



# Primary syndrome with

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## INTRODUCTION

To mention headache in connection with sexual activity may bring a smile to the lips of the uninformed. However, any headache coinciding with sexual activity can be frightening, distressing, disabling and anything but amusing to those who have experienced it. Even though in ancient times Hippocrates described a headache resulting from 'immoderate venery' [cited in Adams (1848)], it was not until the 1970s that attention was drawn to a benign form of headache occurring during sexual activity (Lance 1974; Martin 1974; Paulson & Klawans 1974).

In the first systematic description of the disorder, 21 patients were reported (Lance 1976). One subgroup had pain which evolved slowly, possibly due to excessive muscular contraction of the neck and jaw muscles. A second, larger group of patients experienced sudden onset of pain shortly before, at the moment of, or shortly after orgasm. Another publication described three patients with a third type whose pain resembled that of the headache following lumbar puncture, perhaps resulting from a tear of the dura during sexual intercourse leading to low cerebrospinal fluid pressure (Paulson & Klawans 1974). The first edition of the International Headache Society classification differentiated these three types of

# Primary headache types associated with sexual activity

headache associated with sexual activity (types 1–3, respectively). However, the low-pressure type is so rare, and in any event should be regarded as a symptomatic headache, so in the second edition only the first two types are defined (Table 1) (Headache Classification Committee 1988). Since the first systematic descriptions of headache associated with sexual activity, several case series and case reports have been published, and by 1986 there were 110 published cases: 24% type 1; 69% type 2; and 7% type 3 (Johns 1986).

**Table 1** International Headache Society diagnostic criteria for primary headache associated with sexual activity (Headache Classification Subcommittee 2004)

## Diagnostic criteria

### 4.4.1 Pre-orgasmic headache (type 1)

- A Dull ache in the head and neck associated with awareness of neck and/or jaw muscle contraction fulfilling criterion B.
- B Occurs during sexual activity and increases with sexual excitement.
- C Not attributed to another disease.

### 4.4.2 Orgasmic headache (type 2)

- A Sudden severe ('explosive') headache fulfilling criterion B.
- B Occurs at orgasm.
- C Not attributed to another disease.

Note: On first onset of orgasmic headache it is mandatory to exclude conditions such as subarachnoid haemorrhage and cervical arterial dissection.

## EPIDEMIOLOGY

The frequency of headache associated with sexual activity is unknown. In the only population-based epidemiological study, the lifetime prevalence was about 1% with a wide confidence interval, similar to the frequency of benign cough headache and benign exertional headache (Rasmussen & Olesen 1992). Very likely, the frequency is underestimated because patients often feel too embarrassed to report intimate details about their sexual activities. We estimate that patients with headache associated with sexual activity account for about 1% of all headache patients who are referred to our supraregional headache clinics.

## PATHOPHYSIOLOGY

A study of a single family with four affected sisters suffering from headache associated with sexual activity type 2 has been published, but apart from this there is no evidence that sex headache is primarily genetic (Johns 1986).

Owing to the similarities in the nature of the headache, it has been suggested that type 1 is related to muscle contraction headache or tension-type headache (Lance 1976; Silbert *et al.* 1991; Pascual *et al.* 1996).

Some emphasize the similarity of type 2 to primary exertional headache and postulate that a transient increase in intracranial pressure due to a Valsalva manoeuvre during coitus plays a pivotal role (Calandre *et al.* 1996; Queiroz 2001). In three patients, segmental spasm of cerebral arteries has been observed in the days after the headache; however, the same changes were still present even months after the first angiogram in two of them

(Silbert *et al.* 1989; Kapoor *et al.* 1990; Valenca *et al.* 2004). Moreover, in a series of nine patients no abnormalities of the cerebral vessels were revealed by angiography (Lance 1976).

Recently, the first experimental studies have been published: patients with headache associated with sexual activity type 2 have impaired cerebrovascular autoregulation. Their cerebral vessel walls had impaired vasodilatation in response to a decrease in pH compared with healthy subjects and with migraineurs (Evers *et al.* 2003).

Other authors emphasize a pathophysiological relationship between headache associated with sexual activity type 2 and migraine (Johns 1986; Silbert *et al.* 1991), and postulate a release of vasoactive substances such as neurokinins, serotonin and catecholamines (Martin 1974; Paulson & Klawans 1974; Silbert *et al.* 1991). In favour of this idea is the fact that patients with headache associated with sexual activity type 2 have a similar loss of cognitive habituation to patients with migraine (Frese *et al.* 2003a). During visual event-related potential testing, both are characterized by potentiation instead of habituation of stimulation-evoked cortical responses. Furthermore, comorbidity of migraine, or of a family history for migraine, with headache associated with sexual activity type 2 has been shown in several case series (Johns 1986; Silbert *et al.* 1991; Pascual *et al.* 1996; Frese *et al.* 2003b). Nonetheless, any definite link between migraine and the headache associated with sexual activity is still somewhat uncertain.

In contrast to previous assumptions (Mann *et al.* 1982; Akpunonu & Ahrens 1991), arterial hypertension is not thought to be a major risk factor for headache associated with sexual activity (Frese *et al.* 2003b). However, patients with type 2 headache do show a higher increase of arterial blood pressure under physical stress (Evers *et al.* 2003).

## CLINICAL FEATURES

The following description of the typical clinical features of headache associated with sexual activity is mainly based on a recent case series which included the largest cohort of patients with both subtypes (Frese *et al.* 2003b). The operational diagnostic criteria of the International Headache Society are given in Table 1 (Headache Classification Subcommittee 2004).

The mean age of patients presenting for the first time with headache associated with sexual activity is about 40 years (Table 2). However, the onset

**Table 2** Comparison of demographic and clinical features between type 1 ( $n = 11$ ) and type 2 ( $n = 40$ ) headache associated with sexual activity (Frese *et al.* 2003b)

	Type 1	Type 2
Age at onset (years $\pm$ standard deviation)	40 $\pm$ 10	34 $\pm$ 10
Sex ratio (female to male)	2 : 9	11 : 29
Unilateral pain	36%	33%
Occipital/diffuse localization	82%	75%
Throbbing quality	36%	50%
Duration of severe pain (median)	30 min	30 min
Duration of milder pain (median)	1 h	4 h
Arterial hypertension	27%	15%
Comorbid migraine	9%	30%
Comorbid exertional headache	9%	35%
Comorbid tension-type headache	55%	43%

has two peaks: the first between the 20th and 24th years of life, and a second broader peak between the 35th and 44th years of life (Frese *et al.* 2003b). Whether these peaks are caused by changes in sexual activity or by something inherent in the patients is unknown. The male to female ratio is about 3–4 to 1 (Lance 1976; Silbert *et al.* 1991; Østergaard & Kraft 1992; Pascual *et al.* 1996).

Pain is occipital or diffuse in three-quarters of the patients, bilateral in two-thirds and unilateral in one-third. The quality is dull in about one-half, throbbing in one-half, and stabbing in about one-half of the patients (patients could give more than one quality). The duration of pain varies widely. The median duration is 30 min (range 1 min to 24 h). Most patients have severe pain for less than 4 h. None has severe pain lasting longer than 24 h, but many do have longer lasting milder pain. The median duration of the milder pain is 4 h. Accompanying symptoms such as nausea and dizziness are rare but do occur (Frese *et al.* 2003b).

Extramarital sexual activity, as well as exotic sexual habits, have been said to be precipitants for headache associated with sexual activity. In fact, the vast majority of patients develop their headaches during sexual activity with their usual partner or during masturbation. The risk does not seem to increase when the partner or setting changes. Many patients can abort or ease their headache by stopping sexual activity, or by taking a more passive role during sexual activity (Frese *et al.* 2003b).

Normally, the patients are healthy and do not have any concomitant vascular disease. Howev-

er, about two-thirds also suffer from other headache disorders such as episodic tension-type headache (36%), benign exertional headache (30%), migraine (26%) and chronic tension-type headache (10%) (Frese *et al.* 2003b).

Type 1 occurs in about one-fifth of patients and type 2 in about four-fifths. No patient is known with both type 1 and type 2 on different occasions. There are no large differences in mean age at onset, sex ratio, localization and quality, and duration of pain between the subtypes (Table 2). As expected, by definition, there is a significant difference in the onset time related to orgasm: median onset time in type 1 is 150 s before orgasm, while in type 2 it is exactly with or up to 5 s before orgasm. As a consequence, many more patients with type 1 headache can terminate this pain by stopping sexual activity early (Frese *et al.* 2003b).

## DIAGNOSIS

The term 'benign' defines a primary headache syndrome not caused by any intracranial disorder (Lance 1992). Subarachnoid haemorrhage (SAH) and cervical arterial dissection must be ruled out when headache associated with sexual activity occurs for the first time. SAH, for example, occurs during sexual activity in about 4% to 12% of all cases and so can easily be misdiagnosed as a benign headache associated with sexual activity (Locksley 1969; Lundberg & Osterman 1974). Cerebral or brain stem infarction at the time of orgasm has been reported (Lance 1976; Martinez *et al.* 1988), as has one patient with symptomatic orgasmic headache due to an intraventricular arachnoid cyst, the headache disappearing after removal of the cyst (Lasoasa 2003).

Attempts have been made to define 'red flags' which indicate a serious underlying pathological condition (Lundberg & Osterman 1974; Queiroz 2001; Frese *et al.* 2003b). Vomiting, decreased level of consciousness, meningism, motor, sensory or visual disturbances, or severe pain persisting for more than 24 h are *not* features of primary headache associated with

sexual activity. These symptoms require immediate diagnostic work-up. On the other hand, nausea, dizziness and mood disturbances are compatible with the diagnosis of an idiopathic headache.

Therefore, whenever headache associated with sexual activity appears for the first time, a CT scan of the brain and, if negative, a lumbar puncture should be performed to exclude SAH, even if the presentation seems typical for headache associated with sexual activity. If the history and/or the neurological examination suggest arterial dissection, in particular of the vertebral artery, the diagnosis has to be excluded by ultrasound examination and/or MRI.

## PROGNOSIS

The prognosis of headache associated with sexual activity is excellent. In the majority, the headache appears in a bout of some weeks or months and disappears without any specific treatment (Silbert *et al.* 1991; Østergaard & Kraft 1992). The number of attacks within one bout ranges from 2 to 50 (Frese *et al.* 2003b). Sometimes a bout recurs. About one-quarter of patients suffer from continued attacks without longer remissions. Prognostically there is no significant difference between types 1 and 2 (Frese *et al.* 2004).

## MANAGEMENT

Headache associated with sexual activity can be very frightening, and so explanation, counselling and reassurance are the most important therapeutic interventions (Clifford Rose & Petty 1982; Lance 1991). Education and information

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**Table 3** Pragmatic management of headache associated with sexual activity

## Management steps

## Step 1: Patient education and advice

- inform about the benign nature and good prognosis of the headache
- remain sexually inactive as much as possible during the bout of headaches
- more passive role during sexual activity

## Step 2: Short-term prophylaxis

- indomethacin, oral, 25–100 mg 30–60 min before sexual activity

## Step 3: Long-term prophylaxis

- propranolol, oral, 20–80 mg tid
- metoprolol (100–200 mg/day) or diltiazem (180 mg/day) if propranolol fails, is not tolerated, or is contraindicated

Note: prophylactic treatment should be tapered off after 3 months maximum to check for spontaneous remission.

about the benign nature and good prognosis are crucial. Further management is based on experience and case series, not on any randomized trials (Table 3).

There is no acute drug treatment for the headache. Analgesics (ibuprofen, diclofenac, paracetamol, acetylsalicylic acid) given after the onset of headache are of limited or no value in nearly all patients (Frese *et al.* 2004).

In several cases, recurrence has been reported when patients resumed sexual activity shortly after an attack (Lance 1976; Porter & Jankovic 1981; Edis & Silbert 1988; Kim 1992). Therefore it seems reasonable to advise the patients to remain sexually inactive as much as possible until they are completely free of symptoms. Another potentially helpful tip is to try a physically more passive role during sexual activity. More patients with type 1 than type 2 can terminate the pain by stopping sexual activity early, or by being more passive during the sexual activity (Frese *et al.* 2004).

If changing the sexual habits is not sufficient to avoid the headache, short-term prophylaxis can be tried. Indomethacin in an oral dose between 25 and 100 mg given 30–60 min prior to sexual activity has been successful in some patients, whereas ibuprofen and diclofenac have not (Silbert *et al.* 1991; Pascual *et al.* 1996; Frese *et al.* 2004). Ergotamine and benzodiazepines have also been tried but with no benefit in most patients (Paulson & Klawans 1974; Lewis 1976; Nutt 1977; Porter & Jankovic 1981).

For patients with longer lasting bouts, or with repeated attacks, longer term prophylactic treatment with propranolol can be successful (between 60 and 240 mg daily) (Porter & Jankovic 1981; Johns 1986; Edis & Silbert 1988; Pascual *et al.* 1996; Frese *et al.* 2004). However, there

**EDITORIAL COMMENT:  
WHAT'S IN A NAME?**

Maybe it is embarrassment but the name of this syndrome does keep moving around. When I started neurology it was called 'benign coital headache', but when neurologists could pluck up the courage to ask the relevant question the name broadened to 'benign orgasmic cephalgia' which has a fine ring to it. Now we have a name written by committee, 'Headache associated with sexual activity', which is a real mouthful. Some, including me, prefer the Anglo-Saxon simplicity of 'sex headache'. No doubt more changes are on the way, but for now the authors prefer 'headache associated with sexual activity' and we will leave it at that.

**CONCLUSIONS**

- Primary headache associated with sexual activity is an idiopathic and benign headache disorder precipitated by sexual activity.
- There are two main types: a pre-orgasmic headache which gradually increases with sexual excitement (type 1), and a more common orgasmic headache which occurs explosively at orgasm (type 2).
- The most important differential diagnosis is subarachnoid haemorrhage which has to be ruled out by investigation.
- Normally, the headaches appear in bouts lasting some weeks or months and disappear without any specific treatment.
- For patients with longer lasting bouts, short-term prophylaxis with indomethacin or long-term prophylaxis with propranolol are therapeutic options.
- More widespread knowledge about the existence and nature of this headache disorder will help patients to confide in their doctors and so improve their care.

are also reports of treatment failure with propranolol (Paulson & Klawans 1974; Silbert *et al.* 1991; Evans & Pascual 2000; Frese *et al.* 2004). Some success has been reported with metoprolol, atenolol and diltiazem (Silbert *et al.* 1991; Akpunonu & Ahrens 1991; Frese *et al.* 2004). Any prophylactic treatment should be stopped after a few months because the headache is highly likely to have resolved.

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