Writer's Cramp

Synonyms: focal hand dystonia, task-specific dystonia of writing, writing dystonia

Dystonia is an involuntary, sustained muscle contraction with abnormal postures. Focal dystonia affects only one body part. Writer’s cramp is a type of focal dystonia that is specific to the task of writing and is the most common focal primary dystonic disorder. Typically it begins with an abnormally tight grip while writing, progressing to increasing difficulty with the task. Excessive muscle spasms may progress proximally, leading to abduction of the arm. Although historically thought to be psychogenic, it is now recognised as a type of dystonia, and consists of both a sensory impairment with reduced spatial sensitivity, and a motor abnormality. Functional MRI suggests that there is a defect in inhibitory control that may explain the unintended activation of muscles and resulting abnormal movements. Cerebellar abnormalities have been detected, as well as abnormal dopaminergic transmission in the basal ganglia, but it is not understood whether these are the cause or the effect of writer's cramp. The main pathophysiological mechanisms explaining the disorganisation of the sensory-motor system appear to be loss of inhibition, aberrant neural plasticity, and defective learning-based sensory-motor integration.

Epidemiology

It is difficult to know the true incidence of the condition, as those affected may not seek medical attention. It is thought to be most prevalent between the ages of 30 and 50 with a mean age of onset of 38. There is a slight male preponderance but females tend to present slightly earlier. It is likely that the condition is becoming less frequent as writing with the hand is increasingly replaced by use of a keyboard.

Risk factors

5-20% give a family history of this or a similar condition and it is highly likely that there is a genetic component in these cases. Indeed, a genetic defect has been found in some patients. A case control study identified that:

- Cases had a college or university degree more frequently than controls.
- The risk of writer’s cramp increased with the time spent writing each day.
- It was also associated with an abrupt increase in the writing time during the year before onset.
- Head trauma with loss of consciousness and myopia were both associated.

The study found no association with peripheral trauma, left-handedness, constrained writing, writing in stressful situations or the choice of writing tool.

Presentation

History

- Prolonged periods of writing cause cramping, aching and inco-ordination of the hand. As the condition progresses, the duration required to produce these symptoms reduces.
- There is often an exaggeration of the normal semi-flexed position of the fingers, but hyperextension of the distal interphalangeal joint of the index finger. There may also be hyperflexion or extension of the wrist with supination or pronation.
- Symptoms may become exaggerated with attempts to write and the hand may even dart across the page with a sudden jerk.
- One third of patients have a tremor in the affected arm or hand while writing or when the arm is outstretched.
Examination
- There may be very subtle findings like mild dystonic postures developing, either spontaneously or with movement, and reduced arm swing on walking.
- Neurological examination reveals no abnormality.
- Observe the patient writing. Dystonic postures should be apparent.

Differential diagnosis[6]
- Parkinson's disease causes difficulty with writing but with micrographia and distinct physical signs.
- If other dystonias are found, there should be suspicion of a more general dystonia syndrome and neurological referral should be considered.
- Neurological signs suggest a different diagnosis, such as multiple sclerosis or Wilson's disease.
- Compartment syndrome of the forearm causes cramp and may need to be excluded if discomfort is a predominant feature.

Related conditions
- Primary writing tremor is a variant of writer's cramp with large-amplitude tremor only during writing.[7] Dystonic posturing is unusual with this condition.
- Musicians can also get focal hand dystonia if they have practised for many hours a day.

Investigations
- The diagnosis is essentially clinical. Functional assessments such as the Arm Dystonia Disability Scale (ADDS) or Writer's Cramp Rating Scale (WCRS) may be useful in some patients, as may kinematic analysis of handwriting movements (kinematics = the study of motion).[8]
- Electromyography (EMG) may show simultaneous contraction of agonists and antagonists.[9]
- Nerve conduction studies may be required to exclude a trapped nerve.
- An MRI scan may be indicated if a structural lesion is suspected.

Management[6]
- Reducing the amount of writing that is done is basic to an overuse syndrome. Use of a keyboard instead of a pen may help.
- There may be some benefit from using a wider pen or an attachment to make it wider.
- Training to write with the hand and using a modified pen grip are sometimes beneficial, as may be behavioural techniques, such as auditory grip force feedback (using an auditory signal to indicate grip strength).[10]
- Transcutaneous electrical nerve stimulation has been shown to have a significant beneficial effect.
- Treatment aimed at facilitating interdigit separation of digits 1, 2 and 3 may be beneficial by promoting beneficial spasticity.[11]
- A small study of 13 patients has shown that retraining the brain with fitness activities, task practice, learning based memory and sensorimotor training can lead to sustained improvements.[12]
- Botulinum toxin is a common symptomatic treatment, normalising writing in half of patients and producing a partial benefit in 10% with a mean length of effect of six months. It is time-consuming and its long-term effects, including loss of function due to weakness, remain controversial.[3]
- A number of drugs, especially anticholinergics and L-dopa, have been used but there have been no randomised controlled trials of any medications.
- Thalamic deep brain stimulation has been helpful in some patients.[13, 14]
- In exceptional cases, stereotactic nucleus ventro-oralis thalamotomy may be of value.[15]

Prognosis
Prognosis is variable, as is response to treatment. Remissions are uncommon, and symptoms can progress to the other hand. Some general features that are associated with poor prognosis include secondary dystonia, tremor, and long-duration or progressive symptoms.[1]

Further reading & references
- The Dystonia Society
- Guidelines on diagnosis and treatment of primary dystonias; European Federation of Neurological Societies (2010)
13. Deep brain stimulation for tremor and dystonia (excluding Parkinson's disease); NICE Interventionsal Procedure Guidance, August 2006

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