Progression of dystonia: learning from distorted feedback?

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Progression of idiopathic focal dystonia may be explicable on the basis of abnormal muscle spindle properties during muscle fatigue

Development of dystonia of the hand may be devastating to the professional musician or sportsperson. The results of EMG guided botulinum toxin treatment of arm dystonia may be disappointing or associated with unacceptable adverse effects, and the clinical skills required to undertake it are not universally available.

Our understanding of the natural history and pathophysiology of focal dystonias remains incomplete. Dystonia is not spasm or over activity of a single muscle or muscle group. In this issue of *J Neurol Neurosurg Psychiatry*, Rosset-Llobet and colleagues¹ describe the characteristics of dystonia in 101 musicians (*see page 949*). They highlight that dystonia commonly affects multiple overlearned activities or progresses from disrupting a single skill to several related manual tasks. Sixty per cent of the musicians in their study reported difficulties playing a second instrument. In half, the dystonia playing the second instrument appeared after a delay (in other words, the condition progressed). Progression is not unusual in other focal dystonias.²

The physiology of movement control is intimidatingly complex. As basal ganglia lesions can sometimes produce dystonia, most attempts to model the physiology of idiopathic focal dystonia are based around a supposition of abnormal basal ganglia functioning. These models are inadequate in idiopathic focal dystonia

where structural pathology in the central nervous system has not been demonstrated. Why might this be?

Learning a musical instrument (and writing) involves imprinting memories of limb movement sequences, and the relationship between them, by repetition. Ia afferent activity from muscle spindles encodes position, velocity, force and load information, as well as enabling the subject to adapt for muscle fatigue.³ This process is so refined that a musician can play instruments with different touch, vary playing posture and continue performing for hours without losing subtlety of musical phrasing and delicacy of touch. Writers and musicians characteristically use their hands and learn movement sequences in the fatigued or partially fatigued state (over 5 h a day practice in Rosset-Llobet's study). The learning of motor subroutines, such as those required for writing or playing a musical instrument, requires constancy of the relationship between muscle spindle activity and limb or digit function as muscles fatigue, otherwise estimation of position, velocity of movement and force would become corrupted as a muscle is used repetitively.

Widespread abnormalities of muscle spindle function have been demonstrated in dystonic subjects,⁴ and this may represent an endophenotypic marker for predisposition to dystonia. In contrast with normal individuals where the function remains constant, dystonic subjects

show an *increase* in muscle spindle afferent functionality with muscle fatigue.⁵ These properties may reflect inherited abnormalities of muscle spindle elasticity and thixotropy. A change in muscle spindle function with fatigue poses a problem for motor learning. In the subject with the dystonic endophenotype, motor subroutines learned in the fatigued state are based on distorted muscle spindle afferent information and are therefore inappropriate for the unfatigued state, resulting in the affected muscles moving with inappropriate force when initiating the motor subroutine in the unfatigued state. As the individual attempts to compensate by increasing opposing muscle activity, repetitive use of the hand in the dystonic posture facilitates learning of other corrupt motor subroutines, hence progression of the condition.

The relationship between peripheral and central abnormalities in dystonia, how motor programmes may become fixed or unlearned and how the many genetic predispositions with low penetrance may interact with the endophenotype described above to produce symptomatic dystonia is intriguing.

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