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Involuntary Thumb Flexion on Neurological Examination: An Unusual Form of Upper Limb Dystonia in the Faroe Islands

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Abstract

Background: The prevalence of dystonia varies worldwide. A prior report suggested a high prevalence of focal dystonia in the Faroese population, possibly reflecting a founder effect. During standardized neurological examination as part of an ongoing neuroepidemiologic study in the Faroe Islands, we noted an unusual phenomenon of thumb flexion during repetitive hand movements in a subset of subjects and sought to define its phenomenology.

Methods: We requested commentary from a panel of dystonia experts regarding the phenomenology of the movements. These experts reviewed the videotaped neurological examination.

Results: Among the experts, dystonia was the leading diagnosis. Alternate causes were considered, but deemed less likely.

Discussion: Diagnosis of dystonia requires careful clinical assessment and consideration of associated features. We report a novel form of dystonia, not previously described to our knowledge, in this isolated population. Further studies of dystonia prevalence in the Faroe Islands are merited to characterize its burden in this population and its specific clinical characteristics.

Keywords: Dystonia, focal dystonia, Faroe Islands, thumb flexion, phenomenology


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Conflicts of Interest: The authors report no conflicts of interest.

Ethics statement: This study was performed in accordance with the ethical standards detailed in the Declaration of Helsinki. The authors’ institutional ethics committee has approved this study and all patients provided written informed consent.
Introduction

The Faroe Islands are an archipelago in the North Atlantic Ocean, with a population of 51,540.1 Regarding ancestry, genetic studies suggest discrepant origins of female and male populations, with the female population apparently deriving from predominantly British Island ancestry and the male population from Scandinavian.3 A high degree of inbreeding among this geographically isolated population has been reported.2,4 Due to its genetically and culturally homogeneous population, as well as its nationalized healthcare system, it has been a site for the study of several complex diseases. Increased prevalence of several diseases, including Parkinson’s disease, has been reported, in comparison to neighboring nations.

The prevalence of isolated focal dystonia has previously been reported as ranging from 82 (33–98) per million in Japan to 312 (251–384) per million in Iceland, in service-based studies. In a 2015 service-based study, Joensen reported a relatively high prevalence of isolated focal dystonia (including cervical dystonia, writer’s cramp, oromandibular dystonia, and laryngeal dystonia) in the Faroe Islands, noting 29 confirmed cases among the Faroese population (48,100), a prevalence of 602 (395–873) per million, with a particularly high prevalence of cervical dystonia at 478 (332–728) per million. The prevalence reported by Joensen matches or exceeds that reported among other European or North American populations in service-based assessments.5,10–12 That author proposed a possible contribution from founder effects. Population-based studies of dystonia prevalence are few in number, and generally report prevalence estimates that are higher than those reported by service-based studies, as ascertainment of cases is more complete in the former. A prevalence of 439 (284–648) per million was reported in a population-based study in eastern India, utilizing neurological examination of a subset screened positive via initial questionnaire.13 The one population-based study that utilized a movement disorder neurologist to review all cases via videotaped neurological examinations, in northern Italy, reported the highest prevalence of all: 7,320 (3,190–15,640) per million, likely partly reflecting greater recognition of dystonic features by the subspecialty trained physician.

Consistent assessment of dystonia prevalence requires consensus regarding phenomenology. We present an unusual pattern of thumb activation (Video 1) noted during neurological examination of numerous subjects in an ongoing population-based study of essential tremor (ET) in the Faroe Islands. To our knowledge, the phenomenology has not been reported previously, although the impression of two movement disorder neurologists (C.Y.K. and E.D.L.) was that it represented a form...
of dystonia. To gain further diagnostic clarity, we sought the independent commentary of four internationally renowned dystonia experts regarding the phenomenology and its differential diagnosis.

Methods

Subjects were selected from an ongoing study of ET in the Faroe Islands. Recruitment and study procedures have been outlined in previous publications. In brief, we used a two-phase population-based study design with an initial screening and an in-person clinical evaluation of a subset of the screening population. The screening group consisted of 3,000 randomly selected Faroese individuals aged 40 years and older, of whom 1,334 (44.5%) returned spirals and questionnaires. In initial calculations, which included the projected proportion that would screen positive as well as the expected proportion of these that would have ET, we concluded that 3,000 screened participants would yield approximately 100 ET cases for further analyses.

In the second phase of the study, a subsample of 282 individuals was invited to participate in an in-person clinical evaluation, of whom 227 (80.5%) accepted and completed the in-person clinical evaluation. The standardized in-person evaluation included a 10-minute videotaped examination performed by a trained nurse (E.H.E.). Subjects were examined for the presence of dystonia of the eyelids, mouth, speech (with limited assessment of swallowing), neck, arm, trunk, and leg in accordance with the Burke–Fahn–Marsden (BFM) dystonia rating scale (movement scale). Detailed assessment for dystonia was performed through review of views of the face, neck, trunk, and extremities: while seated, standing, and walking (including turning); with posture (arms suspended in front of body and in “wing-beating” position); and with multiple tests of action (finger taps, hand-opening/closure, finger-to-nose, pressing/drinking/lifting water with a spoon, and alternating toe-heel taps). Handwriting was videotaped and reviewed. Audio recordings of sustained phonation and speech were also reviewed to assess for dysphonia.

Demographic and clinical questionnaires and videotaped neurological examinations were reviewed by E.D.L.; diagnoses of ET (n = 27), dystonia (n = 2), and Parkinson’s disease (n = 0) were assigned according to published diagnostic criteria. During video review of motor examinations, a recurring phenomenon was noted during performance of hand-opening/closure as included in the UPDRS motor assessment, specifically a tendency to flex the thumb beneath (rather than over) the fingers during hand-opening/closure, which occurred bilaterally. This phenomenon was noted in 76 of 227 screened (33.5%), although the majority of subjects performed hand-opening as expected, with the thumb flexed over the fingers. Among these 76 subjects, we did not note cervical dystonia, blepharospasm, or writer’s cramp.

We sought to define the phenomenology of thumb flexion in this setting. We selected a clear and representative series of four subjects demonstrating the phenomenon (Video 1, subjects 2–5) and one who did not (Video 1, subject 1). We then invited an international panel of dystonia experts from diverse training lineage to comment independently on the phenomenology. All commentators have published extensively on the clinical characteristics and phenomenology of dystonia. We requested they opine on the phenomenology of the thumb movement and its differential diagnosis, specifically whether the finding might reflect dystonia or other causes.

Ethical considerations

All study procedures were approved by the local ethical review committee of the Faroe Islands and by the Institutional Review Board at Yale University, with participation on a voluntary basis. Signed informed consent was obtained from each enrollee.

Expert commentary

Dr. Defazio

Subjects 2–5 manifested impairment of finger movements during performance of the hand closure–opening task. Video recordings showed that thumb movements are patterned, repetitive, and sometimes sustained, and seem to be triggered by voluntary contraction of hand muscles during the hand closure and opening task. These findings would suggest a diagnosis of dystonia. No sensory trick was evident or checked for. In the absence of a sensory trick, experts recommend actively excluding clinical features related to conditions mimicking dystonia, that is, features that would be expected to be absent in dystonia. In this regard, the main clinical conditions possibly mimicking “thumb dystonia” may be orthopedic and rheumatologic diseases leading to abnormal “fixed” thumb postures; lower motor neuron disease/myopathy inducing weakness in the muscles antagonizing the abnormal posture; upper motor neuron disease inducing spasticity and weakness of muscles causing the abnormal posture; chorea inducing nonpatterned and nonrepetitive involuntary movements; and tics associated with ability to mentally suppress the spasms.

As far as I can see:

- the involuntary thumb posture is not fixed, which would exclude orthopedic or rheumatologic diseases
- there is no weakness of muscles antagonizing the abnormal thumb posture, which would exclude lower motor neuron diseases/myopathy
- there is no weakness of muscles causing the abnormal posture, which would exclude spasticity
- involuntary thumb movements are patterned and repetitive, which would exclude chorea
- ability to mentally suppress the spasms needs to be checked for to exclude dystonic tics. It is worth noting that dystonic tics sometimes have intermediate features between dystonia and tics.

Summary. Overall, the aforementioned considerations make a diagnosis of dystonia likely for the thumb flexion movement/posture seen in these subjects. Further supporting this view, movements of fingers other than the thumb also seem to be slightly impaired during the execution of the hand closure and opening task. In subject 2, the third and the fourth fingers of left hand seem to be in adduction; in subject 4, the fifth finger of both hands seems to be in abduction; and in subject 5, flexion movements of the fifth left-sided finger seem to be limited.
Thumb Flexion on Neurological Examination

The hand in those people looks extremely similar more affected than the other. Most other fingers appeared more normal varying degrees. It was bilateral, but sometimes one hand appeared of release of the thumb muscles. The folding involved both the distal thumb extension) appeared to be delayed or hesitant in many of the first digit, a phenomenon that seemed to occur when finger flexion (as described in these patients).

Another way to approach this is to ask: “what are potential alternative diagnoses?” The closest possibility is the cortical thumb: a phenomenon wherein the thumb is adducted into the hand when the person makes a fist. This is normal in newborns, but persistent cortical thumb is a sign of brain damage, most often reported with static encephalopathy after birth injury. The hand in those people looks extremely similar to the hands in the patients presented here. There is very little literature about the cortical thumb phenomenon and most of it is about the cortical thumb in infants. I could find no reference about whether such people have difficulty abducting the thumb out of the posture (as described in these patients).

Summary. In summary, I think this is dystonia, but the phenomenon does force you to think more carefully about when to diagnose dystonia.

Dr. Greene

The thumb movements in this video meet the standard, core requirements for dystonia: involuntary sustained muscle contractions causing abnormal postures. Unfortunately, not all such movements are thought to be dystonic (e.g., decorticate posturing) and not all dystonic movements meet these criteria (e.g., blepharospasm without sustained eye closure). If the movements are task specific or if people have sensory tricks that reduce the posture, then it makes it more likely the movements represent dystonia. We do not know whether these patients have tricks. Task specificity in this case would be to see if there is thumb flexion or electrical activity in the adductor or opponens pollicis of the hand when the person flexes digits 2–5 but not when they use digits 2–5 to grab an object (or perhaps some particular object). It would also be interesting to know whether the adductor or opponens pollicis in one hand has electrical activity when the opposite hand closes. This is often the case in dystonia involving one hand, but may also occur in people without dystonia.

Another way to approach this is to ask: “what are potential alternative diagnoses?” The closest possibility is the cortical thumb: a phenomenon wherein the thumb is adducted into the hand when the person makes a fist. This is normal in newborns, but persistent cortical thumb is a sign of brain damage, most often reported with static encephalopathy after birth injury. The hand in those people looks extremely similar to the hands in the patients presented here. There is very little literature about the cortical thumb phenomenon and most of it is about the cortical thumb in infants. I could find no reference about whether such people have difficulty abducting the thumb out of the posture (as described in these patients).

Summary. In summary, I think this is dystonia, but the phenomenon does force you to think more carefully about when to diagnose dystonia.

Dr. Jinnah

All affected subjects had a similar problem, with some minor variations among individuals. Although the normal pattern is to fold the fingers under the thumb on hand closure in this task, all subjects showed intermittent but patterned folding of fingers over the thumb. An occasional variation was incomplete folding of the tip of the thumb against the first digit, a phenomenon that seemed to occur when finger flexion happened faster than thumb flexion. Indeed, hand opening (especially thumb extension) appeared to be delayed or hesitant in many of the cases, either due to failure of the extensors, or excessive flexion and lack of release of the thumb muscles. The folding involved both the distal interphalangeal joint of the thumb and the more proximal joints to varying degrees. It was bilateral, but sometimes one hand appeared more affected than the other. Most other fingers appeared more normal in both flexion and extension.

I believe these movements are abnormal. They are not likely to be variations of normal behavior, and I am not aware of any cultural or regional “habit” that would explain it. However, it would be important to know what the examiner was demonstrating to the patients to be sure they were not trying to copy an incorrect instruction for this task.

The movements have some partial resemblance to the “simian hand” which typically results from median nerve damage with weakness of thumb abduction. However, the problem in the videos is not precisely the same as the simian hand because the abduction movement is possible, just not occurring when rapid repetitions are conducted. Also, the thenar eminence seemed to be quite generous in all cases, and this muscle normally shrinks in the simian hand. Another differential to consider is the “cortical thumb” or “thumb in palm” or “fisting” posture of newborns. This is normal in newborns, but its persistence beyond a year of age is abnormal and thought to be a sign of upper motor neuron injury. In view of these considerations, it could be useful to ensure that the neurological examination confirms normal strength in the hand muscles, and potentially confirms with electromyography (EMG).

Summary. The intermittent but patterned movements, in the absence of any clear neuromuscular cause, could indeed be a dystonia, although I am not aware of other patients with this particular pattern.

Dr. Tijssen

This is an interesting phenomenon. What I see is a repeated flexion and adduction of the thumb when flexing the fingers. The first thought that came to my mind was whether this was the way people in the Faroe Islands learn to flex their hands for whatever reason. A second thought was whether there are anatomical changes in such a way that adduction and flexion of the thumb is coactivated.

Summary. Looking at all the videos, though, I think that dystonia is also an option, as one can classify what we see as an abnormal movement leading to an abnormal position. I wonder whether there are mirror movements if the person moves the other hand. I cannot judge that on the video and that would further support dystonia. I also wonder whether it is task specific or if it also occurs with other activities.

Discussion

In the present series, uniformity of interview and examination across subjects allowed the recognition of a recurrent phenomenon in a subgroup of subjects, not previously described to our knowledge. The hand-opening and closure task, not necessarily performed during clinical neurologic examination of idiopathic dystonia in the absence of signs of parkinsonism, was here performed systematically. The videotaping of the examination allowed for repeated review for clarification by a senior movement disorder neurologist, to review subtle movement phenomena. Our goal was to define the phenomenology of this specific examination finding, thumb flexion during hand-opening and closure, noted in a subgroup of subjects during a structured neurological examination.

We specifically questioned whether the movement might reflect dystonia. Current Movement Disorder Society (MDS) consensus criteria define dystonia as “characterized by sustained or intermittent muscle
contractions causing abnormal, often repetitive, movements, postures, or both. Dystonic movements are typically patterned, twisting, and may be tremulous. Dystonia is often initiated or worsened by voluntary action and associated with overflow muscle activation.\(^7\) As mentioned by our experts, additional associated features may aid in its diagnosis, specifically the presence of mirror dystonia and/or sensory tricks alleviating the dystonic movement. As mentioned, features supportive of a diagnosis of dystonia here include the movement’s repetitive, patterned quality, as well as its apparent activation with a voluntary task. The movement was not noted during other manual tasks included in our examination. Although assessment for motor overflow is limited, as noted, movements of other fingers display subtle posturing. We lack additional history regarding subjective features or presence of sensory trick, which might confirm a diagnosis of dystonia; however, their absence would not exclude it, particularly given the phenomenon’s subtlety. Although sensory tricks are common in cranial and cervical dystonia, they are not common in limb or generalized dystonia, and they often abate with time.

Based on the observed phenomenology of recurrent thumb flexion during hand-opening and closure, all expert commentators agreed that underlying dystonia was the leading diagnosis (Table 1). Orthopedic or rheumatologic causes were felt unlikely. Several alternate neurologic or culturally influenced causes were mentioned.

Weakness of thumb extension due to lower motor neuron (median nerve) injury was considered as a possible cause; that is, an underlying weakness of thumb extension rather than overactivation of flexion. Confrontation testing was not performed; however, antigravity movements of thumb extension are demonstrated during hand-opening by all subjects, with the exception of the left hand of subject 4, in which thumb extension is not captured. As mentioned, no atrophy of the thenar eminence is apparent in any subjects (including subject 4). Possible anatomical variation resulting in coactivation of thumb flexion and adduction during finger flexion was also proposed. Coactivation of thumb flexors upon activation of finger flexors has been reported after central injury in stroke survivors.\(^28,29\) Synkinesis due to aberrant reinnervation following lower motor neuron injury, most commonly facial nerve, is well established. These phenomena are not described in the absence of neurological injury to our knowledge, although remain possible.

Two commentators noted similarities to the phenomenon of the cortical thumb in their differential diagnoses. Cortical thumb, or persistent adduction of the thumb, sometimes in association with persistent fisting of the hand with enclosure of the thumb by the other fingers, is considered a possible marker of corticospinal injury if persistent in infants,\(^30\) although it is present in a large portion (30–60%)\(^31,32\) of healthy neonates. As noted, characterization of cortical thumb, particularly regarding associated features, in adulthood is limited, although other corticospinal signs might be expected in association, not specifically assessed for here.

In the limited literature, dystonic tics are characterized by relatively slow, briefly sustained abnormal posture.\(^33\) They have been suggested to share features with dystonia, including an alleviating sensory trick.\(^34–36\) As mentioned, assessment for subjective features, including premonitory urge, discomfort with suppression of and subjective relief with performing the movement, might contribute to diagnostic clarification, reported in 72.7% in one case series of specifically adult-onset dystonic tics; fluctuation in severity, and co-occurrence with other clonic or vocal tics, may also aid in diagnosis of dystonic tic.\(^37\) Evidence from two small case series suggests a predominance of cranial–cervical over limb dystonic tics.\(^34,37\) Notably, no dystonic tics involving the upper limbs were reported in these series, although the diagnosis is not excluded here. Reports regarding prevalence and phenomenology of tic disorders in adults are limited,\(^38\) but suggest isolated limb tics, without other associated motor or vocal tics, are rare. As noted, motor behaviors can be affected by cultural convention. Culturally bound syndromes including Latah\(^39\) and the phenomenon of the jumping Frenchmen of Maine,\(^40\) characterized by exaggerated startle response followed by stereotyped vocal and motor behaviors, occur in isolated communities and are widely regarded as culturally influenced.\(^39–42\) We cannot here exclude culturally specific conventions in hand closure. However, the subtle asymmetries in execution between hands (e.g., in subjects 2, 4, and 5 [Video 1 and legend]) might be unexpected if the movement’s execution was influenced primarily by cultural convention. Furthermore, local investigators verified no such convention of hand movements in the Faroe Islands and verified no instructions were provided to perform hand movements with the thumb flexed.

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<th>Table 1. Leading Diagnosis and Differential Diagnosis by Author (Diagnoses Deemed Excluded by an Author Are Not Included Here)</th>
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In conclusion, the clinical diagnosis of dystonia relies on both careful assessment of phenomenology and associated features. We present a subtle but recurrent phenomenon, thumb flexion during hand closure and opening, not previously reported to our knowledge. Although assessment for associated confirmatory or exclusionary features is here limited, all experts agreed dystonia was the leading diagnosis based on video review. We thus propose the phenomenon as a previously unreported form of dystonia.

The substantial portion of subjects displaying the described thumb flexion is intriguing and suggests its prevalence is unlikely to be negligible, particularly in the setting of a previously reported high prevalence of focal dystonia in the Faroese population in Joensen’s service-based study. While other novel focal dystonias among population isolates have not been reported to our knowledge, forms of genetic dystonia have been reported with population-specific predominance. The combined dystonia, X-linked dystonia parkinsonism (Lubag syndrome), is perhaps the most well-known example, primarily affecting Filipino males with highest prevalence in the Province of Capiz on Panay Island. If a high dystonia prevalence is supported among the Faroese, we suspect a possible founder effect in this genetically homogenous and geographically isolated population. Further population-based studies of dystonia prevalence in the Faroe Islands are underway to characterize clinical features and disease burden, with prevalence of the present entity to be specifically characterized.

References


