Headache Medicine

What's new in SUNCT and SUNA? An umbrella review of the last 5 years

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Introduction
Short-lasting unilateral neuralgiform headache attacks with conjunctival injection and tearing (SUNCT) and short-lasting unilateral neuralgiform headache with autonomic symptoms (SUNA) are trigemino-autonomic headaches characterized by rapid episodes of pain associated with autonomic symptoms. They are rare headaches with limited literature available.

Objective
To do an overview of reviews on SUNCT and SUNA from the last 5 years to update information on epidemiology, pathophysiology, and treatment.

Methods
A literature search was conducted using the DeCS/MeSH descriptors "Short-lasting unilateral neuralgiform headache attacks with conjunctival injection and tearing," "SUNCT," "short-lasting unilateral neuralgiform headache with autonomic symptoms," and "SUNA." Boolean operators "OR" were used for the acronyms and "AND" between the rest in the PubMed database. Inclusion criteria for articles were reviews published in English, between 2019 and 2023.

Results
A total of 31 articles were found according to the search, of which 11 met the inclusion criteria. Regarding epidemiology, SUNCT/SUNA has an estimated prevalence ranging from 6.6 to 100 cases per 100,000, with an estimated annual incidence of 1.2 cases per 100,000. The onset age varies between 40-70 years, with an average age of 48, and rare pediatric cases. The male-to-female ratio is 1.5:1. Approximately 90% of SUNCT/SUNA patients have the chronic form of the disease, with attacks lasting for more than 1 year without remission periods or with remission periods lasting less than 1 month.
Concerning pathophysiology, it is not fully understood but involves dysfunction that, possibly related to variations in nociceptive receptors and abnormal production of substance P and CGRP, make the trigeminovascular system more sensitive to painful stimuli. These abnormal stimuli are transmitted to the trigeminal ganglion and the trigeminal caudate nucleus in the medulla, forming the trigeminocervical complex. This abnormal transmission intensifies signaling in the pain processing pathway and activates the trigeminal reflex, leading to stimulation of the facial parasympathetic system and the emergence of autonomic symptoms. Additionally, the hypothalamus, especially its posterior region mediated by orexin, can modulate both systems and their interaction, influencing the development of an attack.
Regarding treatment, first-line drugs for prophylactic treatment are antiepileptics, especially lamotrigine, which has a favorable response rate of up to 64% and is safe during pregnancy.Topiramate and carbamazepine were effective in reducing pain in a significant number of patients.For acute pain episodes, the preferred drug is intravenous lidocaine, which has shown pain-free rates of up to 80%. In terms of surgical treatments, microvascular decompression of the trigeminal nerve (MVD) is the only open surgical option that has been effective in SUNCT/SUNA patients but should only be considered in cases with evidence of vascular compression. Neuromodulation treatments have shown promising results, such as deep brain stimulation (DBS) in the posterior hypothalamus area and occipital nerve stimulation.

Conclusion
SUNCT/SUNA primarily affects men over 40 years of age and has a chronic nature. Its pathophysiology is not well understood but involves aberrant activations of pain pathways and the autonomic system mediated by a dysfunctional hypothalamus. Treatment is based on prophylaxis of attacks, especially with lamotrigine, and new methods of neuromodulation are emerging, showing encouraging outcomes.These conditions have limited literature available, particularly in terms of epidemiological studies and double-blind treatment studies, highlighting the need for further research.

Keywords: Umbrella Review; SUNCT; SUNA.