Camptocormia: Pathogenesis, Classification and Response to Therapy

Shaheda Azher, MD and Joseph Jankovic, MD

Parkinson's Disease Center and Movement Disorders Clinic, Department of Neurology, Baylor College of Medicine, Houston, Texas

Abstract

Initially considered a psychogenic disorder, camptocormia, an abnormal posture with marked flexion of thoracolumbar spine, which abates in the recumbent position, is becoming an increasingly recognized feature of parkinsonian and dystonic disorders. Prior reports were limited by sample size, short follow-up, and paucity of data on response to therapy. We report fifteen patients evaluated in our Parkinson's Disease Center and Movement Disorders Clinic diagnosed with camptocormia. In addition to detailed neurological assessment all patients were videotaped. The mean age was 66.9 ± 15.9 years, duration from onset of neurological symptoms to development of camptocormia was 7.2 ± 7.6 years, and the mean duration of camptocormia was 3.9 ± 3.1 years. Of the 15 patients, eleven (73.3%) had Parkinson's disease (PD). The diagnosis in the other patients included: abdominal dystonia [3] and Tourette's syndrome [1]. All eleven patients with PD received levodopa, with minimal or no improvement in their camptocormia. Eight patients received botulinum toxin type A (BTX) injections into the rectus abdominus, with notable improvement in four. One patient underwent bilateral subthalamic nucleus deep brain stimulation for PD, but there was no improvement in camptocormia. Based on our series and a thorough review of the literature we classified camptocormia into the following nine categories: 1. Parkinsonism, 2. Dystonia, 3. Spine abnormalities, 4. Brain injury, 5. Stroke, 6. Neuromuscular disorders, 7. Psychogenic, 8. Misc., and 9. Idiopathic. Camptocormia is a heterogeneous disorder with multiple etiologies and variable response to systemic and local therapies.

Introduction

Camptocormia (from the Greek "kamptos" = bend and "kormos" = trunk), also referred to as "bent spine syndrome", is characterized by abnormal posture of the trunk with marked flexion of the thoracolumbar spine which increases during walking and abates in the recumbent position [Figure 1]. Rarely reported in the literature, camptocormia was first described by the English physiologist and surgeon Brodie in 1818 and again in 1837 [1,2]. Until quite recently, camptocormia was assumed to be a psychogenic disorder described as a conversion reaction during World War I and II in male military recruits and soldiers who were unable to cope with the stress of combat preparation and unpleasant military life, perhaps promoted by stooped posture when walking in the trenches. The French neurologists, Souques and Rosanoff-Saloff, drew attention to this disorder in their report of 16 cases in 1915 [3].

Subjects and Methods

We reviewed the medical records and videos of patients diagnosed with camptocormia in our Parkinson's disease and Movement Disorders Clinic between 1980 and 2004. The diagnosis of camptocormia was made if the patients exhibited marked (45° to 90°) flexion of their thoracolumbar spine that increased during walking and markedly abated or disappeared in the recumbent position. We excluded patients with isolated neck flexion ("head drop syndrome") [4-7], but we did include patients with the combination of camptocormia and head drop. In addition to complete neurological examination, patients with PD were rated on the Unified Parkinson's Disease Rating Scale.

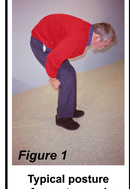


Figure 1

Typical posture of camptocormia

Table 1. Camptocormia: Demographics (N = 15)

Clinical variables [a]	
Sex	9 M, 6 F
Presence of family Hx of PD	5 (33.3)
Mean age at initial visit, yrs	61.9 ± 17.9
Mean age at onset of neurological symptoms, yrs	53.9 ± 18.1
Latency between initial symptom and camptocormia, yrs	7.2 ± 7.6
Duration of camptocormia, yrs	3.9 ± 3.1
Presence of sensory trick	4 (26.6)
Presence of abdominal contractions	5 (33.3)
Type of predisposed / Comorbid condition	
Fall/back surgery	5 (33.3)
Sleep disorder — sleep apnea and REM behavior disorder	4 (26.6)
Presence of RLS	1 (6.7)
Presence of ET and dementia	2 (13.3)

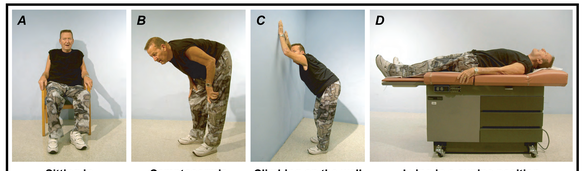
[a] Percent in parenthesis
PD = Parkinson's disease; RLS = Restless leg syndrome; ET = Essential tremor

Results

Fifteen patients (9M, 6F) with camptocormia were included in this series [Tables 1]. Their mean age was 66.9 ± 15.9 years, mean duration of neurological symptoms was 11.1 ± 7.8 years, and the duration of camptocormia was 3.9 ± 3.1 years (range: 1–13 years). All patients exhibited flexed posture when they were either standing or ambulating, but were able to sit erect, and extend their trunk fully when "climbing up the wall" with hands touching the wall, or lying down in supine position. Four patients described a powerful pulling sensation associated with palpable contraction of the rectus abdominus. Eleven of the fifteen (73.3%) patients had clinical diagnosis of probable idiopathic PD based on the NIH criteria [8]. Of the remaining four patients, one had abdominal dystonia with initial presentation of stooped posture, one had abdominal dystonia after disc surgery, one had abdominal dystonia producing flexion of the trunk secondary to syringomyelia, and one had Tourette's syndrome manifested by dystonic tic. Except for syringomyelia between C7 and T7 in one patient, no specific neuroimaging abnormalities were found. All eleven patients with PD were treated with levodopa with good response in their motor symptoms, but minimal or no effect on the camptocormia [Table 2]. Eight patients received BTX injections into rectus abdominus muscles with moderate to marked improvement in their posture lasting for about three months after each injection. One patient underwent bilateral STN DBS for PD associated with disabling levodopa-related motor complications, but there was no improvement in camptocormia.

Illustrative Case Reports

CASE #6 (VIDEO SEGMENT 1) This 66-year-old right-handed Caucasian man presented with progressive stiffness, rigidity and inability to stand straight since age 58. He dated the onset of his symptoms to a fall complicated by a fracture of his right humerus. He also complained of back pain radiating to his right leg attributed to spinal stenosis at L3-L4 and L4-L5, confirmed by imaging studies. On examination he had marked rigidity in all limbs, bradykinesia and camptocormia with his spine bent to 90° on walking. When he was sitting in a chair he maintained erect posture, but once he started walking, he immediately assumed markedly flexed posture. He was able to climb the wall to an erect posture and lie down flat in a supine position. Prior to our evaluation he received clonazepam (0.5 mg/d), levodopa (600 mg/d), pramipexole (6 mg/d) and a course of IVIG for presumed "stiff-peson syndrome" with no improvement in his symptoms. He also received BTX injections in his lumbar paraspinal muscles without any benefit. He was subsequently placed on a high dose of levodopa (1500 mg/d), but he developed severe fluctuations in his blood pressure associated with nausea and the individual levodopa dosages were decreased and given at more frequent intervals which resulted in resolution of the adverse effects and marked improvement in his rigidity and bradykinesia, but no change in his posture [Figure 2].



Sitting in an erect position | Camptocormia | Climbing on the wall | Lying in a supine position

Figure 2

Table 2. Treatment Outcome (N = 15)

Therapy		n	%	Response to CC
Pharmacological treatment:	Anticholinergics	3	20.0	1-Partial response
	Amantadine	4	26.6	NR
	Selegiline	3	20.0	NR
	Dopamine agonists	9	60.0	NR
	Levodopa	8	53.3	NR
	Muscle relaxants	4	26.6	NR
	Anti tremor medications	3	20.0	NR
	Tetrabenazine	2	13.3	1-Responded
	Epidural injections	1	6.6	NR
	Intrathecal medications	1	6.6	NR
Non-pharmacological therapy:	Massage therapy/Stretching Exercises	3	20.0	NR
	Back brace	1	6.6	NR
Botulinum toxin injections		8	53.3	4 -Responded
Surgery:	DBS (BSTN)	1	6.6	NR
	Bilateral pallidotomy	1	6.6	NR
	Back surgery (after onset of CC)	1	6.6	NR

CC = Camptocormia; NR = No response; DBS = Deep brain stimulation; BSTN = Bilateral subthalamic nucleus

CASE #10 (VIDEO SEGMENT 2) This 61-year-old Caucasian woman underwent extensive lumbar surgery for herniated L4-L5 disc when she was 55 years old. Three months post surgery she developed pain and spasms involving her lower back, abdomen and groin, present only on standing. Over the years her symptoms progressed and were manifested chiefly by marked contractions of abdominal muscles, particularly the rectus abdominus, associated with a pulling sensation and bending of her trunk forward. Unable to walk without assistance, the patient was able to straighten her back by stepping onto a chair (sensory trick). She was treated with multiple medications including baclofen, clonazepam (60 mg/d), gabapentin (1800 mg/d), and tetrabenazine (75 mg/d) with no improvement in her symptoms. She received BTX injections into rectus abdominus, continuous intrathecal baclofen and fentanyl infusion without noticeable improvement in her symptoms.

Table 3. Clinical Characteristics and Treatment Outcome of All Reported Patients

Source	Year	Sex	Age	Etiology/Predisposing Factors	Treatment [*Response (n)*]
Lazare	1970	1F	38	Conversion disorder; Acute onset	Psychotherapy [*GR*]
Carter	1972	1M	22	Conversion disorder; Back pain	Muscle relaxants and PT [*NR*]; Release from active duty [*Complete relief*]
Soreff	1983	1M	29	Conversion disorder, triggered by sciatica; 90° CC	Epidural anesthesia [*PR*]; PT [*NR*]
Rosen	1985	1F	31	Psychogenic; S1 radiculopathy	Epidural block [*NR*]; Psychotherapy [*GR*]
Massa	1989	1M	18	Mood disorder; Remote Hx of foot ball injury	Psychotherapy and amitriptyline [*GR*]
Miller	1990	6-N/A	22	Conversion disorder; Prior Hx of back injury	Psychotherapy [*GR (1); Lost for follow up (5)*]
Perez-Sales	1990	1M	36	Conversion disorder; Prior Hx of head injury in the army	Carbamazepine [—]
Sinel	1990	1M, 1F	53	Psychogenic; Normal EMG and myelogram	Psychotherapy [—]
Laroche	1995	5M, 22F	69	Primary muscular disease	— [—]
Ehrenstein	1996	1F	75	Myopathy	Prednisolone [*NR*]
Abdulhadi	1996	1M	47	CC as a kinematic abnormality due to excessive anterior pelvic tilt	PT [*GR*]
Zwecker	1998	1M	49	Paraneoplastic syndrome of Non-Hodgkin's lymphoma	PT, hydrotherapy, dorso-lumbar corset, and chemotherapy [*NR*]
Djaldetti	1999	2M, 6F	66	PD	L-dopa started soon after onset [*GR (4)*]; L-dopa initiation delayed [*NR (4)*]
Nieves	2001	1M, 1F	72	Right putaminal infarct (1) Right lenticular hemorrhagic infarct (1)	L-dopa [*NR (1)*]; PT [*GR (1&2)*]
Van Garpen	2001	1M	57	Amyotrophic lateral sclerosis; Diffuse muscle cramps	— [—]
Friedman	2001	1M	75	PD (CC as form of dystonia)	L-dopa [*NR*]
Wunderlich	2002	1M	63	PD; CC with pain s/p enteral infection; Focal myositis	Steroids [*GR*]
Nandi	2002	1M		PD	Bilateral GPi DBS [*GR*]
Slawek	2003	1F	49	PD (CC as form of dystonia)	L-dopa, pergolide, muscle relaxants, NSAIDS [*NR*]; Right pallidotomy [*GR*]
Schäbitz	2003	4M	66	MSA (1); PD (2); ET and PD (1)	L-dopa, bromocriptine, pergolide, pramipexole, amantadine, and bilateral STN [*NR*]; L1 S1 spine surgery [*GR (1)*]
Inzelberg	2003	1F		Autosomal recessive juvenile parkinsonism; Parkin gene; Truncal dystonia	L-dopa [*NR*]
Holler	2003	2M	66	PD (CC as form of dystonia)	L-dopa, gabapentin, clonazepam, baclofen, and ropinirole [*NR*]
Azher	2004	9M, 6F	67	PD (11); TS (1); Abdominal dystonia (3)	L-dopa [*PR or NR (11)*]; BTX injections [GR (*4/8*)]; STN DBS [*NR (1)*]

GR = Good response; PR = Partial response; NR = No response; CC = Camptocormia; PT = Physical therapy; Hx = History; PD = Parkinson's disease; EMG = Electromyography; L-dopa = Levodopa; DBS = Deep brain stimulation; GPi = Globus pallidus internus; STN = Subthalamic nucleus; NSAIDS = Non steroidal anti-inflammatory drugs; MSA = Multiple systems atrophy; ET = Essential tremor; BTX = Botulinum toxin injections; TS = Tourette Syndrome

Discussion

Our series of 15 patients who satisfy the diagnostic criteria for camptocormia, marked flexion of the thoracolumbar spine most prominent on standing and walking and relieved in supine position, draws attention to the a broad spectrum of musculoskeletal and neurological etiologies of this disorder. Although PD accounted for 11 of our 15 (73.3%) patients, this may represent a selection bias as the patients were referred to our Parkinson's Disease Center and Movement Disorders Clinic. Camptocormia may represent one end of the spectrum of PD-related abnormal postures ranging from striatal hand or foot deformity on one end of the spectrum to scoliosis and camptocormia on the other end of the spectrum [9]. Indeed, camptocormia may be the presenting or most disabling feature of PD and other parkinsonian disorders [10-12]. While flexion of the neck (rather than camptocormia) is typically present in patients with multiple system atrophy [13], neck extension is more characteristic of progressive supranuclear palsy. Djaldetti at al. [10] first reported camptocormia in eight patients with idiopathic PD and suggested that "bent spine" might represent an action dystonia of the trunk, precipitated or exacerbated in standing position or with walking, resulting from striatal, especially putaminal, damage. Later, Holler et al [11] added two cases of camptocormia in PD and again suggested that it may be a form of dystonia. Dystonia without parkinsonism, present in our cases (4/11) has been also described as a possible cause of camptocormia [14]. It is interesting to note that five patients in our study had a history of injury or surgery to their back prior to onset of their symptoms suggesting the possibility that their camptocormia was peripherally induced [15,16]. Four patients in our study had palpable contraction of the rectus abdominus muscles and improved with BTX. In addition to movement disorders (PD, dystonia, Tourette's syndrome), camptocormia may be associated with a variety of other etiologies [Table 3]. The study of cases of secondary camptocormia associated with structural lesions in the brain or spinal cord may provide important insights into the pathogenesis of this disorder. For example, the observation that camptocormia may be associated with lenticular lesions suggests that the striatum and pallidum play an important role in the maintenance of axial posture [17].

Conclusion

Despite earlier reports attributing the disorder to hysteria, malingering or other psychogenic causes [18-20] and despite a relatively high frequency of psychogenic disorders in our clinic [21], none of our patients with camptocormia were thought to be of psychogenic origin. Based on a thorough review of the literature we classified our and reported cases into nine categories based on their etiology [Table 4].

Table 4. Classification of Camptocormia

1. Parkinsonism:	a. Idiopathic parkinson's disease [10,12]
	b. Multiple system atrophy
	c. Autosomal recessive juvenile parkinsonism (parkin mutation) [22]
	d. Post encephalitic parkinsonism [23,24]
	e. Atypical parkinsonism
2. Dystonia	
Primary dystonia [14]	
Secondary dystonia	a. Associated with parkinsonism [25]
	b. Associated with structural lesions in the brain or spinal cord [26]
3. Spine abnormalities	
4. Brain injury	
5. Stroke [17]	
6. Neuromuscular:	a. Focal myopathy [12,27,28]
	b. Amyotrophic lateral sclerosis [29,30]
	c. Inclusion body myositis [31]
	d. Nemaline myopathy
7. Psychogenic [18,20,32]	
8. Miscellaneous:	a. Drug Induced [33]
	b. Paraneoplastic [34]
	c. Tourette's syndrome (Azher et al, 2004)
9. Idiopathic	

References

(Please refer to the Reference Handout.)

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