Cluster headache: Epidemiology, clinical features, and diagnosis

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INTRODUCTION — Cluster headache belongs to a group of idiopathic headache entities, the trigeminal autonomic cephalalgias (TACs), all of which involve short-lasting, unilateral, severe headache attacks and typical accompanying autonomic symptoms [1]. Cluster headache is the most prominent of these entities. The concept of the TACs is useful for clinicians seeking a pathophysiologic understanding of the primary neurovascular headaches and a rational therapeutic approach to treating or preventing these headaches. (See "Pathophysiology of the trigeminal autonomic cephalalgias").

This review focuses on the epidemiology, clinical features, and diagnosis of cluster headache. Treatment is discussed separately. (See "Cluster headache: Acute and preventive treatment").

PATHOPHYSIOLOGY — The pathogenesis of cluster headache is complex and remains incompletely understood. The most widely accepted theory is that primary cluster headache is characterized by hypothalamic activation with secondary activation of the trigeminal-autonomic reflex, probably via a trigeminal-hypothalamic pathway (show figure 1). Another theory holds that neurogenic inflammation of the walls of the cavernous sinus obliterates venous outflow and thus injures the traversing sympathetic fibers of the intracranial internal carotid artery and its branches. (See "Pathophysiology of the trigeminal autonomic cephalalgias").

EPIDEMIOLOGY — The prevalence of cluster headache is <1 percent and mostly affects men [2-5]. In a meta-analysis of 16 population-based epidemiologic studies, the following observations were reported [5]:

- The lifetime prevalence of cluster headache for adults of all ages was 124 per 100,000 (95% CI 101-154), or approximately 0.1 percent
- The one year prevalence of cluster headache was 53 per 100,000 (95% CI 26-95)
- The overall male to female ratio was 4.3:1

There is some evidence that the male preponderance in cluster headache is decreasing, particularly in patients with headache onset after 1960 or 1970 [6-8]. The probable explanation is improved understanding of the pathophysiology of this syndrome and consequently a higher acceptance and awareness leading to more frequent diagnosis. However, this finding has not been confirmed in other studies [3,5].

Some studies have noted an apparently high incidence of head trauma in patients with cluster headache. As an example, one study of 374 patients found a history of head injury prior to

cluster headache onset in 15 percent [9]. However, such an association does not establish a cause-and-effect relationship. In addition, the average time between head injury and cluster headache onset (10 years) seems too long to support a causative role.

Genetics — Before 1990, cluster headache was not thought of as an inherited disorder. However, the importance of considering genetic factors in the etiology is highlighted by the following observations:

- In a report of monozygotic twins, both had cluster headache [10].
- Epidemiologic studies have reported a family history of cluster headache in 5 to 20 percent of patients with cluster headache [3,11-13]. Compared with the general population, the risk of cluster headache for first-degree relatives was increased by 14 to 39-fold, and for second-degree relatives by two to eight-fold [11-13].

The evidence of increased familial risk supports the hypothesis that cluster headache has a genetic component, at least in some families.

Results of a complex segregation analysis in one study suggested that an autosomal dominant gene may play a role in cluster headache inheritance in some families [14], although there is also evidence for autosomal recessive or multifactorial inheritance in others [2]. Future studies must take into account that cluster headache can start between the age of 7 [15] and 83 [16] and that the distinction between affected and unaffected individuals is clearly provisional.

Another issue concerns evidence that up to 85 percent of patients with cluster headache are also chronic cigarette smokers [9]. Quitting smoking has no effect on the disease. However, smoking may be a risk factor for the development of cluster headache, possibly on the basis of a genetic predisposition [17]. A study of clinically-detected cluster headache showed a marked decline in the incidence of cluster headaches between 1979-81 and 1990-91 in Olmsted County, Minnesota, during a time when the incidence of smoking declined in the population [18].

CLINICAL FEATURES — Cluster headache is characterized by attacks of severe orbital, supraorbital, or temporal pain, accompanied by autonomic phenomena [1]. The stereotypical attacks may strike up to eight times a day and are relatively short-lived. Cluster headache is strictly unilateral, and the symptoms remain on the same side of the head during a single cluster attack. However, the symptoms can switch to the other side during a different cluster attack (so-called side shift) in approximately 15 percent of cases [19].

In contrast to migraineurs, patients with cluster are restless and prefer to pace about or sit and rock back and forth. The attacks of cluster headache can be so vicious that patients may commit suicide if the disease is not diagnosed or treated [20].

Autonomic symptoms — The unilateral autonomic symptoms, such as ptosis, miosis, lacrimation, conjunctival injection, rhinorrhea, and nasal congestion, occur only during the pain attack and are ipsilateral to the pain. These symptoms are indicative of both parasympathetic hyperactivity and sympathetic impairment. In some patients, the signs of sympathetic paralysis (miosis and ptosis) persist indefinitely [21], but intensify during attacks. Sweating and cutaneous blood flow also increase on the painful side, particularly in areas of sympathetic deficit [22,23].

About 3 percent of all patients lack autonomic symptoms [24]. In rare cases, sympathetic

disturbances persist on the previously-affected side of the face in patients whose cluster headache has switched sides [25].

Circadian periodicity — Another clinical landmark of the cluster headache syndrome is the circadian rhythmicity of the relatively short-lived (15 to 180 minutes) painful attacks. In the episodic form, attacks occur daily for some weeks followed by a period of remission. In the chronic form, attacks occur without significant periods of remission. On average, a cluster period lasts 6 to 12 weeks while remissions can last up to 12 months or longer.

The episodic form is the most common, affecting 80 to 90 percent of patients with cluster headache. It is characterized by periods of attacks (clusters or bouts) and periods of remission. In a bout, patients may experience one to eight attacks per day, and bouts may last from seven days to 12 months [1]. When not in a bout, patients are usually asymptomatic.

The chronic form of cluster headache lacks remissions and is diagnosed after a year without remission, or if remission has lasted less than 30 days. Chronic cluster may arise de novo (primary chronic cluster headache) or evolve from the episodic type (secondary chronic cluster headache).

DIAGNOSIS — Cluster headache, in its typical form, is unmistakable. The diagnosis is exclusively a clinical task based upon a compatible history and diagnostic criteria from the second edition of the International Classification of Headache Disorders (ICHD-2) [1].

No single instrumental examination is able to define, ensure, or differentiate idiopathic headache syndromes [26]. Nevertheless, neuroimaging is suggested to exclude a cranial lesion in patients with suspected cluster headache. (See "Secondary cluster headache" below).

Diagnostic criteria — For the diagnosis of cluster headache, the ICHD-2 requires at least five headache attacks fulfilling the following criteria (show table 2) [1]:

- Severe or very severe unilateral orbital, supraorbital, and/or temporal headache attacks, which last untreated for 15 to 180 minutes. During part (but less than half) of the time course of the cluster headache, attacks may be less severe, less frequent, or of shorter or longer duration.
- The headache is accompanied by at least one of the following symptoms:
  - Ipsilateral conjunctival injection or lacrimation
  - Ipsilateral nasal congestion and/or rhinorrhea
  - Ipsilateral eyelid edema
  - Ipsilateral forehead and facial sweating
  - Ipsilateral miosis and/or ptosis
  - A sense of restlessness and agitation
- The attacks have a frequency from one every other day to eight per day.
- The history and physical and neurologic examinations do not suggest any other disorder, and/or such a disorder is ruled out by appropriate investigations, or such disorder is present but attacks do not occur for the first time in close temporal relation to the disorder.

Attacks fulfilling all but one criteria for cluster headache are diagnosed as probable cluster headache.

The ICHD-2 diagnostic criteria for episodic and chronic cluster headache are as follows [1]:

- Episodic cluster headache is diagnosed when at least two cluster periods lasting seven days to one year are separated by pain free periods lasting one month or longer.
- Chronic cluster headache is diagnosed when attacks occur for more than one year without remission or with remission for less than one month.

Neuroimaging — Occasionally, patients with atypical or even typical clinical features of cluster headache are found to have a potential secondary cause for headache, such as a structural brain lesion. (See "Secondary cluster headache" below).

Thus, for the initial diagnosis of patients with suspected cluster headache, including those with typical features, we suggest neuroimaging with a cranial CT scan or a cranial MRI study to exclude abnormalities of the brain and pituitary gland.

We recommend neuroimaging for patients with an abnormal neurologic examination or those with clinical suspicion for a pituitary abnormality.

Laboratory investigations — Other laboratory examinations, including electrophysiologic testing (eg, evoked potential, electroencephalography) and examination of the cerebrospinal fluid, are not helpful.

DIFFERENTIAL DIAGNOSIS — The differential diagnosis of cluster headache involves consideration of headache syndromes that manifest with strictly unilateral, brief but frequent attacks (show table 1). Such syndromes include the following:

- Paroxysmal hemicrania
- Short-lasting unilateral neuralgiform headache with conjunctival injection and tearing (SUNCT syndrome)
- Trigeminal neuralgia (see "Trigeminal neuralgia")
- Primary stabbing headache (see "Primary stabbing headache")
- Headache associated with an underlying intracranial lesion (ie, secondary cluster headache)

As noted earlier, cluster headache, paroxysmal hemicrania and SUNCT syndrome belong to a group of idiopathic headache entities, the trigeminal autonomic cephalalgias (TACs), that involve activation of trigeminovascular nociceptive pathways and reflex cranial autonomic activation [1]. All of the TACs have two features in common:

- Brief, unilateral, severe headache attacks
- Typical autonomic accompanying symptoms

The TACs differ in attack duration, frequency, and rhythmicity [27] and in the intensity of pain and autonomic symptoms (show table 1). They also differ with regard to treatment options.

Paroxysmal hemicrania — Paroxysmal hemicrania is a rare condition with a female predominance. It is estimated that the paroxysmal hemicranias comprise about 3 to 6 percent of all the trigeminal autonomic cephalalgias. The headache usually starts between the ages of

20 and 40. (See "Paroxysmal hemicrania: Clinical features and diagnosis").

The paroxysmal headache attacks, the character and localization of the pain, and the autonomic symptoms are very similar to those observed in cluster headache (show table 1). However, typical attacks of paroxysmal hemicrania are shorter (2 to 30 minutes) and more frequent (more than five attacks per day) than cluster attacks. In addition, the autonomic symptoms are often less severe than in cluster headache. About 80 percent of patients with paroxysmal hemicrania have the chronic form, while the remaining 20 percent have the episodic form. (See "Paroxysmal hemicrania: Clinical features and diagnosis").

The most important criterion for the diagnosis of paroxysmal hemicrania is the complete response to indomethacin (show table 4). Within one week (often within three days) after the initiation of indomethacin at an adequate dose, the attacks disappear and this effect is maintained long-term. (See "Paroxysmal hemicrania: Clinical features and diagnosis", section on , and see "Paroxysmal hemicrania: Treatment and prognosis").

SUNCT syndrome — The name of this syndrome (short-lasting unilateral neuralgiform headache attacks with conjunctival injection and tearing) describes its typical clinical features. SUNCT syndrome is rare, although its true frequency is unclear. SUNCT may be a subset of another proposed headache called short-lasting unilateral neuralgiform headache attacks with cranial autonomic symptoms (SUNA). (See "SUNCT and SUNA headache syndromes: Clinical features and diagnosis").

SUNCT syndrome is characterized by very short (5 to 240 seconds) attacks with neuralgiform pain quality and severe intensity. The attacks occur at a frequency of 60 (3 to 200) per day on average, are strictly unilateral (periorbital), and are often triggered by touching, speaking, or chewing. When triggered, SUNCT attacks generally do not have a refractory period. The autonomic symptoms are mostly restricted to lacrimation and conjunctival injection. The diagnostic criteria are listed (show table 3). Distinct episodic and chronic forms of SUNCT syndrome are yet to be recognized in formal classifications, but both types occur. (See "SUNCT and SUNA headache syndromes: Clinical features and diagnosis").

The most important differential diagnosis for SUNCT is classic trigeminal neuralgia. In trigeminal neuralgia, unlike in SUNCT syndrome, autonomic symptoms are not prominent and triggered attacks have a clear refractory period [28]. (See "Trigeminal neuralgia").

Secondary cluster headache — Associated cranial lesions have been reported in patients with clinical attacks that resemble cluster headache, although a causal relationship is often uncertain. One review identified 31 reports of patients with a clinical picture suggesting one of the trigeminal autonomic cephalalgias, including 21 thought to have cluster headache, who turned out to have an associated structural lesion [29]. These lesions included the following:

- Intracranial large artery aneurysms
- Meningiomas
- Brain arteriovenous malformations
- Pituitary macroadenomas
- Recurrent nasopharyngeal carcinoma
- Metallic foreign body in the maxillary sinus
- Aspergilloma in sphenoid sinus
- Benign posterior fossa tumor
- Cavernous hemangioma

In each case, the headache improved after treatment of the lesion, suggesting but not proving a causal role for the lesion [29]. Given that the headaches were often without atypical features, the results of this study suggest that all patients with symptoms of cluster headaches should have neuroimaging to exclude a cranial lesion.

SUMMARY AND RECOMMENDATIONS

- The pathogenesis of cluster headache is complex and remains incompletely understood. The most widely accepted theory is that primary cluster headache is characterized by hypothalamic activation with secondary activation of the trigeminal-autonomic reflex, probably via a trigeminal-hypothalamic pathway (show figure 1). (See "Pathophysiology" above).

- The lifetime prevalence of cluster headache is <1 percent, and the disorder mainly affects men. (See "Epidemiology" above).

- Accumulating evidence supports the hypothesis that cluster headache has a genetic component, at least in some families. (See "Genetics" above).

- Cluster headache is characterized by attacks of severe unilateral orbital, supraorbital, or temporal pain, accompanied by autonomic phenomena. Unilateral autonomic symptoms are ipsilateral to the pain and may include ptosis, miosis, lacrimation, conjunctival injection, rhinorrhea, and nasal congestion. Attacks usually last 15 to 180 minutes. In the episodic form, attacks occur daily, usually one to eight times a day for some weeks, followed by a period of remission. The chronic form of cluster headache lacks sustained remissions. (See "Clinical features" above).

- Cluster headache, in its typical form, is unmistakable. The diagnosis is exclusively a clinical one (show table 2). (See "Diagnosis" above).

- Occasionally, patients with atypical or even typical clinical features of cluster headache are found to have a potential secondary cause, such as a structural brain lesion. For patients with suspected cluster headache, we suggest neuroimaging with a cranial CT scan or a cranial MRI study to exclude abnormalities of the brain and pituitary gland. We recommend neuroimaging for patients with an abnormal neurologic examination or those with clinical suspicion for a pituitary abnormality. (See "Neuroimaging" above and see "Secondary cluster headache" above).

- The differential diagnosis of cluster headache involves consideration of other unilateral, brief but frequent headaches (show table 1), including the following:

  - Paroxysmal hemicrania
  - Short-lasting unilateral neuralgiform headache with conjunctival injection and tearing (SUNCT syndrome)
  - Trigeminal neuralgia
  - Primary stabbing headache
  - Headache associated with an underlying intracranial lesion (ie, secondary cluster headache)

(See "Differential diagnosis" above).

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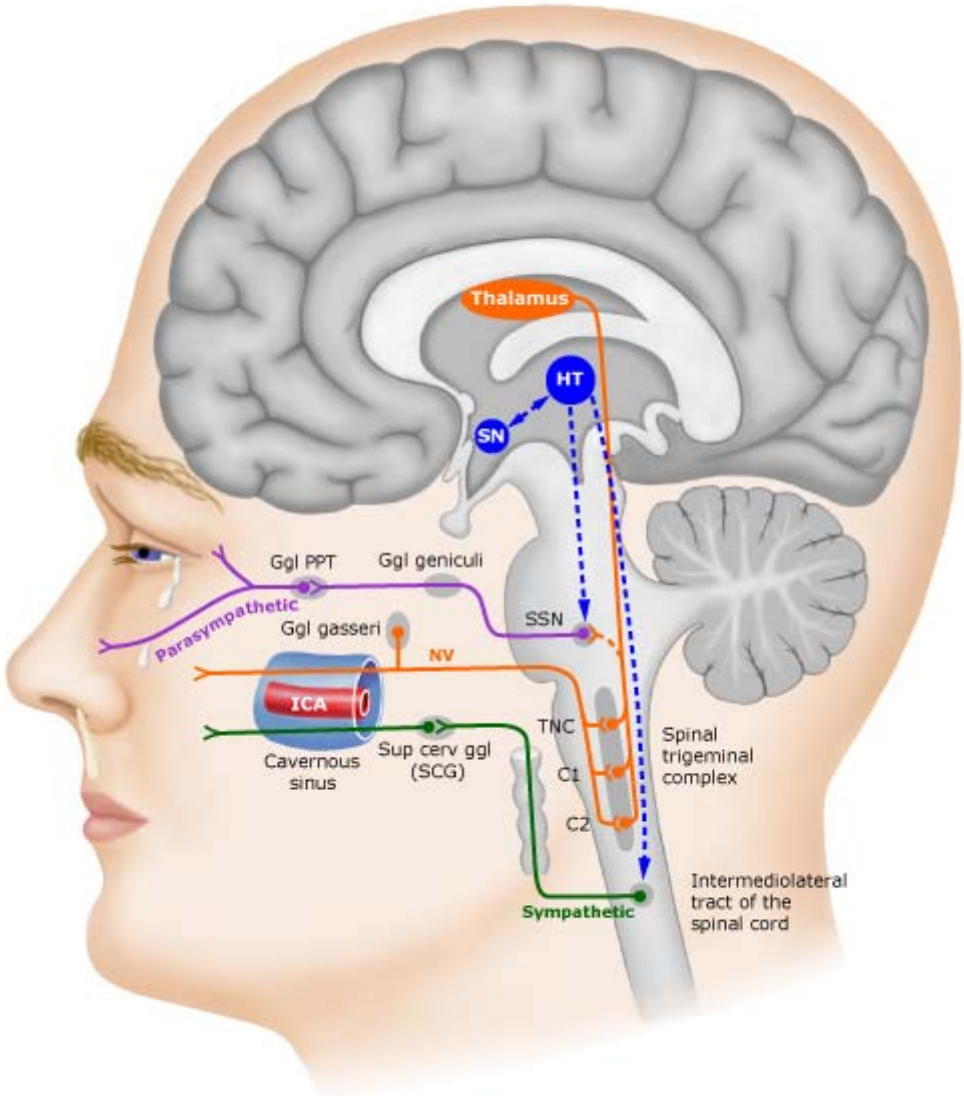
GRAPHICS

Pathogenesis of cluster headache



Schematic model showing most of the putative actors in CH pathogenesis. Pain afferents from the trigeminovascular system synapse on the trigeminocervical complex (TNC), and then project to the thalamus and lead to activation in cortical areas known to be involved in pain transmission. Either a direct influence of the hypothalamus or a reflex activation of the parasympathetic outflow from the superior salivatory nucleus (SSN) predominately through the pterygopalatine (sphenopalatine) ganglion, leads to the parasympathetic symptoms ipsilateral to the pain. A third-order sympathetic nerve lesion, thought to be caused by vascular changes in the cavernous sinus loggia with subsequent irritation of the local plexus of nerve fibers, results in a partial Horner's syndrome. The key site in the CNS for triggering the pain and controlling the cycling aspects is in the posterior hypothalamic grey matter region, modulated by phase-shifting in the suprachiasmatic nuclei.

Abbreviations: Ggl = ganglion, HT = hypothalamus, ICA = internal carotid artery, NV = trigeminal nerve, PPT = pterygopalatine, SCG = superior cervical ganglion, SN = suprachiasmatic nucleus, SSN = superior salivatory nucleus, TNC = trigeminal nucleus caudalis

Modified from: May A. Cluster headache: pathogenesis, diagnosis, and management. Lancet 2005; 366:847.

ICHD-2 diagnostic criteria for cluster headache

A. At least five headache attacks fulfilling criteria B through D:
B. Severe or very severe unilateral orbital, supraorbital and/or temporal headache attacks, which last untreated for 15 to 180 minutes. During part (but less than half) of the time course of the cluster headache, attacks may be less severe, less frequent, or of shorter or longer duration.
C. The headache is accompanied by at least one of the following symptoms:
1. Ipsilateral conjunctival injection or lacrimation
2. Ipsilateral nasal congestion and/or rhinorrhea
3. Ipsilateral eyelid edema
4. Ipsilateral forehead and facial sweating
5. Ipsilateral miosis and/or ptosis
6. A sense of restlessness and agitation
D. The attacks have a frequency from one every other day to eight per day
E. History and physical and neurologic examinations do not suggest any other disorder, and/or such a disorder is ruled out by appropriate investigations, or such disorder is present but attacks do not occur for the first time in close temporal relation to the disorder.
Episodic cluster headache: at least two cluster periods lasting seven days to one year separated by pain free periods lasting one month or longer.
Chronic cluster headache: attacks occur for more than one year without remission or with remission less than one month.
Probable cluster headache: attacks fulfilling all but one criteria for cluster headache.

Headache Classification Subcommittee of the International Headache Society. The International Classification of Headache Disorders: 2nd edition. Cephalalgia 2004; 24 Suppl 1:9.

Clinical features of the trigeminal autonomic cephalalgias

	Cluster headache	Paroxysmal hemicrania	SUNCT
Sex (female:male)	1:3 to 1:7	1:1 to 2.7:1	1:1.5
Pain			
Type	Stabbing, boring	Sharp, stabbing, throbbing	Burning, stabbing, sharp
Severity	Excruciating	Excruciating	Severe to excruciating
Site	Orbit, temple	Orbit, temple	Periorbital
Attack frequency	1 every other day to 8 per day	1 to 40 a day (>5 per day for more than half the time)	3 to 200 per day
Duration of attack	15 to 180 minutes	2 to 30 minutes	5 to 240 seconds
Autonomic features	Yes	Yes	Yes (prominent conjunctival injection and lacrimation)
Migrainous features (nausea, photophobia or phonophobia)	Yes	Yes	Rare
Alcohol trigger	Yes	Occasional	No
Cutaneous triggers	No	Rare	Yes
Indomethacin effect	None	Absolute	None
Abortive treatment	Sumatriptan injection or nasal spray Oxygen	Nil	Nil
Prophylactic treatment	Verapamil Methysergide Lithium	Indomethacin	Lamotrigine Topiramate Gabapentin

SUNCT: short-lasting unilateral neuralgiform pain with conjunctival injection and tearing.

Diagnostic criteria for paroxysmal hemicrania

A. At least 20 attacks fulfilling B-D
B. Severe unilateral orbital, supraorbital, or temporal pain lasting 2 to 30 minutes
C. Headache is accompanied by at least one of the following:
1. Ipsilateral conjunctival injection and/or lacrimation
2. Ipsilateral nasal congestion and/or rhinorrhea
3. Forehead and facial sweating
4. Ipsilateral eyelid edema
5. Ipsilateral forehead and facial sweating
6. Ipsilateral miosis and/or ptosis
D. Attacks have a frequency >5 per day for more than half the time, although periods with lower frequency may occur
E. Attacks are prevented completely by therapeutic doses of indomethacin*
F. Not attributed to another disorder
Episodic paroxysmal headache
Description: occurs in periods lasting seven days to one year separated by pain free periods lasting one month or more
Chronic paroxysmal headache
Description: attacks occur for more than one year without remission or with remissions lasting less than one month

* To rule out an incomplete response, indomethacin should be used in a dose of ≥150 mg daily orally or rectally, or ≥100 mg by injection. Smaller doses are often sufficient for maintenance.

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Diagnostic criteria for SUNCT and SUNA

Diagnostic criteria for SUNCT (short lasting unilateral neuralgiform headache attacks with conjunctival injection and tearing)
A. At least 20 attacks fulfilling criteria B-E
B. Attacks of unilateral, orbital, supraorbital, or temporal stabbing or pulsating pain last 5 to 240 seconds
C. Pain is accompanied by ipsilateral conjunctival injection and lacrimation
D. Attacks occur with a frequency from 3 to 200 per day
E. Not attributed to another disorder
Diagnostic criteria for SUNA (short lasting unilateral neuralgiform headache attacks with cranial autonomic symptoms)
A. At least 20 attacks fulfilling criteria B-E
B. Attacks of unilateral orbital, supraorbital or temporal stabbing pain lasting from 2 seconds to 10 minutes
C. Pain is accompanied by one of:
1. Conjunctival injection and/or tearing
2. Nasal congestion and/or rhinorrhea
3. Eyelid edema
D. Attacks occur with a frequency of ≥1 per day for more than half the time
E. Not attributed to another disorder
Episodic SUNA:
SUNA attacks occurring for seven days to one year with pain free intervals longer than one month
Chronic SUNA:
At least two attack periods last seven days to one year separated by remission periods of less than one month (untreated)

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