From collar to coccyx: Truncal movement disorders - a clinical review

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Abstract

Background: Movement disorders affecting the trunk remain a diagnostic challenge even for experienced clinicians. However, despite being common and debilitating, truncal movement disorders are rarely discussed and poorly reviewed in the medical literature.

Objectives: To review common movement disorders affecting the trunk and provide an approach for clinicians based on the truncal region involved (shoulder, chest, diaphragm, abdomen, pelvis, and axial disorders). For each disorder, clinical presentation, aetiologic differential diagnosis, and 'clinical clues' are discussed.

Conclusion: This review provides a clinically-focused, practical approach to truncal movement disorders, which will be helpful for physicians in everyday practice.

Introduction

Despite being common, debilitating, and sometimes critical in deciphering disease aetiology, truncal movement disorders are rarely discussed and poorly reviewed in the medical literature. The trunk makes up over 35% of body surface area, yet it is seldom formally examined in clinical movement disorder practice. Even when abnormal trunk movements are spotted, their significance may be unclear. In this review, we provide a practical, clinically focused approach to movement disorders of the trunk. We divide our discussion into a number of different sections, based on the involved truncal region, an approach that we find most useful in clinical practice (see figure 1). Common and important differential diagnoses are discussed in the text, while a more exhaustive list of diagnostic considerations is given in the supplementary tables.

Shoulder movement disorders

Motor tics rank high on the differential diagnosis of hyperkinetic shoulder movements, particularly in children or young adults. They may be simple (affecting single muscle groups, for example, causing intermittent elevation) or complex (resembling purposeful, goal-directed behaviour) (1) and generally manifest along a rostro-caudal gradient (preferentially involving the face, neck, and shoulders). Patients frequently have neuropsychiatric comorbidities such as attention deficit-hyperactivity disorder (ADHD), obsessive compulsive disorders (OCD), and autism spectrum disorders (ASD) (2). Motor tics may exhibit suppressibility, unpleasant premonitory urges, and relief of unpleasant sensations upon the performance of the tic. These features, along with identification of other motor (eye blinks, ocular, facial movements) or vocal (sniffing, throat clearing) tics, help to seal the diagnosis (2). New onset of tics in adulthood is unusual and requires

exclusion of secondary causes, particularly structural brain disease, neurodegenerative disorders (Huntington's disease, neuro-acanthocytosis, Wilson's disease, and brain iron accumulation disorders), and illicit drug use (2).

Dystonia accounts for a large proportion of hyperkinetic shoulder movements in mid-life, where involvement of shoulder elevators such as the levator scapulae or trapezius, generally as part of a craniocervical dystonic syndrome, can produce either tonic or phasic shoulder movements. Clinical clues include patient demographics (isolated craniocervical dystonia being most common in females in the 5th and 6th decades of life), hypertrophy of involved muscles, the presence of a geste antagoniste, and associated dystonia in contiguous body regions (generally the neck) (3). Isolated shoulder dystonia is rare, mainly occurring as a task-specific phenomenon following overuse (4) or after a local trauma (5). Task-specific shoulder dystonia tends to affect young males (often in their 30s), and it is described primarily in highly-skilled sports people (e.g., golfers, billiard, and petanque players) and musicians but can occur in other instances following repetitive upper-limb activities (5). Fixed dystonic shoulder posturing should raise the question of functional dystonia.

Asymmetric parkinsonism, most commonly idiopathic Parkinson's Disease (PD), is the principal cause of shoulder hypokinesia. This manifests as slowed shoulder elevation on the affected side (reduced shoulder shrug to command) and is usually accompanied by other cardinal motor features (tremor, bradykinesia, rigidity) (6). Conversely, PD patients treated with levodopa can also develop hyperkinetic levodopa-induced dyskinesia affecting the shoulder region, particularly as 'peak-dose' phenomena. However, such movements are rarely isolated to the shoulder, and the diagnosis is usually evident from the history.

Two further hyperkinetic shoulder movement disorders are worth mentioning because, though rare, they are both associated with treatable neurological disorders.

- *Faciobrachial dystonic seizures (FBDS)* are semiologically distinct, immune-mediated focal seizure events characteristically encountered in leucine-rich glioma inactivated-1(LG1)-antibody associated limbic encephalitis (7). Typically manifesting as brief (< 3 seconds), frequent (median > 50 / day), coordinated contractions of the face, shoulder, and less frequently leg (though they can involve the upper limb only). Their presence often precedes the development of other disease features (confusion, amnesia, seizures, hallucinations, sleep disturbances, and hyponatremia). Early treatment with immunotherapy is critical in order to achieve seizure control (FBDS respond poorly to antiepileptic agents) and optimize these patients' outcomes (7).

- *Tonic spasms (TS)* are brief (seconds), paroxysmal dystonic-like spasms which occur in the setting of spinal inflammatory lesions, most commonly multiple sclerosis (MS) (8), but also others like neuromyelitis optica-spectrum disorders (NMOSD) (9). The upper limbs are most commonly affected, reflecting predilection for cervical cord involvement in MS. In contrast to primary dystonia, TS are painful and often exhibit a striking response to carbamazepine (8).

Chest movement disorders

Isolated thoracic movement disorders are incredibly rare. The pectoral muscles can be involved in *neuromyotonia*, a peripheral nerve hyperexcitability syndrome producing continuous rippling muscle activity, classically associated with anti- contactin-associated protein 2 (CASPR2) antibodies (10) (11) (12). Neuromyotonia –the presence of which can be confirmed on electromyography (EMG)- may be isolated or part of either Isaac's

syndrome (associated with muscle stiffness and hyporeflexia) (11) or Morvan's syndrome (associated with dysautonomia and headache and encephalopathy) (11).

Neuromyotonia must not be confused with fasciculations, which can be benign, but may reflect denervation and, if seen in the pectoral region, especially alongside muscle wasting, should raise suspicion of an anterior horn cell disease. They can be differentiated both on neurophysiological grounds and clinically, with fasciculations being brief, random, and fleeting muscle twitches in contrast to the continuous rippling seen in neuromyotonia.

Stimulus-sensitive chest muscle jerks might occur in stiff person syndrome (SPS) and progressive encephalomyelitis with rigidity and myoclonus (PERM) (13). Stiffness, slow voluntary movements, muscle hypertrophy, and lumbar hyperlordosis associated with continuous motor unit activity on EMG of paraspinal muscles suggest SPS. In contrast, severe stiffness accompanied by ataxia, dysautonomia, and psychiatric features are seen in PERM (13).

Diaphragmatic movement disorders

Diaphragmatic movement disorders can be broadly divided into tonic, clonic, and dyskinetic movements and often present with unusual abdominal movements or respiratory issues (see supplementary table 3.). The diagnostic work-up for diaphragmatic movement disorders should include abdominal examination at rest (when lying, sitting, or standing), during specific tasks (e.g., finger tapping), and with deep breathing, as deep inspiration and tasks may interrupt diaphragmatic tremor, thus giving a clue to the diagnosis (14). Real-time evaluation of both hemidiaphragms using diaphragmatic videofluoroscopy, EMG, and/or ultrasonography is also suggested (14). Alongside this, features of functional (distractibility, suggestibility, entrainment) or tardive (exposures to

dopamine receptor blocking drugs, akathisia, stereotypies, oro-bucco-lingual dyskinesia) movement disorders should be sought.

Tonic diaphragmatic spasm is rare and usually seen in the setting of specific toxidromes - in particular strychnine poisoning- or infections such as tetanus and furious rabies (15) (16) (17). In these conditions, spasms often begin in the jaw (trismus), face (risus sardonicus), and neck, spreading later to the chest, back, and abdomen (often producing striking opisthotonus). Severe, sometimes life-threatening laryngeal and/or diaphragm spasm may ensue. Diaphragmatic spasms may also be encountered in patients with generalised dystonia syndromes (e.g., dystonia-torsin family 1 member A -DYT-*TOR1A*-) in whom dystonic storms can be triggered by factors such as medication changes, infections, surgical procedures, or sudden discontinuation of deep brain stimulation (18).

The most common *clonic diaphragmatic movement, singultus* (hiccup), is a universal experience. It results from disruption within the hiccup reflex arc, comprising afferent (vagus nerve), central (upper spinal cord and brainstem), and efferent (phrenic nerve) limbs. Hiccups can be triggered by irritation at any of these points. While most often a benign, self-terminating condition, hiccups can occasionally become persistent (>2 days) or intractable (>1 month); persistent singultus should be investigated to rule out secondary causes. The list of potential culprits is vast (19). Important differential diagnoses not to miss include brainstem (especially area postrema) vascular or inflammatory lesions, electrolyte disturbances or cardiac diseases (19). Chronic neurological and gastrointestinal disorders also predispose to pathologic singultus. For instance, 20% of patients with PD report recurrent hiccups; interestingly, dopaminergic transmission is involved in the 'hiccup arc', perhaps explaining this phenomenon and the occasional reports of hiccups induced by dopaminergic agonists (19).

Other rare clonic diaphragmatic movements include *diaphragmatic tremor* and *diaphragmatic flutter*. Both manifest with unusual abdominal movements. They are differentiated from singultus by their high frequency - 4-60 Hz(tremor) and 60-200 Hz(flutter)- and absence of inspiratory sound. Espay et al. define diaphragmatic tremor as persistent, synchronous, rhythmic diaphragmatic contraction under some voluntary control, mainly without respiratory compromise or functional disability (14). In contrast, diaphragmatic flutter is an involuntary, uni- or bi-diaphragmatic myoclonic diaphragmatic contraction, which produces semi-rhythmic jerking of the abdominal wall and often associates with respiratory distress or abdominal wall pain (20) (21). Diaphragmatic tremor may sometimes have a tardive or functional aetiology (20). In contrast, diaphragmatic flutter often results from brainstem lesions, mainly vascular, involving the dentatorubroolivary connections (21).

Respiratory Dyskinesia is usually a tardive phenomenon manifesting as dyspnoea, uncoordinated breathing patterns, huffing, grunting, or forced inspiration against a closed glottis (22). A history of dopamine receptor blocking drug use, as well as other features compatible with tardive syndromes, suggest the diagnosis. Diaphragmatic dyskinesia can also be observed as a levodopa-induced phenomenon in patients with PD. In this instance, a clear temporal association with levodopa intake is useful in making the diagnosis (23).

Abdominal movement disorders

Choreic abdominal movements, known as “Belly dancer’s dyskinesia” (BDD), refer to undulating rhythmical movements of the abdominal wall causing distressing circular rotatory umbilical motion (21). These abdominal movements, which can be transient or continuous, may superficially resemble those seen with diaphragmatic disorders but differ

from these in that they are generated from the abdominal wall proper and have a sinuous, flowing nature as opposed to the jerky movements triggered by diaphragmatic contractions (21). Half of BDD cases follow local trauma or surgical procedures on the abdomen, often without any other abnormalities on examination. Functional disorders should be considered in these patients (24), especially if movements disappear during sleep. Drug-induced BDD has also been reported most commonly either as tardive events or levodopa-induced phenomena in patients with PD (25). Rarely, *abdominal myoclonus* may be observed, generally secondary to propriospinal myoclonus (see below), or extremely rarely, due to epileptic events (abdominal motor seizures) (26) (see supplementary table 4). The abdomen may also be involved in *stiff-person spectrum disorders*, producing board-like abdominal rigidity, usually alongside other characteristic features of these disorders (see below).

Pelvic movement disorders

Pelvic movement disorders are rare, but if present, are almost commonly the result of dopamine receptor blocking drug administration. *Tardive syndromes* may comprise various stereotypies including pelvic thrusting or rocking, often alongside other tardive phenomena (27). Equally, acute drug-induced dystonic reactions can involve the pelvis- these "tortipelvic crises" produce acute involuntary contractions of the abdominal wall, hip, and pelvic muscles (28). Functional pelvic movement disorders should also be considered particularly in the setting of thrusting/copulatory movements (29). Finally, *restless genital syndrome* is an unusual syndrome of intrusive and unwanted persistent genital or clitoral arousal in the absence of sexual desire. It is diagnosed when a person meets the criteria for genital arousal disorder alongside either restless legs syndrome (RLS) and/or overactive bladder or urethral hypersensitivity. It worsens while sitting, improves while

walking, and seems related to small-fiber sensory neuropathy of the dorsal nerve of the clitoris. In some cases, just like RLS, it may respond to dopamine agonist (DA) therapy (30).

Axial truncal movement disorders

Most axial movement disorders are ones of tonic postural abnormality, with one important exception-propriospinal myoclonus.

Postural disorders are classified according to the dominant direction of movement into camptocormia (from the Greek, meaning "to bend" -*κάμπτω, kamptō*- and "trunk" -*κόρμος, kormos*-), opisthotonus (from the Greek, “tension” -*Τάση, tonos*- coming from “behind” -*όπιστο, opistho*-) and pleurothotonus or Pisa syndrome (also from the Greek, “tension” -*Τάση, thotonus*- coming from “sideways” -*πλευρόθεν, pleuro*-).

Important features to assess during their evaluation include:

- Current and previous medication exposure- the development of camptocormia or Pisa syndrome in patients with PD may be associated with DA therapy (31). Moreover, abnormal truncal postures are frequent manifestations of tardive syndromes.
- Whether the abnormal movement is fixed or mobile- fixed postural abnormality may point to a structural spinal disease or a functional aetiology.
- Triggering and relieving factors: camptocormic posturing should improve significantly upon lying down. In contrast, spinal deformities (vertebral fractures, ankylosing spondylitis, acquired degenerative spondyloarthropathy) and functional

movements usually persist. Overflow muscle activity, e.g., during ambulation, may point to a dystonic disorder.

- Presence of other neurological signs: neurological signs, particularly parkinsonism, muscle weakness and/or atrophy (suggesting anterior horn cell diseases or myopathy), and hyperekplexia (32).

Camptocormia

Camptocormia describes a marked (>45°) forward flexion of the thoracolumbar spine in the sagittal plane when standing or walking, which improves with supine positioning (31). It is generally encountered as a part of neurodegenerative disorders, dystonic syndrome, or muscular disorders (33) (34).

The commonest cause of camptocormia is PD, wherein the risk may be influenced by sex, disease duration, previous DA exposure, and ethnicity (35). Camptocormia may also occur in atypical parkinsonian and other neurodegenerative disorders (36).

Isolated axial dystonic camptocormia is rare. It most commonly affects men in their 5th decade, with forward flexion being the dominant phenotype. Clues to a primary dystonic aetiology include overflow muscle activity (causing worsening with action, e.g., walking or exercising) and the presence of a sensory *geste* (37). Most cases are refractory to oral pharmacotherapies, but a response to botulinum toxin or deep brain stimulation may be seen (38).

Paraspinal muscle weakness secondary to inherited/acquired myopathies, anterior horn cell disease, or neuromuscular junction disorders may also produce camptocormia. Compensatory movements to overcome truncal weakness in such instances have been

misconstrued as sensory tricks, leading to misdiagnoses as axial dystonia (39). Clinical clues include weakness of other muscles, difficulty extending the trunk when prone, difficulty performing a 'sit up', or family history of muscle disease. Spinal imaging, paraspinal EMG, and occasionally muscle biopsy may be necessary where there is diagnostic uncertainty (32).

Structural and degenerative spinal diseases may produce forward truncal flexion, though, in contrast to 'true' camptocormia, this will not improve with supine positioning (32). (See expanded differential diagnosis on Supplemental Table 4).

Opisthotonus

Opisthotonus is a critical clue in movement disorder practice, as it carries a limited differential diagnosis that can be further distilled by considering the timing of onset, predominant spinal segment affected, and presence of other neurological and/or systemic signs.

Acute onset (over hours/days) should raise concern for infectious, drug-induced, and functional aetiologies. Tetanus and rabies are the most common infectious causes. In tetanus, jaw and neck spasms are often the first symptoms, but progression to global back spasms resulting in a fixed back-arching often occurs (15). In rabies, the spasms are intermittent and provoked by water ingestion (17). Drug-induced acute dystonic reactions most commonly occur in young men following the administration of dopamine antagonists. Risk factors include previous acute dystonic reactions and recent cocaine use. In addition to truncal posturing, oculogyric crises are also characteristic. 'Awake convulsions', characterised by episodic muscle spasms without loss of consciousness, occur within 15-30 minutes of strychnine exposure (18).

A more subacute/chronic evolution over weeks or months suggests neurodegenerative, autoimmune, or tardive aetiologies (40). The predominant location of the posturing may give a further clue. The predominant involvement of the upper back and neck (retrocollis) often occurs in tardive dystonia (41). Conversely, SPS predominantly affects the lumbar segments, leading to inability to bend forward and touch the toes and an exaggerated lumbar crease. In contrast to dystonia, SPS postures are fixed, do not improve upon recumbency, do not demonstrate sensory *gestes,* and have other associated features, including hyperekplexia and "freezing-like episodes" while walking (41). Additionally, neurophysiologic testing shows continuous motor unit activity and enhanced exteroceptive reflexes; anti-glutamic acid decarboxylase (anti-GAD) or anti-glycine antibodies are frequently detected (42).

Opisthotonic posturing is a classic manifestation of disorders of neurodegeneration with brain iron accumulation (NBIA) but can also be encountered in other isolated and combined dystonia syndromes. Additional oromandibular dystonia, pyramidal signs, and cognitive decline are important red flags for NBIA, especially pantothenate kinase 2 (*PANK2)* and phospholipase A2 group VI (*PLA2G6)* (40).

Pleurothotonus

Pleurothotonus or Pisa Syndrome is defined as >10° lateral spinal flexion upon standing, relieved almost completely by passive mobilization or supine positioning, especially common in multiple system atrophy (MSA) (31) (43). Roughly 10% of PD patients will also develop Pisa syndrome, with DA use being a risk factor. Tardive pleurothotonus is commonly described, and rarely it can be an isolated phenomenon without discernible cause (34).

Notably, all three postural truncal abnormalities, though classically opisthotonus, can be a manifestation of functional neurological disease. In this case, it often appears suddenly, sometimes triggered by physical and/or emotional events, demonstrates variability in phenomenology, distractibility (44) (45), and is more likely to be fixed than mobile (45).

Propriospinal myoclonus

Propriospinal myoclonus is a peculiar disorder characterised by painless, arrhythmic, usually flexor (but occasionally extensor) jerks which begin axially, usually in thoracic spinal segments, and propagate rostro-caudally to the neck, hips, and knees (46). Roughly half of the cases will demonstrate some position dependency, with the usual pattern being worsening of jerks when lying down or sitting (28) (46). Jerks may be provoked by tactile stimuli on the chest or abdomen, by eliciting tendon reflexes, but generally not in response to auditory stimuli (the latter being more suggestive of brainstem myoclonus). Middle-aged men are most commonly affected, and the majority of cases have a functional aetiology. The rare organic cases result from spinal cord pathology (most commonly thoracic disc herniations), and spinal imaging is therefore recommended in all cases. Acute onset, distractibility, variability, spread of jerks to the face, disappearance during sleep, and the presence of Bereitschaftpotentials (BP) on jerk-locked back-averaging of electroencephalography-electromyography (EEG-EMG) recordings are further clues to a functional aetiology (46).

Movement disorders of the back

Rarely, movement disorders can be restricted to other regions of the back. These may occur following spinal instrumentation or peripheral nerve trauma and assume diverse phenotypes, including ‘dancing dorsal quadrilaterals’, peri-scapular movements, and

various jerky, sinuous or tremulous movements of serratus anterior, latissimus dorsi, or trapezius following local nerve injuries (47) (48) (49).

Conclusion

Truncal movement disorders are a neglected area of movement disorder practice, which has received little traction in the field. Yet, a systematic approach to their evaluation, focusing on movement disorder phenomenology, the portion of the trunk affected, and the presence or absence of other diagnostic 'clues' (supplemental tables 1-4), can rapidly narrow the differential diagnosis. Age of onset is always an important consideration. Some disorders are particularly prevalent in young individuals, such as tics and inherited metabolic disorders, while others are primarily the remit of older age-degenerative diseases, tardive syndromes. The tempo of progression is also important to ascertain. Acute or subacute onset suggests infectious, toxic, autoimmune, or sometimes functional aetiologies, while slow progression suggests degenerative disorders. Tardive aetiologies enter the differential diagnosis of many abnormal truncal movements; a history of exposure to dopamine receptor blocking medications, either through prescription, poisoning, traditional or herbal medicines, must always be sought. Similarly, functional movement disorders should always be kept in mind, especially when other suggestive clues (distractibility, entrainability, history of joint hypermobility, and/or postural orthostatic-tachycardia syndrome) are present when this is the most common cause of the syndrome, e.g., propriospinal myoclonus, or when the presentation is incongruent with organic diseases. Such clinical approaches will help clinicians rapidly narrow diagnostic possibilities and apply diagnostic work-up, and in some cases, treatment, accordingly.

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Legends

Figure 1: Summary of common truncal movement disorders and clinical clues suggesting the diagnosis.

SUPPLEMENTAL TABLE 1: Disorders causing hyperkinetic shoulder movements.

SUPPLEMENTAL TABLE 2: Differential diagnosis in thoracic movement disorders.

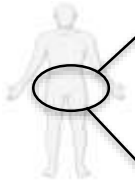
SUPPLEMENTAL TABLE 3: Differential diagnosis of diaphragmatic, abdominal, and pelvic movement disorders.

SUPPLEMENTAL TABLE 4: Differential diagnosis of axial truncal movement disorders.

SHOULDER MOVEMENT DISORDERS

Diagnosis	Clinical Clues
Tics	Primary: Young (<18y);male; comorbid OCD/ADHD; Other motor/verbal tics (blinking, throat clearing) Secondary: Adult onset; associated neurodegenerative disease
Dystonia	Male aged 30-40y (isolated, task specific);Female aged 40-50y(craniocervical) ; geste antagoniste; overflow muscle activity
Parkinsonism	Tremor, bradykinesia, rigidity; non-motor symptoms
FBDS	Cognitive decline; temporal lobe MRI signal change;LGI1 antibody +ve
Tonic Spasm	Inflammatory spinal cord disease; Carbamazepine responsive

PELVIC MOVEMENT DISORDERS

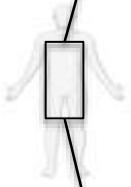


Diagnosis	Clinical Clues
Tardive pelvic thrusting	Other tardive features often present - perioral dyskinesia; akathisia; dystonia. History of neuroleptic exposure
Restless genital syndrome	Genital arousal disorder +restless legs syndrome and/or overactive bladder or urethral hypersensitivity; improves when walking/worse when seated

ABDOMINAL MOVEMENT DISORDERS

Diagnosis	Clinical Clues
Singultus (hiccups)	Clonic movement; inspiratory sound; Pathological if persistent (>2 days)
Diaphragmatic tremor	Frequency 4-60 Hz; rhythmic; some voluntary control; no respiratory compromise may be functional
Diaphragmatic flutter	Frequency 60-200 Hz; arrhythmic; involuntary; respiratory compromise
Respiratory dyskinesia	Uncoordinated breathing, huffing, grunting; tardive phenomenon
Tonic diaphragmatic spasm	Generally part of severe generalised dystonic syndrome (genetic, toxic, infectious);respiratory compromise
Belly dancer's dyskinesia	Sinuous, circular umbilical motion; may be functional or drug-induced

AXIAL MOVEMENT DISORDERS



Diagnosis	Clinical Clues
Camptocormia	Generally associated with parkinsonism, usually PD
Opisthotonos	Organic causes: Genetic isolated/combined generalised dystonia syndromes-DYT1, NBIA, inherited metabolic disease; tardive syndromes; acute drug-induced/infective dystonic reactions Common manifestation of functional disorders
Pleurothotonos	Frequently seen as tardive phenomenon; within parkinsonian disorders, characteristically seen in MSA, or in association with dopamine agonist therapy
Propriospinal myoclonus	Truncal jerks(usually flexor);worse when lying down; usually functional(Bereitschaftspotential); spinal imaging mandatory to outrule structural lesion
Stiff person syndrome	Hyperekplexia; truncal + limb stiffness;enhanced exteroceptive reflexes; continuous muscle fibre activity
Axial Dystonia	Usually males in 50s-60s; overflow muscle activity +/- sensory geste
Back movement disorders	Jerky/tremulous/sinuous movements of dorsal quadrilaterals, latissimus dorsi, serratus anterior etc. often following spinal procedures