Abstracts from the Literature

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Reviewer's note: During the past several years, the second most common topic in the performing arts medicine literature has been focal dystonia. Dozens of scientific papers have been published in a variety of journals, and frequent news items regarding afflicted instrumentalists have appeared in both artistic and lay periodicals. For this issue, I have reviewed a representative selection of recent scientific papers on various aspects of focal dystonia in musicians and nonmusicians. The information contained herein is both intriguing and useful, not only as background but also in the treatment of this frustrating and perplexing disorder.

Altenmüller E: Focal dystonia: advances in brain imaging and understanding of fine motor control in musicians. *Hand Clin* 2001;19:525–538.

If there is one article in the literature that should be required reading for health professionals and afflicted musicians alike, this seems to be the primary candidate. Not only does the author present a thorough and clearly explained review of focal hand dystonia, its pathophysiology, and treatment options, but he begins with a most clear and cogent description of the anatomy and physiology of brain areas involved in making music. Neuroimaging techniques are discussed here as well, including the advantages and disadvantages of each. Following this discussion of the organizations of motor systems is a review of the investigations on the differences between the brains of musicians and nonmusicians. Some of the papers mentioned here have been reviewed in a previous *Abstracts* column (*MPPA* 2004;19:187–189). Perhaps the most thought-provoking section comes next, covering the brain's sensorimotor plasticity and sensorimotor learning. Although many of the mechanisms of sensorimotor learning and processing during music performance have yet to be clarified, the author makes a convincing case for the theory that focal dystonia as a cortical sensorimotor mislearning syndrome may be caused by abnormal plasticity. Why dystonia cannot be overcome easily by retraining of appropriate sensorimotor patterns may be due to the strong linkage of musicians' movements to emotions (musician personality and the frequent coexistence of phobias and panic attacks) and the limbic system. Altenmüller concludes with a detailed description of available treatment options and an excellent bibliography (current to 2001).

Rosenkranz K, Williamon A, Butler K, et al: Pathophysiological differences between musician's dystonia and writer's cramp. *Brain* 2005;128:918–931.

Not all patients with focal hand dystonia have a strict history of excessive hand use; patients with musician's dystonia (MD) spend hours daily focused on instrumental practice, whereas many patients with writer's cramp (WC) have a history of normal hand use. The authors tested whether 7 patients with MD and 6 patients with WC had different pathophysiologic deficits by examining the spatial pattern of sensorimotor organization in the motor cortex. Focal vibration of a single hand muscle produced sensory input, while the excitability of corticospinal outputs to the vibrated and other hand muscles was evaluated with transcranial magnetic stimulation. In healthy nonmusicians, vibration increased the amplitude of motor-evoked potentials and decreased the short-latency intracortical inhibition in the vibrated muscle, while having the opposite effect on the nonvibrated muscles. The pattern of sensorimotor integration was abnormal in both MD and WC subjects, but the nature of the deficit differed between them. Whereas vibration had little effect on cortical excitability on WC, it strongly reduced short-latency intracortical inhibition in all hand muscles, irrespective of spatial organization, in MD. The data are consistent with a model in which musical practice in healthy musicians leads to beneficial changes in organization of the motor cortex, but in MD these progress too far and begin to interfere with movement rather than assist it. The fact that sensory input had no effect on motor output in patients with WC suggests that sensory information from the hand may play a smaller role in provoking pathologic changes in WC than in MD.

Garraux G, Bauer A, Hanakawa T, et al: Changes in brain anatomy in focal hand dystonia. *Ann Neurol* 2004;55:736–739.

The authors note that no consistent anatomic abnormality has been reported in focal hand dystonia (FHD). They used voxel-based morphometry to analyze high-resolution magnetic resonance images in 36 patients with unilateral FHD (31 with writer's cramp and 5 with musician's dystonia) and in 36 controls. Findings included a significant bilateral increase in gray matter volume in the hand representation area of the primary somatosensory cortex and to a lesser extent in the primary motor cortices. These findings are con-

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sistent with disordered cortical representation of the hand in the primary sensorimotor cortical region (S1M1), as suggested by functional studies. The anatomic nature of the changes strongly emphasizes the relevant and critical role played by these regions in the pathophysiology of FHD. Despite the absence of direct evidence, the authors believe that bilateral disturbances in S1M1 of patients with primary FHD support the notion that both hemispheres might be originally affected by genetic and/or epigenetic factors that render patients with dystonia more vulnerable to environmental factors, such as repetitive stereotyped motions. The presence of anatomic changes in the perirolandic cortex for the unaffected hand and for the affected side suggests that these disturbances may be, at least in part, primary.

Blood AJ, Flaherty AW, Choi J-K, et al: Basal ganglia activity remains elevated after movement in focal hand dystonia. *Ann Neurol* 2004;55:744–748.

Relatively little is known about the role of the basal ganglia (BG) in focal hand dystonia (FHD). Unilateral and bilateral finger-tapping tests were administered to 8 patients with FHD and to 5 matched controls. Functional magnetic resonance imaging showed persistence of BG activity after the tasks had ended. Effects of bilateral tapping on BG activity during rest blocks showed greatest increases in the left putamen and right globus pallidum and were duplicated bilaterally in those patients doing unilateral tapping. The effects were observed also after tapping with the nondystonic hand, possibly suggesting an underlying malfunction or dystonic pathophysiology of the motor system, rather than a correlate of dystonic postures induced by performing the tapping task. The authors suggest also that inhibitory control of the BG may be faulty in FHD. Over time, this effect may contribute to the development or expression of other neural abnormalities observed in FHD, such as altered cortical maps and increased cortical excitability. They state that the increases observed in "resting" activity may mask BG activity in standard imaging contrast analyses.

Sohn YH, Hallett M: Disturbed surround inhibition in focal hand dystonia. *Ann Neurol* 2004;56:595-599.

Surround inhibition (SI), or suppression of excitability in an area surrounding an activated neural network, is a physiologic mechanism to focus neuronal activity and to select appropriate neuronal responses. Disturbances in SI could account for various movement disorders. The authors tested the functional operation of SI in the motor cortex of 7 patients with focal hand dystonia (FHD) and 7 matched controls. Transcranial magnetic stimulation was set to be triggered by self-initiated voluntary flexion of the index finger. During this movement, motor-evoked potential amplitudes from the little finger muscle were significantly suppressed in healthy subjects but enhanced in dystonic patients. This finding suggests that the operation of SI is impaired in patients with FHD. Alterations in various cortical inhibitory mechanisms have been reported in patients with FHD. However, these changes may not represent changes in basal ganglia function specifically related to dystonia (see previous abstract) but rather may be nonspecific in that they are also observed in various other neurologic problems of both central and peripheral origin. The authors' data support the idea that disturbed SI is a principal pathophysiologic mechanism of dystonia.

Hirata Y, Schulz M, Altenmüller E, et al: Sensory mapping of lip representation in brass musicians with embouchure dystonia. *Neuroreport* 2004;15:815–818.

Embouchure dystonia is characterized by involuntary and uncontrollable muscle movements of the mouth, face, and jaw. Brass players affected with this condition have difficulty in forming an embouchure, including involuntary jaw movements and lack of lip coordination. There are both parallels and dissimilarities between embouchure dystonia and focal hand dystonia. This study investigated whether embouchure dystonia also is related to a cortical disorder or abnormality. The somatosensory homuncular representation of 8 normal and 8 dystonic subjects was compared using magnetoencephalography and gap detection sensitivity of the lips. All dystonic subjects were former professional brass instrumentalists. Relative to controls, the patient's digit representations, and especially the thumb, were shifted in a lateral direction toward the lip representational zone. Patients' upper lips showed decreased sensitivity compared with their lower lips, an asymmetry that was absent in controls. This infers that there probably is a close relationship between decreased upper lip sensitivity and occurrence of embouchure dystonia, although the actual cause of the decrease is not yet known; several theories are posited. Because brass players often stimulate hand and mouth synchronously, the simultaneous stimulation could produce an altered hand-and-mouth relationship in the somatosensory cortex, which might lead to favoring development of dystonia.

Currà A, Agostino R, Dinapoli L, et al: Impairments of individual finger movements in patients with hand dystonia. *Mov Disord* 2004;19:1351–1357.

To study the pathologic kinematic pattern of finger movements in dystonic patients and to determine whether dystonia impairs individual more than nonindividual finger movements, the authors analyzed repetitive finger-thumb oppositions through a three-dimensional motion analysis system. Nine controls and 9 patients with hand dystonia or hand and other involved areas comprised the study group. During the tasks, normal and dystonic subjects performed finger flexions more rapidly than extensions and invariably paused longer before extension than before flexion. Patients were slower and paused longer than controls in both individual and nonindividual oppositions. During individual finger movements, patients were disproportionately slow during extension and in the pause before extension. This

bradykinesia indicates that dystonia impairs individual more than nonindividual finger movements, including these oppositions. Owing to the high degree of motor cortex activation required for extending the fingers during individual oppositions, the authors attribute the observed kinematic abnormalities to underactivation of the primary motor cortex during movement in focal hand dystonia.

Jabusch H-C, Vauth H, Altenmüller E: Quantification of focal dystonia in pianists using scale analysis. *Mov Disord* 2004;19:171–180.

Focal hand dystonia (FHD) in pianists causes loss of instrumental skills and provokes irregularities in playing. It has been difficult to quantify this disorder. This study compares 8 professional pianists with FHD with 8 healthy professional pianists using a newly developed Scale Analysis technique as well as other methods. Key velocities and timing parameters were measured. In 5 of the dystonic pianists, follow-up examinations were done after treatment with botulinum toxin A. In affected hands, significantly higher mean standard deviations of timing parameters were seen compared with healthy pianists' hands. After treatment, significant improvements in performance parameters were monitored by Scale Analysis. Data from this method correlated with those of the Arm Dystonic Disability Scale scores. The authors conclude that Scale Analysis is a precise and effective tool for quantification of FHD in pianists. It is independent of rating methods and allows reliable follow-up examinations during treatment.

Schuele S, Jabusch H-C, Lederman RJ, et al: Botulinum toxin injections in the treatment of musician's dystonia. *Neurology* 2005;64:341–343.

Many treatment options for focal task-specific dystonia do not seem to produce sustained benefit. This study reports on 84 consecutive musicians with focal task-specific dystonia from a German clinic treated with botulinum toxin A. Three musicians had embouchure dystonia. Mean treatment duration was 23 mos, and the number of injections per musician ranged from 2 to 29 (average, 7.4). Treatment outcome was assessed by subjective estimation of playing before and after treatment and self-rating of treatment response. Fifty-eight musicians experienced improvement from the injection treatment, 38 of whom stated that treatment led to a noticeable improvement in their performance ability. Six instrumentalists indicated that their dystonia had improved sufficiently after an average of 4.4 injections that they no longer required treatment, while 30 reported long-term benefits in their ability to perform. On the negative side, 26 of the 84 said they had no response to the injections or played worse following them, including the 3 wind instrumentalists with embouchure dystonia.