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From Pen to Pain: Exploring the Challenges of Writer's Cramp with Treatment - A Narrative Study

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Abstract: ***Introduction: Writer's Cramp (WC) is a prevalent yet enigmatic form of dystonia, characterized by co-contraction of antagonist muscles during writing, alongside involuntary flexion or extension of fingers and/or wrist. Methodology: This study employed a narrative review approach utilizing articles sourced from Google Scholar. The search parameters were restricted to articles published from 2015 until the conclusion of 2023. Discussion: With a prevalence ranging from 3.8 to 80 per million individuals, WC is considered an orphan disease, typically manifesting around the age of 38. It falls under the umbrella of dystonia, which are broadly categorized into primary and secondary forms based on etiology. Primary dystonia, often genetically influenced or of unknown cause, are further classified by age of onset. Childhood-onset dystonia typically begin in lower limbs and spread, while adult-onset dystonia usually initiates in the upper body and may spread depending on the site of onset. While the basal ganglia have long been implicated in the pathogenesis of dystonia, establishing a direct relationship between dysfunction within this brain region and dystonic symptoms remains challenging. Some studies suggest a deficit of dopamine in the basal ganglia may contribute to dystonia development. Nevertheless, neurosurgical interventions targeting the thalamus and globus pallidus have shown promise in alleviating symptoms, highlighting the intricate neural circuitry involved. Summarizing current treatment options for WC, including pharmacotherapy, botulinum toxin injections, physical therapy, and neurosurgical interventions. Conclusion: This abstract provides a comprehensive overview of WC, shedding light on its clinical characteristics, and ongoing efforts on its treatment Understanding these complexities is crucial for advancing diagnostic and therapeutic approaches in the management of this debilitating condition. Future prospective: This article synthesizes current knowledge on WC, focusing on its epidemiology, clinical presentation, and its treatment. The scope of an article on Writer's Cramp (WC) encompasses various aspects related to the disorder, aiming to provide a comprehensive understanding on its treatment.***

Keywords: writer's cramp, dystonia, Botulinum toxin

1. Introduction

Writer's Cramp (WC) is the most common task specific form of dystonia. With a prevalence of 3.8–80/1,000,000 people it ranks among the orphan diseases. The mean age of onset is 38 years. Clinically, patients present with co-contraction of antagonist muscles during writing. In some cases, involuntary flexion of one or several fingers and/or the wrist are the main complaints. Others show extension of their fingers and/or the wrist during writing or the abnormal posture may be accompanied by tremor.[1]

In general, dystonia's may be classified etiologically into primary dystonia's, in which dystonia is the main sign and the cause is genetic or unknown; and secondary dystonia's, in which dystonia may be one of several disease manifestations and the cause may be identifiable. Primary dystonia is further classified based on age of onset. Childhood-onset dystonia (<28 years of age) usually starts in the lower limbs, trunk or upper extremities and frequently spreads to the rest of the body. Adult-onset usually begins in the upper half of the body with a risk of spread to other body parts depending upon the anatomic site of onset. Dystonia also can be classified by body part affected as focal (one body part), segmental (two or more contiguous body parts), multifocal (two non-contiguous areas), hemi dystonia, or generalized.[2]

Even when the etiology of the disease is known, and the age of diagnosis and distribution of symptoms on the patient's body have been determined, it is difficult to establish the pathophysiology behind this disorder. For years, it has been believed that the primary cause of writer's cramp is a dysfunction within the basal ganglia; however, finding a direct relationship between a dysfunction of the basal ganglia and the manifestation of the symptoms of dystonia is difficult. Some studies have shown that a deficit of dopamine in the basal ganglia may lead to the development of dystonia. However, there are neurosurgical procedures within the thalamus (thalamotomy) and the posterior-ventral part of the globus pallidus (pallidotomy) that can alleviate the symptoms of primary dystonia. [3]

The standard treatment includes botulinum toxin (BoNT) injections into affected muscles. However, BoNT injections are not always helpful and may cause a number of side effects. As alternative treatment approaches training programs have been developed. [1]

Objectives

- To assess the Challenges of Writer's Cramp disorder.
- To assess the physical therapy, occupational therapy, and medications to improve hand coordination and muscle control.

2. Methodology

- **Search Strategy:** A narrative search was conducted in PubMed, Scopus, google scholar and Web of Science using keywords such as 'writer's cramp', 'dystonia', 'Botulinum toxin'. Reference lists of selected articles were hand-searched for additional relevant studies.
- **Inclusion Criteria:** Articles published in English between January 1980 and April 2024 were included. Studies focusing on writer's cramp - a focal dystonia, covering etiology, diagnosis and treatment, were considered.

- **Data Extraction:** Relevant information on writer's cramp was extracted. Key concepts, methodologies, findings, and implications were documented.
- **Data Analysis:** Qualitative analysis using thematic and narrative synthesis methods was performed. Themes related to the writer's cramp, ethiology, diagnosis, treatment and future directions identified and synthesized.
- **Dissemination:** Findings will were be shared through medical journals, online health platforms, social media groups focused on neurology or movement disorders, conferences or seminars, and collaborations with healthcare professionals for articles or educational materials. Creating informative content, sharing personal experiences, and engaging with relevant communities can help spread awareness and understanding of the disorder.

3. Discussion

Etiology

Early reports attributed writer's cramp to psychiatric conditions and neuroses, but recent evidence refutes this claim. Writer's cramp is primarily considered a physical disorder rather than a psychiatric one. Prolonged and repetitive hand use is a commonly reported trigger for writer's cramp. Approximately 5% of patients with writer's cramp have a family history of a similar condition, such as an intronic variant in the ARSG gene, may play a role in writer's cramp. DYT1, a genetic mutation, is a rare cause of writer's cramp. DYT6, DYT7, DYT13, and abnormalities linked to Chromosome 18 have been found in patients with task-specific dystonia.

Diagnosis:

The purpose of the physical exam on task-specific dystonia is to confirm diagnosis, identify the specific triggers of the dystonia, determine the muscles involved in the movement and exclude other potentially confounding conditions. The key features of the history include identification of the precipitating actions that lead to the dystonic movement.

History and Examination:

When evaluating task-specific dystonia (such as writer's cramp), several important steps guide the diagnosis. First, identify the specific actions or tasks that trigger dystonic movements. Elicit detailed information about the characteristics of these movements, including their type and severity. Consider any factors that mitigate or worsen dystonia. During the examination, observe dystonia both at rest and during relevant tasks. Additionally, assess any behavioural adaptations that the patient may use. While routine electromyogram (EMG) is not typically useful for diagnosis, multiple wire EMGs during dystonic activation can help identify the affected muscles.

Electromyography (EMG):

Electromyography (EMG) plays a crucial role in diagnosing focal dystonia like writer's cramp. By analysing muscle activity, EMG reveals cocontraction of agonist and antagonist muscles, along with abnormally prolonged bursts of activity. These findings strongly suggest focal dystonia. When evaluating patients with diagnostic uncertainty, multiple wire EMGs during dystonic activation can help identify the affected muscles. Overall, EMG provides valuable insights for accurate diagnosis and management.

Rating Scales:

When evaluating writer's cramp, various rating scales are utilized for research and therapeutic assessment. These include general dystonia scales (such as the Burke-Fahn-Marsden Scale or Unified Dystonia Rating Scale) with subsections for arm dystonia. Additionally, specific scales like the Arm Dystonia Disability Scale (ADDS) and Writer's Cramp Rating Scale (WCRS) focus on writer's cramp. Computer-based and kinematic writing analyses objectively quantify writing speed, pressure, and stroke fluency. However, there is no consensus on the optimal rating scales or outcome measures for writer's cramp intervention studies.

Differential Diagnosis:

Differential diagnosis involves ruling out other conditions like non-task related dystonia, parkinsonism-associated dystonia, carpal tunnel syndrome, neuropathies, and psychogenic movement disorder. Accurate diagnosis is essential for effective management and personalized treatment strategies.

Treatment

Treatments for writer's cramp include: 1) oral medications (table 1), 2) botulinum toxin injections, 3) non-pharmacological therapies, and 4) surgical procedures. It is important to discuss the goals and expectations of treatment with the patient (e.g., improved writing abilities and functional improvement and decreased discomfort, spasms, abnormal postures, and/or tremor), along with the person's demands for writing tasks, as these may influence the selection of treatment modality

1) Oral medications:

Oral medications used to treat writer's cramp are similar to those medications used for other types of dystonia, namely, those with anticholinergic, dopaminergic, benzodiazepines, GABAergic, and dopamine depleting actions. Treatment effects have been limited and variable, with about 10-20% reporting benefit such as with anticholinergics. Systemic effects and side effects frequently limit the efficacy of these medications. There are no randomized, controlled trials for oral medication treatments of writer's cramp.

Table 1: Different pharmacological treatment used in focal hand dystonia patients

Drug	Mechanism of action	Dosage	Side effects
Trihexphenidyl	Anticholinergic	2-80 mg in 3 divided dosage	Drowsiness, confusion, memory difficulty
Baclofen	Gamma-aminobutyric acid $(GABA)_b$ autoreceptor agonist	30-120 mg	Constipation, drowsiness, decrease neck and trunk control
Clonazepam	Benzodiazepines	1-6 mg	Drowsiness, sedation
Injection lidocaine with ethanol	Local anesthetic agent	5-10 mL of 0.5% lidocaine	Local irritation
Injection botulinum toxin	Interferes with the release of acetylcholine from the presynaptic terminal	Dosages different for individual muscles	Postinjection weakness

2) Botulinum toxin injections:

Currently, botulinum toxin has the best evidence for treating focal dystonia. Approximately 20–90% of patients experience symptomatic improvement with 2 Botulinum toxin - A. However, its benefit can be limited by muscle tonus reduction acting on the muscle spindle.

Botulinum toxin is the main modality of treatment in patients of Focal dystonia. Different types of botulinum toxins are available [Table 2]. Most of the therapeutic trials on focal dystonia have been with botulinum toxin type A (BONT-A). Botulinum toxin is given intramuscular and acts at the neuromuscular junction by inhibiting the release of acetylcholine. It gets internalized in the presynaptic axon where it cleaves SNARE (soluble NSF attachment protein receptor) protein, resulting in inhibition of acetylcholine exocytosis. Botulinum toxin has also been thought to have central effects. Studies in animal models show that botulinum toxin undergoes retrograde and transynaptic transport to affect the spinal cord and brain. Many studies have been conducted to assess the efficacy of botulinum toxin in Focal dystonia, with the majority of studies being focused on writer's cramp.

Table 2: Types of botulinum toxin used in focal hand dystonia patients

Type	Brand	Molecular weight	pH	Formulation	FDA approved	Available in India
Type A	Botox® (OnabotulinumtoxinA; Allergan Inc, Irvine, CA, USA)	900 kDa	7.4	Vacuum dried and reconstitution with normal saline	Yes, 1989 (hemifacial spasm, blepharospasm, strabismus, cervical dystonia, upper extremity spasticity, hyperhidrosis, detrusor over activity, chronic migraine, cosmetic)	Yes
	Dysport® (AbobotulinumtoxinA; Ipsen Ltd, Slough, UK)	L complex (600 kDa) and M complex (300 kDa)	7.4	Freeze dried reconstitution with normal saline	Yes, 2009 (cervical dystonia and cosmetic)	Yes
	Xeomin® (IncobotulinumtoxinA; Merz Pharmaceuticals GmbH, Frankfurt am Main, Germany)	150 kDa	7.4	Powder and reconstituted with normal saline	Yes, 2010 (cervical dystonia, blepharospasm, frown lines)	No
Type B	RimabotulinumB (Solstice Neurosciences Inc., Malvern, PA, USA;marketed in United States and Canada as Myobloc® Injection, as Myobloc® Injectable Solution in Korea, and as NeuroBloc® in the EU, Norway, and Iceland)	700 kDa	5.6	Myobloc: Available as injectable solution	Yes, 2000 (cervical dystonia)	No

3) Non-pharmacological therapies:

A variety of non-pharmacological treatments for writer's cramp and focal hand dystonia have been proposed, including physical therapy, occupational therapy, adaptive devices, sensory retraining, intensive practice, immobilization, and neurosurgical and neuro modulatory stimulation techniques. As seen with sensory tricks, different types of pens (typically thicker barrels and points) or modified grips may help with writer's cramp. Various hand devices have been developed to increase comfort and effectiveness of writing, as seen in 5 cases treated with a hand orthosis who demonstrated improvement in their writing ability.

4. Surgical Procedures

1) Neurosurgical Procedures:

Thalamotomy and deep brain stimulation (DBS) have been explored for writer's cramp. Thalamotomy improved or resolved symptoms, but transient neurological deficits were observed in some patients. DBS targeting different brain regions (nucleus ventrooralis [Vo], ventral intermediate nucleus [Vim], or both) showed benefits, with thalamic stimulation being more effective than pallidal stimulation. Further research is needed to understand the impact of DBS on dystonia pathophysiology and neural plasticity.

2) Non-invasive Stimulation:

Repetitive transcranial magnetic stimulation (rTMS) and transcranial direct current stimulation (tDCS) are non-invasive approaches. Decreasing cortical excitation may alleviate dystonic symptoms. rTMS targeting the primary motor cortex or premotor cortex improved handwriting and reduced writing pressure. Studies also targeted the primary somatosensory cortex, showing subjective and objective improvement in writing with real rTMS. However, optimal dose frequencies and further research are still needed. The cerebellum plays a role in dystonia via connections to the basal ganglia, thalamus, and motor cortex. Anodal cerebellar tDCS improved handwriting kinematics by reducing stroke frequency and pen pressure.

5. Conclusion

The study highlights the Challenges of Writer's Cramp for understanding the complexities of writers cramp it is crucial for advancing diagnostic and therapeutic approaches in managing this condition. This review highlights significant advancements in understanding writer's cramp etiology, diagnosis, and treatment approaches. While diagnostic tools have improved, therapeutic interventions offer hope for symptom management and enhancing overall well-being. Looking ahead, targeted strategies focusing on writer's cramp. addressing current challenges through collaborative efforts and technological advancements will be pivotal in managing writer's cramp. ultimately leading to better outcomes for affected individuals.

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