

BALANCE AND GAIT DISORDERS

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Abstract

Neurological disorders of gait, balance and posture are both debilitating and common. Adequate recognition of these “disorders of axial mobility” is important as they offer useful diagnostic clues in patients with an uncertain clinical diagnosis, often early in the course of neurological disorders. Medical teaching programs typically take classical clinical presentations as the starting point, presenting students with a representative constellation of features that jointly characterize a particular axial motor syndrome. However, this is not the way patients usually present to a physician in clinical practice. Particularly in early disease stages, patients may display just one or at best only a few abnormal signs of gait, balance or posture. Importantly, each of these signs is never pathognomonic for any specific disorder, but comes with an associated differential diagnosis. In this video-supported viewpoint, we offer a new diagnostic approach, taking the presenting signs as the starting point for a focused differential diagnosis and a tailored search into the underlying neurological syndrome.

Background

Careful recognition of neurological gait and balance disorders is very important in everyday clinical practice, for a variety of reasons. First, gait and balance disorders impose significant disability for affected individuals. One main concern is the development of recurrent falls, which can lead to injuries, and which are associated with a reduced survival (1). For example, it is increasingly recognised that freezing of gait is the most important cause of falls in patients with parkinsonism, presumably because the feet suddenly get stuck while the subject is attempting to move forward (2). For most patients, freezing of gait is at least partially treatable, but recognition of freezing of gait in the clinical examination room can be very difficult. Second, difficulties with walking and maintaining balance force patients to reduce their physical activities, and physical inactivity is in turn associated with a host of negative consequences, including a worsening of disease symptoms, development of osteoporosis, sleep disorders, and a reduced survival (3). The tendency to become physically less active is compounded by a fear of falling, which commonly develops in patients with gait and balance disorders, even among those who have never sustained actual fall before (4;5). Third, specific features of gait and balance can offer important diagnostic clues in patients with an uncertain clinical diagnosis. For example, the development of postural instability and recurrent falls within the first year of disease onset in patients with a hypokinetic-rigid syndrome suggest the presence of a form of atypical parkinsonism, such as progressive supranuclear palsy or multiple system atrophy (6-8). Changes in posture can also offer important diagnostic information (9). For example, a disproportionate antecollis in patients with a hypokinetic rigid syndrome suggest the presence of multiple system atrophy, or perhaps a side-effect of dopamine receptor agonists in patients with Parkinson's disease. Importantly, changes in gait typically develop early on in the course of many neurological disorders, as is seen in for example patients with hereditary cerebellar ataxia (10). Another example is the asymmetrically reduced arm swing that may present as an early feature in patients with Parkinson's disease (11). Recent work has even demonstrated that subtle changes in gait can present in carriers of mutations in the dominant *LLRK2* Parkinson gene who are otherwise clinically asymptomatic (12). Finally, gait slowing is another early – but nonspecific – marker of underlying neurological dysfunction (13;14) and a predictor of mortality (15;16).

To improve the recognition of gait and balance disorders, several classification schemes have been proposed (17;18). In case of gait disorders, it is a common approach to define the type of gait disorder according to the main presenting feature. Examples include an ataxic gait, a dystonic gait, or a waddling gait. The typical way of teaching students and residents about such gait disorders is to present a typical phenotype, and to list all the possible features associated with this specific type of neurological gait disorder. However, this is not the way patients usually present to the doctor in clinical practice. Indeed, patients may display only one or at most a few abnormal gait or balance signs, and some of these may fit with different types of neurological disorders. In this syllabus and in our video presentation, we therefore offer a new approach to disorders of gait, balance and posture, taking typical presenting features as the starting point for recognising the associated neurological syndrome, and for building a differential diagnosis. So, as an example, rather than summarising the list of gait features associated with a functional gait disorder (which can include a scissoring gait), we will discuss the differential diagnosis of a patient presenting with a scissoring gait (which includes a functional gait disorder, but also various other neurological conditions with a very different cause, prognosis and treatment). Our presentation will be organised around Tables 1-3 that are presented below, and which lists the most important presenting features of gait, balance and postural disorders. The tables also list a set of suggested tests that can be used to elicit the appropriate gait and balance features. The use of standardized rating scales can be helpful in this regard, as these serve as a guide to perform several of the most important clinical tests. Examples include generic scales, e.g. the Tinetti Mobility Index (19), and disease-specific scales, e.g. the Rating Scale for Gait Evaluation in PD (20). During the presentation, we will present illustrative videos of these various features, encouraging the audience to first define the key presenting gait feature, and to then work up the associated differential diagnosis. We will also emphasize and illustrate the appropriate performance of the most important gait and balance tests.

References

- (1) Wenning GK, Ebersbach G, Verny M, Ray Chaudhuri K, Jellinger K, Mckee A, et al. Progression of falls in postmortem-confined parkinsonian disorders. *Mov Disord* 1999;14:947-50.
- (2) Nutt JG, Bloem BR, Giladi N, Hallett M, Horak FB, Nieuwboer A. Freezing of gait: moving forward on a mysterious clinical phenomenon. *Lancet Neurol* 2011 Aug;10:734-44.
- (3) Speelman AD, van de Warrenburg BP, van NM, Petzinger GM, Munneke M, Bloem BR. How might physical activity benefit patients with Parkinson disease? *Nat Rev Neurol* 2011 Sep;7:528-34.
- (4) Scheffer AC, Schuurmans MJ, van DN, van der HT, de Rooij SE. Fear of falling: measurement strategy, prevalence, risk factors and consequences among older persons. *Age Ageing* 2008 Jan;37:19-24.
- (5) Mak MK, Pang MY. Fear of falling is independently associated with recurrent falls in patients with Parkinson's disease: a 1-year prospective study. *J Neurol* 2009 Oct;256(10):1689-95.
- (6) Litvan I, Agid Y, Calne D, Campbell G, Dubois B, Duvoisin RC, et al. Clinical research criteria for the diagnosis of progressive supranuclear palsy (Steele-Richardson-Olszewski

- syndrome): report of the NINDS-SPSP international workshop. *Neurology* 1996 Jul;47:1-9.
- (7) Wenning GK, Ben-Shlomo Y, Magalhaes M, Daniel SE, Quinn NP. Clinical features and natural history of multiple system atrophy. An analysis of 100 cases. *Brain* 1994;117:835-45.
 - (8) Wenning GK, Geser F, Krismer F, Seppi K, Duerr S, Boesch S, et al. The natural history of multiple system atrophy: a prospective European cohort study. *Lancet Neurol* 2013 Mar;12(3):264-74.
 - (9) Doherty KM, van de Warrenburg BP, Peralta MC, Silveira-Moriyama L, Azulay JP, Gershanik OS, et al. Postural deformities in Parkinson's disease. *Lancet Neurol* 2011 Jun;10:538-49.
 - (10) Jacobi H, Hauser TK, Giunti P, Globas C, Bauer P, Schmitz-Hubsch T, et al. Spinocerebellar ataxia types 1, 2, 3 and 6: the clinical spectrum of ataxia and morphometric brainstem and cerebellar findings. *Cerebellum* 2012 Mar;11:155-66.
 - (11) Lees AJ. When did Ray Kennedy's Parkinson's disease begin? *Mov Disord* 1992;7:110-6.
 - (12) Mirelman A, Gurevich T, Giladi N, Bar-Shira A, Orr-Urtreger A, Hausdorff JM. Gait alterations in healthy carriers of the LRRK2 G2019S mutation. *Ann Neurol* 2011 Jan;69:193-7.
 - (13) Ashburn AM, Stack E, Pickering RM, Ward C. Predicting fallers in a community-based sample of people with Parkinson's disease. *Gerontology* 2001;47:277-81.
 - (14) Verghese J, Wang C, Lipton RB, Holtzer R, Xue X. Quantitative gait dysfunction and risk of cognitive decline and dementia. *J Neurol Neurosurg Psychiatry* 2007 Sep;78:929-35.
 - (15) Cooper R, Kuh D, Hardy R. Objectively measured physical capability levels and mortality: systematic review and meta-analysis. *BMJ* 2010;341:c4467.
 - (16) Afilalo J, Eisenberg MJ, Morin JF, Bergman H, Monette J, Noiseux N, et al. Gait speed as an incremental predictor of mortality and major morbidity in elderly patients undergoing cardiac surgery. *J Am Coll Cardiol* 2010 Nov 9;56:1668-76.
 - (17) Nutt JG, Marsden CD, Thompson PD. Human walking and higher-level gait disorders, particularly in the elderly. *Neurology* 1993;43:268-79.
 - (18) Snijders AH, van de Warrenburg BP, Giladi N, Bloem BR. Neurological gait disorders in elderly people: clinical approach and classification. *Lancet Neurol* 2007 Jan;6:63-74.
 - (19) Tinetti ME. Performance-oriented assessment of mobility problems in elderly patients. *J Am Geriatr Soc* 1986;34:119-26.
 - (20) Thomas M, Jankovic J, Suteerawattananon M, Wankadia S, Caroline KS, Vuong KD, et al. Clinical gait and balance scale (GABS): validation and utilization. *J Neurol Sci* 2004 Jan 15;217:89-99.

Table 1. Abnormalities that can be observed during sitting, during rising from sit-to-stance, and during quiet upright stance. The second column lists the various signs that can be observed during these portions of the neurological exam. The third column lists a few suggestions for the differential diagnosis that is associated with each of these signs, without claiming to be comprehensive.

Examination	Abnormalities that can be observed	Suggestions for the differential diagnosis
Sitting	Leaning sideways	<ul style="list-style-type: none"> • Vertebral column deformities • Pisa syndrome <ul style="list-style-type: none"> ○ Drug-induced (e.g. dopamine receptor antagonists) ○ Associated with neurodegeneration (Alzheimer’s disease, PD, atypical parkinsonism) • Pusher syndrome (unilateral thalamic stroke) • Trunk dystonia (including idiopathic Pisa syndrome)
	Drifting backwards	<ul style="list-style-type: none"> • PD, atypical parkinsonism (PSP) • Higher-level gait disorders • Trunk dystonia (opisthotonus), often drug-induced (dopamine receptor antagonists)
	Forward dropping of the head (disproportionate antecollis)	<ul style="list-style-type: none"> • MSA • ALS • Myasthenia gravis • Polymyositis • Focal posterior cervical myositis • Drug-induced (dopamine receptor agonists, amantadine)
	Backward dropping of the head (retrocollis)	<ul style="list-style-type: none"> • PSP • Cervical dystonia • Young-onset PD • Drug-induced (dopamine receptor antagonists)
Arising from a chair	Feet not brought under the body while arising – inappropriate strategy for arising	<ul style="list-style-type: none"> • Higher-level gait disorders
	Slow, or requiring multiple attempts, or need to push off with	<ul style="list-style-type: none"> • Aspecific; e.g. proximal weakness, PD, higher-level gait disorders

	arms	
	Reckless rising ('rocket sign')	<ul style="list-style-type: none"> • PSP
	Shaking of the legs upon standing (negative orthostatic myoclonus)	<ul style="list-style-type: none"> • Idiopathic orthostatic myoclonus • Survivors of postanoxic encephalopathy • Frontal vascular lesions • Atypical parkinsonism • Alzheimer's disease • Normal pressure hydrocephalus
	Legs wide apart while arising	<ul style="list-style-type: none"> • Sensitive but aspecific, e.g. vestibular ataxia, sensory ataxia, cerebellar ataxia, atypical parkinsonism, chorea
Quiet standing (eyes open)	Normal to narrowed base of support	<ul style="list-style-type: none"> • PD • Spastic paraparesis
	Widened base of support	<ul style="list-style-type: none"> • Sensitive but aspecific, e.g. vestibular ataxia, sensory ataxia, cerebellar ataxia, atypical parkinsonism, chorea
	Progressive instability	<ul style="list-style-type: none"> • Orthostatic tremor • Negative myoclonus • Functional
	Excessive spontaneous sway	<ul style="list-style-type: none"> • Dyskinesias (PD, Huntington's disease, etc.) • Higher-level gait disorders • Functional
	Leaning or drifting sideways	<ul style="list-style-type: none"> • See sitting examination
	Excessive trunk flexion, that persists when lying down	<ul style="list-style-type: none"> • Vertebral column deformities
	Excessive trunk flexion, disappears when lying down	<ul style="list-style-type: none"> • Trunk or hip weakness (myasthenia gravis, motoneuron disease, myopathies) • Camptocormia (PD, atypical parkinsonism)

PD=Parkinson's disease; MSA=multiple system atrophy; ALS=Amyotrophic lateral sclerosis; PSP=progressive supranuclear palsy

Table 2. Abnormalities that can be observed during gait. The second column lists the various signs that can be observed during these portions of the neurological exam. The third column lists a few suggestions for the differential diagnosis that is associated with each of these signs, without claiming to be comprehensive.

Element of gait	Abnormalities that can be observed	Suggestions for the differential diagnosis
Gait initiation	Difficulty starting	<ul style="list-style-type: none"> • PD, atypical parkinsonism • Primary progressive freezing • Other causes of lower-body parkinsonism (e.g. normal pressure hydrocephalus, vascular parkinsonism)
Gait maintenance	Sudden episodes of inability to step, particularly when turning or when passing through doorways (freezing of gait)	<ul style="list-style-type: none"> • PD, atypical parkinsonism • Primary progressive freezing of gait • Other causes of lower-body parkinsonism (e.g. normal pressure hydrocephalus, vascular parkinsonism)
Gait termination	Inability to stop (festination)	<ul style="list-style-type: none"> • PD, atypical parkinsonism • Other causes of lower-body parkinsonism (e.g. normal pressure hydrocephalus, vascular parkinsonism)
Base width	Narrowed base of support	<ul style="list-style-type: none"> • PD • Spastic paraparesis (e.g. hereditary spastic paraplegia)
	Widened base of support	<ul style="list-style-type: none"> • Aspecific, e.g. atypical parkinsonism, cerebellar ataxia, sensory ataxia, vestibular ataxia, higher-level gait disorders. Very wide base of support in some functional disorders
	Scissoring of the legs	<ul style="list-style-type: none"> • Spastic paraparesis (e.g. juvenile cerebral palsy) • Dystonia • Higher-level gait disorder • Huntington's disease (scissoring due to chorea) • Compensation for freezing of gait (e.g. PD) • Functional
	Unable to walk in a straight line, sideways	<ul style="list-style-type: none"> • Unilateral vestibular ataxia, unilateral cerebellar ataxia (consistently

	deviation (veering of gait)	<p>veering into direction ipsilateral to the lesion)</p> <ul style="list-style-type: none"> • Functional (often veering from side to side)
Step length, step height and cadence	Reduced step height	<ul style="list-style-type: none"> • PD, parkinsonism • Foot drop due to: <ul style="list-style-type: none"> • Neuropathy of common fibular nerve or sciatic nerve. • L5 radiculopathy • Charcot Marie Tooth disease • Muscular dystrophy • Stroke or multiple sclerosis (causing distal weakness)
	Small steps	<ul style="list-style-type: none"> • Sensitive but aspecific sign: <ul style="list-style-type: none"> ○ Caused by underlying pathology: common in PD, atypical parkinsonism and normal pressure hydrocephalus ○ Compensation for perceived instability, e.g. functional gait
	Irregular step size (cadence)	<ul style="list-style-type: none"> • Ataxia (cerebellar, vestibular) • Higher-level gait disorders • Chorea
	Reduced stance phase on the affected side (limping)	<ul style="list-style-type: none"> • Pain (antalgic gait); e.g. to trauma or arthritis • Functional
Arm swing	Unilaterally reduced arm swing	<ul style="list-style-type: none"> • Hemiparesis • Dystonia • PD • Orthopaedic or rheumatologic disorders affecting the elbow or shoulder joint
	Bilaterally reduced arm swing	<ul style="list-style-type: none"> • PD, parkinsonism • Dystonia • Orthopaedic or rheumatologic disorders affecting the elbow or shoulder joint
	Excessive arm swing	<ul style="list-style-type: none"> • Chorea • Levodopa-induced dyskinesias • Normal pressure hydrocephalus (in order to compensate for the poor

		<ul style="list-style-type: none"> body propulsion) • Hypotonia • Functional
	Tremor appearing in hand during walking	<ul style="list-style-type: none"> • PD, parkinsonism
	Posturing with the arm	<ul style="list-style-type: none"> • PD, dystonia • Secondary to shoulder/neck injury • Alien limb phenomenon • Functional disorders
Movement fluidity	Dropped foot, lifting the leg higher than normal (<i>steppage</i> gait)	<ul style="list-style-type: none"> • Neuropathy of common fibular nerve or sciatic nerve. • L5 radiculopathy • Charcot Marie Tooth disease • Muscular dystrophy/myopathy • Stroke or multiple sclerosis (causing distal weakness) • Foot dystonia (causing a functional lengthening of the leg)
	Knees giving way (buckling of the knees)	<ul style="list-style-type: none"> • Functional • Quadriceps weakness (limb-girdle myopathy, inclusion body myositis) • Increased knee extension moment because of pes equines (e.g due to upper motor neuron syndrome) • Cataplexy • Negative myoclonus • Dystonia
	Locking of the knees	<ul style="list-style-type: none"> • Cerebellar ataxia • Severe arthrosis/arthritis of the knees • Compensation for quadriceps weakness (e.g. limb-girdle myopathy, IBM)
	Pelvis drop at side of the swing leg, resulting in alternating lateral trunk movements (waddling gait, bilateral Trendelenburg gait)	<ul style="list-style-type: none"> • Bilateral proximal leg and hip girdle muscle weakness. Common in muscular dystrophy (e.g. LGMD, FSHD, Duchenne's muscular dystrophy) and metabolic myopathy (e.g. thyroid myopathy) • Bilateral L5 radiculopathy resulting in weakness of m. gluteus medius • Secondary to joint instability (as in

		Ehlers-Danlos syndrome)
	Stiff gait	<ul style="list-style-type: none"> • Many disorders, including myotonia, dystonia, stiff man (or leg) syndrome
	Bizarre gait pattern	<ul style="list-style-type: none"> • Chorea (including levodopa-induced dyskinesias) • Dystonia • Higher-level gait disorders • Functional
	Twisting or turning in of the foot (foot dystonia)	<ul style="list-style-type: none"> • Young onset PD (exercise-induced dystonia) • (Cerebrovascular) basal ganglia lesion • Corticobasal degeneration • Primary dystonia • Functional
Gait speed	Slow	<ul style="list-style-type: none"> • Aspecific, perceived imbalance, PD, higher level gait disorders, functional gait disorders
	Fast, safe	<ul style="list-style-type: none"> • Vestibular syndromes • Orthostatic tremor
	Fast, unsafe	<ul style="list-style-type: none"> • PSP • Alzheimer's disease • Frontal disinhibition

FSHD=facioscapulohumeral dystrophy; LGMD= limb girdle muscular dystrophy; PD=Parkinson's disease; PSP=progressive supranuclear palsy

Table 3. Additional tests that can be used to elicit the appropriate gait and balance features.

Test	Function	Implications
Quiet standing (eyes closed; Romberg test)	Helps differentiate	Increased sway, or unable to stand independently, can be observed in sensory ataxia (sensory neuropathy) Involuntary movements, as is drug-induced dyskinesias or chorea Excessive sway corrected by distraction is observed in case of anxiety / functional disorders
Response to external perturbation <ul style="list-style-type: none">• Pull test• Push-and release	Detection of symptoms	Patients with postural instability respond with more than two steps, or take no steps at all
Walking (eyes closed)	Helps differentiate	Deviation to one side is seen in unilateral vestibular ataxia, and unilateral cerebellar ataxia
Turning of the head during gait	Helps differentiate	Worsening of gait is seen in vestibular ataxia
Walking backward compared to walking forward	Helps differentiate	Discrepancy with walking forward is seen in dystonia (suggesting task-specificity) and functional gait disorders (suggesting inconsistency)
Regular walking compared to running	Helps differentiate	Better running than regular walking can be seen in dystonia, PD, and functional gait disorders
Tandem walking	Helps differentiate	Tandem walking without side steps is preserved in PD, but impaired in atypical parkinsonism, ataxia and other problems with mediolateral instability
Rapid 360 degree turns on the spot	Detection of symptoms	Frequently evokes freezing of gait
Walking with short steps rapidly	Detection of symptoms	Frequently evokes freezing of gait