

Focal Hand Dystonia affecting Musicians

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You will find a full list of all references used to produce this lecture in the last few slides of this presentation.

Playing-Related Musculoskeletal Disorders (PRMDs)

“Pain, weakness, numbness, tingling, or other symptoms that interfere with (their) ability to play (their) instrument at the level (they) are accustomed to.” (Zaza et al., 1998)

Impact of PRMDs on Players

(Caldron et al., 1986)

Musicians reported their loss of facility in playing in terms of:

- Loss of speed,
- Loss of control of major motions,
- Loss of control of fine motions**,
- Loss of power (forte)**,
- Loss of finger span,
- Other.

Focal Hand Dystonia

(Bejjani et al. 1996, Elbert et al. 1998, Lederman 1998, Lederman 2002, Brandfonbrener et al. 2004, Frucht 2004)

- Painless motor disorder.
- Loss of fine motor control/coordination of individual finger movements.
- Usually involving 3rd to 5th digits.
- Seen in pianists, wind players, guitarists, string players, pipers.
- Thought to be associated with many hours of daily practice.
- Affecting between 5% and 14% of musicians.

Focal Hand Dystonia

(Bejjani et al. 1996, Elbert et al. 1998, Pantev et al. 2001)

- Related to other conditions associated with prolonged performance of rapid, alternating, and/or forceful movements of the digits (typing, data processing...).
- Movements of some fingers of a hand become involuntarily linked to those of others.
- No clear cause, no physiological mechanism determined (idiopathic).
- Treatments: temporary and palliative.

Alterations in Cortical Representation

(Elbert et al., 1995)

- Study on 9 string players: 6 violin-, 2 cello-, 1 guitar-players.
- 6 non-musicians as controls.
- Left and right hands studied.
- Left hand:
 - Digits 2 to 5 are involved in fingering the strings.
 - This involves considerable manual dexterity and enhanced sensory stimulation.
 - Thumb not involved in fingering, but grasps the neck of the instrument: small shifts in position and pressure.

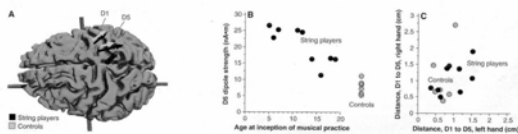


Alterations in Cortical Representation

(Elbert et al., 1995)

- Somatosensory stimulation (light superficial pressure) to fingers D1 and D5, non painful stimulation.
- Magnetic source imaging (magnetoencephalography).
- Significant shift of the cortical representation of the left hand fingers of musicians towards the midline (region corresponding to the palm of the hand) compared to controls and compared to right hand.
- Medial shift greater for D5 than for D1.
- Strength of the response also increased for the left hand of musicians (total neuronal activity: more nerve cells activated).

Increased Cortical Representation of the Left Hand Fingers in String Players (Elbert et al., 1995)

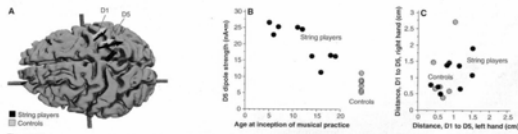


Alterations in Cortical Representation

(Elbert et al., 1995)

- Amount of increase in somatosensory cortical representations of left-hand finger D5 in musicians is dependent on the age at which the musicians started to play their instrument.
- Greater for those who started their instrument earlier: CNS reorganisation or maturation.

Increased Cortical Representation of the Left Hand Fingers in String Players (Elbert et al., 1995)



Conclusions: Use-dependent CNS Plasticity (Elbert et al., 1995)

- Cortical territory occupied by the representation of the left hand digits in string players has expanded.
- String players exhibit a use-dependent enlargement of portions of the somatosensory map in cortical representational zones of the digits of the left hand, which are used intensively to play the instrument.

Focal Hand Dystonia Prevalence per Instrument

(Tubiana & Chamagne 2000, Brandfonbrener et al. 2004, Frucht 2004)

- Piano: right hand mainly.
- Guitar: right hand more often than left.
- Violin and viola: left hand mainly.
- Cello: left hand mainly.
- Flute: left hand mainly.
- Oboe and clarinet: right hand more often than left.

Focal Hand Dystonia



(Sakai, 2006)



(Tubiana & Chamagne, 2000)

Focal Hand Dystonia



(Ackermann & Adams, 2005)



(Tubiana, 2000)

Focal Hand Dystonia Neurological Changes

(Hallett, 1998)

- Brain lesions can cause dystonia: responsible sites include basal ganglia, brainstem, thalamus, putamen.
- Dystonia can be hereditary: genetic linkage in Segawa disease (progressive dystonia).
- Dystonia occurring in Parkinson's Disease as a result of dopamine pharmacology.
- Dystonia can be produced behaviourally when synchronous sensory input leads to remapping of the receptive fields in the cortex and subsequently to a movement disorder.

Focal Hand Dystonia Neurological Changes

(Hallett 1998, Lim et al. 2001, Charness et al. 2004)

- Deficient reciprocal inhibition at spinal cord and brainstem levels: co-contraction of antagonist muscles.
- EEG studies: movement-related cortical potentials (MRCPs) show a reduced amplitude of the negative slope component (Bereitschaftspotential) associated with the preparation and initiation of movement, suggesting deficient motor inhibition. Bereitschaftspotentials precede self-paced movement and are generated in the Motor Cortex (SMA / M1).

Focal Hand Dystonia Neurological Changes

(Hallett 1998, Lim et al. 2001, Charness et al. 2004, Hallett 2004)

- EEG studies: contingent negative variation (CNV) shows deficient late negativity with hand movements in patients with writer's cramp.
- The CNV is the EEG potential that appears between a "warning" and a "go" stimulus. The CNV is thought to be generated by the basal ganglia, putamen, (subcortical) and the SMA and M1 (cortical).
- This suggests abnormal motor preparation and loss of inhibition in cortical processing.

Focal Hand Dystonia Neurological Changes

(Hallett 1998, Ibanez et al. 1999)

- PET studies: positron emission tomography measuring regional cerebral blood flow (rCBF).
- Abnormal suppression of rCBF in writer's cramp patients in the sensorimotor cortex contralaterally, premotor cortex bilaterally, cingulate cortex, supplementary motor area (SMA).
- These observations are consistent with the concept of reduced inhibition. Decreased inhibition in the sensory cortex could drive excessive motor output.
- Abnormal output of the basal ganglia?

Focal Hand Dystonia Neurological Changes

(Ikoma et al. 1996, Chen et al. 1997, Hallett 1998, Hallett 2004)

- TMS studies: transcranial magnetic stimulation measuring motor evoked potentials (MEPs). Increased excitability of motor cortex in patients with dystonia.
- Threshold intensity for the production of MEPs at rest was unchanged. With increased stimulus intensity, there was an abnormal increase in the MEP amplitude in patients compared with normals.
- In patients with writer's cramp when using double pulses at longer intervals with the muscle under study either at rest or contracted, Chen et al. found a deficiency only in the symptomatic hand and only with background contraction. Silent period following MEP was also shorter, suggesting deficiency in inhibition.

Focal Hand Dystonia Neurological Changes

(Hallett 1998, Lim et al. 2001, Hallett 2004)

- Basal Ganglia (BG): evidence that dystonia is associated with functional disturbances of the BG.
- BG organised to work in a centre-surround mechanism.
- Direct pathway (center): inhibitory synapses from **Striatum** → **Globus Pallidus** → **Thalamus**. Therefore a net excitatory pathway. Then Thalamus has excitatory effect on cortex to produce desired movement.
- Indirect pathway (surround): excitatory synapses from **Subthalamic Nucleus** → **Globus Pallidus**. Therefore a net inhibitory pathway. Suppresses unwanted movements.

Focal Hand Dystonia Neurological Changes

(Hallett 1998, Lim et al. 2001, Hallett 2004)

A reduction in the influence of the indirect pathway in the basal ganglia would lead to overactivity of the direct pathway, i.e. inhibition of the Globus Pallidus, and its influence on the thalamus would act to increase excitation of the cortex, and may explain the dystonic movements.

Focal Hand Dystonia Neurological Changes

(Byl et al. 1996, Elbert et al. 1998, Pantev et al. 2001)

Motor disturbances and distortions of receptive fields and representational zones of the digits in area 3b of the somatosensory cortex in monkeys during frequent repetition of grasp over time: breakdown, reordering and emergence of new cortical zones.

Focal Hand Dystonia Neurological Changes

(Byl et al. 1996, Elbert et al. 1998, Pantev et al. 2001)

- Disordered cortical representation of the digits in blind individuals who read Braille with 3 fingers simultaneously for many hours on a daily basis.
- Asynchronous stimulation leads to separation.

Focal Hand Dystonia Neurological Changes

(Elbert et al., 1998)

Extensive simultaneous stimulation of the digits and other types of prolonged, unusual types of sensory input can produce a use-dependent reorganisation of digital receptive fields.

Focal Hand Dystonia Neurological Changes

(Bara-Jimenez et al. 1998, Bara-Jimenez et al. 2000, Charness et al. 2004)

- Abnormalities in sensory maps: fusion of the cortical representation of the digits in the primary sensory cortex of patients with focal hand dystonia, as well as inversion of the position of the thumb and little finger.
- Abnormal temporal discrimination among dystonic subjects, correlated with the degree of severity of dystonia.
- Decreased ability for spatial discrimination in dystonic subjects (gap detection and localisation tests).
- Theoretically, spatial processing abnormality may be the result of deficient temporal processing, since sensory stimuli that occur separated in time may appear simultaneous. This could explain the increase in receptive field size and decreased spatial localisation ability.

Focal Hand Dystonia Study Protocol

(Elbert et al., 1998)

- Study involving 8 musicians affected by dystonia, 8 unaffected musicians, 9 non-musicians as controls.
- Somatosensory stimulation (light pressure).
- Tactile stimulation all digits of both hands.
- Magnetic source imaging (magnetoencephalography).
- Fusion of cortical representations of the digits of the dystonic hand.

Focal Hand Dystonia Study Results

(Elbert et al., 1998)

- Reduced distance between the representational zones of the digits in primary somatosensory cortex for the affected hand of dystonic musicians.
- Fusion of cortical digital receptive fields.
- Fusion also occurred in the cortex opposite the non-dystonic hand in 4 of 7 musicians studied.

Fusion of Cortical Representations

(Elbert et al., 1998)

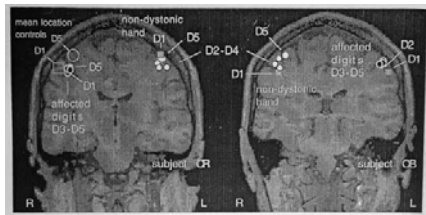


FIG. 2. A coronal MRI section through the supplementary cortex of 2 musicians CR and CB with focal hand dystonia who are presented the distal locations of digits 1-5 (D1-D5) resulting from anatomical stimulation. The large open symbols in the right hemisphere of subject CR indicate the mean location of digits for D1 and D5 in normal control subjects.

Focal Hand Dystonia

Study Results

(Elbert et al., 1998)

- Fusion also occurred in the cortex opposite the non-dystonic hand in 4 of 7 musicians studied.
- Some string players who change the hand used for fingering the strings after developing focal hand dystonia, also develop dystonia in the hand to which they switch.

Focal Hand Dystonia Study Discussion

(Elbert et al., 1998)

- “Chicken and egg situation” !
- Is cortical digital fusion a causal factor in the genesis of focal hand dystonia?
- Or did the dystonia (resulting from some other causes) produce the cortical fusion?

Focal Hand Dystonia Conclusion

(Elbert et al., 1998)

- If cortical digital fusion in response to repetitive finger movement is a causal factor in the development of dystonia, it would be possible that individuals who are susceptible to the development of focal dystonia are those who have a prior tendency toward cortical digital fusion.
- Use-dependent cortical plasticity.

Focal Hand Dystonia Treatment Options

(Elbert et al. 1998, Byl 2004)

If cortical digital fusion is the cause of dystonia:

- Intervention to break apart the fusion may be effective.
- Sensory relearning: extensive practice in tactile discriminations with individual fingers.

Focal Hand Dystonia Treatment Options

(Elbert et al. 1998, Byl 2004)

- Dystonia is a disturbance of motor production:
 - Extensive practice in making discrete individual finger movements.
 - In combination with constraint-induced movement therapy.

Focal Hand Dystonia Sensory Motor Retuning

(Candia et al. 1999, Candia et al. 2002)

- 11 professional musicians:
 - Affected by task-specific focal hand dystonia.
 - 6 piano players, 2 guitar players.
 - 3 wind players: 2 flutists and 1 oboist.
- Follow-up:
 - 3-25 months in piano and guitar players.
 - 0-4 months in wind players.
- Measurements:
 - Dystonia Evaluation Scale.
 - Smoothness of movement: spectral analysis output from a dexterity-displacement device that continuously recorded digital displacement during finger movements.

Dystonia Evaluation Scales

(Candia et al. 1999, Tubiana & Chamagne 2000)

- 0: As bad as at its worse
- 1: Slightly improved
- 2: Moderately improved
- 3: Almost normal
- 4: Normal
- 0: Unable to play
- 1: Plays several notes but stops because of blockage or lack of facility
- 2: Plays short sequences without rapidity and with unsteady fingering
- 3: Plays easy pieces but is unable to perform more technically challenging pieces
- 4: Plays almost normally but difficult passages are avoided for fear of motor problems
- 5: Returns to concert performances

Focal Hand Dystonia Sensory Motor Retuning

(Candia et al. 1999, Candia et al. 2002)

- Immobilisation by splints of one or more fingers (compensatory fingers) other than the dystonic finger.
- Repetitive exercises of dystonic finger (left free) in coordination with one or more of the other fingers for 1.5 to 2.5 hours per day over 8 days.
- Continue at home, with splint, for 1 hour per day for one year. Gradual increase of repertoire practice periods without splint.

Focal Hand Dystonia Sensory Motor Retuning

(Candia et al. 1999, Pantev et al. 2001, Candia et al. 2002)

- Each of the pianists (except one non compliant) and guitarists improved in spontaneous repertoire performance without splint at the end of treatment. 4 returned close to the level of performance they had before the onset of dystonia.
- Neuroimaging results showed normalisation of the cortical representational maps for subjects successfully treated.

Focal Hand Dystonia Sensory Motor Retuning

(Candia et al. 1999, Pantev et al. 2001, Candia et al. 2002)

The 3 wind players did not improve:

- Finger-mouth coordination not addressed in the protocol: fusion of digits and mouth cortical representational zones? Need to include exercises to differentiate between digit and mouth: finger movement while simultaneously blowing in the instrument.
- Constant pressure from thumb to hold the instrument and maintain orientation may hinder relearning.
- Posture of the digits and nature of fingering?

Focal Hand Dystonia Conclusion

(Byl et al. 1996, Elbert et al. 1998, Pantev et al. 2001, Candia et al. 2002, Byl 2004)

- Neuronal networks are particularly plastic, and are able to alter their architecture and function to afferent input throughout life.
- Cortical reorganisation and plasticity can therefore be maladaptive as in focal hand dystonia among professional musicians who train intensively.
- Behavioural techniques focusing on movement (Sensory Motor Retuning) and based on recent research on neuroplasticity may be effective.

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