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.

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Focal Hand Dystonia

(Elbert et al. 1998, Lederman 2002, Schuele & Lederman 2003, Brandfonbrener et al. 2004, Frucht 2004, Lim et al. 2004, Frucht 2009, Altenmuller & Jabusch 2010)

- 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.

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Risk Factors

(Altenmüller & Jabusch, 2010)

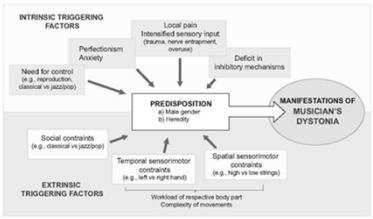

FIGURE 4. The possible interplay between predisposition and intrinsic and extrinsic triggering factors in the manifestation of musician's dystonia. (Adapted from Jabusch H, Altenmüller E. Adv Cogn Psychol 2006; 2:207-220.)

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Flute Player – Day 1

(Berque et al., 2007)

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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.

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Focal Hand Dystonia
Neurological Changes
(Hallett 1998, Lim et al. 2001, Charness et al. 2004)

- 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).

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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.

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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?

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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.

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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.

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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.

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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.

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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.

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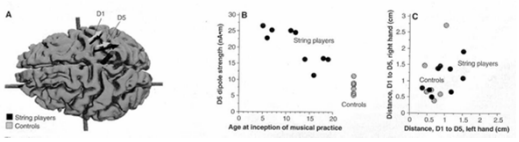
Increased Cortical Representation of the Left Hand Fingers in String Players (Elbert et al., 1995)



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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.

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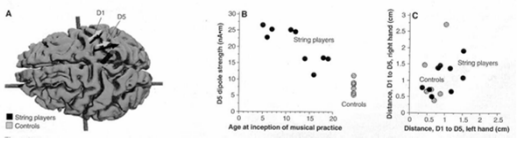
Increased Cortical Representation of the Left Hand Fingers in String Players (Elbert et al., 1995)



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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.

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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.

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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).

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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.

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Fusion of Cortical Representations
(Elbert et al., 1998)

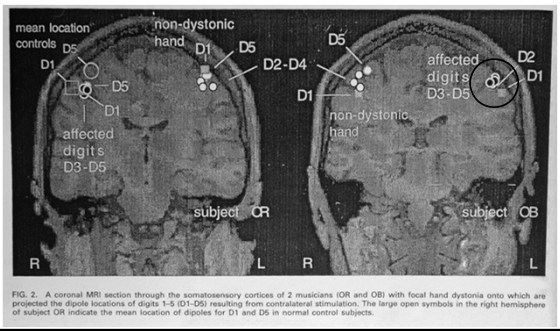

FIG. 2. A coronal MRI section through the somatosensory cortices of 2 musicians (OR and OB) with focal hand dystonia onto which are projected the dipole locations of digits 1–5 (D1–D5) resulting from contralateral stimulation. The large open symbols in the right hemisphere of subject OR indicate the mean location of dipoles for D1 and D5 in normal control subjects.

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?

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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.

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Focal Hand Dystonia
Neurological Changes
(Chen & Hallett 1998, Elbert et al. 1998, Hallett 1998, Bara-Jimenez et al. 2000, Charness 2004, Hallett 2004, Lin & Hallett 2009, Altenmuller & Jabusch 2010)

- 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.

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

- Oral anticholinergic medication: Trihexiphenedyl.
- "Botox": Botulinum Toxin injections.
- Limitations: side-effects, dosage, leakage into adjacent muscles, patient's response.

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FHD – Management Strategies

- 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).

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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.

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Berque et al. (2010)

A Combination of Constraint-induced Therapy and Motor Control Retraining in the Treatment of Focal Hand Dystonia in Musicians

Patrice Berque, BSc (Hons), MCSP, Heather Gray, MSc, MCSP, Cassandra Harkness, BSc, MCSP, and Angus McFadyen, PhD

Focal hand dystonia (FHD) in musicians is a painless task-specific motor disorder characterized by an involuntary loss of control of individual finger movements. The aim of this study was to investigate the effects of an innovative behavioural therapy intervention, aimed at normalising movement patterns, in musicians affected by FHD. Methods: Eight musicians volunteered to take part in this retraining protocol. Intensive constraint-induced therapy and motor control retraining at slow speed were the interventions. Video recordings of the subjects playing two pieces were used for data analysis. The Frequency of Abnormal Movements scale (FAM), the change in metronome speed achieved during motor control retraining, and two ordinal dystonia evaluation scales were chosen as outcome measures. It was hypothesised that there would be significant differences in the FAM scores and metronome speeds over a 12-month period. Results: For the main outcome measure, the FAM scale scores, the two-factor repeated measures ANOVA revealed a very significant decrease in the number of abnormal movements per second of instrumental playing over the 12-month period ($F = 6.32$, $df = 7$, $p < 0.003$). Tukey's post-hoc tests carried out for the FAM scores revealed that significant changes occurred after 8 months of therapy. Discussion: These results suggest that a combination of constraint-induced therapy and specific motor control retraining may be a successful strategy for the treatment of musicians' FHD. Furthermore, the results suggest that retraining strategies may need to be carried out for at least 8 months before statistically significant changes are noted. *Med Probl Perform Art* 2010; 25:139-153.

LITERATURE REVIEW

untary loss of control and coordination of individual finger movements.[1–4] It is a disorder associated with a sudden or insidious deterioration of sensorimotor skills which, in most cases, only occurs in the context of instrument playing.[5–7] Involuntary spasms, cramping sensations, abnormal hand posture, finger curling, loss of coordination during specific fingerings, fingers sticking on the keys of the instrument, irregularities in rhythm and tempi are common findings.[3,5,8–11] It more often involves digits 3, 4, and 5 (D3 to D5) of the hand[1,5,11,8] and is thought to be related to the intense and prolonged practice of rapid, alternating, and highly precise finger movement patterns.[2,5,11,8] The condition can be disabling enough to curtail a professional career.[2,5,12,6]

Prevalence Amongst Musicians

FTSD has been estimated to affect between 5% and 14% of musicians consulting performing arts clinics in the US.[5,13,1] This would give an estimated prevalence of 0.2% to 0.5% in the population of professional musicians.[5,4,8] Focal hand dystonia (FHD) occurs much more frequently in males than females. One study[11,8] revealed that 73% of instrumentalists affected were men, two others reported 80%[14] and 83%[1]. Symptoms usually begin in the third or fourth decade.[4,5,10,11]

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.

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Berque et al. (2013)

A Combination of Constraint-Induced Therapy and Motor Control Retraining in the Treatment of Focal Hand Dystonia in Musicians

A Long-term Follow-up Study

Patrice Berque, BSc (Hons), MCSP, Heather Gray, Prof D, MSc, MCSP, and Angus McFadyen, PhD

Focal hand dystonia (FHD) in musicians is a painless, task-specific motor disorder characterized by involuntary loss of control of individual finger movements. The aim of this study was to investigate the long-term effects of a combined behavioral therapy intervention aimed at normalizing finger movement patterns. METHODS: Eight musicians with FHD had taken part in the 1-year study involving intensive constraint-induced therapy and motor control retraining at slow speed as the interventions. Four of those subjects volunteered to take part in this 4-year follow-up. A quasi-experimental, repeated measures design was used, with 9 testing sessions over 4 years. Video recordings of the subjects playing two pieces were used for data analysis. The Frequency of Abnormal Movements scale (FAM) was the main outcome measure. It was hypothesized that there would be significant differences in FAM scores achieved over the 4-year period. RESULTS: The results from the ANOVA revealed a significant decrease, by approximately 80%, in the number of abnormal movements for both pieces over the 4-year period ($F = 7.85$, $df = 8$, $p < 0.02$). Tukey's post hoc test revealed that significant improvements occurred after 6 months of therapy (p values between $p < 0.001$ and $p < 0.044$). Although the results were not significant between month 12 and follow-up at year 4, the trend revealed that the progress achieved during the first year of intensive retraining was maintained at year 4. CONCLUSIONS: A 1-year period of intensive task-specific retraining may be a successful strategy with long-term, lasting effects for the treatment of musician's FHD. Results suggest that retraining strategies may need to be carried out for at least 6 months before statistically significant changes are noted. *Med Probl Perform Art* 2013; 28(2):33–46.

Focal task-specific dystonia (FTSD) affecting musicians' hands (also termed focal hand dystonia, FHD) is a task-specific, painless motor disorder characterized by an involuntary loss of control and coordination of individual finger movements, accompanied by abnormal co-contractions of agonist and antagonist muscles.[1–4] In most cases, it only occurs in the context of instrument playing, more often involves digits 3, 4, and 5 (D3 to D5) of the hand, and is thought to be related to the intense and prolonged practice of highly skilled movement patterns. The hand that performs the most complex movement patterns on the instrument is usually affected: the right hand of pianists, the right hand of guitarists, the left hand of flautists, and the left hand of string players.[5–7]

FTSD has been reported to affect 0.2% to 0.5% of professional musicians, and males are much more affected than females.[4,8,10–13] The condition can be disabling enough to curtail a professional career.[10,13]

LITERATURE REVIEW

Neurophysiology of Focal Dystonia

Extensive research on the neurophysiology of FTSD has been

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Subjects

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

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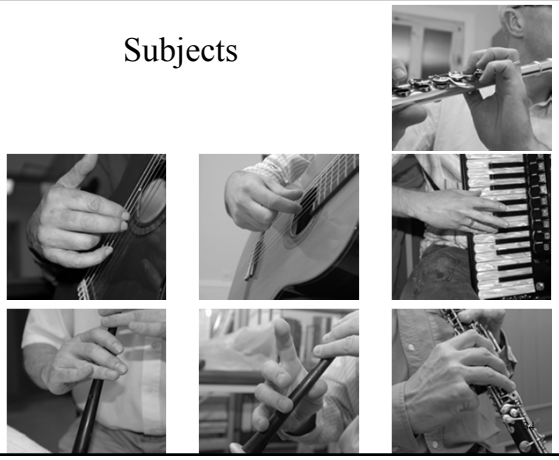
Subjects



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).

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Tubiana & Chamagne Scale

TCS	Stage Definition
Stage 0	Unable to play
Stage 1	Plays several notes but stops because of blockage or lack of facility
Stage 2	Plays short sequences without rapidity and with unsteady fingering
Stage 3	Plays easy pieces but is unable to perform more technically challenging pieces
Stage 4	Plays almost normally but difficult passages are avoided for fear of motor problems
Stage 5	Returns to concert performances

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Arm Dystonia Disability Scale

ADDS	Stage Definition
Stage 0	No dystonia
Stage 1	Mild difficulty playing
Stage 2	Moderate difficulty playing
Stage 3	Marked difficulty playing

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Hypothesis

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

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Study Design

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

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Constraint-Induced Therapy

(Berque et al., 2010)

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Constraint Induced Therapy

(Berque et al., 2007)

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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.

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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).

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Peterson et al. (2013)

VIEWS & REVIEWS

Rating scales for musician's dystonia

The state of the art

David A. Peterson, PhD
Patrice Berque, BSc
Hans-Christian Jabusch, MD
Eckart Altenmüller, MD, MA
Steven J. Frucht, MD

ABSTRACT

Musician's dystonia (MD) is a focal adult-onset dystonia most commonly involving the hand. It has much greater relative prevalence than non-musician's focal hand dystonias, exhibits task specificity at the level of specific musical passages, and is a particularly difficult form of dystonia to treat. For most MD patients, the diagnosis confirms the end of their music performance careers. Research on treatments and pathophysiology is contingent upon measures of motor function abnormalities. In this review, we comprehensively survey the literature to identify the rating scales used in MD and the distribution of their use. We also summarize the extent to which the scales have been evaluated for their clinical utility, including reliability, validity, sensitivity, specificity to MD, and practicality for a clinical setting. Out of 135 publications, almost half (62) included no quantitative measures of motor function. The remaining 73 studies used a variety of choices from among 10 major rating scales. Most used subjective scales involving either patient or clinician ratings. Only 25% (18) of the studies used objective scales. None of the scales has been completely and rigorously evaluated for clinical utility. Whether studies involved treatments or pathophysiologic assays, there was a heterogeneous choice of rating scales used with no clear standard. As a result, the collective interpretive value of these studies is limited because the results are confounded by measurement effects. We suggest that the development and wide-spread adoption of a new clinically useful rating scale is critical for accelerating basic and clinical research in MD. *Neurology*® 2013;81:589–598

GLOSSARY

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.

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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, $\alpha = 0.05$.

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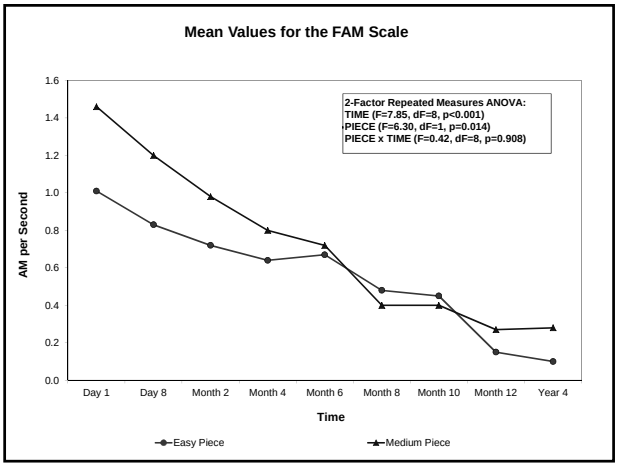

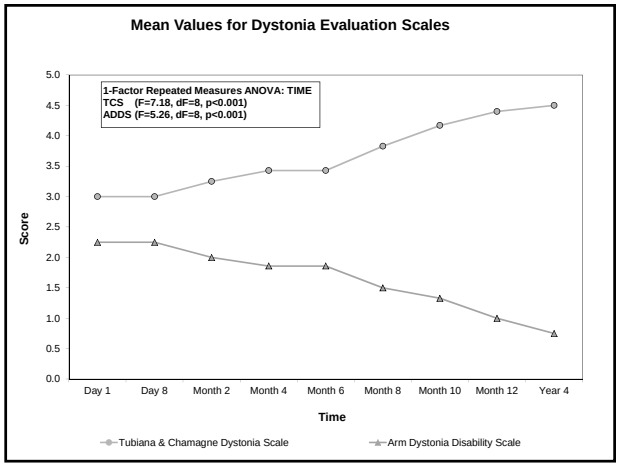

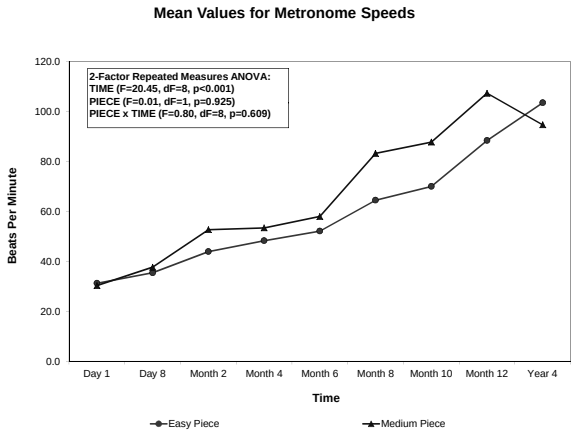

Practice Profile

(adapted from Ackermann & Driscoll, 2010)

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

Flute Player – Month 10

(Berque et al., 2008)

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Flute Player – Year 4

(Berque et al., 2011)

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Limitations

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

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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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Co-authors

- Heather Gray, Senior Lecturer, GCU.
- Cassandra Harkness, Hand Therapist Physiotherapist, Canniesburn, GRI.
- Angus McFadyen, Reader in Health Statistics, GCU.

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