mitochondrial DNA in CADASIL pedigrees compared with healthy controls, and of muscle mitochondrial abnormalities in several CADASIL patients.⁵

In conclusion, our report demonstrates that a NCSE may mimic an ischaemic stroke or a prolonged migraine aura in patients with CADASIL. We propose that EEG should be performed in CADASIL patients with acute neurological deficits, particularly when no acute ischaemic alterations are found in DWI. In addition, evaluation of anticonvulsants as possible prophylactic treatment is warranted in a larger series of CADASIL patients.

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Prevalence of cluster headache in Germany: results of the epidemiological DMKG study

Cluster headache is a primary headache disorder diagnosed according to the criteria of the International Headache Society.¹ Previous epidemiological studies have shown prevalence rates of 0.056–0.38%. These were lifetime prevalence rates which are subject to recall bias. No population based prevalence rates for Germany have been published to date. Despite the use of common criteria, sociocultural influences on the classification of cluster headache may be significant. Thus it is important to compare epidemiological study results from different countries to elucidate such relations. We conducted a population based survey on the prevalence of cluster headache in Germany which was part of a larger epidemiological study on the prevalence of different headache types and which was coordinated by the German Migraine and Headache Society (DMKG).

Methods

A sample of 3425 inhabitants of the city of Dortmund, Germany, aged 25–75 years, was randomly selected and invited to participate in a larger epidemiological study in 2005. In total, 2291 individuals (67%) responded, 979 subjects by answering a standardised question-naire and 1312 through participation in a face to face interview. This analysis was restricted to the latter group. A standardised set of questions was used to assess headache symptoms, characteristics and time pattern.

For cluster headache classification, we chose a two step approach and recontacted all participants who fulfilled the following criteria:

- untreated headache attacks shorter than 6 h,
- unilateral headache,
- at least one of following accompanying symptoms: lacrimation, rhinorrhoea, ptosis, reddening of the face or restlessness during headache.

The second contact was done by a neurologist trained in headache diagnosis. This expert made a final diagnosis according to the criteria of the International Headache Society.

Results

In total, 33 subjects (2.5%) fulfilled the criteria for a personal interview by the neurologist (17 males, mean age 55 years, mean age at headache onset 33 years). A diagnosis of cluster headache was made in two subjects. In the other individuals, the diagnoses were sinusitis (two subjects) and migraine without aura (one subject); the remaining subjects fulfilled the criteria of episodic tension-type headache. They stated unspecific autonomic symptoms such as warmness and reddening in the whole head or feeling hot in the body in the first interview which were revealed as not fulfilling the criteria for trigemino-autonomic cephalalgias in the second interview.

The two cluster headache patients represent a 12 month prevalence of 0.15% (95% CI 0.01 to 0.55). Both patients were male. One was 37 years old, with cluster headache onset at the age of 22 years. The other was 55 years, with cluster headache onset at the age of 37 years. Both suffered from episodic cluster headache with one bout per year and a bout duration of 6–10 weeks. In both patients, daily right sided attacks occurred with typical autonomic features. Only one of the two patients was informed about the diagnosis and treated his attacks with oxygen.

 Table 1
 Previous epidemiological studies on the incidence or prevalence of cluster headache

Study	Country	Results
This study	Germany	0.15% (12 month prevalence)
Ekbom <i>et al</i> 2006 ⁶	Sweden	0.15% (lifetime prevalence)
Torelli <i>et al</i> 2005 ⁴	Italy	0.28% (lifetime prevalence)
Sjaastad and Bakketeig 2003 ³	Norway	0.38% (lifetime prevalence)
Tonon <i>et al</i> 2002 ²	San Marino/Italy	0.056% (lifetime prevalence)

No other neurological or medical disease was apparent in either patient. Interestingly, both had a migration background, with their parents having migrated to Germany.

Discussion

The prevalence in this study falls within the range of the few cluster headache prevalence rates reported in previous studies. They centre around a prevalence of 0.1%, as shown in table $1.^{2-5}$ We can also confirm the male preponderance of this disorder. It is important to note that the results of several epidemiological studies on cluster headache are more consistent when compared, for instance, with epidemiological studies on migraine. This supports the assumption that cluster headache is a biological disorder with little influence of ethnic or socioeconomic factors.

Our prevalence rate transforms to an estimation of approximately 120 000 cluster headache cases in Germany in the year 2005. This high value contrasts with the number of patients treated for cluster headache in Germany. Based on the German DRG registry in 2005, only 716 German patients were treated as inpatients for cluster headache. Thus underdiagnosis of this headache disorder is likely.

A limitation of our study was that we may have overlooked individual patients with rare cluster headache types, such as bilateral cluster headache, cluster headache bouts not remembered by the patient and cluster headache without autonomic features. However, as these cases are very rare, we believe that our prevalence rate is valid and realistic. Another limitation was the low number of subjects in the sample, resulting in a large confidence interval and making it impossible to define subgroups. This is a problem with all rare diseases. Therefore, additional epidemiological studies on large samples of cluster headache sufferers are warranted.

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