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What's in a name? Conundrums common to the task-specific disorders

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Ethical Compliance Statement

Informed patient consent was not necessary for this work. We confirm that we have read the Journal’s position on issues involved in ethical publication and affirm that this work is consistent with those guidelines
Task-specificity is a fascinating feature of a subset of movement disorders. In this issue of Movement Disorders in Clinical Practice, Prasad et al., describe a highly selective writing deficit for a single symbol in Bengali: “Letter specific dysgraphia: A silent stutter” bringing to the forefront a number of absorbing questions.¹

For example, how do we define whether a disorder is task-specific? Frequently we use clinical markers as our gold standard; subjective impairment described during history taking or an abnormality of movement observed during clinical examination. However, such markers are likely to have a different sensitivity to experimentally derived markers. For example, modern motion capture technology can record movement with a spatial accuracy and frequency beyond that consciously perceived by the human brain or witnessed by the human eye. Experimentally, task-specific dystonia is associated with abnormal neurophysiological responses (plasticity, inhibition) and these markers are sampled using experimental paradigms removed from the affected task’s context. Therefore, task-specificity appears to be a relative task-specificity. That is, a particular task is preferentially affected and causes disability to the patient. A gradient of subtle abnormalities for other tasks is likely to be present and skills most similar to the affected task may be the most affected, as their required neuronal networks are more closely related. Experimentally, we are also able to tap into more generalised changes within the nervous system which may be causally related to the pathophysiology or epiphenomena.

Task-specificity is a feature of a range of neurological disorders. Many clinical examples can be conceptualised as an interaction between the network encoding the task and the network affected by the disorder. Occasionally, focal lesions selective for an essential feature of task performance can lead to a task-specific deficit. A task-specific presentation of a diffuse disease processes is more dependent on a threshold effect; once a critical amount of dysfunction to the skill network has been induced by the disease process, a deficit will become apparent. Difficulty with more complex motor skills with high performance requirements or unique task features subserved by an otherwise ‘silent’ area of the brain may therefore unmask an insidious disease processes as only a small burden of disease may be symptomatic. The sensitivity of the individual to the deficit may also vary depending on influences such as attentional monitoring of the task and the extent the action is required for daily living. Correspondingly, writing dystonia and writing tremor are recognised presentations of the genetic and classically generalised dystonia DYT1 dystonia²,⁴ and difficulty styling hair has
been described as a presenting feature of a retired hairdresser that then developed apraxia and neuroimaging findings suggestive of a neurodegenerative process.

Considering the different functions of the neuronal network required for task performance can also help guide our assessment and management. Prasad et al. described a deficit writing a spiral symbol common to a Bengali letter and number. It would be interesting to know whether this deficit was also present across other muscle effectors e.g. if the symbol was still difficult to transcribe with the foot. This would suggest a higher level problem in motor planning. If solely in the hand the deficit of motor control is likely to be encoded at ‘lower’ levels of motor control which define the spatio-temporal dynamics of the arm muscles activated when writing this symbol.

For the isolated task-specific disorders, we then need to select the most appropriate diagnostic label. Prasad et al., selected the descriptor dysgraphia:

Upon writing the Bengali alphabet an abnormality was observed while the patient wrote the letter ‘‘bengali t’… the abnormality was restricted to the initial segment of the letter, wherein a rapid jerky movement of the hand with increased activity of the wrist extensors and flexors was observed.

How is this different to task-specific tremor and is this a variant of dystonia? Similarities are certainly seen as task specificity for a single letter or number has also been described in writer’s cramp yet the aetiology of the motor deficit is debated. Any attempt to answer such questions will be deeply rooted in the semantics of medical terminology and drilling down into the meaning of diagnostic labels is at times difficult. Medical language is considered an extended natural language, one that has emerged naturally around clinical observations. The naming of clinical diseases is iteratively updated through the interaction of clinical reasoning and new findings on the underlying causes in experimental science. This contrasts with formal languages used in computer programming, for example, which are characterised by defined semantic rules. Medical language is therefore characterised by a lack of precise semantics and we should be humble to the limitations of our classification systems constructed on such foundations. This is particularly relevant as diagnostic labels are usually embedded within their own unique literature and if not mindful we can un-knowingly overlook shared disease mechanisms and treatment options. A task-specific dystonia was considered by the authors,
but excluded, as presumably no overt abnormality of posture was seen. However, broader definitions of task-specific dystonia have been proposed in which a loss of motor control (with or without abnormal posturing) for a task is the main feature. The uniting task-specificity may be the most important descriptive label as it hints that there are shared aetiological substrates. Whether the disorder is most representative of a dysgraphia, tremor and/or dystonia can be debated.

Similarly, as so enticingly phrased in the paper’s title, is letter specific motor dysfunction a silent stutter, that is, a problem with initiation and fluency of the hand sequence? Indeed, both language and skilled action are uniquely developed skills in humans. Specifically, humans have a greater capacity than non-human primates to learn new sequences of movement in addition to genetically preconditioned stereotypical motor programs such as walking, climbing or swallowing. These higher motor programs involve flexibly compiled serial orders of movements for communication (e.g. speech, sign-language), everyday tool use (e.g. handwriting, tying shoelaces), or artistic expression (e.g. dance, musical performance)\(^{10}\). Both domains also share a common network of key neuroanatomical structures including the premotor cortices and Broca’s area\(^{10}\). These features have fuelled the idea of an evolutionary link between skilled action and language in humans\(^{10,11}\). It is therefore interesting to consider whether the reported case is an example of stutter outside the speech domain.

There are therefore many reasons why we should continue to dwell on the mechanisms underlying task-specificity and rationalise the language that we use. Task-specific disorders affect only a fragment of the broad repertoire of human movement but occupy an important and intriguing part of our clinical caseload.

References


