AN APPROACH TO FUNCTIONAL MOVEMENT DISORDERS

Anthony E. Lang

University of Toronto Ontario, CANADA

This lecture will provide an overall approach to patients presenting with functional movement disorders (FMD). The notes below will simply highlight the general topics covered with a series of tables summarizing the Phenomenology seen in patients with FMD, the clues to the History and Physical Examination, the approach to the Laboratory Evaluation (largely special testing of movement disorders patients), the Classification of FMD and the approach to the Delivery of the Diagnosis and the Management of FMD.

Table 1 * Approach to Varied Phenomenology of Functional / Psychogenic Movement Disorders

Combined data on psychogenic movement disorder seen at several movement disorders centers.^a

Abnormal movement	Toronto Western Hospital ^b	NewYork Columbia Presbyterian Medical Center	Cleveland Clinic Florida ^d	Marshfield Clinic ^e	Baylor College of Medicine ^f	Total
	(%)	(%)	(%)	(%)	(%)	(%)
Dystonia	89(31.9)	82(54)	43(26.5)	22 (24)	89(39)	325(33.8)
Tremor	91(32.6)	21(14)	61(37.6)	33 (35.8)	127(55.7)	333(34.7)
Myoclonus	51(18.3)	11(7)	8((4.9)	22 (24)	30(13.2)	122(12.7)
Gait disorder	16(5.7)	14(9)	17(10.5)	11 (11.9)	7(3.1)	65(6.7)
Parkinsonism	14(5)	3(2)	5(3.0)	4 (4.3)	6(2.6)	32(3.3)
Tics	3(1.1)	2(1)	9(5.5)	-	15 (6.6)	29(3.0)
Other	15(5.4)	19(13)	19(11.7)	-	-	53(5.5)
Total	279	152	162	92	274	959

* Table taken from Sa DS, Galvez-Jimenez N, Lang AE. Psychogenic Movement Disorders. In: RL Watts, D. Strandaert, JA Obeso, editor(s). Movement Disorders. 3rd ed. New York: McGraw-Hill; 2012. p. 975-1003.

- a. Data collection varied from listing the predominant movement as in (b), (d) and (e) to all movements observed (c)
- Baik JS, Lang AE. Gait abnormalities in psychogenic movement disorders. Movement Disord 2007; 22(3):395-399
- c. Williams DT, Ford B, Fahn S. Phenomenology and psychopathology related to psychogenic movement disorders. Adv Neurol 1995; 65:231-258.
- d. Galvez-Jimenez, N, unpublished data (Cleveland Clinic Florida (CCFla) 1998 2008)
- e. Sa, Daniel. Unpublished data. Collected July 2006 December 2008
- f. Thomas M, Jankovic J. Psychogenic Movement Disorder: diagnosis and management. CNS Drugs 2008; 18:437-452.

Table 2. Approach to the History: Clues Suggesting a Functional / Pychogenic Etiology

Abrupt onset (symptoms often maximal at that time)

Static course

Spontaneous remissions/cures

Paroxysmal symptoms (generally non-kinesigenic)^a

Psychiatric comorbidities^b

Secondary gain (often not apparent)

Risk factors for conversion disorder^c

Psychological stressors^d

Multiple somatizations/undiagnosed conditions

Employed in allied health professions (infrequent)

^aSeparation from organic paroxysmal dyskinesias can be challenging, particularly if they occur infrequently with prolonged symptom-free periods

^bPsychiatric diseases can also coincide with organic illness or present as part of the organic movement disorder

^cSexual and physical abuse, trauma

^dOften initiated by injury (often minor) or motor vehicle accident associated with litigation or compensation

Adapted from Pringsheim T, Chen R, Lang AE. Psychogenic Movment Disorders. In: Freund H.J., Jeannerod M, Hallet M, Leiguarda R, editors. High-order motor disorders - from neuroanatomy and neurobiology to clinical neurology. Oxford: Oxford University Press, 2005: 397-412.

Table 3. Approach to the Examination: Clues Suggesting a Functional / Pychogenic Etiology

Movement inconsistent

Variability over time (frequency, amplitude, direction/distribution of movement)

Distractibility reduces or resolves^a, attention increases

Selective disability

Entrainment (especially with tremor)

Movement incongruous with organic movement disorders

Mixed (often bizarre) movement disorders

Paroxysmal attacks (including pseudoseizures)

Precipitated paroxysms (often suggestible/startle)

Suggestibility^b

Effortful production or deliberate slowness (without fatiguing) of movement

Self-inflicted injury (caution: tic disorders)

Delayed and excessive startle response to a stimulus

Burst of verbal gibberish or stuttering speech^c

False (give away) weakness

Non-anatomical sensory loss or spread of movement

Certain types of abnormal movements common in individuals with PMD^d

Functional disability out of proportion to examination findings Therapeutic responses: Unresponsive to appropriate medications. Response to placebos. Remission with psychotherapy.

^a Distractibility should be tested both with mental and motor tasks. Although most often organic movement disorders are not suppressed, organic tics or akathisia can be suppressible, and recently it was shown that diaphragmatic tremor was suppressed by simple motor tasks, perhaps by interference with central nervous system circuitry (Espay AJ, Fox SH, Marras C, Lang AE, Chen R. Isolated diaphragmatic tremor - Is there a spectrum in "respiratory myoclonus"? Neurology 2007; 69(7):689-692)

^b For example, application of pressure with finger or tuning fork may reduce symptom. Repeated, reliable induction of the paroxysmal events by suggestion may be very helpful. However, with paroxysmal symptom, suggestibility and placebo trial may not be helpful unless repeated reversals with placebo are documented when symptom otherwise is frequent and attacks are prolonged.

^c Particularly if the entire word is repeated (typically broken up into syllables, each repeated), rather than the initial syllable.

^d Such movements include dystonia that begins as a fixed posture (particularly if abrupt onset, painful, and early contractures are seen); bizarre gait; twisting facial movements that move mouth to one side or the other (organic dystonia of the facial muscles usually does not pull the mouth sidewise).

Adapted from Pringsheim T, Chen R, Lang AE. Psychogenic Movment Disorders. In: Freund H.J., Jeannerod M, Hallet M, Leiguarda R, editors. High-order motor disorders - from neuroanatomy and neurobiology to clinical neurology. Oxford: Oxford University Press, 2005: 397-412.

Table 4: Approach to the Laboratory Investigation of Possible Functional / Psychogenic Movement Disorders

	Peripheral Electro- physiology	Central Electro- physiology	Imaging	Comments
Tremor	+++	+	+	Combinations needed ^a ; Routine Dx
Myoclonus	+++	+++	-	Combinations ^b ; Routine Dx
Parkinsonism	_	_	++ ^c	Routine; Caution
Dystonia	_	? - ^a	+ ^e	? Diagnostic utility

- Not useful; + Possibly useful; ++ ? Diagnostic; +++ Diagnostic

All testing must be interpreted cautiously and in context of clinical features

a. Schwingenschuh P, Saifee TA, Katschnig-Winter P, Macerollo A, Koegl-Wallner M, Culea V, Ghadery C, Hofer E, Pendl T, Seiler S, Werner U, Franthal S, Maurits NM, Tijssen MA, Schmidt R, Rothwell JC, Bhatia KP, Edwards MJ. Validation of "laboratory-supported" criteria for functional (psychogenic) tremor. Mov Disord. 2016 Apr;31(4):555-62

b. Hallett M. Neurophysiologic studies of functional neurologic disorders. In: Functional Neurological Disorders. Eds M. Hallett, J, Stone, A. Carson Elsevier, Amsterdam, 2016, pp.61-72

c. Evaluation of pre-synaptic nigrostriatal dopamine system with PET (e.g., F-dopa) or SPECT (e.g., DAT scan)

d. See Quartarone A, Rizzo V, Terranova C, Morgante F, Schneider S, Ibrahim N, Girlanda P, Bhatia KP, Rothwell JC. Abnormal sensorimotor plasticity in organic but not in psychogenic dystonia. Brain. 2009 Oct;132(Pt 10):2871-7. However, see Sadnicka A, Hamada M, Bhatia KP, Rothwell JC, Edwards MJ. A reflection on plasticity research in writing dystonia. Mov Disord. 2014 Jul;29(8):980-7 for an alternative/revised opinion.

e. Schrag AE, Mehta AR, Bhatia KP, Brown RJ, Frackowiak RS, Trimble MR, Ward NS, Rowe JB. The functional neuroimaging correlates of psychogenic versus organic dystonia. Brain. 2013 Mar;136 (Pt 3):770-81

Traditional Classification of Degrees of Certainty in	Proposed revision of Classification of Degrees of		
Diagnosis**	Certainty in Diagnosis*		
1. Documented ^a :	1. Documented (as in original)		
- Remittance with suggestion, physiotherapy,			
psychotherapy, placebos, "while unobserved"			
2. Clinically established ^a :	2a. Clinically established plus Other features (as		
- Inconsistent over time / Incongruent with clinical	in original)		
condition	2b. Clinically established <i>minus</i> Other features		
+	- Unequivocal clinical features incompatible with		
- Other manifestations: other "false" signs, multiple	organic disease with no features suggesting		
somatizations, obvious psychiatric disturbance	another underlying neurological or psychiatric		
	problem		
	1+2a+2b = Clinically Definite		
3. Probable:	3. Laboratory Supported Definite		
- Inconsistent/Incongruent - no other features ^b	- Electrophysiological evidence proving a		
- Consistent/Congruent + "false" neurological signs	psychogenic movement disorder (primarily in		
c	cases of psychogenic tremor and psychogenic		
- Consistent/Congruent + multiple somatizations ^c	myoclonus)		
4. Possible ^c :	4. Possible		
 Consistent/Congruent + obvious emotional 	- Movement disorder demonstrates clinical		
disturbance	features (or electrophysiology) that are strongly		
	suggestive but not diagnostic of a psychogenic		
	cause		

a Subsequently Fahn and his colleagues proposed combining categories 1+2 under "Clinically Definite" (Williams DT, Ford B, Fahn S. Phenomenology and psychopathology related to psychogenic movement disorders. In: Weiner WJ, Lang AE, editors. Behavioural Neurology in Movement Disorders. New York: Raven Press, 1994: 231-257.)

b Problem: Patients with unequivocal psychogenic movement disorders (for example, completely distractible or fully entrainable tremor) lacking other psychogenic features (e.g. false neurological signs, multiple somatizations etc.) can only be classified as "probable".

c Problem: Most patients fulfilling these criteria have organic movement disorders with additional psychiatric disturbances (i.e., with "functional overlay")

** From: Fahn S, Williams PJ. Psychogenic dystonia. Adv Neurol 1988; 50:431-455. Problem: does not take into account the possibility of confirming that the movement disorder is psychogenic with additional electrophysiological testing

* Taken from Gupta A, Lang AE. Psychogenic movement disorders. Curr Opin Neurol 2009; 22(4):430-436.

Table 6. Approach to the Delivery of the Diagnosis to a Patient with a Functional / Psychogenic MovementDisorder

- 1. Emphasize that the symptoms are real rather than feigned useful to emphasize that the diagnosis does NOT impy that the patient is "doing it on purpose" or that they are "crazy"
- 2. Use the "functional" signs in presenting and explaining the diagnosis to patient and family (certainty of diagnosis, difference from other forms of the same movement disorder, need for brain to focus on maintaining the movement disorder (e.g., distratability) or non-physioloigcal aspect of the sign (e.g., Hoover's sign).
- 3. Use the analogy of software (functional) vs hardware (structural) problem
- 4. State the diagnosis confidently and also tell them what they don't have (brain tumor, Stroke, MS, Parkinson's, etc)
- 5. Emphasize that Functional Neurological Disorders are *common* and potentially fully reversible; refer the patient and family to the Website: <u>www.Neurosymptoms.com</u>
- 6. Avoid unnecessary tests and emphsize that no further investigations are indicated (if you are uncertain about the diagnosis do not present the diagnosis until you are certain; any degree of uncertainty → keep an open mind and follow patient with *repeated examinations* and further testing as necessary)
- 7. Communicate the diagnosis to other care providers emphasizing the positive / definitive nature of the diagnosis avoid uncertainty and avoid negative diagnostic terminology (i.e., what the diagnosis isn't or is inconsistent with (medically unexplained symptoms)).

Table 7. Approach to the Treatment of Functional / Psychogenic Movement Disorders

- 1. No adequate randomized controlled trials
- 2. Best outcomes with short interval between onset, diagnosis and treatment
- 3. Best administered in multimodal fashion (*experienced teams*) → markedly variable outcomes
- 4. Psychotherapy, CBT, Rehabilitation, "Motor reprogramming", Activity, Antidepressants, Hypnosis, Acupuncture, rTMS, TENS, Botulinum toxin (dystonia, palatal tremor), Therapeutic sedation, others
- 5. Use of "functional" signs in presenting Dx and initiating treatment showing patient ability to change the characteristics of the movement (e.g., tremor change frequency or complete suppression)^a
- 6. In the case of a combination of a functional MD superimposed on an organic MD often (but not always) the former results in the predominant disability. Therefore one usually needs to concentrate on managing the functional disorder first rather than expending efforts managing the underlying organic component

a. Stone J, Edwards M. "Trick or Treat?" Neurology. 2012 Jul 17;79(3):282-4.