

# The challenge of diagnosing focal hand dystonia in musicians

J. Rosset-Llobet<sup>a\*</sup>, V. Candia<sup>b\*</sup>, S. Fàbregas i Molas<sup>a</sup>, D. Dolors Rosinés i Cubells<sup>a,b,c</sup> and A. Pascual-Leone<sup>d,e</sup>

<sup>a</sup>*Institut de Fisiologia i Medicina de l'Art-Terrassa, Terrassa, Barcelona, Spain;* <sup>b</sup>*Collegium Helveticum, University of Zurich and ETH Zurich, Zurich, Switzerland;* <sup>c</sup>*Servei de Cirurgia Ortopèdica i Traumatologia, Fundació Althaia, Unitat Asistencial de Sant Joan de Deu, Manresa, Barcelona, Spain;* <sup>d</sup>*Center for Noninvasive Brain Stimulation, Beth Israel Deaconess Medical Center, Harvard Medical School, Boston, MA, USA;* and <sup>e</sup>*Hospital de Neurorehabilitació, Institut Guttmann, Barcelona, Spain*

## Keywords:

diagnosis, focal dystonia, hand problems, motor disorders, musicians' disorders

Received 21 July 2008

Accepted 18 February 2009

**Background and purpose:** To most clinicians, medical problems in musicians, particularly those concerning focal hand dystonia, constitute an unfamiliar domain difficult to manage. The latter can importantly influence diagnostics and the course of treatment. The purpose of this study was to enlighten the issue and to identify possible problems in diagnosing musicians' cramp within the Spanish medical community.

**Methods:** We used a brief questions' catalog and clinical histories of 665 musicians seen at our clinic for performing artists. We analyzed patients' diagnosis records in 87 cases of focal hand dystonia (13.1%). In so doing, we surveyed previous diagnoses and diverse treatments prescriptions prior to referral to our clinic.

**Results:** Referrals came primarily from orthopaedists and neurologists. The 52.9% arrived at our clinic without a diagnosis or a suspicion of suffering from focal dystonia. The most frequently attempted diagnoses other than musicians' dystonia included nerve compression, tendonitis and trigger fingers. Commonly prescribed treatments included rest, various surgical procedures, physiotherapy and oral anti-inflammatory medication.

**Conclusions:** This data depicts the diagnostic challenges of medical professionals may encounter when confronted with musician's focal dystonia.

## Introduction

Hand dystonia, also called musician's cramp, is particularly common amongst musicians, affecting one out of 200 musicians and 13% of the patients seen in performing artists' clinics [1]. Focal hand dystonia can be devastating to a musician's career. Focal dystonia is usually a painless sensorimotor condition that affects the coordination of the hand and fingers (see Fig. 1) but it can also act upon motor coordination of orofacial muscles in wind players. Characteristically, focal hand dystonia begins at the later stages of musical education, usually during the last academic years or during the course of full-time professional exercise. Concomitant to periods of intensified practice and professional stress (e.g. concerts or exams), patients typically notice uncontrolled movements, movement difficulty or ten-

sion in the most exercised body part [2]. Symptoms tend to remain exclusively movement-specific, at least at the very early stages of the disturbance (see [3] for a more extensive discussion on secondary disturbances in musicians' dystonia). Remarkably, symptoms only manifest during real attempts to play the affected musical instrument and not during the mere imitation of the involved movement. Moreover, some affected guitarists may experience dystonic problems only during attempts to play *classical* and not whilst playing the electric guitar [4]. Men are more often affected than women [2,4,5].

It is perhaps the result of its bizarre appearance, its association to stressors, and its relatively marked context-specificity that misconceptions about the origins of focal dystonia still arise. Focal hand dystonia, a sensorimotor disorder of neurological origin with no important if any association to psychopathology, was once considered a psychiatric disturbance [6–8]. Today, neuroscience research in animals and humans suggests that prolonged repetitive work may develop into dysfunctional brain plasticity, and that this may be importantly involved in the development of focal hand

Correspondence: Jaume Rosset-Llobet, Ctra de Montcada 668, E08227 Terrassa, Barcelona, Spain (tel.: +34 937844775; e-mail: info@institutart.com).

\*These authors contributed equally to this work.



**Figure 1** The figure shows one of the most common symptoms in focal hand dystonia in musicians: finger flexion whilst attempting to play a musical instrument, in this case the guitar. Note the involuntary and exaggerated flexion of the middle finger.

dystonia [9–13]. This is plausible, as it has been demonstrated that the repetition of skilled movements has the potential to trigger the functional reorganization of brain areas associated to these kinds of behaviors in both, animals and humans [9,14–19]. Recent work suggests that the role of a genetic predisposition in musicians' dystonia shouldn't be completely neglected [20,21].

After 124 years, as Gower's original description of the disorder [22] and 25 years posterior to the seminal report by Sheehy and Marsden in 1982 [7], a considerable amount of studies on focal hand dystonia have been published. Apparently, the dissemination of this knowledge to practitioners is still rudimentary and may account for the diagnostic challenge and the associated treatment delays. Therefore, in this report, we attempt to assess diagnosis, prescriptions and treatment histories in a series of sufferers who were ultimately diagnosed with focal hand dystonia. We note that the main focus of this report is on the presence of symptoms compatible with musicians' dystonia irrespective of its severity. The latter will be therefore not considered here.

Our fundamental goal is to contribute to the further dissemination of knowledge regarding a common musicians' pathology amongst the general medical community, including those involved in neurorehabilitation.

## Materials and methods

Diagnosis was based on screening of clinical histories and a comprehensive neurological examination. In those cases in which it was considered necessary, a magnetic resonance imaging (MRI) of the central nervous system was requested. The main criteria for a positive diagnosis of musicians dystonia were (i) clear signs of abnormal hand and fingers weakness or abnormal hand manoeuvres and positions during instrumental playing, (ii) symptoms absence or amelioration whilst performing imitative and homologous motor movements without playing the affected musical instrument (e.g. tapping the same affected finger sequence on a table), (iii) normal electromyography (EMG) and nerve conduction velocity (NCV) of the affected extremity, (iv) negative copper and ceruloplasmine analysis, (v) normal sensibility, coordination, reflexes and absence of sustained pain in the affected extremity (vi) no history of central nervous system pathologies and (vii) normal MRI of the brain when this examination was requested. To assess potential nerve conduction irregularities underlying the overt symptoms, we performed EMG and NCV of the affected extremity in 65 patients.

For the assessments, we collected data containing: records of clinicians previously visited, their diagnoses and the treatments they prescribed, as well as prior treatments (see Table 1). All patients were informed about the purposes of the assessment and agreed to freely participate in the clinical inquiries. The ethics committee of the Institut de Fisiologia i Medicina de l'Art-Terrassa approved the present study.

## Results

Over the course of 4 years (2001–2005), we evaluated and treated 665 musicians. Based on the criteria described in Materials and methods, 87 musicians with

**Table 1** Questions catalog

Previously, did you consult another/other medical specialist(s) concerning your hand problems?
Regarding your hand problems, which was the medical specialty of the physician you visited at first?
Which was the first diagnosis related to your hand complains?
Which were the treatment(s) that (was) were recommended?
Did your practitioner recommend you to consult another medical specialist?
Regarding your hand problems: what was the specialty of the other practitioner(s) you have consulted?
Did you consult another kind of health professionals?
Which were their specialties (physical therapy, osteopaths, acupuncture, other)?

**Table 2** Number of consultations for hand problems

Physicians	Number of consultations (%)	Diagnosis of focal dystonia (%)
Orthopaedists	69 (34.5)	5 (7.2)
Neurologists	54 (27)	38 (70.4)
General practitioners	31 (15.5)	1 (3.2)
Others	46 (23)	5 (10.9)
Total	200	49 (24.5)

focal hand dystonia were diagnosed, representing 13.1% of the total patients' population. EMG and NCV assessment of the affected extremity of 65 patients revealed 12 of them (18.5%) presenting some form of neuropathy (six radial, five median and one C6 radiculopathy). None of these neuropathies showed a clinical correlation with the observed symptoms.

Prior to their first contact with our clinic, these 87 musicians generated a total of 200 medical consultations that specifically concerned hand problems (mean of consultations = 2.3, SD = 1.8, range 1–6). A total of 218 consultations with specialists other than physicians were reported (mean of consultations = 2.5, SD = 1.3, range 0–7). Table 2 lists the different specialists, who were consulted.

The first physicians being consulted [nearly twice as frequently as general practitioners, who were consulted in 18 cases (20.7%)] were orthopaedists (30 cases = 34.5%). At their first consultation, 13 patients (14.9%) visited our institute, 11 (12.6%) consulted non-medical professionals; three patients consulted a neurologist (3.4%), whilst the remaining 12 (13.8%) requested help from several other medical professionals. Most musicians consulted more than one physician, thus the total number of given treatments reached 125. Amongst the most frequently consulted, non-physicians were physiotherapists (46 cases; 21.1%), acupuncturists (31 cases; 14.2%) and homoeopathists (19 cases; 8.7%).

Slightly more than half of the patients (52.9%) arrived at our clinic without a diagnosis or a suspicion of suffering from focal dystonia. The average time between the appearance of the first symptoms and a clear diagnosis of focal dystonia was 2.1 years (SD = 1.7, range 0–38). According to patients' records, the most frequently observed diagnoses amongst orthopaedists and neurologists were nerve compression, tendonitis and trigger fingers (see Table 3).

In 29 cases (23.2%), rest or reduction of playing time was suggested. In 16 cases (12.8%), surgery was prescribed, whilst in 15 (12%) physiotherapy, and in another 14 cases (11.2%), botulinum toxin injections were recommended. In 13 cases (10.4%), anti-inflammatory drugs were given. The 50 different treatments prescribed by orthopaedists included surgical interventions in 12

**Table 3** Orthopaedists' and neurologists' diagnoses

	n (%)
<b>Orthopaedists' diagnosis</b>	
Nerve compressions	16 (23.2)
Don't know	11 (15.9)
Tendonitis	11 (15.9)
Trigger finger	8 (11.6)
Normal	5 (7.2)
Focal dystonia	5 (7.2)
Muscular overuse	4 (5.8)
Others	9 (13)
Total	69
<b>Neurologists' diagnoses</b>	
Parkinson's diseases	1 (1.9)
Dystonia	38 (70.3)
Tendonitis	3 (5.6)
No diagnosis/don't know	4 (7.4)
Psychological	1 (1.9)
Nerve compressions	5 (9.3)
Congenital tremor	1 (1.9)
Myotonia	1 (1.9)
Total	54

cases (24%), rest or immobilization in 11 patients (22%), physiotherapy in another 10 cases (20%), and oral anti-inflammatory drugs in eight (16%) occasions, vitamin B in four instances (8%) whilst in the remaining two cases (4%), they recommended infiltrations with corticoids. Orthopaedists referred patients to other colleagues in three cases (6%). Only six patients underwent surgery following medical prescription. Surgical interventions targeted two trigger fingers, two Fröhshe arcades, two ulnar nerve entrapments at the elbow level, one cervical disk hernia and one carpal tunnel syndrome (surgery procedures include one musician who was surgically treated three times for cervical disk hernia, nerve decompression of the radial and the ulnar nerve, respectively) (see Table 3).

## Discussion

We analyzed 87 cases of focal hand dystonia and screened previous clinical records, diagnoses and treatments prior to referral to our clinic. In this series, musicians went through several clinical and non-clinical evaluations including various medical professionals until they ultimately received a final diagnosis of focal dystonia. Orthopaedists and neurologists were the physicians consulted the most. Several diagnoses were attempted.

In our sample, orthopaedists correspond to more than one-third of all specialists and therapists being consulted, and several musicians consulted an orthopaedist first. This trend might be grounded in (i) the local characteristics of the symptoms (initially a single

finger may be affected [23]), (ii) the absence of what patients may consider archetypal of neurological problems (numbness, tremor, weakness, tingling sensation, etc.), but also in the fact that patients with long standing history of dystonia may have some motor impairment in music-unrelated movements as well, especially if the dystonia is severe [3], (iii) and alternatively, because pain and inflammation secondary to the effort of playing with hand dystonia have developed, irrespective of the fact that pain is not the primary complaint of affected musicians [24] or (iv) the interaction of some or all of these factors. In this series, orthopaedists, in their majority, and an important number of neurologists did not diagnose focal dystonia. These results strongly suggest that dissemination of scientifically gathered knowledge on focal hand dystonia in musicians is still insufficient. Bearing in mind that these two specialties are those consulted at most, it is probable and even likely that the situation would be even worse if other clinicians were considered. Whilst looking for possible reasons, it is important to bear in mind that the number of cases is very low in the general community. This fact imposes clear difficulties to those involved in the diagnosis process. Moreover, the condition is not stable over time, and diagnosis is more prone to face difficulties at the beginning of symptoms. On the other hand, musicians tend to seek help for their hand problems after a considerable time period, when their own attempts to remediate their problems have been unsuccessful, and these attempts generally worsen the condition [4]. Therefore, symptoms may be comparatively evident by the time of first consultation.

Slightly more than half of those musicians seeking help for their hand problems did not receive the final diagnosis of focal hand dystonia in advance to their contact with our clinic, and that might be attributable to the concomitant – and not necessarily dystonia-related symptoms and complaints. For example, flexor nodes and pain (the latter as a reaction to manual pressure to different zones of the forearm) have been a common finding in musicians consulting upper limb specialists [25]. Therefore, these clinical signs may have been present, contributing to complicate the diagnosis of the main pathology. Moreover, in some instances, musicians' description of their symptoms can be very vague. In addition, patients' descriptions of the form '...the finger stays in the palm of the hand...' are common and can be very suggestive leading practitioners to interpret the symptoms as a mechanical, rather than a neurological problem. These facts hold for standard clinical assessments, but also for assessments during instrumental playing (see Fig. 1). Consequently, a clinical observation during instrumental playing manoeuvres is mandatory in the case of focal dystonia

and critical for the diagnosis of musician's pathologies in any case.

Another factor that hampers a diagnosis of focal hand dystonia, might be the overestimation, in some reports, of nerve compressions in association to musicians' cramp [26,27]. Indeed, we found evidence of nerve compression in only a few cases. We concede that nerve conduction blocks may well represent a precipitant for musicians' dystonia to occur, but we consider their appearance might also result from changes in work routine or even focal, peripheral trauma not directly associated with dystonia [28]. Interestingly, decompression surgery did not alleviate the dystonic symptoms in any of the assessed patients. Accordingly, the guitarist who underwent surgery several times did not experience any change in symptoms. Therefore, nerve compression does not seem to be necessarily associated to dystonic symptoms, and, in our experience, these signs do not represent an important proportion in musicians' cramp. Most importantly, surgery does not appear to be an option in the case of focal dystonia in musicians, even though it might well be suitable as a treatment in other pathologies of musicians' hands.

Many patients received prescriptions mostly aimed at resolving inflammatory processes (e.g. physiotherapy, oral anti-inflammatory drugs, infiltrations and systemic corticoids). In line with the use of such medications, a period of rest was the preferred treatment orthopaedists prescribed. A discussion on treatment options would be out of the scope of the present report, but two recent works treatise the topic [20,29].

Whilst in writer's cramp, mirror dystonia (dystonia occurring in the affected hand when using the non-affected hand in a specific task) can be observed in about 45% of cases [30–32], whether this clinical feature may offer certain help when diagnosing musicians' dystonia is open to inquiry. Mirror dystonia does not necessarily implies movements which are exactly the 'mirror' of the observed abnormal postures [30,32], and this might be a limitation considering the rather specific symptoms in musicians' cramp [24]. Whilst some other diagnostic tools like, e.g. grip tests have been used in the past to assess musicians' cramp, and in clinical practice they might be simple, safe and rapid its sensitivity and specificity remains to be statistically determined [33].

Despite significant advances in the understanding of the pathophysiology of focal hand dystonia, its cause still remains unclear. In addition, the contribution of greater pathophysiological insight into the development of effective treatments for the disorder has been sparse [2]. Many clinicians still consider focal dystonia to be an intractable clinical burden, and the rudimentary knowledge of focal dystonia amongst physicians, is paralleled by poor understanding of symptoms amongst

patients themselves. These factors may interact, pushing musicians to frequently oscillate between one specialist and another, and to also search for advice and treatment from non-physicians, which accounted for more than twice as many consultations as those for physicians. This might explain the important proportion of searches for alternative treatments observed in this series of affected musicians as well.

Consequently, when a musician complains on hand or embouchure problems (mainly slowness of movements, loss of movement and posture control, tension, rigidity or weakness whilst playing), a diagnosis of focal dystonia should be considered as being potentially causative, bearing in mind that at least one of 10 musicians will finally present the condition [24]. Overseeing these factors added to the above-mentioned rudimentary dissemination of knowledge of musicians' disorders, and in particular of focal dystonia, can critically contribute to diagnostic hindrances. Last but not least, a rapid diagnosis will ultimately contribute to diminishing the obvious medical costs currently resulting from focal dystonia.

Because this series of patients is not free of regional bias, it may be that our results represent a local phenomenon and cannot be generalized to other countries. Nevertheless, we believe that the data we present was at least of heuristic value to reflect on a potentially common problem also in other countries, something that deserves further investigation.

## Acknowledgements

Research was supported by the Fundació Ciència I Art, the Cogito Foundation, and a grant from the National Institutes of Health (K24 RR018875). The authors thank Dr Randolph Nudo, Department of Molecular and Integrative Physiology and the Landon Center on Aging of the University of Kansas for important suggestions.

## References

- Nutt JG, Muentner MD, Melton LJ III, Aronson A, Kurland LT. Epidemiology of dystonia in Rochester, Minnesota. *Adv Neurol* 1988; **50**: 361–365.
- Schuele S, Lederman RJ. Long-term outcome of focal dystonia in string instrumentalists. *Mov Disord* 2004; **19**: 43–48.
- Rosset-Llobet J, Candia V, Fabregas S, Ray WJ, Pascual-Leone A. Secondary motor disturbances in 101 patients with musician's dystonia. *J Neurol Neurosurg Psychiatry* 2007; **78**: 949–953.
- Rosset-Llobet J, Fabregas i Molas S, Rosines i Cubells D, Narberhaus Donner B, Montero i Homs J. [Clinical analysis of musicians' focal hand dystonia. Review of 86 cases]. *Neurologia* 2005; **20**: 108–115.
- Brandfonbrener AG, Robson C. Review of 113 musicians with focal dystonia seen between 1985 and 2002 at a clinic for performing artists. *Adv Neurol* 2004; **94**: 255–256.
- Grafman J, Cohen LG, Hallett M. Is focal hand dystonia associated with psychopathology? *Mov Disord* 1991; **6**: 29–35.
- Sheehy MP, Marsden CD. Writers' cramp—a focal dystonia. *Brain* 1982; **3**: 461–480.
- Jabusch HC, Muller SV, Altenmuller E. Anxiety in musicians with focal dystonia and those with chronic pain. *Mov Disord* 2004; **19**: 1169–1175.
- Pascual-Leone A, Amedi A, Fregni F, Merabet LB. The plastic human brain cortex. *Annu Rev Neurosci* 2005; **28**: 377–401.
- Hirata Y, Schulz M, Altenmuller E, Elbert T, Pantev C. Sensory mapping of lip representation in brass musicians with embouchure dystonia. *Neuroreport* 2004; **15**: 815–818.
- Elbert T, Candia V, Altenmuller E, *et al.* Alteration of digital representations in somatosensory cortex in focal hand dystonia. *Neuroreport* 1998; **9**: 3571–3575.
- Candia V, Wienbruch C, Elbert T, Rockstroh B, Ray W. Effective behavioral treatment of focal hand dystonia in musicians alters somatosensory cortical organization. *Proc Natl Acad Sci USA* 2003; **100**: 7942–7946.
- Byl NN, Merzenich MM, Jenkins WM. A primate genesis model of focal dystonia and repetitive strain injury: I. Learning-induced dedifferentiation of the representation of the hand in the primary somatosensory cortex in adult monkeys. *Neurology* 1996; **47**: 508–520.
- Plautz EJ, Milliken GW, Nudo RJ. Effects of repetitive motor training on movement representations in adult squirrel monkeys: role of use versus learning. *Neurobiol Learn Mem* 2000; **74**: 27–55.
- Nudo RJ, Milliken GW, Jenkins WM, Merzenich MM. Use-dependent alterations of movement representations in primary motor cortex of adult squirrel monkeys. *J Neurosci* 1996; **16**: 785–807.
- Kleim JA, Barbay S, Nudo RJ. Functional reorganization of the rat motor cortex following motor skill learning. *J Neurophysiol* 1998; **80**: 3321–3325.
- Kleim JA, Barbay S, Cooper NR, *et al.* Motor learning-dependent synaptogenesis is localized to functionally reorganized motor cortex. *Neurobiol Learn Mem* 2002; **77**: 63–77.
- Classen J, Liepert J, Wise SP, Hallett M, Cohen LG. Rapid plasticity of human cortical movement representation induced by practice. *J Neurophysiol* 1998; **79**: 1117–1123.
- Pascual-Leone A, Nguyet D, Cohen LG, Brasil-Neto JP, Cammarota A, Hallett M. Modulation of muscle responses evoked by transcranial magnetic stimulation during the acquisition of new fine motor skills. *J Neurophysiol* 1995; **74**: 1037–1045.
- Jankovic J, Ashoori A. Movement disorders in musicians. *Mov Disord* 2008; **23**: 1957–1965.
- Schmidt A, Jabusch HC, Altenmuller E, *et al.* Dominantly transmitted focal dystonia in families of patients with musician's cramp. *Neurology* 2006; **67**: 691–693.
- Gowers W. *A Manual of Diseases of the Nervous System*, 2nd edn. Darien, CT: Reprinted by Hafner Publishing Co., 1883.
- Candia V, Schafer T, Taub E, *et al.* Sensory motor retuning: a behavioral treatment for focal hand dystonia

- of pianists and guitarists. *Arch Phys Med Rehabil* 2002; **83**: 1342–1348.
24. Lederman RJ. Neuromuscular and musculoskeletal problems in instrumental musicians. *Muscle Nerve* 2003; **27**: 549–561.
  25. Nourissat G, Chamagne P, Dumontier C. [Reasons why musicians consult hand surgeons]. *Rev Chir Orthop Reparatrice Appar Mot* 2003; **89**: 524–531.
  26. Ross MH, Charness ME, Lee D, Logigian EL. Does ulnar neuropathy predispose to focal dystonia? *Muscle Nerve* 1995; **18**: 606–611.
  27. Charness ME, Ross MH, Shefner JM. Ulnar neuropathy and dystonic flexion of the fourth and fifth digits: clinical correlation in musicians. *Muscle Nerve* 1996; **19**: 431–437.
  28. Frucht S, Fahn S, Ford B. Focal task-specific dystonia induced by peripheral trauma. *Mov Disord* 2000; **15**: 348–350.
  29. Jankovic J. Treatment of dystonia. *Lancet Neurol* 2006; **5**: 864–872.
  30. Sitburana O, Jankovic J. Focal hand dystonia, mirror dystonia and motor overflow. *J Neurol Sci* 2008; **266**: 31–33.
  31. Merello M, Carpintiero S, Cammarota A, Meli F, Leiguarda R. Bilateral mirror writing movements (mirror dystonia) in a patient with writer's cramp: functional correlates. *Mov Disord* 2006; **21**: 683–689.
  32. Jedynak PC, Tranchant C, de Beyer DZ. Prospective clinical study of writer's cramp. *Mov Disord* 2001; **16**: 494–499.
  33. Pesenti A, Priori A, Scarlato G, Barbieri S. Transient improvement induced by motor fatigue in focal occupational dystonia: the handgrip test. *Mov Disord* 2001; **16**: 1143–1147.