Focal Hand Dystonia in Musicians

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"If I don’t practise for one day, I know it; if I don’t practise for two days, the critics know it; if I don’t practise for three days, the audience knows it"

Ignacy Jan Paderewski.

Focal Hand Dystonia


• Painless motor disorder.
• Involuntary loss of fine motor control and coordination of individual finger movements.
• Deterioration of sensorimotor skills, task-specific.
• Usually involving 3rd to 5th digits.
• Estimated prevalence of less than 1% of the population of professional musicians.

Risk Factors

(Altenmüller & Jabusch, 2010)

• 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

• Deficient reciprocal inhibition at spinal cord, brainstem, and cortical 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

• 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

• 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

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.
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.
- Cortical territory occupied by the representation of the left hand digits in string players has expanded.

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)

• 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
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
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).

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)

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
Treatment Options
(Byl et al. 1996, Elbert et al. 1998)

If cortical digital fusion is the cause of dystonia:
– Intervention to break apart the fusion may be effective.

Focal Hand Dystonia
Neurological Changes

• Reduced inhibition and increased excitation at spinal cord, brainstem, and cortical levels, leading to excessive motor output with overflow into inappropriate muscles.
• This would explain co-contraction of agonist and antagonist muscles observed in FHD.
• Altered sensory perception and maladaptive cortical plasticity.
• Impaired sensorimotor integration.

Focal Hand Dystonia
Management Strategies
(Lim et al., 2001; Schuele et al., 2005; Jabusch & Altenmüller, 2006)

• Oral anticholinergic medication: Trihexyphenidyl.
• “Botox”: Botulinum Toxin injections.
• Limitations: side-effects, dosage, leakage into adjacent muscles, patient’s response.

FHD – Management Strategies
(Lim et al., 2001; Schuele et al., 2005; Jabusch & Altenmüller, 2006)

• Limb immobilisation (Priori et al., 2001);
• Learning-based sensory training (Byl et al., 2009);
• Sensory retraining – Braille reading (Zeuner et al., 2002);
• Proprioceptive retraining (Rosenkranz et al., 2009);
• Constraint-induced therapy (Candia et al., 2002);
• Motor Control Retraining – “Slow-Down Exercise” (Sakai, 2006).

AIMS

• Investigate the effects of a combined behavioural therapy over a 12-month period in musicians affected by FHD:
  – Constraint-induced therapy.
  – Motor control retraining (Slow-Down Exercise).
• Subsidiary aim: reliability study of the outcome measure: FAM scales.
AIMS

- Investigate the long-term effects of a combined behavioural therapy in musicians affected by FHD, 3 years after completion of the initial 12-month study = 4-year follow-up:
  - Constraint-induced therapy.
  - Motor control retraining (Slow-Down Exercise).

- Subsidiary aim: reliability study of the outcome measures: ADDS, TCS scales.

Subjects

<table>
<thead>
<tr>
<th>Instrument</th>
<th>Dystonia</th>
<th>Side</th>
<th>Onset</th>
<th>Compliance</th>
</tr>
</thead>
<tbody>
<tr>
<td>Guitar 1</td>
<td>D3, D4, D5</td>
<td>R</td>
<td>2006</td>
<td>95%</td>
</tr>
<tr>
<td>Guitar 2</td>
<td>D3, D4, D5</td>
<td>R</td>
<td>1982</td>
<td>76%</td>
</tr>
<tr>
<td>Flute 1</td>
<td>D4, D5</td>
<td>L</td>
<td>2002 (D5)</td>
<td>95%</td>
</tr>
<tr>
<td>Flute 2</td>
<td>D4, D5</td>
<td>R</td>
<td>2004</td>
<td>95%</td>
</tr>
<tr>
<td>Piper 1</td>
<td>D5</td>
<td>R</td>
<td>2005</td>
<td>77%</td>
</tr>
<tr>
<td>Piper 2</td>
<td>D3, D4</td>
<td>R</td>
<td>1995</td>
<td>40%</td>
</tr>
<tr>
<td>Oboe</td>
<td>D4, D5</td>
<td>R</td>
<td>2006</td>
<td>88%</td>
</tr>
<tr>
<td>Accordeon</td>
<td>D3, Wrist, D2, D4</td>
<td>R</td>
<td>2005</td>
<td>N/A</td>
</tr>
</tbody>
</table>

Outcome Measures

- 2 test pieces: easy and medium difficulty;
- Frequency of Abnormal Movements (FAM) scale (Spector & Brandfonbrener, 2005);
- 2 ordinal Dystonia Evaluation Scales:
  - Tubiana & Chamagne Scale (TCS),
  - Arm Dystonia Disability Scale (ADDS);
- Change in metronome speed achieved during Slow-Down Exercise (Sakai, 2006).
Arm Dystonia Disability Scale

<table>
<thead>
<tr>
<th>ADDS</th>
<th>Stage Definition</th>
</tr>
</thead>
<tbody>
<tr>
<td>Stage 0</td>
<td>No dystonia</td>
</tr>
<tr>
<td>Stage 1</td>
<td>Mild difficulty playing</td>
</tr>
<tr>
<td>Stage 2</td>
<td>Moderate difficulty playing</td>
</tr>
<tr>
<td>Stage 3</td>
<td>Marked difficulty playing</td>
</tr>
</tbody>
</table>

Hypothesis

Significant differences in Frequency of Abnormal Movement Scale scores and metronome speeds would be achieved between testing sessions over time for both pieces.

Study Design

- Repeated Measures Design: subjects tested at Day 1, Day 8, then every 2 months;
- Standardised protocol;
- Standardised metronome speed for each piece.

Constraint-Induced Therapy

(Berque et al., 2010)

Home Protocol

- Week 1: constraint-induced therapy only. 2 hours per day;
- Constraint-induced: ½ hour to 1 hour per day;
- Slow-Down Exercise: ½ hour per day;
- Free playing: ½ hour per day for motivation and compliance.
- No monitoring of subjects between Month 12 and Year 4.
Outcome Measures – Reliability
(Spector & Brandfonbrener 2005, Spector & Brandfonbrener, 2007)

• Lack of evaluation of the clinical utility of common outcome measures used in studies on FHD:
  – TCS never evaluated for reliability.
  – ADDS evaluated in one study only (Spector & Brandfonbrener, 2005).
  – FAM developed by Spector & Brandfonbrener and evaluated in their study (Spector & Brandfonbrener, 2005).

Outcome Measures – Reliability Tests
ICC Model (2,1)

• Frequency of Abnormal Movement Scale:
  – Intra-rater: ICC = 0.985 – 0.999, \( p < 0.001 \), narrow C.I. (0.985 – 1.000).
  – Test-retest: ICC = 0.739 – 0.996, majority with \( p < 0.001 \), wider C.I. but robust model.

• TCS + ADDS – Ordinal Scales:
  – Intra-rater: 0.700 – 1.000, \( p < 0.001 \).
  – Inter-rater: 0.760 – 0.900, \( p < 0.003 \).
  – Fairly narrow to reasonable C.I.

Statistical Analysis – FAM Scores

• Diagnostics performed: normality and homoscedasticity confirmed.

• Two-factor parametric repeated measures analysis of variance ANOVA, with “number of abnormal movements per second” as dependent variable.

• Tukey’s post-hoc test where applicable.

• Level of significance, \( \alpha = 0.05 \).
**Mean Values for Metronome Speeds**

![Graph showing mean values for metronome speeds over time.](Image)

**Practice Profile**
(adapted from Ackermann & Driscoll, 2010)

<table>
<thead>
<tr>
<th>Question</th>
<th>S. 1</th>
<th>S. 2</th>
<th>S. 3</th>
<th>S. 4</th>
</tr>
</thead>
<tbody>
<tr>
<td>On average, how many days per week did you practise your specific exercises?</td>
<td>5</td>
<td>4</td>
<td>4</td>
<td>6</td>
</tr>
<tr>
<td>On average, how many practice sessions would you normally do per day for your specific exercises?</td>
<td>1</td>
<td>1</td>
<td>1</td>
<td>2</td>
</tr>
<tr>
<td>How long have your average practice sessions been for your specific exercises:</td>
<td></td>
<td></td>
<td></td>
<td></td>
</tr>
<tr>
<td>- Less than 15 minutes?</td>
<td>√</td>
<td>√</td>
<td>√</td>
<td></td>
</tr>
<tr>
<td>- Between 15 minutes and half an hour?</td>
<td>√</td>
<td></td>
<td></td>
<td></td>
</tr>
<tr>
<td>- Between half an hour and one hour?</td>
<td></td>
<td></td>
<td></td>
<td>√</td>
</tr>
</tbody>
</table>

**Flute Player – Month 10**
(Berque et al., 2008)

**Flute Player – Year 4**
(Berque et al., 2011)

**Limitations**

- No control group;
- Small sample;
- Missing data for the medium difficulty piece;
- Two strategies were used.

**Clinical Recommendations**

- A 1-year retraining protocol may lead to long-term benefits for musicians with FHD;
- Progress maintained with only 15 to 30 minutes of daily specific practice;
- Intensive retraining to be carried out for more than 6 months;
- The FAM scale is a useful and valid clinical tool;
- The ordinal scales showed good to very good intra- and inter-reliability.
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