What is the evidence for the diagnostic validity of "psychogenic dystonia"?

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INTRODUCTION

- Dystonia is a clinically defined syndrome without pathognomonic features.
- Patients with dystonic movements and atypical or hysterical features have been described for some time.
- In the late 1970s, Dr. Stanley Fahn and colleagues at Columbia U. invented a diagnosis, “psychogenic dystonia,” to describe these patients (Table 1).
- There are generally accepted validation criteria for any new diagnosis (Table 2).

SPECIFIC AIM

- Does “psychogenic dystonia” meet the criteria listed in Table 2 for validity of diagnosis?

METHODS

- MEDLINE search for studies of dystonia
- Followed references cited in available journal articles or chapters
- Used validation criteria of Robins and Guze (1970)

TABLE 1. "Psychogenic dystonia" defined

Fahn & Williams classification (1988)

- Documented psychogenic movement disorder
  - symptoms completely relieved by psychotherapy, suggestion or placebo, *or*
  - no signs when surreptitiously observed
- Clinically established psychogenic disorder
  - inconsistent over time *or*
  - "incongruent with a classical movement disorder" *and* any of the following:
    - other psychogenic signs, e.g. false weakness
    - multiple somatizations
    - an obvious psychiatric disturbance
    - disappearance with distraction
    - apparently deliberate slowness of movement
- Probable psychogenic disorder
- Possible psychogenic disorder

Table 2. Robins-Guze validity criteria

- clinical description
- laboratory studies
- delimitation from other disorders
- follow-up study
  (including response to treatment)
- family study

RESULTS (clinical description)

- Some of the defining criteria are vague, *e.g.* "inconsistent over time" or "obvious psychiatric disturbance."
- Fahn et al. have provided substantial descriptive data regarding these patients.
- There is no information regarding the reliability of this diagnosis.

RESULTS (laboratory studies)

- There are no pertinent laboratory studies (*e.g.* H-reflex, PET sensorimotor response, DYT-1 gene frequency).
- Some psychometric data are available for (presumably typical) dystonia but there are no direct comparisons with results in "psychogenic dystonia" patients.

RESULTS (delimitation from other disorders)

- There are no studies directly comparing typical dystonia and "psychogenic dystonia," including no comparisons of:
  - frequency of defining clinical features such as inconsistency over time, response to distraction, prior conversion symptoms, personality, etc.
  - natural history, including spontaneous remissions
  - response to placebo or other treatment
  - family history
- Two older studies do give relevant empiric data on similar patients (Meares 1971, Tibbets 1971)

RESULTS (follow-up study / tx response)

- Limited clinical follow-up data has been reported.
- The recovery rate is said to be ≈25%-58%, excluding malingerers.
- There are no reported clinical trials of any intervention.

RESULTS (family study)

- There are no pertinent family studies.

CONCLUSIONS

- There is inadequate data at present to support the validity of the diagnosis "psychogenic dystonia."
- Fahn and colleagues have drawn attention to an important group of patients who are difficult to diagnose and treat.
- Fahn et al. have provided rich clinical descriptive material which may be helpful in designing future studies.

DISCUSSION (1)

- Questions about "psychogenic dystonia" (Table 3) led our group not to accept the approach of Fahn et al. without validation.
- There are at least two reasonable, nonexclusive, next steps:
  - Gather further validation data regarding "psychogenic dystonia" as defined by the Columbia group.
  - Define an approach to these patients that does not require this unproven diagnosis.

TABLE 3. Problems with the Columbia approach

- **Face validity:** Placebo response or comorbid psychiatric illness doesn't prove etiology. Why call idiopathic dystonia "organic"? What does "psychogenic" mean?
- **Clinical utility:** Does this diagnosis help the patient? ("conversion disorder" has a poor follow-up record).
- **Confidence *vs* knowledge:** It is awkward at best to tell patients "we know what causes your symptoms" when in truth we don't.
- **"First, do no harm":** Misdiagnosis may hurt the patient.
- **Ignores** prior empiric research on somatization.
- **Placebo response:** Typical patients can respond or even remit (≈10%) and "psychogenic" patients often do not.

(See also Quotes page.)

Quotes

Definition: "Psychogenic dystonia is difficult to diagnose since there are no laboratory tests to establish the diagnosis of organic idiopathic dystonia." (Fahn, 1994) "Despite a professional lifetime of working with patients with dystonia, I am still surprised by seeing new variants of the condition." (Marsden, 1995) "The bizarre nature of the initial symptoms and their exaggeration in periods of stress, as well as their variability in certain settings not infrequently lead to a diagnosis of hysteria. ... The extreme variability of the natural history of this disorder makes evaluation of the effects of various treatment measures difficult to assess." (Fahn, 1984) "The most important point to remember in recognizing and treating the hysterical patient is that the diagnosis is *not* made by exclusion. There must be *positive* evidence demonstrating that the dysfunction is functional in origin rather than organic. *It is not a diagnosis of exclusion*." (Weintraub, 1983 [emphasis in original])

Remission: "Most organic movement disorders rarely remit spontaneously and completely except for tics, tardive dyskinesia, infectious … and drug-induced reactions, and, rarely, essential myoclonus. … On occasion [dystonia totally remits.]" (S. Fahn, 1994) "I emphasize the small but definite chance of spontaneous remission." (C. D. Marsden, 1995) Spontaneous remission found in many prior outcome studies of (presumably typical) dystonia: Meares (1971) 8 of 32 torticollis; Marsden & Harrison (1974) 7 of 42 torsion dystonia (5 children generalized dystonia); Cooper et al (1976) 10 of 226 primary dystonia; Jayne et al (1984) 10 of 26 torticollis; Jahanshahi & Marsden (1988) 8 of 100 torticollis (total 43/426 = 10%).

Placebo response: "Dystonic movements have been reported to be effectively controlled for varying periods by … biofeedback techniques." (Fahn, 1984) Placebo responses were reported in 9%-17% of clinical trials of botulinum toxin in (presumably typical) dystonia (Tsui et al, 1986; Gelb et al, 1989; Lorentz et al, 1991; Jankovc and Orman, 1987; Truong et al, 1991; Cole et al, 1995). "Oculogyric crises, dystonic conjugate spasms of the eye, were often 'contagious' in a ward with post-encephalitics. When one patient would go into crisis it could set off other patients on the same ward, even when they had never suffered them before. The crises could also be triggered by the sight of certain people, hypnotism or emotional trauma and resolved by simple suggestion or injections of distilled water" (from Delbeke & van Bogaert, 1928). On the other hand, in patients with "psychogenic dystonia," Fahn & Williams say "placebo treatment by itself usually failed" (1988, p. 442).

Psychological symptoms: "Obvious psychiatric disturbances [are a] clue that suggests a psychogenic movement disorder." (Marsden, 1995) "Dystonia generally produces lifelong disability. Few are not emotionally scarred by their illness." (Marsden, 1995)

"First, do no harm": "A diagnosis of a psychogenic disorder can create emotional trauma to the patient and his/her family." (Fahn, 1994)

Confidence vs knowledge: "When initially informing the patient about the diagnosis … state firmly that he/she has a movement disorder … and that 'it is caused by the mind controlling the body. Pent-up emotions [produce] these abnormal movements.' … Do not convey uncertainty." (Fahn, 1994)

Miscellaneous: "It is possible that conversion reactions occur during sleep." (Fahn, 1994) "The psychiatrist cannot make the diagnosis of a psychogenic movement disorder. That diagnosis can only be made by a neurologist. … If the psychiatric consult does not detect psychopathology, this could be in error." (Fahn and Williams, 1988) "It's either psychiatric or organic." = "It's either psychogenic or neurologic." = "It's either raining or it's Tuesday." (Rogers, 1992)

DISCUSSION (2 – a research strategy)

- List clinical features emphasized by Fahn et al., those tested in the empiric studies of Tibbets and of Meares, and others
- Prospectively gather this information and psychiatric diagnostic information, and follow patients over time
- Controlled, blinded treatment studies of both placebo and other treatments (as ethically appropriate)

DISCUSSION (3 – our clinical approach)

Treat patients according to whether *movements* are:

- typical for dystonia
  - treat same as other dystonia patients, *regardless* of psychological symptoms
  - offer treatment for comorbid illnesses including psychiatric illness if any
- possible dystonia but with unusual features (*e.g.*, atypical, dramatic, stress-related, placebo-responsive)
  - tell patient that his/her symptoms are atypical for dystonia if this is true
  - continue conservative neurologic and psychiatric follow-up and continue the search for valid diagnoses (including neurologic illness or somatization d/o)
  - offer adjunctive treatments as appropriate without claiming etiologic significance (psychotherapy, botox, P.T./O.T., treatment of comorbid illness)
  - use placebos only with the patient's informed consent
- clearly not dystonia
  - don't call it a movement disorder; be comfortable with "undiagnosed"
  - management as above for "possible dystonia"

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