The Brighter Side of Music in Dystonia

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Objective: To report a patient with genetically proven DYT1 dystonia who shows dramatic improvement in symptoms while playing the piano.

Design: Case study.

Setting: Sobell Department for Motor Neuroscience and Movement Disorders, Institute of Neurology, University College London, England.

Patient: A 49-year-old right-handed male civil servant.

Main Outcome Measures: The patient was videotaped, and electromyographic activity was recorded from the splenius capitis, sternocleidomastoid, and orbicularis oculi muscles, while he was (1) at rest, (2) playing an electric piano with auditory feedback, and (3) playing an electric piano without auditory feedback (ie, when the sound of the piano is turned off).

Results: At baseline, the patient had generalized dystonia with prominent upper limb, neck, and facial involvement. While he was playing the piano, there was an instant and almost complete improvement in dystonia symptoms. The improvement was also noticeable when he played the piano without auditory feedback. There was a significant reduction in electromyographic activity for all recorded muscles when he played the piano, compared with his baseline electromyographic activity.

Conclusion: This is a unique case of “paradoxical” improvement in dystonia symptoms with activity (ie, playing a piano), in contrast to the typical worsening of dystonia symptoms with activity. We discuss the possible mechanisms underlying this phenomenon.


One of the most intriguing features of primary dystonia is the variability of abnormal muscle activity relative to the context in which movement is attempted (eg, the exquisite task specificity of focal hand dystonia or the phenomenon of the geste antagonistique).1 We present a unique case of an amateur pianist with genetically proven DYT1 dystonia who shows dramatic improvement in generalized dystonia symptoms while playing piano.

Methods

A 49-year-old right-handed male civil servant first noticed abnormal neck movements with head turning to the right at 13 years of age. Abnormal postures spread to affect the face, hands, and trunk, and a diagnosis of primary generalized dystonia was made at 15 years of age. Treatment with tetrabenazine and anticholinergics produced a moderate benefit. A thalamotomy was performed when he was 18 years of age, but he had a limited response to the procedure. Genetic analysis revealed 904_906delGAG in the TOR1A gene.

His “musical” history started at the age of 7 years when he began taking weekly piano lessons, practicing 20 minutes each day. At 14 years of age, he started playing the church organ. Presently, he continues to play piano as a hobby and the organ for a church congregation. He has always noticed clear and sustained reduction of dystonia while playing, and he enjoys playing because it relieves his symptoms. He has noticed that other manual tasks, including writing and typing, have not improved his symptoms.

The patient was videotaped while he was (1) at rest, (2) playing an electric piano with auditory feedback, and (3) playing an electric piano without auditory feedback (ie, when the sound of the piano is turned off). To measure the muscle activity during each of the 3 conditions, we recorded electromyographic activity from neck muscles and orbicularis oculi muscles (eAppendix).

Results

At baseline, he had generalized dystonia with prominent upper limb involvement, jerky torticollis to the right, and blepha-
rospasm (segment 1 of video, http://www.archneurol.com). There was no improvement in dystonia symptoms when he was listening to music (segment 2 of video) or imagining playing. While he was playing the piano, there was an instant and almost complete improvement in generalized dystonia symptoms (segment 3 of video). The improvement was still present, although less striking, when he played the piano without auditory feedback (segment 4 of video). However, dystonia symptoms returned within minutes after he stopped playing (segment 5 of video). There was no significant improvement in dystonia symptoms when he was writing (segment 6 of video). As expected, there was a significant reduction in electromyographic activity for all recorded muscles when he played the piano, compared with his baseline electromyographic activity (eTable).

The classical association between dystonia and music occurs when professional musicians develop dystonia while performing, typically in the body part with the greatest technical and spatiotemporal demands required by the instrument. Rather at odds with this typical task-specific emergence of dystonia, our patient displays an almost complete resolution of generalized dystonia while playing the piano.

One might consider this phenomenon to be similar to a sensory trick. There is complex afferent input and integration across sensory modalities while playing a musical instrument, and this input, similar to the simple sensory input provided by a typical sensory trick, may act to “reset” abnormal dystonic motor output. Auditory feedback appears to be an important component of this process in our patient because the level of improvement was reduced when auditory feedback was masked. However, an important difference from typical sensory tricks is that, in our patient, dystonia also improved in distant body parts not engaged in playing. This effect persisted throughout the time of playing and for a short period thereafter.

This patient’s experience is quite unique and very different from most of the patients with primary generalized dystonia, in whom dystonic movements and postures get worse with action. Fahn coined the term paradoxical dystonia to describe unusual patients like ours in whom dystonia symptoms get better with certain actions. For example, in blepharospasm, speaking, whistling, humming, or chewing may improve dystonic eye spasms. Fahn described rare patients who purposefully and constantly performed particular actions in an attempt to relieve their symptoms, including a woman with “paradoxical” torticollis who became “a victim” to ceaseless knitting, as her cervical dystonia would remit when she knitted. Similarly, our patient reports that he often plays for the relief from his dystonic symptoms.

The mechanism underlying the paradoxical improvement of dystonia with voluntary movement has not been investigated, perhaps because this phenomenon is rather exceptional. Nevertheless, the occurrence of paradoxical dystonia suggests that motor programs for some voluntary activities remain preserved in generalized dystonia and may be performed in a nondystonic manner. As to why this occurs only occasionally and only with some specific actions and not with others, we can merely speculate. In our patient, who is an amateur musician, playing the piano or church organ was the sole activity that relieved his symptoms, and in this context we suggest that there is a relation between his playing skills (which he had developed in childhood, even before the onset of clinical dystonia) and the improvement seen in his dystonia symptoms. The proposed neural networks involved in both musical performance and primary dystonia are complex, and there are many common substrates. It is possible that, in our patient, the improvement of dystonia depends on a modulation of the “dystonic” brain network that occurs when he plays the piano. Fludeoxyglucose F18–positron emission tomographic studies have disclosed an abnormal metabolic pattern affecting the posterior putamen/globus pallidus, cerebellum, and supplementary motor area in DYT1 dystonia. These areas overlap with regions of increased activity during music playing in healthy musicians, where activation of the basal ganglia, lateral cerebellar hemispheres, and supplementary motor area are also described. Competing activation of these key areas during musical performance may disrupt the abnormal “dystonia network” and, by changing activation patterns within the sensorimotor network, can result in a near-normal motor performance during piano playing and the disappearance of dystonia symptoms in other body parts. However, to our knowledge, this case is a unique example of dystonia improving with piano playing, and it is not known whether other musicians affected by generalized dystonia report a similar experience. Curiously, similar effects of music on motor performance have been recognized in musicians with other basal ganglia disorders, including Tourette syndrome and parkinsonism, which suggests that, by performing highly skilled and well-consolidated motor activities (such as piano playing), some individuals are able to overcome abnormal motor output related to basal ganglia dysfunction.

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Author Contributions: Dr Kojovic had full access to all the data in the study and takes responsibility for the integrity of the data and the accuracy of the data analysis. Study concept and design: Kojovic, Pareés, Sadnicka, Rothwell, Edwards, and Bhatia. Acquisition of data: Kojovic, Pareés, Sadnicka, Kassavetis, and Rubio-Agusti. Analysis and interpretation of data: Kojovic, Saifee, and Bolognina. Drafting of the manuscript: Kojovic, Pareés, and Sadnicka. Critical revision of the manuscript for important intellectual content: Kassavetis, Rubio-Agusti, Saifee, Bologna, Rothwell, Edwards, and Bhatia. Statistical analysis: Kojovic. Administrative, technical, and material support: Kassavetis. Study supervision: Rothwell and Bhatia.
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REFERENCES


Announcement

Archives of Neurology will publish a special theme issue in March 2013 on Genomics/Genetics and Epigenetics. We invite the submission of papers as Neurological Reviews, Clinical Trials, Original Contributions, Case Reports, Images in Neurology, and Research Letters. Papers submitted by September 1, 2012, will have the best opportunity to be considered for this theme issue.