Primary Writing Tremor (PWT) is a type of task-specific tremor which occurs only while writing (Type A PWT) or assuming a writing position of the hand (Type B PWT). There is a considerable overlap of clinical features between PWT and writer’s cramp which creates difficulty in diagnosing this condition in the clinic. PWT usually affects the dominant hand and is typically 5–7 Hz in frequency, worsened by anxiety, temporarily relieved by alcohol and associated with reduced writing speeds. There are a variety of hypotheses about the phenomenology of PWT (regarding whether it is a variant of essential tremor, focal dystonia or an independent entity). Unlike writer’s cramp, PWT shows normal reciprocal inhibition of H reflex, does not exhibit excessive EMG activity in proximal muscles, and on fMRI shows underactivation of cingulum and overactivation of primary motor and supplementary areas. There are no randomised controlled trials currently for the treatment of PWT. Treatment modalities available are: medical treatment, botulinum toxin, surgical management (including DBS) as well as adaptive strategies and occupational therapy.

Keywords: Dystonia, essential tremor, primary writing tremor, writer’s cramp

INTRODUCTION

Primary Writing Tremor (PWT) is a type of task-specific tremor which occurs only while writing or assuming a writing position of the hand, and not during other tasks. The etiology of this tremor is debated, as well as the efficacy of the various treatment modalities used currently. Even though this is one of the most common task-specific tremors seen, there is considerable overlap of clinical features between PWT and writer’s cramp which creates difficulty in diagnosing this condition in the clinic. This article aims to review the phenomenological spectrum of PWT, the neurophysiological aspects of this tremor as well as the therapeutic interventions available.

SEARCH STRATEGY

To review the possible phenomenological spectrum, neurophysiological aspects and treatment modalities of PWT, a PubMed search using a variety of search terms was made in November 2020. We also reviewed the references of these articles and the final bibliography was based on the relevance of the review.

PHENOMENOLOGY OF PRIMARY WRITING TREMOR—A VARIANT OF ESSENTIAL TREMOR OR WRITER CRAMP OR AN INDEPENDENT ENTITY?

PWT tends to occur around the sixth decade of life, unlike writer’s cramp which begins 15–20 years earlier. The mean age is 50.1 years. It is usually a sporadic disease, although some authors have shown that some cases are inherited in an autosomal dominant pattern. A history of trauma to the dominant hand may be present in a minority of cases. One study also reported the presence of ‘essential tremor (ET)’, PWT, and writer’s cramp in the same family.

PWT is often classified into two types: Type A and Type B. Type A refers to task-induced tremor where the tremor starts occurring only when the person begins to write. The tremor also occurs when drawing, however, it is not present on rest or when doing other fine activities like sewing. Type B refers to positionally sensitive tremor where the tremor occurs as soon as the person’s hand assumes the position of writing; they need not have to actually write.

This tremor is usually non-progressive in nature and primarily involves the dominant hand, however, in some cases it may eventually become less task dependent with time and may progress to the non-dominant hand also. The tremor is usually worsened by anxiety, writing quickly, and exercise. The tremor is 5–7 Hz in frequency and is pronation-supination or flexion-extension in nature.

Patients with PWT also have significantly reduced writing speeds, which may be further worsened by anxiety. The tremor is usually bilateral, although the non-dominant hand may be involved in some cases. The tremor is usually temporarily relieved by alcohol and associated with reduced writing speeds. There are a variety of hypotheses about the phenomenology of PWT (regarding whether it is a variant of essential tremor, focal dystonia or an independent entity). Unlike writer’s cramp, PWT shows normal reciprocal inhibition of H reflex, does not exhibit excessive EMG activity in proximal muscles, and on fMRI shows underactivation of cingulum and overactivation of primary motor and supplementary areas. There are no randomised controlled trials currently for the treatment of PWT. Treatment modalities available are: medical treatment, botulinum toxin, surgical management (including DBS) as well as adaptive strategies and occupational therapy.

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speeds with their dominant hand, although the non-dominant hand does not exhibit this reduction.\textsuperscript{11} Usually, it is equally difficult to write letters and numbers, however, there have been reports that it is more difficult to write numbers as opposed to letters.\textsuperscript{16}

There are a variety of hypotheses about the phenomenology of PWT. Some say it is a type of task-specific focal dystonia of the hand, others argue it is a type of ET, and yet others believe it is a separate disorder, different from both ET and dystonia.

There is mounting evidence in recent years that PWT could be a variant of task-specific focal dystonia of the hand (FHD) [Table 2].\textsuperscript{7,11-13} FHD is characterized by excessive contractions of muscles of the hand leading to abnormal posturing and also in some instances, unintentional movements. Researchers who argue that PWT is a variant of task-specific focal dystonia point out its task-specific nature, the unilateral and focal involvement of only one limb (unlike the bilateral, progressive involvement of both limbs seen in ET), as well as the efficacy of treatment used for dystonic disorders in this tremor.\textsuperscript{10,11} Patients with PWT do not usually show abnormal posturing of their hands, however, this is debated. It was shown that EMG findings in PWT were consistent with subtle posturing, but it was not visible grossly because of the severe tremor.\textsuperscript{7} Another study discussed a case in which dystonic posturing of the wrist was visible, in addition, they also showed the presence of a mirror tremor, that is, writing with the left hand produced a mirror tremor on the right hand also.\textsuperscript{8} However, as discussed in more detail later, there are significant differences in neurophysiological studies between PWT and writer’s cramp.

There are researchers who argue that PWT is a variant of ET [Table 3].\textsuperscript{1,12-17} ET has a strong familial linkage and is characterized by bilateral tremor of the upper limbs, head, or voice. Similar to ET, reports of patients with PWT also suggest considerable relief of symptoms after drinking alcohol.\textsuperscript{1,16,18} Both ET and PWT have similar frequencies (between 4 and 8 Hz).\textsuperscript{1} However, symptoms of PWT are usually non-progressive in severity (especially in Type A PWT) and appear primarily during writing (a skilled task), unlike ET which can usually be suppressed during skilled tasks, and become increasingly severe with time.\textsuperscript{1,19}

There are also researchers who argue that PWT is neither a form of ET nor a type of focal dystonia. [Table 4].\textsuperscript{20-22} One study showed that intracortical and spinal excitability are abnormal in writer’s cramp and in some cases of ET, but normal in patients with PWT, suggesting that PWT is distinct

| Table 1: PubMed search (November 2020) related to primary writing tremor |
|-----------------------------------|-----------------|-----------------|
| Search term                                      | Results obtained | Articles included |
| (Primary writing tremor) AND (Pathophysiology)                | 45              | 18              |
| (Primary writing tremor) AND (Etiology)                 | 28              | 2               |
| (Primary writing tremor) AND (Electromyography)           | 13              | 1               |
| (“Primary Writing Tremor”) AND (MRI)                  | 2               | 1               |
| (“Primary Writing Tremor”) AND (Neuroimaging)           | 2               | 1               |
| (“Primary Writing Tremor”) AND (Neuromodulation)        | 11              | 0               |
| (Primary writing tremor) AND (Writer’s Cramp)           | 34              | 2               |
| (“Primary Writing Tremor”) AND (Treatment)             | 32              | 5               |
| (Primary writing tremor) AND (Botulinum toxin)      | 14              | 3               |
| (Primary writing tremor) AND (Deep brain stimulation)   | 7               | 3               |
| (Primary writing tremor) AND (Writing devices)          | 6               | 1               |
| (Primary writing tremor) AND (Surgical treatment)       | 8               | 1               |

| Table 2: Results highlighting similarities between primary writing tremor and focal hand dystonia |
|--------------------------------|-----------------|-----------------|
| Study                         | Number of patients in study | Technique/Modality used to assess disease | Result highlighting similarity with focal hand dystonia |
| Ravits 1985\textsuperscript{11}  | 4                  | Electromyography | Showed that PWT patients responded positively to anticholinergic treatment. |
| Elble 1990\textsuperscript{7}  | 5                  | Electromyography | Showed that PWT patients responded positively to anticholinergic treatment. |
| Byrne 2005\textsuperscript{9}  | 1                  | Transcranial Magnetic Stimulation | Using TMS, they showed that the motor map corresponding to the first dorsal interosseous muscle in PWT patients before treatment with Botulinum toxin was initially displaced posterolaterally, and returned to a normal position after treatment with low dose Botulinum toxin A |
| Singer 2006\textsuperscript{9}  | 5                  | Electromyography | Improvement in both clinical findings as well as EMG studies after treatment with low dose Botulinum toxin A |
| Schreglmann 2015\textsuperscript{9} | 1                  | Clinical Assessment | Showed presence of abnormal posturing and presence of a mirror tremor in PWT, similar to mirror dystonia seen in focal hand dystonias. |

PWT: Primary writing tremor; EMG: Electromyography
Table 3: Results highlighting similarities between primary writing tremor and essential tremor

| Study                 | Number of patients in study | Technique/Modality used to assess or treat disease | Result highlighting similarity with focal dystonia of hand |
|-----------------------|----------------------------|--------------------------------------------------|---------------------------------------------------------|
| Ohye 1982[17]         | 1                          | Stereotactic selective thalamotomy                | Successfully treated a PWT patient with stereotactic selective thalamotomy |
| Bain 1995[1]          | 21                         | Electromyography Accelerometry                    | Showed that propranolol was beneficial in 4 out of 12 patients, and primidone in 3 out of 4 PWT patients. Alcohol also improves symptoms in 33% of cases. |
| Jimenez-Jimenez 1998[16] | 1                       | Clinical Assessment                                | Showed reduction in symptoms in PWT after alcohol consumption |
| Minguez-Castallenos 1999[14] | 1                       | Thalamic DBS                                       | 86% improvement in tremors following Thalamic DBS |
| Racette 2001[13]      | 1                          | Thalamic DBS Accelerometry                         | Near complete response of Thalamic DBS in PWT |
| Lyons 2013[15]        | 1                          | Thalamic DBS Accelerometry                         | Complete resolution of symptoms following thalamic DBS |
| Jhunjhunwala 2017[12] | 10                         | MRI Voxel based Morphometry Diffusion tensor imaging | Showed significant atrophy in cerebellum in PWT similar to essential tremor |

DBS: Deep Brain Stimulation; MRI: functional Magnetic Resonance Imaging

Table 4: Results suggesting primary writing tremor is a separate entity

| Study/year | No. of patients in study | Technique/Modality used to assess or treat disease | Result suggestive of Primary Writing Tremor (PWT) being a separate entity |
|------------|--------------------------|---------------------------------------------------|--------------------------------------------------------------------------|
| Berg 2000[23] | 3                        | Functional MRI                                    | Cases of PWT show activation patterns in brain which share aspects of both ET and writer’s cramp. |
| Mondugno 2002[20] | 7                      | EMG, TMS                                          | Normal short latency ICI in PWT unlike ET and writer’s cramp |
| Battista 2015[22]       | 1                        | Levodopa/Carbidopa                                | Showed improvement in tremor following Levodopa/ Carbidopa, which is not seen in ET and writer’s cramp |

TMS: Transcranial Magnetic Stimulation; ICI: Intracortical Inhibition

from writer’s cramp and to a less extent from ET also.[20] Some researchers also suggested a scheme to highlight the relationship of some PWT cases to generalized essential tremor and others to generalized dystonia.[23]

Thus, further studies involving a greater number of subjects are required to understand the phenomenology of PWT better.

**Pathogenesis of Primary Writing Tremor**

The exact pathophysiology of PWT is still controversial. While recent research has shed some light into the neurophysiology of PWT, a lot of work still remains to be done in order to obtain a clearer picture. A few important aspects are detailed below:

**Insights from electromyography**

Electromyography (EMG) has been conducted in several studies on PWT, and a diverse set of results have been obtained. In healthy people, the activity of writing produces periods of rhythmic EMG activity in the range of 4–7.7 Hz.

In patients with PWT, rhythmic EMG activity is obtained of similar frequency and magnitude, however, it was more sustained in nature and not easily broken down into periods.[11] In addition, patients with PWT did not have an excessive overflow of EMG activity into the proximal muscle such as the biceps or triceps compared to normal people. This is unlike writer’s cramp where there was sharply increased EMG activity in proximal muscles. The pattern of EMG activity shows considerable variation among the patients with PWT. One study reported three out of seven cases to have an alternating pattern of EMG activity (i.e., there is alternate flow of activity into the agonist and antagonist muscles) while the other four had co-contractions.[20] On the other hand, another study with 21 cases reported alternating (9 cases), alternating/extensor (2 cases), extensor only (4 cases), co-contracting (3 cases) as well as indeterminate (3 cases) patterns.[11] Yet another study also reported alternating pattern of EMG in the antagonist muscles evoked by stretch of pronator teres.[11]

**Reciprocal inhibition of H reflex**

The H reflex, or Hoffmann’s reflex, refers to the reflex contraction of muscles after low intensity (below motor threshold) electrical stimulation of Ia sensory afferents (higher intensities would induce a direct motor response) present in the nerves innervating the muscles. It is a bit similar to the deep tendon reflexes used clinically (for instance the knee jerk reflex), however, the stimulation here is an electrical stimulation of the nerve and not mechanical, and the H reflex does not involve the muscle spindle itself.

It was found by Hoffmann that normally the magnitude of the H reflex is decreased on contraction of the antagonist muscle (after stimulation of the antagonist peripheral nerve). This is referred to as reciprocal inhibition of H reflex. This reciprocal inhibition is found to be reduced in dystonia and is used as a sensitive (but not specific) method for detecting abnormality in dystonia.[24]
Reciprocal inhibition of the median nerve H reflex was found to be normal in patients with PWT by multiple studies unlike writer’s cramp, where it was found to be reduced.[1,20,25] In PWT patients, the study found no differences in the size of H reflex responses obtained before or after radial nerve stimulation for any conditioning-test shock interval between -5 and + 75 ms from both patients and controls.[1] However, in case of writer’s cramp there was a significant difference at conditioning-test intervals of 20, 30, as well as 50 ms. Thus, this lends more credence to the theory that PWT might not be related to dystonia.

**Intracortical inhibition**

Intracortical inhibition (ICI) is one of the most popular techniques used to study the electrophysiology of the brain in many neurologic diseases.

The technique relies on transcranial magnetic stimulation (TMS) which is a unique modality first used in 1986 to explore the electrophysiology of the brain.[26] It uses small pulses of magnetic field to transfer the corresponding electric field to the brain (thus bypassing the high resistance skull) to stimulate the excitatory synaptic inputs. When two such impulses are given, specifically a conditioning impulse and a test impulse, separated by a predetermined amount of time (referred to as the interstimulus interval, or ISI), then it was found that depending on the ISI, the conditioning impulse would either facilitate or inhibit the intracortical circuits. An ISI of 1-7 ms caused inhibition, while an ISI of 10–30 ms would cause facilitation.

There is evidence to suggest that reduced inhibition of intracortical circuits could contribute to the development of PWT. It was seen that short latency ICI was reduced in the dominant hand of a patient with PWT.[20] This is similar in some ways to focal dystonia. However, in focal dystonia, this reduction was bilateral, while in PWT it occurred only with respect to the dominant hand. However, on the other hand, another study showed that intracortical and spinal excitability are abnormal in writer’s cramp and in some cases of ET, but normal in patients with PWT, suggesting that PWT is distinct from writer’s cramp and to a less extent from ET also.[20]

Other TMS-based methods might also shed some light into the pathophysiology of PWT. One study was performed using TMS to measure changes in duration of silent period.[27] Normally, TMS of the motor cortex during tonic muscle contraction has been shown to produce a motor evoked potential, or MEP, followed by a silent period in the electromyogram, because of central inhibitory mechanisms. The study showed a significant shortening of this silent period during near maximum voluntary contraction bilaterally in six PWT patients compared to seven healthy controls. However, during ordinary writing, no such difference was found.

**Insights from neuroimaging**

Studies done using functional magnetic resonance imaging (fMRI) have revealed the roles of different neuroanatomical structures in the pathophysiology of PWT to a considerable extent. In particular, the cerebellum and cingulum appear to be involved heavily, along with some areas of the frontal lobe. The tremor was shown to be associated with increased activity of the cerebellum bilaterally, along with bilateral activation of the parietal lobule (which they speculate to be a result of increased neuronal activity at the cortical level), contralateral premotor area (area 6), as well as ipsilateral prefrontal area.[21] On the other hand, another study showed that the cerebellum as well as the cingulate motor area showed less activity in cases of PWT while there was increased activity of primary and supplementary motor areas.[28]

A recent study that used MRI as well as advanced image processing techniques such as voxel-based morphometry and diffusion tensor imaging found that in patients with PWT there was predominantly gray matter atrophy in frontal lobe as well as the cerebellum, along with white matter changes in the frontal lobe and cingulum.[13]

Another interesting study was performed recently using graph-based neural network analysis on resting state fMRI and structural MR imaging to study the resting state functional brain connectivity.[29] In this analysis, the brain is first modeled as a connection of various nodes representing different brain regions and edges which represent the functional connection between them. Two parameters- “clustering coefficient” signifying the degree of local connectivity of every node to all the other nearby nodes and “intermodal path length” (signifying the distance between two nodes and hence the efficiency of information transfer between them) were used to assess small world brain connectivity. After the analysis, it was found that altered small world brain connectivity (decreased clustering coefficient and increased path length) was seen in PWT, particularly in the bilateral medial cerebellum. Since the cerebellum is one of the major components involved while writing, it is possible that decreased connectivity between the cerebellum and the other regions of the brain might be involved in the pathogenesis of PWT.

**Treatment of Primary Writing Tremor**

There are no randomized controlled trials or comparative studies done for the treatment options for PWT and hence there are no specific guidelines for their treatment.[30] The treatment of these tremors is symptomatic.[31] The treatment modalities available currently are: medical treatment, botulinum toxin (BoNT), surgical management (lesional surgery, magnetic resonance guided focussed ultrasound, deep brain stimulation), as well as adaptive strategies and occupational therapy.[30,32] Other modalities like TENS (transcutaneous electrical nerve stimulation) which has improved outcome in writer cramp has not been proven to be beneficial and it actually worsens the tremor.[33]

**Medical treatment**

Many drugs via oral route have been tried for the treatment of PWT, namely propranolol, primidone, diazepam, topiramate, as well as anticholinergics like trihexyphenidyl
and alcohol [Table 5]. The PWT patients do not appear to respond as successfully to the typical anti-tremor medications as patients having ET and Parkinson’s associated tremor.[34]

According to one study only four out of 12 subjects had an improvement in their writing tremor after administering propranolol.[1] Another study used both intravenous and oral propranolol and four out of five patients receiving intravenous propranolol had obvious improvements in tremor bursts measured via EMG.[5] In the same study, six patients took 120–240 mg oral propranolol and all of them reported improvement in writing tremor. Primidone exhibited improvement in three out of four patients in one study[1] but only mild and unsatisfactory benefits in another study with 18 patients.[34] There was a reported improvement in four out of 12 and one out of three patients respectively with anticholinergics like trihexyphenidyl.[1,7]

Researchers in one study concluded that one patient on trihexyphenidyl (up to 6 mg/day) responded moderately but discontinued after 1 year because of side effects like constipation, dry nose, and throat.[10] 10 patients who had oral benzodiazepines only experienced mild and unsatisfactory improvement in tremor.[34] Overall about 60% of patients treated with medical therapy reported that they obtained some benefit from it.[34]

**Botulinum toxin**

BoNT is given intramuscularly and acts at the neuromuscular junction by inhibiting the release of acetylcholine. It has been found that motor maps in cortex of PWT patients are shifted posteriorly and there is evidence that BoNT can reverse this shifting.[9]

One study had four patients (two males and two females) with mean age 53.4 ± 14.9 years and mean age of tremor onset 48.4 ± 15.5 years. All of them were administered BoNT type A under electromyographic guidance in 2–3 sessions (dose 10u–12.5u). One patient reported marked improvement while the other three patients experienced moderate improvement in tremor at 1-year follow-up.[10]

Another study described moderate to marked improvement in 17 patients in which BoNT was used.[34]

Researchers in one study reported symptomatic improvement in two patients after administration of BoNT (200u intramuscular) for 4–5 months[1] [Table 6].

**Surgical management of primary writing tremor**

**A. Lesional surgery**

Surgery is opted in patients resistant to medical treatment. Possible side effects of surgery include confusion, weakness, speech, and balance problems. In one study, three patients with PWT refractory to medical treatment were selected to undergo stereotactic selective thalamotomies of ventral intermediate nucleus.[17] Under local anesthesia, they used Leskell’s stereotactic apparatus to introduce two semimicroelectrodes that could record electrical activity and thus help delineate subcortical
structures based on their characteristic neural activities and help decide the final point for the therapeutic lesion. Thereafter, these electrodes were replaced with coagulation electrodes and the lesion was made. The first patient, a 47-year-old male clerk who had gradual worsening of PWT and nervousness associated with writing underwent a series of two operations in a period of 13 days; his writing became normal thereafter. He experienced recurrence of symptoms in 1 year and got a third surgery done after 2.5 years after which his writing became completely legible without tremors and there were no complaints at follow-up after 2.5 years of third surgery. The second patient was a 36-year-old male schoolteacher who had PWT since fifth grade. Stereotactic thalamotomy was performed and resulted in complete disappearance of symptoms. Similar results were obtained with thalamotomy in the third patient, a 49-year-old male who had tremor on writing for 10 years. None of the patients experienced any motor, sensory, or mental side effects from surgery.

B. Magnetic resonance guided focussed ultrasound (MRgFUS)

It is a new upcoming modality which is considered when medical treatment or BoNT fails to improve patient outcome. It is an incision-less intracranial ablation procedure in which the key nuclei of the thalamus, namely ventral intermediate and ventralis oralis posterior nuclei are disrupted. These nuclei are a part of the motor cortex-basal ganglia-thalamus-cortex loop and thalamo-cerebellum pathway and are suspected to be dysfunctional in task-specific movement disorders. Their ablation leads to excellent symptomatic benefit in patients. Meng described a 60-year-old right-handed male patient with a history of writing tremors and spasms for 20 years which worsened over past 8 years with a clinical rating scale for tremor (CRST) score of 30. This patient was then treated with MRgFUS ablation and immediate benefit was reported. On follow-up at 1, 3, and 6 months the benefit was sustained.[35]

As of now, there are no comparative studies to support MRgFUS over stereotactic radiosurgery or deep brain stimulation. This modality clearly has some advantages; the effect is immediate, and as the procedure is non-invasive, there is no risk of intracranial infections or hemorrhage. On the other hand, it has its own shortcomings as well; specifically, an increased chance of developing sensory disturbances and disequilibrium. The long-term effectiveness of this modality is not known as well. Hence, further investigation is required to prove the benefit of this modality in PWT.

C. Deep brain stimulation

Deep brain stimulation (DBS) is a surgical procedure through which electrodes are implanted in the targeted region of the brain. They deliver electric impulses that block or change the abnormal neuronal activity causing the symptoms. DBS has various applications in movement disorders, chronic pain, epilepsy, psychiatric illness, etc., DBS that involves the thalamus (Thalamic nucleus ventralis intermedius or Vim and posterior subthalamic area) has been tried in patients with PWT with notable success.[13,15,34,36] [Table 7]. In one study, a 40-year-old male patient with type B PWT since the age of 16 years who only had partial response to drugs like propranolol and benzodiazepine and made unsuccessful efforts at learning to write with his left hand finally underwent DBS of left Vim nucleus at an amplitude of 2.2 volts with pulses of 60 ms width and frequency of 130 Hz. His clinical tremor rating scale score declined from 27 (17.3% of maximum score) to 4 (2.56% of maximum score) with no functional disturbances.[37] At 1 year follow-up he continued to have the same improvement in his tremor.[14] Another study reported a 66-year-old female patient with 6-year history of medical treatment resistant PWT causing disability with her occupation. Left vim nucleus DBS was performed with stimulation parameters: amplitude 1.5 volts, pulse width 60 ms, and rate 180 Hz. She reported complete amelioration of tremor and

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### Table 6: Studies reporting botulinum toxin in the treatment of primary writing tremor

| Study                  | Number of patients | Gender ratio | Dose of botulinum toxin | Outcome                                                                 | Follow up |
|------------------------|--------------------|--------------|-------------------------|------------------------------------------------------------------------|-----------|
| Bain 1995[1]           | 2                  | --           | 200U                    | Both patients improved.                                                 | 4-5 months|
| Papapetropoulos 2006[10]| 4                  | 1:1          | 10-12.5U                | One patient reported 3 and other 3 reported 2 on a scale of 0, no effect; 1, mild improvement; 2, moderate improvement; 3, marked improvement | 1 year    |
| Ondo 2011[34]          | 17                 | --           |--                       | Benefit 1.9±1.0 on a 0-4 scale (0=no benefit, 1=mild but not satisfactory benefit, 2=moderate benefit, 3=marked benefit, 4=complete tremor cessation) | --        |

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### Table 7: Studies reporting deep brain stimulation in the treatment of primary writing tremor

| Study by                          | Number of patients | Nuclei stimulated | Results                        |
|-----------------------------------|--------------------|-------------------|--------------------------------|
| Minguez-Castellanos 1999[21]      | 1                  | Thalamic nucleus ventralis intermedius | 86% improvement                |
| Racette 2001[20]                  | 1                  | Thalamic nucleus ventralis intermedius | Nearly complete control        |
| Blomstedt 2009[35]                | 1                  | Posterior subthalamic area              | Complete control               |
| Lyons 2011[21]                    | 1                  | Thalamic nucleus ventralis intermedius | Complete control               |
| Ondo 2011[36]                     | 5                  | ventral intermediate (VIM) thalamic nucleus | 3.4±0.5 n=5                    |

*Mean efficacy based on a 0-4 scale (0=no benefit, 1=mild but not satisfactory benefit, 2=moderate benefit, 3=marked benefit, 4=complete tremor cessation)
The advantages of DBS over lesional surgery are that it is a minimally invasive and reversible procedure, and its settings can be customized as per patient’s clinical status. Side effects are generally minimal and reversible including headache, infection, risk of brain haemorrhage (<1%) and paresthesias in face and limbs.[14]

Adaptive strategies and occupational therapy

Writing devices

Writing devices have shown improvements in writer’s cramp and hence have been tried for the treatment of PWT. The principle behind these devices is the substitution of action made by distal muscles (which are normally used when writing and are affected in PWT) with proximal muscles, that is, elbow and shoulder muscles which are unaffected while writing.

In one study, nine patients with PWT were selected and seven out of them had failed medical treatment.[38] These patients were asked to write and draw spirals with and without the orthotic devices. Their writing and drawing scores with and without the device were compared and showed excellent improvement.[38]

As medical therapy in most of the candidates in the above study failed, they became candidates for surgical modalities, but considering the tremendous benefit provided by these writing devices, these can be considered instead of surgery. These devices have advantages like immediate benefit, cost-effectiveness, easy usage, and better compliance when compared with oral medications, BoNT injections, and surgery.

Conclusion and future research

PWT is one of the more common task-specific tremors seen in the clinic. Yet the current understanding about the disease is very limited. The etiology of this disorder is debated and requires further research. On the one hand, it has been shown to exhibit focal dystonic features and improve with the administration of BoNT, thus putting it within the broader spectrum of dystonia, the definition of which in 2013, by a consensus committee of the Movement Disorders Society, was amended to include tremors as a possible phenotype of dystonia. On the other hand, it has also been shown to improve after treatment with drugs such as primidone and beta blockers, as well as thalamic DBS, suggesting a possible link with ET. Yet other features of this disorder differ considerably from either of the two above-mentioned spectra.

Hence, further research with more statistical power is sorely needed to clearly elucidate the pathophysiology of this tremor. This will possibly help in putting together a management protocol for this disorder, which at this moment also lacks the necessary evidence to strongly support a particular line of medical (or surgical) treatment. Thus, studies are also required to compare the efficacy of the different drugs and new upcoming modalities such as MRgFUS, and even the relative efficacy of DBS of different parts of the brain, in particular the Vim nucleus of the thalamus (which has shown promise), and internal Globus Pallidus, which has traditionally been the target for other dystonic disorders.

Roles of the authors

Abhigyan Datta and Nitya Batra did the literature search and wrote the first draft. Sanjay Pandey contributed in manuscript preparation by writing the first draft, review, and critique.

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Conflicts of interest

There are no conflicts of interest.

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