Focal dystonia in musicians: phenomenology, pathophysiology and triggering factors

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Background: Musician’s dystonia is a task-specific movement disorder that manifests itself as a loss of voluntary motor control in extensively trained movements. In many cases, the disorder terminates the careers of affected musicians. Approximately, 1% of all professional musicians are affected. The pathophysiology of the disorder is still unclear. Findings include: (i) reduced inhibition in different levels of the central nervous system, (ii) maladaptive plasticity, e.g. in the somatosensory cortex and in the basal ganglia and (iii) alterations in sensorimotor processing.

Methods: Review of the literature.

Results: Epidemiological data demonstrated a higher risk for those musicians who play instruments requiring maximal fine-motor skills. For instruments where workload differs across hands, focal dystonia appears more often in the more intensely used hand. In psychological studies, musicians with dystonia had more perfectionist tendencies than healthy musicians. These findings strengthen the assumption that behavioural factors may be involved in the etiology of musician’s dystonia. Hereditary factors may play a greater role than previously assumed.

Conclusions: We propose a heuristic model that may explain the relatively high incidence of focal dystonia in musicians. This model assumes the coactions between a predominantly genetically determined predisposition and intrinsic and extrinsic triggering factors.

What is musician’s dystonia?

Focal dystonia in musicians, also known as musician’s cramp or musician’s dystonia, is a task-specific movement disorder which presents itself as muscular incoordination or loss of voluntary motor control of extensively trained movements whilst a musician is playing the instrument. For those who are affected, focal dystonia is highly disabling and in many cases the disorder terminates musical careers [1].

Musician’s dystonia may be classified according the task specifically involved. For example, embouchure dystonia may affect coordination of lips, tongue, facial and cervical muscles and breathing in brass and wind players, whereas pianist’s cramp and violinist’s cramp may affect the control of finger, hand, or isolated arm movements. According to recent estimates, one percent of all professional musicians are affected [1]. In contrast, in the general population, prevalence of focal dystonias, including writer’s cramp, blepharospasm and cervical dystonia, is estimated as 29.5 per 100 000 in the USA and 6.1 per 100 000 in Japan [2,3].

Typically, musician’s dystonia occurs without pain, although muscle aching can present after prolonged spasms. Lack of pain distinguishes it from repetitive strain injury or occupational fatigue syndrome. It is important to make this distinction bearing in mind that prolonged pain syndromes may lead to symptomatic dystonia, possibly because of the degradation of sensory receptive fields in the somatosensory cortex [4]. The loss of muscular coordination is frequently accompanied by a co-contraction of antagonist muscle groups. For example, in pianist’s cramp, the co-activation of wrist flexor and wrist extensor muscles is frequently observed. In Fig. 1, typical postures of dystonic finger and embouchure movements in musicians are shown.

What causes musician’s dystonia?

The etiology of musician’s dystonia is not completely understood but is probably multifactorial. It may develop in individuals with a family history of dystonia.
and may be related to alterations in the basal ganglia circuitry [6]. Most studies of musician’s dystonia reveal abnormalities in three main areas: (i) reduced inhibition in the motor system at cortical, subcortical and spinal levels (ii) reduced sensory perception and integration; and (iii) impaired sensorimotor integration (for recent reviews on this topic see also [7,8]. The latter changes are mainly believed to originate from dysfunctional brain plasticity. Such a dysfunctional plasticity has been described in the sensory thalamus [9]. Disorganized motor somatotopy could be found in the putamen of patients suffering from writer’s cramp [10]. Finally, there is growing evidence for an abnormal cortical processing of sensory information as well as degraded representation of motor functions in patients with focal dystonia. In monkeys, repetitive movements induced symptoms of focal hand dystonia and a distortion of the cortical somatosensory representation [11], suggesting that practice-induced alterations in cortical processing may play a role in focal hand dystonia.

Indeed, using somatosensory-evoked potential technology, it was demonstrated that in the somatosensory cortex, the topographical location of sensory inputs from individual fingers overlap more in patients with writer’s cramp than in healthy controls [12]. Similar observations have been made using magnetoencephalography [13]: Elbert et al showed that there is an overlap of the representational zones of the digits in primary somatosensory cortex for the affected hand of musicians with dystonia compared with the representations of the digits in non-musician control subjects.

A clinical sign, which emphasizes the important role of sensory-motor integration in the pathophysiology of musician’s dystonia, is the ‘sensory trick’ phenomenon. This phenomenon is known from patients with cervical dystonia: touching the face contralateral, but also ipsilateral to the direction of head rotation, can reduce or abolish involuntary muscle activity [14]. In a similar way, musicians suffering from dystonia frequently experience marked improvement in fine-motor control when playing with a latex glove, or when holding an object, for example a rubber gum, between the fingers, thus changing the somatosensory input information [15].

Interestingly, in musicians with hand dystonia, an association exists between the instrument group and the localization of focal dystonia. In instruments with different work load, different complexity of movements or different temporospatial precision for both hands, focal dystonia appears more often in the more heavily used hand. Keyboard musicians (piano, organ, harpsichord) and those with plucked instruments (guitar, e-bass) are primarily affected in the right hand. All these instruments are characterized by a higher workload in the right hand. Additionally, guitar playing requires higher temporospatial precision in the right hand compared to the left hand. Bowed string players who have a higher workload and complexity of movements in the left hand are predominantly affected in that hand [1].

Several predisposing factors have been identified, such as male gender [16] as well as a positive family history [17], which might constitute a particular vulnerability or predisposition to musician’s dystonia. A genetic contribution with an autosomal-dominant inheritance has been discussed. Additional extrinsic and intrinsic factors may trigger the manifestation of musician’s dystonia on the basis of a given predisposition. Intrinsic triggering factors are, for example,
suggest that musicians with dystonia demonstrate an activity of the brain became obvious. These changes of the error-related oscillatory performance, changes of the error-related oscillatory circuits. In fact, in an electroencephalography study in dorsolateral, orbitofrontal and motor frontostriatal circuits. In the context of brain adaptation, it is intriguing that recent findings point at a different origin of musician's dystonia when compared to other focal dystonias. Generally, focal hand dystonia has been suggested to be a maladaptive response of the brain to repetitive performance of stereotyped and attentionally demanding hand movements [11]. However, not all patients with focal hand dystonia have a strict history of excessive hand use. For example; in contrast to professional musicians, many patients with writer's cramp have a history of average hand use. Rosenkranz and collaborators [28] could convincingly demonstrate that these two patient groups have different pathophysiological deficits by examining their sensory-motor integration

Psychological factors may trigger musician’s dystonia

Although musician’s dystonia clearly is a neurological movement disorder, as has been demonstrated by Marsden and Sheehy [20], recent studies have revealed additional, though subtle behavioural dysfunctions in patients with focal dystonia. In two studies, we demonstrated that psychological conditions in musicians with focal dystonia differed from those of healthy musicians [21,22]. Anxiety disorders, and above all social phobias and specific phobias occurred more often in musicians with dystonia. Additionally, musicians with focal dystonia were found to have highly perfectionist tendencies. In the Freiburg Personality Inventory (FPI-R), musicians with dystonia showed more somatic complaints than healthy musicians. Taken together, musicians with dystonia showed a pattern of exaggerated perfectionism, of social phobia and specific phobias that was not seen in healthy musicians or those with chronic pain. In keeping with these results, Bugallo and colleagues [23] demonstrated increased perseveration in the Wisconsin Card Sorting Test, indicative for defective set shifting and higher intensity of obsessive compulsive disorder in patients with focal dystonia. With respect to brain mechanisms involved in triggering dystonia, these findings may reflect a pattern of complex neurophysiological dysfunction involving dorsolateral, orbitofrontal and motor frontostriatal circuits. In fact, in an electroencephalography study in pianists, suffering from dystonia, electrophysiological correlates of such a defective set shifting could be convincingly demonstrated. In comparison with healthy pianists, those with dystonia could not appropriately deactivate the preparatory cortical activation in anticipation of playing a C-major scale, when a No-go condition was introduced requiring them not to play the scale [24]. Similarly, when pianists suffering from focal dystonia were tested with their healthy hand in a task requiring precise error monitoring during piano performance, changes of the error-related oscillatory activity of the brain became obvious. These changes suggest that musicians with dystonia demonstrate an enhanced conscious evaluation of errors which might be related to increased behavioural adjustments. Furthermore, during the feed-forward control processes when playing piano, the interactions between the medial prefrontal cortex and the lateral prefrontal cortex were different from those in healthy pianists. Thus, the coordination between brain regions and the corresponding large-scale integration of anticipatory and evaluative processes seem to be anomalous in patients with musician’s dystonia, even in tasks performed by the ‘healthy’ non-affected hand [Herrojo-Ruiz M, Strübing F, Jabusch HC, Altenmüller E (unpublished data)].

Is musician’s dystonia special?

As has been pointed out, musician’s dystonia has a higher incidence than other task-specific dystonias, e.g. writer’s cramp. The remarkably strong adaptations of the central nervous system to prolonged practice in skilled musicians have been demonstrated by several researchers [for a review see 25]: For example, comparison of the brain anatomy of skilled musicians with that of non-musicians shows that prolonged instrumental practice leads to an enlargement of the hand area in the motor cortex and to an increase in gray matter density corresponding to more and/or larger neurons [26]. These adaptations appear to be particularly prominent in all instrumentalists who have started to play prior to the age of ten and correlate positively with cumulative practice time. It is not only motor areas, however, that are subject to anatomical adaptation. Professional violinists have been shown to possess enlarged sensory areas corresponding to the index through to the small (second to fifth) fingers of the left hand [27], whilst their left thumb representation is no different from that of non-musicians. Again, these effects are most pronounced in musicians who started their instrumental training prior to the age of ten and correlate closely to the lifelong cumulative practice times.

In the context of brain adaptation, it is intriguing that recent findings point at a different origin of musician’s dystonia when compared to other focal dystonias. Generally, focal hand dystonia has been suggested to be a maladaptive response of the brain to repetitive performance of stereotyped and attentionally demanding hand movements [11]. However, not all patients with focal hand dystonia have a strict history of excessive hand use. For example; in contrast to professional musicians, many patients with writer’s cramp have a history of average hand use. Rosenkranz and collaborators [28] could convincingly demonstrate that these two patient groups have different pathophysiological deficits by examining their sensory-motor integration

physical disorders resulting in local pain and/or intensified somatosensory input. Traumatic injuries, nerve-entrapment or overuse injury may also lead to a degradation of sensory-motor representation at several levels of the sensory-motor circuits [18].

Extrinsic triggering factors, according to epidemiological findings [19], are spatial and temporal sensorimotor constraints as well as musical and social constraints typical of the performance situation in classical music.
capacity. They tested the lowering of cortical motor thresholds in response to vibration inputs to the hand. Whereas vibration had little effect on cortical excitability in patients suffering from writer’s cramp, it strongly reduced thresholds in all hand muscles in patients with musician’s dystonia. In the healthy musicians, thresholds were in-between those of the latter two groups. These data are consistent with a model in which musical practice in healthy musicians leads to beneficial plastic adaptation if the motor cortex, i.e. reduction in motor thresholds and increase in motor excitability, whereas in patients suffering from musician’s dystonia these progress too far and begin to compromise movement patterns rather than assist it.

A specific feature of musician’s dystonia therefore seems to be its close link to overuse and over-practice. It is worth mentioning that musician’s dystonia has emerged relatively recently during the early romantic period, when eminent virtuosos like Paganini or Franz Liszt pushed technical demands to new limits. The first proven records of this condition date only back to 1830, when the ambitious pianist and composer Robert Schumann developed a pianist’s cramp, affecting the fine-motor control of the middle finger of his right hand [29]. As a supposedly precipitating factor, Robert Schumann had dramatically increased his piano practice schedules up to 7 h a day to attain the pianistic technique necessary to compete with the eminent virtuosos of the early romantic period. However, besides prolonged practice time and musculoskeletal overuse, psychosocial stressors and personality factors may have contributed to his disorder. According to his diaries and the written testimonials of his friends, Schumann tended to compulsive working behavior, to harsh self-criticism, to anxiety and depression and to frequent and excessive alcohol consumption. Already at this time his physicians discussed whether he presented a psychogenic or a neurological condition [30].

Taking our results on occurrence of anxiety and perfectionism in musicians suffering from dystonia into account, we suggest that additionally specific affective conditions play an important role as a triggering factor. Music performance is strongly linked to the emotions in a way which is not comparable to any other activity in human life. On the one hand, music is the ‘language of emotions’ and is able to communicate positive or negative feelings. On the other hand, instrumental music is based on extreme spatio-temporal sensorimotor precision which can be scrutinized by both the musician and the audience. Therefore, many musicians experience strong and contrary feelings, with the joy of performing alongside the fear of playing wrong notes or the fear of failure which reflects the extremely strict system of reward and punishment in professional musicianship. This double link to the emotions is a unique characteristic of making music [31].

We propose that during onset of musician’s dystonia, this link between music performance and strong emotions may facilitate the establishment of dystonic movement patterns in patients with perfectionism and anxiety. It is possible that stress-induced motor memory consolidation may contribute to fixed dysfunctional movement pattern in this subgroup. The process might start with the occurrence of a dystonic movement for which the cause is unknown. Musicians with an inclination toward anxiety and extreme perfectionism may emphasize the disturbing and threatening element in the occurred wrong movement. This psychological stress might induce the cascade of emotionally induced memory consolidation that has previously been described and applied to different forms of memory and which mainly relies on noradrenergic activation of the basolateral amygdala (BLA) [32]. The primary motor cortex, which is an essential locus of representation of digital-motor sequences, receives a BLA projection [33]. Thus, it may be assumed that consolidation of early dystonic movements as dysfunctional motor programs may be facilitated by a BLA-mediated process in the primary motor cortex.

Undoubtedly, the previous outlined scenario constitutes only an epiphenomenon in a subgroup of patients with the described psychological conditions. Further support for a participation of limbic circuits in the development of focal dystonia comes from the phenomenologies of related focal dystonias, for example in calligraphists, telegraphers, money counters and golfers. Two common features can be observed: (i) the necessity for high precision and (ii) a strong emotional component. The first is related to the nature of the respective activities, the latter originates in the fact that these activities underlie a strict system of reward and punishment. In sports, this is provided by victory and defeat with the resulting advantages and disadvantages, the other activities are performed in professional contexts which mean that people’s income depends on the quality of their work. One can speculate that in all of these activities, an inclination toward anxiety and extreme perfectionism may also foster consolidation of dystonic movement patterns.

Conclusion and consequences

In summary, overuse and compulsive working behavior, spatio-temporal constraints and special psychological conditions including anxiety and extreme perfectionism may trigger the onset of the disorder on the basis of a given predisposition that may be genetically determined. It should be emphasized that the
emotional aspects of music performance, specifically the enormous professional pressure substantially contribute to stress-induced processes that may foster consolidation of dystonic movements. In part, the unyielding reward and punishment frame in the reproductive classical music scene provides a fertile ground for these stresses in musicians. This in turn could explain why, for example, improvising Jazz musicians is much less likely to develop musician’s dystonia. Here, as in many other music cultures, reproduction of the precise musical notation plays only a minor role. Learning is frequently based on imitation, and movements frequently can be selected deliberately, obeying the individual’s anatomical prerequisites.

In Figure 2, we propose a model, specifying the possible co-action between predisposition and intrinsic and extrinsic triggering factors.

With this model, we now have means at hand contributing to the prevention of musician’s dystonia. Preventing dystonia is important, because successful treatment is still a challenge. Many of the available medical approaches are only moderately effective, and other options have yet to be developed. Behavioral therapies and interdisciplinary strategies combining pharmacological and pedagogical methods are promising, but the different approaches need to be evaluated and long-term effects are still unknown (for a review see [34]).

Concerning prevention, exaggerated perfectionism and anxiety as triggering factors should be addressed in the education of musicians. This has to be started at early infancy. From the first lesson on, music educators should strive to create a friendly, supportive atmosphere focusing on creativity, curiosity and playful experiences in the world of sounds. It is not by chance that we commonly speak of ‘playing an instrument’ and not of ‘working an instrument’. Of course, structured, goal-directed learning is a prerequisite of musical mastery. Here, reasonable practice schedules, economic technique, prevention of overuse and pain, mental practice, variations in movement patterns, maintenance of motivation and avoidance of mechanical repetitions and frustration, healthy living habits, warm-ups and cooldown exercises, regular physical exercise, sufficient brakes and sleep are the cornerstones of healthy musical practice.

Finally, the role of the societal constraints should not be neglected. In the last decades, the classical music sector was inundated by CD recordings of peers in the fields. Frequently, these recordings can be regarded as ‘laboratory music’, composed in the tranquility of the studios with the help of tone engineers and electronic ‘remastering’. These recordings are considered as ‘gold standard’. They frequently create unrealistic expectations in listeners and music critics, adding stress to the performers. In addition, our classical music culture reflects the general societal pressures of the developed countries. Highest precision and efficiency are the demands we all are subjected to. In music, this frequently creates an attitude of great artistic accomplishment, which, however, frequently is not nurtured by a sufficient personal expression of emotional experience. The latter, of course, has to be collected somewhere outside the practising room. As a consequence, we, therefore, should correct our expectations and listening habits, replacing the fascination of mere perfection and virtuosity by the joy of emotional communication shared with the audience and the musicians.
Conflicts of interest

None.

References


