acad Neurol 2018;21:S39-44.  [Back to cited text no. 5](https://www.indianjpain.org/article.asp?issn=0970-5333;year=2019;volume=33;issue=2;spage=62;epage=66;aulast=Jose#ft5) [[PUBMED](https://www.medknow.com/crt.asp?prn=5;aid=IndianJPain_2019_33_2_62_264071;rt=P;u=http://www.ncbi.nlm.nih.gov/entrez/query.fcgi?cmd=Retrieve&db=PubMed&list_uids=29720817&dopt=Abstract)]  [[Full text]](https://www.medknow.com/crt.asp?prn=5;aid=IndianJPain_2019_33_2_62_264071;rt=F;u=http://www.annalsofian.org/article.asp?issn=0972-2327;year=2018;volume=21;issue=5;spage=39;epage=44;aulast=Wei) |
| [6.](https://www.indianjpain.org/article.asp?issn=0970-5333;year=2019;volume=33;issue=2;spage=62;epage=66;aulast=Jose" \l "ft6) | Ravishankar K. Classification of trigeminal autonomic cephalalgia: What has changed in international classification of headache disorders-3beta? Ann Indian Acad Neurol 2018;21:S45-50.  [Back to cited text no. 6](https://www.indianjpain.org/article.asp?issn=0970-5333;year=2019;volume=33;issue=2;spage=62;epage=66;aulast=Jose#ft6) |
| [7.](https://www.indianjpain.org/article.asp?issn=0970-5333;year=2019;volume=33;issue=2;spage=62;epage=66;aulast=Jose" \l "ft7) | Schwartz DP, Robbins MS. Primary headache disorders and neuro-ophthalmologic manifestations. Eye Brain 2012;4:49-61.  [Back to cited text no. 7](https://www.indianjpain.org/article.asp?issn=0970-5333;year=2019;volume=33;issue=2;spage=62;epage=66;aulast=Jose#ft7) |
| [8.](https://www.indianjpain.org/article.asp?issn=0970-5333;year=2019;volume=33;issue=2;spage=62;epage=66;aulast=Jose" \l "ft8) | Matharu MS, Boes CJ, Goadsby PJ. Management of trigeminal autonomic cephalgias and hemicrania continua. Drugs 2003;63:1637-77.  [Back to cited text no. 8](https://www.indianjpain.org/article.asp?issn=0970-5333;year=2019;volume=33;issue=2;spage=62;epage=66;aulast=Jose#ft8) |
| [9.](https://www.indianjpain.org/article.asp?issn=0970-5333;year=2019;volume=33;issue=2;spage=62;epage=66;aulast=Jose" \l "ft9) | Chowdhury D. Secondary (Symptomatic) trigeminal autonomic cephalalgia. Ann Indian Acad Neurol 2018;21:S57-69.  [Back to cited text no. 9](https://www.indianjpain.org/article.asp?issn=0970-5333;year=2019;volume=33;issue=2;spage=62;epage=66;aulast=Jose#ft9) [[PUBMED](https://www.medknow.com/crt.asp?prn=9;aid=IndianJPain_2019_33_2_62_264071;rt=P;u=http://www.ncbi.nlm.nih.gov/entrez/query.fcgi?cmd=Retrieve&db=PubMed&list_uids=29720820&dopt=Abstract)]  [[Full text]](https://www.medknow.com/crt.asp?prn=9;aid=IndianJPain_2019_33_2_62_264071;rt=F;u=http://www.annalsofian.org/article.asp?issn=0972-2327;year=2018;volume=21;issue=5;spage=57;epage=69;aulast=Chowdhury) |
| [10.](https://www.indianjpain.org/article.asp?issn=0970-5333;year=2019;volume=33;issue=2;spage=62;epage=66;aulast=Jose" \l "ft10) | May A, Swanson JW, Dashe JF. Cluster headache: Epidemiology, clinical features, and diagnosis. ClusterHeadaches.2009.  [Back to cited text no. 10](https://www.indianjpain.org/article.asp?issn=0970-5333;year=2019;volume=33;issue=2;spage=62;epage=66;aulast=Jose#ft10) |
| [11.](https://www.indianjpain.org/article.asp?issn=0970-5333;year=2019;volume=33;issue=2;spage=62;epage=66;aulast=Jose" \l "ft11) | Garza I, Schwedt T. Hemicrania Continua. UpToDate; 2011.  [Back to cited text no. 11](https://www.indianjpain.org/article.asp?issn=0970-5333;year=2019;volume=33;issue=2;spage=62;epage=66;aulast=Jose#ft11) |
| [12.](https://www.indianjpain.org/article.asp?issn=0970-5333;year=2019;volume=33;issue=2;spage=62;epage=66;aulast=Jose" \l "ft12) | Benoliel R, Sharav Y, Haviv Y, Almoznino G. Tic, triggering, and tearing: From CTN to SUNHA. Headache 2017;57:997-1009.  [Back to cited text no. 12](https://www.indianjpain.org/article.asp?issn=0970-5333;year=2019;volume=33;issue=2;spage=62;epage=66;aulast=Jose#ft12) |
| [13.](https://www.indianjpain.org/article.asp?issn=0970-5333;year=2019;volume=33;issue=2;spage=62;epage=66;aulast=Jose" \l "ft13) | Obermann M, Holle D, Nagel S. Functional neuroimaging in trigeminal autonomic cephalalgias. Ann Indian Acad Neurol 2018;21:S51-6.  [Back to cited text no. 13](https://www.indianjpain.org/article.asp?issn=0970-5333;year=2019;volume=33;issue=2;spage=62;epage=66;aulast=Jose#ft13) |
| [14.](https://www.indianjpain.org/article.asp?issn=0970-5333;year=2019;volume=33;issue=2;spage=62;epage=66;aulast=Jose" \l "ft14) | Benoliel R. Trigeminal autonomic cephalgias. Br J Pain 2012;6:106-23.  [Back to cited text no. 14](https://www.indianjpain.org/article.asp?issn=0970-5333;year=2019;volume=33;issue=2;spage=62;epage=66;aulast=Jose#ft14) |
| [15.](https://www.indianjpain.org/article.asp?issn=0970-5333;year=2019;volume=33;issue=2;spage=62;epage=66;aulast=Jose" \l "ft15) | May A, Leone M, Afra J, Linde M, Sándor PS, Evers S, *et al.* EFNS guidelines on the treatment of cluster headache and other trigeminal-autonomic cephalalgias. Eur J Neurol 2006;13:1066-77.  [Back to cited text no. 15](https://www.indianjpain.org/article.asp?issn=0970-5333;year=2019;volume=33;issue=2;spage=62;epage=66;aulast=Jose#ft15) |
| [16.](https://www.indianjpain.org/article.asp?issn=0970-5333;year=2019;volume=33;issue=2;spage=62;epage=66;aulast=Jose" \l "ft16) | Miller S, Matharu M. Trigeminal autonomic cephalalgias: Beyond the conventional treatments. Curr Pain Headache Rep 2014;18:438.  [Back to cited text no. 16](https://www.indianjpain.org/article.asp?issn=0970-5333;year=2019;volume=33;issue=2;spage=62;epage=66;aulast=Jose#ft16) |
| [17.](https://www.indianjpain.org/article.asp?issn=0970-5333;year=2019;volume=33;issue=2;spage=62;epage=66;aulast=Jose" \l "ft17) | Leroux E, Ducros A. Cluster headache. Orphanet J Rare Dis 2008;3:20.  [Back to cited text no. 17](https://www.indianjpain.org/article.asp?issn=0970-5333;year=2019;volume=33;issue=2;spage=62;epage=66;aulast=Jose#ft17) |
| [18.](https://www.indianjpain.org/article.asp?issn=0970-5333;year=2019;volume=33;issue=2;spage=62;epage=66;aulast=Jose" \l "ft18) | Costa A, Antonaci F, Ramusino MC, Nappi G. The neuropharmacology of cluster headache and other trigeminal autonomic cephalalgias. Curr Neuropharmacol 2015;13:304-23.  [Back to cited text no. 18](https://www.indianjpain.org/article.asp?issn=0970-5333;year=2019;volume=33;issue=2;spage=62;epage=66;aulast=Jose#ft18) |
| [19.](https://www.indianjpain.org/article.asp?issn=0970-5333;year=2019;volume=33;issue=2;spage=62;epage=66;aulast=Jose" \l "ft19) | Gaul C, Diener HC, Müller OM. Cluster headache: Clinical features and therapeutic options. Dtsch Arztebl Int 2011;108:543-9.  [Back to cited text no. 19](https://www.indianjpain.org/article.asp?issn=0970-5333;year=2019;volume=33;issue=2;spage=62;epage=66;aulast=Jose#ft19) |
| [20.](https://www.indianjpain.org/article.asp?issn=0970-5333;year=2019;volume=33;issue=2;spage=62;epage=66;aulast=Jose" \l "ft20) | Tyagi A, Matharu M. Evidence base for the medical treatments used in cluster headache. Curr Pain Headache Rep 2009;13:168-78.  [Back to cited text no. 20](https://www.indianjpain.org/article.asp?issn=0970-5333;year=2019;volume=33;issue=2;spage=62;epage=66;aulast=Jose#ft20) |
| [21.](https://www.indianjpain.org/article.asp?issn=0970-5333;year=2019;volume=33;issue=2;spage=62;epage=66;aulast=Jose" \l "ft21) | Iacovelli E, Coppola G, Tinelli E, Pierelli F, Bianco F. Neuroimaging in cluster headache and other trigeminal autonomic cephalalgias. J Headache Pain 2012;13:11-20.  [Back to cited text no. 21](https://www.indianjpain.org/article.asp?issn=0970-5333;year=2019;volume=33;issue=2;spage=62;epage=66;aulast=Jose#ft21) |
| [22.](https://www.indianjpain.org/article.asp?issn=0970-5333;year=2019;volume=33;issue=2;spage=62;epage=66;aulast=Jose" \l "ft22) | Wolter T, Kaube H. Neurostimulation for chronic cluster headache. Ther Adv Neurol Disord 2012;5:175-80.  [Back to cited text no. 22](https://www.indianjpain.org/article.asp?issn=0970-5333;year=2019;volume=33;issue=2;spage=62;epage=66;aulast=Jose#ft22) |
| [23.](https://www.indianjpain.org/article.asp?issn=0970-5333;year=2019;volume=33;issue=2;spage=62;epage=66;aulast=Jose" \l "ft23) | Gooriah R, Buture A, Ahmed F. Evidence-based treatments for cluster headache. Ther Clin Risk Manag 2015;11:1687-96.  [Back to cited text no. 23](https://www.indianjpain.org/article.asp?issn=0970-5333;year=2019;volume=33;issue=2;spage=62;epage=66;aulast=Jose#ft23) |
| [24.](https://www.indianjpain.org/article.asp?issn=0970-5333;year=2019;volume=33;issue=2;spage=62;epage=66;aulast=Jose" \l "ft24) | Wei DY, Jensen RH. Therapeutic approaches for the management of trigeminal autonomic cephalalgias. Neurotherapeutics 2018;15:346-60.  [Back to cited text no. 24](https://www.indianjpain.org/article.asp?issn=0970-5333;year=2019;volume=33;issue=2;spage=62;epage=66;aulast=Jose#ft24) |
| [25.](https://www.indianjpain.org/article.asp?issn=0970-5333;year=2019;volume=33;issue=2;spage=62;epage=66;aulast=Jose" \l "ft25) | Di Sabato F, Giacovazzo M. Management of cluster headache in the emergency department. J Headache Pain 2005;6:294-7.  [Back to cited text no. 25](https://www.indianjpain.org/article.asp?issn=0970-5333;year=2019;volume=33;issue=2;spage=62;epage=66;aulast=Jose#ft25) |
| [26.](https://www.indianjpain.org/article.asp?issn=0970-5333;year=2019;volume=33;issue=2;spage=62;epage=66;aulast=Jose" \l "ft26) | Kruszewski P, Pareja JA, Caminero AB, Sjaastad O. Cluster headache and SUNCT: Similarities and differences. J Headache Pain 2001;2:57.  [Back to cited text no. 26](https://www.indianjpain.org/article.asp?issn=0970-5333;year=2019;volume=33;issue=2;spage=62;epage=66;aulast=Jose#ft26) |
| [27.](https://www.indianjpain.org/article.asp?issn=0970-5333;year=2019;volume=33;issue=2;spage=62;epage=66;aulast=Jose" \l "ft27) | Rossi P, Tassorelli C, Allena M, Ferrante E, Lisotto C, Nappi G. Focus on therapy: Hemicrania continua and new daily persistent headache. J Headache Pain 2010;11:259-65.  [Back to cited text no. 27](https://www.indianjpain.org/article.asp?issn=0970-5333;year=2019;volume=33;issue=2;spage=62;epage=66;aulast=Jose#ft27) |
| [28.](https://www.indianjpain.org/article.asp?issn=0970-5333;year=2019;volume=33;issue=2;spage=62;epage=66;aulast=Jose" \l "ft28) | Goadsby PJ. Trigeminal autonomic cephalalgias. Continuum (Minneap Minn) 2012;18:883-95.  [Back to cited text no. 28](https://www.indianjpain.org/article.asp?issn=0970-5333;year=2019;volume=33;issue=2;spage=62;epage=66;aulast=Jose#ft28) |
| [29.](https://www.indianjpain.org/article.asp?issn=0970-5333;year=2019;volume=33;issue=2;spage=62;epage=66;aulast=Jose" \l "ft29) | Miller S, Sinclair AJ, Davies B, Matharu M. Neurostimulation in the treatment of primary headaches. Pract Neurol 2016;16:362-75.  [Back to cited text no. 29](https://www.indianjpain.org/article.asp?issn=0970-5333;year=2019;volume=33;issue=2;spage=62;epage=66;aulast=Jose#ft29) |
| [30.](https://www.indianjpain.org/article.asp?issn=0970-5333;year=2019;volume=33;issue=2;spage=62;epage=66;aulast=Jose" \l "ft30) | Ahmed F. Headache disorders: Differentiating and managing the common subtypes. Br J Pain 2012;6:124-32.  [Back to cited text no. 30](https://www.indianjpain.org/article.asp?issn=0970-5333;year=2019;volume=33;issue=2;spage=62;epage=66;aulast=Jose#ft30) |
| [31.](https://www.indianjpain.org/article.asp?issn=0970-5333;year=2019;volume=33;issue=2;spage=62;epage=66;aulast=Jose" \l "ft31) | Sjaastad O. SUNCT syndrome: The materialization of a headache syndrome. Clin Ophthalmol 2008;2:533-43.  [Back to cited text no. 31](https://www.indianjpain.org/article.asp?issn=0970-5333;year=2019;volume=33;issue=2;spage=62;epage=66;aulast=Jose#ft31) |
| [32.](https://www.indianjpain.org/article.asp?issn=0970-5333;year=2019;volume=33;issue=2;spage=62;epage=66;aulast=Jose" \l "ft32) | Romero-Reyes M, Uyanik JM. Orofacial pain management: Current perspectives. J Pain Res 2014;7:99-115.  [Back to cited text no. 32](https://www.indianjpain.org/article.asp?issn=0970-5333;year=2019;volume=33;issue=2;spage=62;epage=66;aulast=Jose#ft32) |
| [33.](https://www.indianjpain.org/article.asp?issn=0970-5333;year=2019;volume=33;issue=2;spage=62;epage=66;aulast=Jose" \l "ft33) | Prakash S, Patel P. Hemicrania continua: Clinical review, diagnosis and management. J Pain Res 2017;10:1493-509.  [Back to cited text no. 33](https://www.indianjpain.org/article.asp?issn=0970-5333;year=2019;volume=33;issue=2;spage=62;epage=66;aulast=Jose#ft33) |
| [34.](https://www.indianjpain.org/article.asp?issn=0970-5333;year=2019;volume=33;issue=2;spage=62;epage=66;aulast=Jose" \l "ft34) | Viana M, Tassorelli C, Allena M, Nappi G, Sjaastad O, Antonaci F. Diagnostic and therapeutic errors in trigeminal autonomic cephalalgias and hemicrania continua: A systematic review. J Headache Pain. 2013;14:14.  [Back to cited text no. 34](https://www.indianjpain.org/article.asp?issn=0970-5333;year=2019;volume=33;issue=2;spage=62;epage=66;aulast=Jose#ft34) |
| [35.](https://www.indianjpain.org/article.asp?issn=0970-5333;year=2019;volume=33;issue=2;spage=62;epage=66;aulast=Jose" \l "ft35) | Argoff CE, Dubin A, Pilitsis J. Pain Management Secrets E-Book. New York, USA: Elsevier Health Sciences; 2017.  [Back to cited text no. 35](https://www.indianjpain.org/article.asp?issn=0970-5333;year=2019;volume=33;issue=2;spage=62;epage=66;aulast=Jose#ft35) |
| [36.](https://www.indianjpain.org/article.asp?issn=0970-5333;year=2019;volume=33;issue=2;spage=62;epage=66;aulast=Jose" \l "ft36) | Baraldi C, Pellesi L, Guerzoni S, Cainazzo MM, Pini LA. Therapeutical approaches to paroxysmal hemicrania, hemicrania continua and short lasting unilateral neuralgiform headache attacks: A critical appraisal. J Headache Pain 2017;18:71.  [Back to cited text no. 36](https://www.indianjpain.org/article.asp?issn=0970-5333;year=2019;volume=33;issue=2;spage=62;epage=66;aulast=Jose#ft36) |
| [37.](https://www.indianjpain.org/article.asp?issn=0970-5333;year=2019;volume=33;issue=2;spage=62;epage=66;aulast=Jose" \l "ft37) | Lund JP, Lavigne GJ, Dubner R, Sessle BJ. Orofacial pain: from basic science to clinical management. Chicago: Quintessence. 2001:300.  [Back to cited text no. 37](https://www.indianjpain.org/article.asp?issn=0970-5333;year=2019;volume=33;issue=2;spage=62;epage=66;aulast=Jose#ft37) |
| [38.](https://www.indianjpain.org/article.asp?issn=0970-5333;year=2019;volume=33;issue=2;spage=62;epage=66;aulast=Jose" \l "ft38) | Leone M, Franzini A, Proietti Cecchini A, Mea E, Broggi G, Bussone G. Deep brain stimulation in trigeminal autonomic cephalalgias. Neurotherapeutics 2010;7:220-8.  [Back to cited text no. 38](https://www.indianjpain.org/article.asp?issn=0970-5333;year=2019;volume=33;issue=2;spage=62;epage=66;aulast=Jose#ft38) |
| [39.](https://www.indianjpain.org/article.asp?issn=0970-5333;year=2019;volume=33;issue=2;spage=62;epage=66;aulast=Jose" \l "ft39) | Zakrzewska JM. Differential diagnosis of facial pain and guidelines for management. Br J Anaesth 2013;111:95-104.  [Back to cited text no. 39](https://www.indianjpain.org/article.asp?issn=0970-5333;year=2019;volume=33;issue=2;spage=62;epage=66;aulast=Jose#ft39) |