

REVIEW ARTICLE

The ‘essentials’ of essential palatal tremor: a reappraisal of the nosology

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Essential palatal tremor (EPT) is an uncommon disorder, distinct from symptomatic palatal tremor (SPT), but characterized by superficially similar rhythmic movements of the soft palate. While the pathophysiology of SPT has been relatively well defined, this is not the case in EPT. Based on an analysis of 103 published cases, we reviewed EPT in the context of other movement disorders with similar features and outline possible pathophysiological mechanisms. Phenomenologically it remains best classified as a tremor. Four major causes, including a central generator, peripheral/mechanical, voluntary/special skill and psychogenic, appear to account for the majority of cases of EPT, although there is considerable overlap in the pathogenic mechanisms underlying these categories. Among the cases reviewed, a large proportion fit into the latter two categories, although there are others where multiple mechanisms are likely at play. Based on our reappraisal, we suggest a change in designation to ‘isolated palatal tremor’, with primary and secondary subtypes. This retains the distinction from SPT and emphasizes the non-uniform, heterogeneous nature of the disorder.

Keywords: essential palatal tremor; nosology; symptomatic palatal tremor; aetiology

Abbreviations: EPT = essential palatal tremor; LVP = levator veli palatini; SPT = symptomatic palatal tremor; PT = palatal tremor; TVP = tensor veli palatini

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Palatal tremor is a rare movement disorder that has garnered much attention given its curious nature. Initially it was most commonly referred to as ‘palatal myoclonus’ but was subsequently renamed ‘palatal tremor’ at the First International Congress of Movement Disorders in 1990 (oral discussion) to acknowledge the continuous, rhythmic nature of the jerks of the soft palate. Prior to a large review by Schenck (1965) there was no distinct classification of palatal tremor, as evidenced by the use of numerous different terms for this movement (Table 1). Schenck recognized two types subsequently named ‘essential palatal tremor’ (EPT) and ‘symptomatic palatal tremor’ (SPT) by Deuschl who also pointed out that EPT is not simply a form fruste of SPT (Table 2) (Deuschl *et al.*, 1990).

In SPT, ear clicks are rarely present, and palatal movements, which rarely bring a patient to medical attention, are part of a constellation of clinical findings, including dysarthria, nystagmus and ataxia. The levator veli palatini muscle, which

is innervated by cranial nerves IX and X, is responsible for the rhythmic movements of the soft palate. Imaging changes in

Table 1 Terms used to describe palatal movements or ear clicks

Brain stem or palatal myorhythmia
Cephalic murmur
Chorea of the soft palate
Focal dyskinesia
Gaumensegelnystagmus
Myoclonies oro-branchio-respiratoires
Myoclonies vélo-pharyngo-laryngo-oculo-diaphragmatiques
Nystagmus du voile
Objective tinnitus
Palatal myoclonus
Palatal tremor
Reflexive rhythmic cramps
Rhythmic myoclonus
Tic
Tinnitus of tubal origin

Table 2 Features distinguishing between symptomatic and essential palatal tremor

	Sex ratio	Age of onset	Presenting complaint	Frequency	Muscles involved	Other clinical features	Presence in sleep	MRI	Causes
EPT (n = 103) ^a	1 : 1 ^b	29.4 ± 16.8 ^b	Ear click ^b	125 ± 70 ^c	Tensor veli palatini; others; never extremity tremor or ocular ^b	None ^b	Possible ^d	Normal ^b	See text for details
SPT (n = 210) ^e	2 : 1	45.1 ± 17.3	Usually not ear click	139 ± 51	Levator veli palatini; frequently extrapalatal	Depends on extent of lesion; often includes ataxia, dysarthria, and nystagmus	Consistent feature	Olivary hypertrophy ± presence of causative lesions	Multiple—typically a lesion in the dentato-olivary pathway

^aFrom this review.

^bSimilar to series reported by Deuschl *et al.* (1990) (n = 77).

^cHigher than reported by Deuschl *et al.* (1990) (107 ± 41).

^dConsidered less common in series by Deuschl *et al.* (1990).

^eFrom Deuschl *et al.* (1990).

SPT include olivary pseudohypertrophy and possibly evidence of the causative lesion in the dentato-olivary tract.

In EPT, on the other hand, palatal movements and ear clicks are the sole clinical manifestation (Deuschl *et al.*, 1990). In fact, in EPT, ear clicks are often the only complaint and can be distressing enough to lead to suicide attempts (Götze, 1957; Hughes, 1965). The clicks are frequently audible to others, as illustrated by one young gentleman who was asked by his fiancée to 'remove the watch from his coat pocket' (Freund, 1956). The sounds have been described as ticking, banging, cracking, popping, clattering, crunching or crackling noises (Götze, 1957), and have even been denoted in musical terms (Vali, 1905). The tensor veli palatini muscle (TVP), which is innervated by cranial nerve V, is generally thought to be the culprit. It is hypothesized that the ear clicks are produced by contractions of the TVP muscle that opens the eustachian tube, causing a sudden decrease of the surface tension within the tube (Poltizer, 1870; Deuschl *et al.*, 1994a). In EPT, all imaging and laboratory investigations are normal (Deuschl *et al.*, 1994b) and, to our knowledge, there are no post-mortem cases of EPT.

Although this classification serves to categorize the majority of cases with palatal movements, especially SPT, and, importantly, highlights the clinical and aetiological differences between the two entities, it is increasingly clear that many cases of EPT do not fit neatly into this scheme. Moreover, the origin and underlying cause of EPT largely remain elusive. Several recent case reports have illustrated potential variable aetiologies of EPT, including even the volitional performance of the movement (Scott *et al.*, 1996; Vieregge *et al.*, 1997; Cho *et al.*, 2001), thus further supporting the notion that it represents a heterogeneous disorder. This prompted us to review 103 EPT cases and to reconsider its classification and aetiology. We have included a select number of case reports in the text to illustrate the proposed theories on the aetiology of EPT, but a summary table of the clinical features of all 103 cases is also available in the Supplementary Material available at *Brain* online.

Review of the EPT literature

We searched the Medline database (via PubMed, a service of the National Library of Medicine's National Center for Biotechnology Information, online at <http://www.ncbi.nlm.nih.gov>) for publications from January 1966 to July 2005, using the search terms 'essential palatal myoclonus', 'idiopathic palatal myoclonus', 'essential palatal tremor', 'idiopathic palatal tremor' and 'objective tinnitus', which resulted in 416 citations. Based on the abstract or paper, we selected 42 publications (69 cases) that had a sufficiently detailed description to be considered as EPT. Criteria for exclusion included cases with acute onset and coexistent brainstem dysfunction or cases with a progressive neurological syndrome. We also excluded articles if there was insufficient information to determine whether they met exclusion criteria. We included articles in English, German and French. With

respect to cases published before 1966, we selected articles on EPT based on a comprehensive review by Schenck (1965). Further cases were ascertained through individual article reference review, resulting in an additional 24 articles (34 cases). We avoided double counts and included a total of 66 articles (103 cases) in our literature review. More than one-half of the cases (62 out of 103) have not been included in modern reviews, including 27 cases from 1867 to 1990 and 35 cases from 1991 to 2005.

Demographic and clinical features of the study sample

Demographic features in our sample were similar to the 77 EPT cases reviewed by Deuschl *et al.* (1990). The male : female ratio was about 1 : 1 (49m, 45f, 9 unspecified); average age of onset was 29.4 ± 16.8 (range: 4–74 years). Movement frequency ranged from 20 to 420 clicks/min (125 ± 70), which is in between the frequency ranges of EPT and SPT reported previously (Deuschl *et al.*, 1990) (Table 2). When documented (31 cases), the movement is equally found to be completely rhythmic (19) or only partially rhythmic or ‘irregular’ (12). Although EPT is generally considered to cease during sleep (Deuschl *et al.*, 1990), persistence in sleep was reported in almost half of the reviewed cases for which this information was available (16 out of 36). In almost one-third, muscles other than the TVP were involved but were typically restricted to the oropharyngeal region. Other areas have been reported, however, and include the periorbital and perioral regions and the masseter and temporalis muscles. About half of the case reports that discussed laterality had unilateral ear clicks (24 out of 47), with the left side predominating (17 left versus 7 right). Distractibility and entrainability were commented on in only eleven cases, five of whom showed one or both of these features. Depending on the author’s specialty (neurology versus ENT), the two most common concurrent features reported were psychiatric (Swartz, 1948; Leventon *et al.*, 1968; Williams, 2004; Samuel *et al.*, 2004) and otolaryngeal (Baker, 1885; Shadle, 1889; Vali, 1905; Götze, 1957).

Phenomenology of EPT

The unique phenomenology of EPT overlaps with, but does not strictly adhere to, the classic concepts of myoclonus or tremor. This difficulty in phenomenological classification is paralleled by the lack of knowledge about the underlying aetiology.

Myoclonus

EPT used to be referred to most commonly as one form of palatal ‘myoclonus’. Myoclonus refers to a brief, involuntary, twitching or ‘shock-like’ contraction of a muscle or muscle group. In EPT, while the movements may be sudden and brief, they are less ‘shock-like’ and more regular and continuous than is typically seen with myoclonus. Furthermore, unlike in myoclonus, the movements can often be altered and

suppressed (Vali, 1905; Stern, 1949; Freund, 1956; MacKinnon, 1968). Individual cases have been reported to respond to drugs influencing GABA (valproate) (Borggreve and Hageman, 1991) and serotonin (sumatriptan) (Scott *et al.*, 1996), both of which are neurotransmitters implicated in various forms of myoclonus. However, these responses have not been replicated (Yokota *et al.*, 1990; Vieregge *et al.*, 1997; Pakiam and Lang, 1999). Based on the phenomenological differences outlined above, EPT was reclassified as a tremor in 1990 (1st Meeting of the MDS 1990).

Tremor

Although the movement in EPT is more consistent with tremor (rhythmic, involuntary, oscillatory movements of a body part) than with myoclonus, there are important differences. First, tremors associated with a specific condition tend to have a relatively constant and characteristic frequency, such as the typical 4–6 Hz tremor in Parkinson’s disease. In contrast, the frequency of EPT is not only highly variable among different patients (ranging from 20 to 420 cycles/min), but may also vary within a single individual (Bogner, 1956; Freund, 1956). As previously mentioned in approximately one-third of the cases where the information was provided the movement was only partially regular or considered ‘irregular’. Secondly, unlike other tremors, EPT is said to be present in sleep in ~50% of the cases. Thirdly, although many patients with early stages of tremor can exert some control over their tremor, suppression by various tricks, as described in EPT (Vali, 1905; Davis and Kirchick, 1950; Roche and Perkin, 1987; Tomkinson *et al.*, 1995), is not a usual feature in tremor disorders. The latter is more reminiscent of dystonia.

Dystonia

Although the movement of EPT is not typical of dystonia, it could be more akin to a pure dystonic tremor. One case was reported to have other, albeit mild, dystonic signs in addition to EPT; however, this is the exception rather than the rule (Tanaka *et al.*, 1984). Some of the manoeuvres performed to suppress the movements in EPT, such as digital pressure behind the mastoid (Engström and Graf, 1952) and adopting certain neck positions (Heller, 1962; Tomkinson *et al.*, 1995; Scott *et al.*, 1996), could be considered sensory tricks. However, the majority of manoeuvres described in EPT may work simply by altering the position and tone of the muscles involved or by changing pressure in the ear canal. Such ‘tricks’ include pushing the palate up with a finger (Poltitzer, 1870), opening the mouth widely (MacKinnon, 1968; Leventon *et al.*, 1968), holding the nose and plugging the ears (Davis and Kirchick, 1950) or performing a Valsalva manoeuvre (Hughes, 1965; MacKinnon, 1968).

Tic

A final consideration when discussing the phenomenology of EPT is a tic disorder. As with dystonia, tics and EPT have only

one feature in common, i.e. an urge to perform the movement. However, while this is an inherent feature of tics, it is only infrequently described in EPT (Götze, 1957; Williams, 2004). Furthermore, continuous rhythmic movements would be exceedingly rare as a form of tic.

Therefore, based on the discussion above, although classification remains imperfect, the hyperkinetic palatal movements are still best characterized as an unusual form of tremor.

Theories on the aetiology of EPT

As mentioned above, theories on aetiology of EPT also remain a matter of vivid debate. Our review of 103 EPT cases suggests four main possible underlying causes. However, many cases either do not fit neatly into these categories or appear to be a consequence of a combination of more than one aetiological factor and these categories are, in fact, not perfectly segregated as will be discussed.

Central generator

Although it is recognized that EPT and SPT are two distinct entities, the similarity of palatal movements has led to the assumption that the underlying generator in both disorders may be related, if not identical. Pseudohypertrophy of the inferior olive is characteristic of SPT, with the lesion responsible situated along the dentato-olivary pathway. Although there is no comparable lesion known to exist in EPT, the inferior olive and adjacent brainstem structures have been implicated based on functional MRI studies (Boecker *et al.*, 1994; Nitschke *et al.*, 2001). These observations might be consistent with a single generator; however, the final pathway differs between the two conditions because the two predominant muscles involved are innervated by different nerves (EPT: TVP/trigeminal nerve; SPT: LVP/glossopharyngeal and vagus nerves). Owing to the anatomy of the TVP, some authors suggest that it is responsive to the same mechanism that underlies physiological clonus (Klien, 1918). Additional support for a central mechanism derives from reported beneficial responses to centrally acting medications (Case 1, Nasr and Brown, 2002; Case 2, Borggreve and Hageman, 1991; Case 3, Cakmur *et al.*, 1997; Fabiani *et al.*, 2000), although admittedly the reported drug trials have not been placebo controlled, and long-term outcomes have not been reported, and thus responses cannot indisputably imply causation. Finally, the movements in palatal tremor are reminiscent of those in gill-breathing vertebrates (Stern, 1949) and disinhibition of a primitive pharyngeal reflex arch has been postulated to result in palatal tremor.

Case 1 (Nasr and Brown, 2002): response to centrally acting medication (lamotrigine)

A 37-year-old male was admitted to the hospital in the context of excessive alcohol intake. He had a history of alcohol abuse for several years. He used alcohol as a self-prescribed medication for palatal tremor and ear clicks which began 9 years

earlier and were very distressing to him. He was placed on alcohol detoxification and prescribed antipsychotics. He was then given lamotrigine for the treatment of palatal tremor. There was gradual improvement in his ear clicks and the frequency of palatal movements became much slower. Upon discharge he began drinking again and stopped the lamotrigine which resulted in the return of his palatal tremor.

Case 2 (Borggreve and Hageman, 1991): response to centrally acting medication (sodium valproate)

A 23-year-old man presented with involuntary audible ear clicks which worsened with stress. Examination revealed rhythmic bilateral and symmetrical movements of the soft palate.

Sodium valproate completely resolved the palatal tremor within a few days. A dose reduction after a period of a few months led to a reappearance of the palatal tremor. Subsequent increases in the dose produced a significant but incomplete improvement.

Case 3 (Cakmur *et al.*, 1997): response to centrally acting medication (flunarazine)

A 16-year-old girl developed ear clicks at the age of 6 years. Examination showed continuous rhythmic involuntary movements of the soft palate. The rest of the examination and work up (including brain MRI) were normal. Sodium valproate decreased the palatal tremor frequency but it was still disturbing to the patient. Flunarazine was started for frequent migraine attacks. After initiation of this drug the palatal tremor disappeared and did not recur when sodium valproate was stopped. However, after discontinuation of flunarazine, the palatal tremor reappeared. Administration of flunarazine resulted in a gradual decrease of the palatal tremor.

Mechanical/peripheral origin

While a central mechanism has garnered much attention in the neurological literature, there is evidence to support a peripheral or mechanical origin of EPT in the otolaryngology literature. Local acute inflammatory changes or chronic hypertrophy of the mucosa of the nasal and oral cavity have frequently been found in EPT patients (Case 4, Shadle, 1889; Bredlau, 1950; Wakata *et al.*, 2002). Accordingly, it has often been reported that the onset of EPT was preceded by an upper respiratory infection (Samuel *et al.*, 2004; Case 6, MacKinnon, 1968; Wakata *et al.*, 2002). In addition, EPT has sometimes been successfully treated with tonsillectomy and other local therapies (Baker, 1885; Vali, 1905; Klein, 1918). EPT can also be influenced both by pressure changes in the ear canal (Leventon *et al.*, 1968; Case 6, MacKinnon, 1968; Case 5, Gupta *et al.*, 1972) and changes in tone and position of pharyngeal muscles (Kadokia and McAbee, 1990). However, this evidence must be interpreted with caution. The presence of local inflammatory changes does not necessarily imply causation, especially in cases, albeit unusual, in which clicks are not immediately apparent (Ross and Jankovic, 2005). Moreover, local therapies do not always 'cure' the ear click

(Bredlau, 1938; Alfaro, 1950), and certainly it is possible that these are chance associations or even placebo responses. Finally, it is hard to imagine how such continuous types of movements could be maintained for prolonged periods in the absence of a central generator. In fact, peripheral causes may serve to switch on a central generator in the same fashion that peripheral insults are known to trigger other centrally mediated movement disorders (Jankovic, 1994).

Case 4 (Shadle, 1889): PT due to nasal inflammation and response to local surgical procedure

A young lady presented with bothersome ear clicks. On examination, ear clicks were audible and rhythmic, and ‘choreic’ movements of the soft palate were noted. The lower and middle nasal conchae were hypertrophied and pressed on the nasal septum causing stenosis of the middle nasal duct. The ear clicks and movements of the soft palate disappeared after adenoidectomy.

Case 5 (Gupta *et al.*, 1972): PT influenced by pressure changes in the ear canal

A 28-year-old female presented with audible clicks and ‘twitchings in her throat’ five months prior to her admission. These were disturbing to her but did not interfere with any daily activities. On examination, rhythmic contractions of the muscles of the soft palate and the lateral pharyngeal wall could be seen. Valsalva manoeuvres and holding of the breath stopped the jerks while breath taking restarted the contractions. Mental diversion by asking the patient to do mental calculations did not have any effect on the muscular contractions. The movements did not change during a 9 month follow-up period.

Case 6 (Case 1 in MacKinnon, 1968): PT following upper respiratory infection and influence of pressure changes in the ear canal

A 35-year-old woman developed ear clicks following a severe cold 11 years previously. The intensity of the ear clicks worsened when she was increasing her altitude by going up in an elevator or airplane. The ear clicks could be stopped by Valsalva manoeuvre, swallowing and opening the mouth widely. Examination revealed bilateral palatal tremor and audible ear clicks.

Voluntary/special skill

There are many patients who can voluntarily elicit the movements seen in EPT (Baker, 1885; Götze, 1957; Schubert and Neuss, 1959; Case 8, Klein *et al.*, 1998). This is particularly evident in individuals with exquisite control over their pharyngeal muscles, such as in musicians who play a wind instrument (personal observation). A nice example of acquired voluntary control is the case of a scuba diver who learned to open and close his eustachian tubes for diving and subsequently could produce ear clicks in either ear independently

as well as in both ears simultaneously (Case 9, Seidman *et al.*, 1999). The ability to voluntarily contract the TVP muscle has been compared with other ‘special skills’ that are present only in a minority of the population, such as ear wiggling (Case 8, Klein *et al.*, 1998). These traits are thought to be hereditary, and voluntary palatal movements identical to EPT have been seen in members of the same family (Case 8, Klein *et al.*, 1998). Furthermore, there are patients in whom the clicks seemingly begin spontaneously, who then learn to modulate frequency and volume or to even elicit and suppress them (Wakata *et al.*, 2002; Case 8, Klein *et al.*, 1998; Case 7, Jacobs *et al.*, 1981). Conversely, in some, the movements start voluntarily but ultimately become a subconscious habit (Klien, 1918). None of the individuals in the preceding discussion is bothered by these movements or clicks, nor do they report an urge to produce them and have no obvious secondary gain. In order to sustain persistent rhythmic movements of the palate it is likely that these individuals have acquired voluntary control over a central generator, further emphasizing the overlap in possible aetiological categories.

Any movement that can be performed voluntarily may also occur as a tic or a psychogenic movement disorder. By definition, such patients would possess the ‘special skill’ to perform the movements which, as indicated above, probably involves activation of a central generator. However, the site of ‘suprasegmental’ drive to such activation would differ in cases of EPT occurring on a completely voluntary basis, as a manifestation of a tic disorder or as a psychogenic movement disorder. The patient’s history, accompanying features, and particularly the patient’s description of the nature of their movements and attitude towards them would distinguish these three categories. Although the movements of EPT could be a manifestation of a tic disorder, prolonged rhythmical movements are exceedingly rare as a form of tics.

Case 7 (Case 2 in Jacobs *et al.*, 1981) spontaneous onset of PT followed by learned voluntary control

A 7-year-old girl noted rhythmic clicks in her ears. On examination 1 year later, she was noted to have rhythmic movements of palatopharyngeal and submental muscles with audible clicks emanating from the ears, which were synchronous with the palatal movements. Six months later the clicks became erratic and by the age of 8 years and 9 months the clicks had ceased entirely. At the age of 9 years she could repeatedly voluntarily induce them ‘by concentrating on her throat’. No other manoeuvre could bring on these movements. Over five years of follow-up, the palatal tremor never recurred spontaneously but she continued to induce it at will.

Case 8 (Case 1 in Klein *et al.*, 1998): familial PT with voluntary control

A 21-year-old gentleman presented with a 1 year history of clicking sounds predominantly in the left ear. He was puzzled

but not disturbed by his condition. On examination there was a rhythmic tremor mainly on the left side of the soft palate and a clicking sound synchronous with the palatal tremor could be heard with a stethoscope placed on the external orifice of the left auditory canal. He could voluntarily switch the sounds on or off by 'thinking' about them and he could modulate the frequency of the ear clicks 'by giving the command "faster" or "slower"'. Mental distraction had no effect. The remainder of the examination (including MRI) was normal. His sister, aged 26 years, also reported the presence of ear clicks. She, too, could voluntarily provoke, stop and modulate them. Her examination revealed rhythmic, bilateral tremor of the soft palate that was induced by the patient by 'concentrating on her throat'. Finally, their father reported a brief time period during which he, too, experienced these movements; however, they were no longer present and he refused examination.

Case 9: (Case 6 in Seidman et al., 1999): acquired voluntary control of PT in a scuba diver

A 39-year-old gentleman learned how to open and close his eustachian tubes while scuba diving 15 years earlier. Since that time he learned to control the contractions and produce audible ear clicks in either ear independently as well as in both ears simultaneously. To produce ear clicks unilaterally he inhaled and to produce bilateral ear clicks he exhaled. He also controlled the frequency of the pulsating muscles. The click was audible to the observer and the patient, and the palatal movements were easily demonstrable. The remainder of the neurological examination was normal.

Psychogenic

On the other hand, the careful examinations documented in many of the patients described as having EPT (see

Supplementary Table) reveals 'volitional' control over the ear clicks (Table 3). In contrast to the 'voluntary' group above, however, they are extremely distressed by them, and secondary gain can often be suspected. In some of these cases, a psychogenic origin has been proposed based on features commonly seen in psychogenic movement disorders (Schwartz, 1948; Götze, 1957; Leventon *et al.*, 1968; Case 11, Williams, 2004) (Table 3). Unfortunately, some features that can be seen in EPT (i.e. distractibility, entrainability and variability) (Sa *et al.*, 2004; Schrag and Lang, 2005) are rarely documented in the literature (Case 12, Vali, 1905; Cho *et al.*, 2001; Case 11, Williams, 2004; Samuel *et al.*, 2004; Case 10, Yokota *et al.*, 1990). As in other psychogenic movement disorders (Sa *et al.*, 2004), acute onset following an emotional stressor (Leventon *et al.*, 1968; Case 10, Samant *et al.*, 1970; Yokota *et al.*, 1990) or a trivial trauma (Schubert and Neuss, 1959; Heller, 1962; Jacobs *et al.*, 1981; Morini *et al.* 2005) is not uncommon. Although patients with psychogenic movement disorders often have an organic model to mimic, this is rarely the case in EPT. However, it has been reported, as in one young woman who developed ear clicks during an emotional crisis and after her neighbour was diagnosed with a symptomatic palatal tremor (Case 11, Williams, 2004). One patient even admitted under superficial anaesthesia that she elicited the noise voluntarily (Götze, 1957). Finally, although the presence of an overt psychiatric condition (Niederwieser, 1938; Pearson and Barnes, 1950; Schenck, 1965; Leventon *et al.*, 1968; Samuel *et al.*, 2004) raises suspicion of an underlying psychogenic origin, experience with other psychogenic movement disorders emphasizes that the presence of underlying causative psychological factors may be extremely difficult to elicit (Schrag *et al.*, 2004). Therefore, lack of these does not rule out a psychogenic origin.

Table 3 Red flags to suggest psychogenic palatal tremor

Psychogenic features	Case references
Distractibility	Pakiam and Lang (1999), Cho <i>et al.</i> (2001), Williams (2004) and Samuel <i>et al.</i> (2004)
Entrainability	Samuel <i>et al.</i> (2004)
Variability (short and long term)	Vali (1905) and Cho <i>et al.</i> (2001)
Involvement of extrapalatal muscles (e.g. tongue, jaw, pharynx) clinically distinct from those seen in SPT (ocular, cranial, axial and limb tremor)	Yokota <i>et al.</i> (1990), Pakiam and Lang (1999) and Cho <i>et al.</i> (2001)
Emotional trigger (acute onset)	Leventon <i>et al.</i> (1968), Samant <i>et al.</i> (1970), Yokota <i>et al.</i> (1990) and Williams (2004)
Trivial trauma preceding onset (acute) (e.g. blunt blow to temple, fall into water, whiplash injury 24 h prior)	Schubert and Neuss (1959), Heller (1962) and Klein <i>et al.</i> (1998)
Response to verbal suggestion or hypnosis	Niederwieser (1938), Schwartz (1948) and Pearson and Barnes (1950)
Response to non-physiological treatment or placebo (e.g. copper pennies on mastoids)	Schwartz (1948) and Yokota <i>et al.</i> (1990)
Long lasting benefit from short-acting drugs (prolonged remission after a single treatment with botulinum toxin)	Varney <i>et al.</i> (1996), Jero and Salmi (2000), Cho <i>et al.</i> (2001) and Kutukcu <i>et al.</i> (2003)
Presence of other psychiatric features	Niederwieser (1938), Pearson and Barnes (1950), Schenck (1965), Leventon <i>et al.</i> (1968) and Samuel <i>et al.</i> (2004)

**Case 10 (Case 2 in Yokota *et al.*, 1990):
response to placebo**

A 16-year-old girl noted ‘twitching’ of her throat that often occurred when she was anxious or excited. The twitching became more frequent over a 2 year period and so a neurological consultation was requested. On examination, bilateral, rhythmic movements of the pharynx, larynx and sometimes masseter muscles were noted. She could not voluntarily induce or stop the movements but they only occurred when she was with somebody else. The movements disappeared immediately after intravenous injection of 10 ml saline as a placebo as well as of 10 mg diazepam. A psychogenic aetiology was suspected. The movements disappeared after it was explained to her that her disease was benign and transient and should not be worried about.

**Case 11 (Williams, 2004): acute onset of
PT following emotional stressor with organic
model to mimic**

A 44-year-old woman presented 3 years after developing an unusual ‘vibration in her throat’. The onset coincided with a period of high anxiety related to the birth of her niece to her sister who had major depression and was in an abusive relationship. Around the same time she became aware of a neighbour who developed true palatal tremor. Initially the tremor was only occasional but within 2 months it became constant, at which time she noticed a clicking in both ears that occurred with the sensation of ‘vibration in her throat’. There was a history of depression and anxiety. The neurological examination was normal except for an audible clicking sound that could be heard by the examiner and could only be elicited with the mouth closed. It was distractible.

**Case 12 (Lang, 2001; Cho *et al.*, 2001):
variable and distractible movements and
striking response of palatal and extrapalatal
movements to a single, local injection**

A 21-year-old male presented with a 2 year history of bilateral ear clicking that he could increase in volume with concentration. He felt as if he was going insane due to the clicking. Examination revealed rhythmical movements although frequency varied quite widely. Contractions of the temporalis, tongue, floor of mouth and mentalis were observed as well. When he spoke or opened his mouth, the contractions diminished or disappeared. Head nodding, present at a slower frequency than palatal movements, disappeared as well. Botulinum toxin was injected into bilateral TVPs and at a 3 month follow-up all involuntary movements (including movements of muscles far from injection sites such as the temporalis as well as head nodding) had ceased.

**Reappraisal of terminology
‘Essential’ palatal tremor**

The term ‘essential’ was first applied to EPT to distinguish it from SPT and to align it with other ‘primary’ or ‘essential’ movement disorders. However, EPT is not analogous to disorders such as ‘essential tremor’ or ‘essential myoclonus’ or ‘primary’ dystonia. When named, these were thought to be idiopathic, often familial disorders. They have well-defined natural histories, and causative genes have been identified in some of them (Zimprich *et al.*, 2001). Conversely, EPT is usually sporadic and its natural history still remains unknown, largely because it is very rare and more heterogeneous than the other ‘essential’ disorders, and because little long-term follow-up is available. Based on our review, it is clear that the natural history is quite variable, ranging from spontaneous remission to a persistent distressing state.

As mentioned earlier, there are a number of cases that cannot be classified into a single aetiology postulated above, either because the mechanism is yet to be identified or because there are multiple mechanisms at play. Indeed, it is difficult to conceive how this type of movement could be maintained for prolonged periods in the absence of a central generator, independent of the presence of any other causative pathogenic factors. This could be triggered by a peripheral insult, or driven consciously and under voluntary control or by other ‘suprasegmental’ centres in the case of palatal tremor occurring as a rare manifestation of a tic disorder or as a psychogenic movement disorder. One example of the combination of a peripheral trigger and psychogenicity is seen in psychogenic cough where a respiratory infection often precedes the onset of the cough but a clear functional component then takes over (Mastrovich and Greenberger, 2002; Bhatia *et al.*, 2002).

Revised classification

Although imperfect, phenomenologically, EPT still remains best classified as a tremor. The aetiology of EPT is heterogeneous with a considerable proportion of cases having ‘voluntary’ control over the movements, be it in the context of a special skill (truly voluntary), a tic disorder (semi-volitional) or a psychogenic condition potentially ranging from a conversion disorder with little conscious volitional component to malingering with full voluntary control (Sa *et al.*, 2004). The current literature does not permit an exact determination of the proportion of cases that would be considered ‘voluntary’ because most case reports do not report the presence or absence of findings that would be helpful in making this determination (Table 4). It is clear, however, that EPT is not an atypical form of SPT but rather represents a distinct hyperkinetic movement disorder, and the classification should reflect this. As different aetiologies of EPT are being elucidated, many cases would probably be included as a ‘symptomatic’ palatal tremor. In the current classification this is not possible. Therefore, we propose three possible alternatives.

Table 4 Important features of EPT to elicit from history and exam

History	Age at onset
	Trigger
	Provoking factors
	Presence in sleep (by history only or documented)
	Voluntary control and tricks
Examination	Psychiatric features
	Frequency
	Variability over time (short and long term)
	Influence of respiratory cycle, breath holding, phonation
	Muscles involved
	Distractibility (complete suppression, change in rhythmicity or frequency)
	Entrainability (takes on entirely new frequency of a requested task, e.g. finger tapping)
Response to treatment	

Because the term 'essential palatal tremor' is already engrained in the literature, we could continue to refer to 'essential' palatal tremor and subdivide it into primary (cause undefined) and secondary EPT (including special skill, tic, psychogenic), recognizing that 'essential' is not akin to the term used in other 'essential' movement disorders. Alternately, because 'symptomatic' palatal tremor is also engrained in the literature as a disorder due to a definable lesion, another possibility is to reclassify current cases of 'EPT' as 'non-symptomatic' palatal tremor. Neither of these alternatives is entirely satisfactory, but we need to recognize that the current system is no longer sufficient to distinguish between different forms of this disorder. Therefore, we would propose to rename EPT 'isolated palatal tremor' (IPT) in order to avoid the described implications of the term 'essential'. The term 'isolated' would indicate the absence of other neurological disturbances or imaging abnormalities (rather than the absence of other muscles involved). It would allow a separation of these cases from SPT and would also allow for subcategories such as primary IPT (cause undefined) and secondary IPT (e.g. special skill, psychogenic). In order to help further refine the classification and permit advances in our understanding of these disorders it will be important for future published cases to carefully specify associated clinical features (Table 4). In conjunction with advances in the fields of neuroimaging, electrophysiology and molecular neurobiology, it will lead to a better aetiological understanding of this heterogeneous condition.

Supplementary data

Supplementary table, electronic file name E-table 1.

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