Camptocormia

Etiology, diagnosis, and treatment response

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Abstract

Background

We sought to determine the etiologies, diagnostic testing, and management of a retrospective cohort of patients with camptocormia evaluated at a single center.

Methods

We reviewed medical records of all adult patients evaluated at Mayo Clinic Rochester with a diagnosis of camptocormia from 2000 to 2014. Demographic and clinical data were abstracted and analyzed.

Results

There were 276 patients (58.0% male), with mean age at presentation of 68.6 (± 12.7) years. An etiology was identified in 98.2%. The most common etiologies were idiopathic Parkinson disease (22.5%), idiopathic axial myopathy (14.1%), and degenerative joint disease without fixed deformity (13.0%). We also identified several rare causes of camptocormia. Investigations included spine imaging, needle and surface EMG, and muscle biopsy. Most patients received physical therapy and orthotic support with limited benefit. Limited improvement of camptocormia was seen where a treatable etiology was identified.

Conclusions

An etiology can be identified in almost all cases of camptocormia. Most cases are due to 3 common disorders: Parkinson disease, axial myopathy, and degenerative joint disease. A diagnostic and treatment algorithm is proposed.

Camptocormia is derived from the Greek words “kamptos,” meaning “bent forward,” and “kormos,” which means “torso.” There is forward flexion of the spine in the upright position that disappears in the supine position, without fixed deformity. It was first described as “bent spine” by Brodi¹ in 1818. The term camptocormie was first used by Rosanoff² in 1915, to describe soldiers with a forward bent posture, attributed to conversion disorder. During the 1940s–1960s, camptocormia was reported in soldiers and patients with psychiatric disease.^{3–5} However, by the 1990s, physicians recognized camptocormia as a manifestation of neurologic and musculoskeletal disorders.⁶ Camptocormia has been described in case reports and case series in association with parkinsonian disorders, myopathies, dystonia, and infrequently functional neurologic disorders.^{7–13} One case series of 63 patients determined paraspinal myopathy as the most common etiology in 64%, and distinguished this from spinal osteoarthritis with spine imaging and muscle biopsy.¹⁴ There is no consensus on the method of quantifying camptocormia, but a forward flexion of greater than 45° with resolution on assuming a supine posture is considered diagnostic. Suggested measurement

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techniques include goniometric analysis and Cobb angle.¹⁵ Despite growing evidence, the most recent version of the ICD-10 puts camptocormia in the same category as conversion disorder.

In this study, we evaluate a large cohort of patients with camptocormia in a tertiary medical center, and present an updated range of etiologies, diagnostic workup, and management.

Methods

We retrospectively identified all patients with a diagnosis of camptocormia at Mayo Clinic Rochester between 2000 and 2014. Search keywords were “camptocormia” and “bent spine syndrome.” We included patients age 18 or older, who had $\geq 45^\circ$ of forward spinal flexion. Patients with minimally stooped posture, lateral flexion, congenital spine deformity, fixed spinal flexion deformity that did not resolve in the supine position, or lack of a definitive diagnosis of camptocormia were excluded. Degree of camptocormia was not measured objectively in all cases and was based on descriptive documentation by the physician. The initial search yielded 412 patients. Of these, 136 patients were excluded, leaving 276 patients included in the final cohort. All patients were evaluated by a spine, orthopedic, neuromuscular, or movement disorders specialist at our institution.

Demographic and clinical data were extracted. Recorded variables included age, sex, etiology, or condition associated with camptocormia as determined by the evaluating physician, diagnostic studies performed (needle EMG, brain and spine MRI, multichannel surface EMG, muscle biopsy), and treatment modalities. Diagnosis was assigned based on impression of the evaluating physician in conjunction with the available diagnostic data. Elevated creatine kinase (CK) was defined as any value above the set reference normal range for the Mayo Medical Laboratories’ photometric assay: 52–336 u/L for adult men and 38–176 u/L for adult women. Data were recorded in a secure Redcap database. Descriptive statistics were calculated using JMP software.

Standard protocol approvals, registration, and patient consent

This study was approved by the Mayo Clinic internal review board/ethics committee. Written informed consent was waived due to the retrospective nature of the study.

Data availability

All data associated with this study are reported in this article. No data will be deposited.

Results

Of 276 patients with camptocormia, 58% were male, with a mean age at presentation of 68.6 (± 12.7) years. Their demographic, clinical, and diagnostic workup features are summarized in table 1.

Etiologies

Table 2 lists the etiologies of camptocormia in these 276 patients. Certain conditions were considered causative, such as myopathy and dystonia, whereas others were thought to be associated with camptocormia by the evaluating physicians. These are discussed below.

CNS and neurodegenerative disorders

Camptocormia was associated with a number of neurodegenerative disorders. The most common was idiopathic Parkinson disease (PD) in 22.5% patients, followed by truncal dystonia (7.3%), multiple system atrophy (5.8%), dementia with Lewy bodies (3.3%), frontotemporal lobar degeneration (1.1%), progressive supranuclear palsy, and Alzheimer disease (0.7% each). Dystonia may occur in isolation without prominent degeneration or as part of a neurodegenerative process. These patients were diagnosed clinically by a movement disorders or behavioral neurology specialist at our center. PD diagnosis was based on clinical features, levodopa responsiveness, and absence of red flags for atypical parkinsonism, and atypical parkinsonism if patients did not meet these criteria.

Neuromuscular disorders

These patients were younger than the other subgroups, with a mean age at diagnosis of 50 years. The most common etiology was idiopathic axial myopathy in 14.1% ($n = 39$). These cases had an isolated or predominantly paraspinal myopathic process, with myopathic features on muscle biopsy or paraspinal EMG. MRI of the spine in revealed paraspinal muscle atrophy and fatty infiltration of varying degrees. Degenerative joint disease in these cases was mild. A muscle biopsy was performed in 22 of these 39 patients at the following locations: thoracic paraspinal ($n = 4$), gluteus maximus ($n = 5$), splenius capitis ($n = 3$), trapezius ($n = 3$), quadriceps ($n = 3$), deltoid ($n = 2$), cervical paraspinal ($n = 1$), and vastus medialis ($n = 1$). The findings were those of nonspecific myopathy, such as muscle fiber splitting, fatty infiltration, and atrophy. Other myopathies and muscular dystrophies associated with camptocormia often presented

Table 1 Clinical features and diagnostic workup of camptocormia patients (n = 276)

	N (%) or mean $\pm$ SD
Male sex	161 (58)
Mean age at diagnosis, y	68.6 $\pm$ 12.7
Family history of camptocormia	22 (7.9)
Concurrent head drop	40 (14.5)
Follow-up available	115 (41.7)
EMG performed	207 (75)
Normal	38 (18.4)
Neuropathic findings	66 (32)
Myopathic findings	98 (47.3)
Neuromuscular junction transmission defect	5 (2.4)
Movement disorders laboratory study	25 (9.06)
Normal	12 (48)
Dystonia	8 (32)
Inconclusive	5 (20)
Muscle biopsy performed	61 (22.1)
Nonspecific myopathic findings	31 (50.8)
Inflammatory findings	7 (11.5)
Mitochondrial myopathy	3 (4.9)
Fatty infiltration	2 (3.8)
Denervation atrophy	16 (26.2)
Elevated creatine kinase^a	39 (14.1)
MRI brain	137 (49.6)
Leukoaraiosis	55 (40.1)
Cerebral atrophy	10 (73)
Leukoaraiosis and cerebral atrophy	37 (27)
Chronic basal ganglia lacunar strokes	4 (2.9)
Pontine/midbrain atrophy	2 (1.5)
Normal	29 (12.2)
MRI cervical/thoracic spine	72 (26.1)
Degenerative joint disease	59 (81.9)
Paraspinal muscle atrophy	7 (9.7)
Narrow spinal canal	1 (1.4)
Syrinx	2 (2.7)
Myelitis	1 (1.4)
Normal	1 (1.4)
MRI lumbosacral spine	187 (67.8)
Degenerative joint pathology	145 (77.5)

Table 1 Clinical features and diagnostic workup of camptocormia patients (n = 276) (*Continued*)

	N (%) or mean $\pm$ SD
Compression fractures	16 (8.6)
Paraspinal muscle atrophy	15 (8.02)
Normal	11 (5.88)

^a Creatine kinase was considered elevated for any value above laboratory reference range.

with more widespread muscle weakness. These included facioscapulohumeral muscular dystrophy (2.5%), inflammatory myopathies (2.1%), inclusion body myositis (1.4%), mitochondrial myopathy (1.1%), myotonic dystrophy (1.1%), dysferlinopathy (0.7%), and rarely nemaline myopathy and central core myopathy, with 1 case of each (0.3%). Amyotrophic lateral sclerosis was diagnosed in 9.4%, myasthenia gravis in 1.8%. Patients undergoing treatment of myasthenia gravis and inflammatory myopathy had partial improvement. All patients were seen by a neuromuscular specialist at our center.

Musculoskeletal disorders

Camptocormia was attributed to severe degenerative joint disease in 13.0% (n = 36). These were distinguished from cases of axial myopathy based on predominance of severe degenerative joint disease and spondylosis in the cervical/thoracic/lumbar spine. These patients had a predominance of neurogenic changes on EMG and denervation atrophy on muscle biopsy as shown in table 3. Most of these patients were evaluated by an orthopedic or spine specialist.

Immune-mediated disorders

There were 7 cases (2.5%) where an autoimmune antibody was identified in the absence of other causes of camptocormia. None was associated with an underlying malignancy. GAD-65 antibody was elevated in 2 patients and the clinical phenotype was consistent with stiff-person syndrome. There was partial improvement of the camptocormia with benzodiazepines and immunotherapy. Two patients had voltage-gated potassium channel (VGKC) antibodies, which were not speciated into contactin associated protein-2 (Caspr-2) or leucine-rich glioma inactivated protein-1 (LGI-1). One patient had an elevated N-type calcium channel antibody. Steroid therapy was trialed unsuccessfully in the cases with elevated VGKC antibody, raising suspicion that these antibodies may be unrelated to the clinical phenomenon.

Miscellaneous/rare

Drug-induced causes were uncommon and included olanzapine 0.7%, levodopa-induced dystonia 0.7%, and steroid related myopathy 0.4%. Improvement was noted after discontinuation of the drug. Other infrequent causes are listed in

Table 2 Etiologies of camptocormia

Diagnostic categories	Specific diagnoses	Patients, n (%)
Neuromuscular and musculoskeletal disorders (48.2%)	Degenerative joint disease	36 (13.0)
	Axial myopathy	39 (14.10)
	Amyotrophic lateral sclerosis	26 (9.42)
	Facioscapulohumeral muscular dystrophy	7 (2.54)
	Inflammatory myopathies	6 (2.17)
	Myasthenia gravis	5 (1.80)
	Inclusion body myositis	4 (1.45)
	Mitochondrial myopathies	3 (1.09)
	Myotonic dystrophy	3 (1.09)
	Dysferlinopathy	2 (0.72)
	Nemaline myopathy	1 (0.36)
	Central core myopathy	1 (0.36)
CNS and neurodegenerative diseases (41.3%)	Idiopathic Parkinson disease	62 (22.50)
	Dystonia	20 (7.25)
	Multiple system atrophy	16 (5.80)
	Lewy body disease	9 (3.26)
	Frontotemporal dementia	3 (1.09)
	Progressive supranuclear palsy	2 (0.72)
	Alzheimer disease	2 (0.72)
Miscellaneous/rare (6.2%)	Conversion disorder	2 (0.72)
	Myoclonus dystonia syndrome	3 (1.09)
	Radiation myopathy	2 (0.72)
	Infectious myelitis	1 (0.36)
	Compressive cervical myelopathy	2 (0.72)
	Postpolio syndrome	2 (0.72)
	Idiopathic/undetermined	5 (1.80)
Immune-mediated (2.5%)	Voltage-gated potassium channel antibody	2 (0.72)
	N-type calcium channel antibody	1 (0.36)
	Anti-CRMP antibody	1 (0.36)
	Ach-R antibody	1 (0.36)
	GAD-65 antibody	2 (0.72)
Drug-induced (1.8%)	Olanzapine	2 (0.72)
	Steroids	1 (0.36)
	Levodopa	2 (0.72)

All causes identified to be associated with camptocormia in this cohort of 276 patients.

table 2. In 5 cases (1.8%), a cause or associated factor was not discovered.

Other clinical features

An associated head drop was seen in 14.5%. This was most commonly seen in patients with axial myopathy (n = 10) or PD (n = 6). Less common causes were multiple system atrophy (n = 4), dystonia (n = 4), degenerative joint disease (n = 3), idiopathic axial myopathy (n = 3), amyotrophic lateral sclerosis (n = 2), myotonic dystrophy (n = 2), Alzheimer disease (n = 1), inflammatory myopathy (n = 1), inclusion body myositis (n = 1), myasthenia gravis (n = 1), facioscapulohumeral muscular dystrophy (n = 1), and Lewy body disease (n = 1). Twenty-two patients (7.9%) had a family history of camptocormia, only observed in patients with facioscapulohumeral muscular dystrophy, myotonic dystrophy, or idiopathic axial myopathy.

Diagnostic evaluation

All patients underwent supplementary testing in addition to clinical examination. The findings are summarized in table 1, whereas table 3 compares findings between the most common subgroups.

Three-quarters of cases (n = 207) underwent EMG. Of these, 18.4% were normal, 47.3% revealed predominantly myopathic findings, 32% had predominantly neurogenic findings, and 2.4% patients had a neuromuscular transmission defect. Of the 39 patients with axial myopathy, 36 had myopathic findings on EMG and 3 had neurogenic findings. Of 36 patients with degenerative joint disease, 32 underwent EMG, which showed predominantly neurogenic changes in 15, myopathic changes in 11, and 6 were normal.

A movement disorders laboratory study with multichannel surface EMG was performed for 9.1% of patients with suspected dystonia. One-third confirmed dystonia, half of the studies were normal, and the rest were inconclusive.

Muscle biopsy was performed in 61 patients with muscle weakness on examination. The biopsy site was selected based on affected muscle by EMG, and rarely by MRI. Biopsy sites included the following: thoracic paraspinals (n = 13), gluteus maximus (n = 9), biceps (n = 6), splenius capitis (n = 6), trapezius/vastus/quadriceps (n = 5 each), triceps (n = 4), deltoid (n = 3), pectoralis (n = 2), and 1 case each of gastrocnemius, cervical, and lumbar paraspinals. Of the 61 biopsies, 2 were normal. The other 59 were abnormal, and the most common findings included nondiagnostic myopathic features with fatty infiltration (n = 33), inflammatory findings (n = 7), mitochondrial myopathy (n = 3), and denervation atrophy (n = 16). Of the 7 patients with inflammatory findings, 6 were eventually diagnosed as inflammatory myopathy. Muscle biopsy was instrumental in diagnosis of a few cases of camptocormia due to central core myopathy, nemaline rod myopathy, and inflammatory myopathy.

Table 3 Most common etiologies of camptocormia, clinical features, relevant diagnostic tests, and response to treatment are summarized

Most common etiologies	Parkinson disease (n = 62)	Axial myopathy, idiopathic (n = 39)	Degenerative joint disease (n = 36)	Neuromuscular disorders (n = 58)
Male, %	74	48.7	48.8	55.0
Mean age at diagnosis, y	68	70	72	50
Diagnostic tests				
EMG	11 Neuropathic changes	All patients had myopathic findings on EMG	15 Patients had neuropathic changes on EMG	Aided specific diagnosis: myasthenia gravis, motor neuron disease, myopathy
	9 Myopathic changes	10 Patients had elevated CPK	11 Patients had myopathic changes on EMG	
	12 Normal	Paraspinal muscle atrophy or spondylosis	5 Had a normal EMG	
MRI brain	Nonspecific findings: leukoaraiosis and atrophy	Noninflammatory myopathy in all cases	Multilevel spondylosis, prior surgeries	Nonspecific
MRI spine	Nonspecific		5 Had muscle biopsy, of which 3 showed denervation atrophy	
Movement laboratory study	6 Movement laboratory studies were done, 5 of which were normal and 1 showed dystonia			
Muscle biopsy	Noninflammatory myopathy			Diagnostic in specific cases of suspected inflammatory myopathy or muscular dystrophy
Treatment responsiveness	Mild variable response to levodopa; poor response to DBS	Mild benefit from PT and orthotic; correction with surgical fixation	Mild benefit with PT and orthotic devices	Variable; based on underlying pathology

Abbreviations: DBS = deep brain stimulation; PT = physical therapy.

Mild CK elevation was found in 39 patients, 31 of whom had myopathic features on muscle biopsy but were not diagnostic of specific disease process. CK elevation was not independently diagnostic.

Neuroimaging included MRI of the brain, cervical, thoracic, and lumbar spine. MRI brain was performed in 49.6% and was normal for age in most cases. The most common findings on spine MRI are summarized in table 1. A syrinx noted in the cervicothoracic spinal cord in 2 patients was deemed to be asymptomatic.

Treatment

The most common causes of camptocormia, test findings, and treatment modalities are summarized in table 4.

Three-quarters of patients were treated with physical therapy, with limited benefit. Almost one-third (30.8%) of patients underwent a trial of levodopa therapy, for idiopathic PD (n = 48), multiple system atrophy (n = 10), Lewy body disease (n = 8), dystonia (n = 15), and other unclear diagnoses, which were eventually found to be non-levodopa-responsive pathologies, such as myopathy (n = 4). In almost

50% of cases associated with idiopathic PD, mild to moderate subjective improvement was documented. Anterior or posterior spinal fixation surgery was performed for 15 patients (5.4%), including degenerative joint disease (n = 7), PD (n = 5), and axial myopathy (n = 3). There was improvement in posture noted in all patients; however, long-term follow-up was limited.

Botulinum toxin was administered in 10 cases with presumed dystonia: 4 had PD, 5 had surface EMG proven dystonic camptocormia, and 1 was subsequently not found to have dystonia on surface EMG. Botulinum toxin produced partial benefit in 6 of the 10. Muscles injected included iliopsoas, rectus abdominis, and transverse abdominis.

Deep brain stimulation surgery was performed for camptocormia in 4 patients with PD. Two patients underwent bilateral globus pallidus interna implantation with no improvement, and 2 underwent bilateral subthalamic nucleus implantation, one of whom had mild benefit.

Patients with myasthenia gravis (n = 5) and inflammatory myopathies (n = 6) were administered steroids and

PD, axial myopathy, and spine degenerative joint disease were the most common etiologies underlying camptocormia

immunosuppressants, with at least partial subjective improvement in almost all patients. However, an objective measure of improvement and the duration of the therapeutic response was poorly documented.

Follow-up at Mayo Clinic was available for 115 (41.7%) of the patients for a median of 8 months (interquartile range 6–12). However, due to incomplete documentation, assessment of response to treatment was limited.

Discussion

In this study, we describe a large cohort of camptocormia patients, and report several novel findings. An underlying etiology can be discovered in almost all cases of camptocormia; hence a thorough evaluation guided by history and examination is warranted. PD, axial myopathy, and spine degenerative joint disease were the most common etiologies underlying camptocormia. There is an opportunity of improving camptocormia when the underlying etiology identified is amenable to treatment. If not, treatment appears to be of limited efficacy.

A wide range of disease processes associated with camptocormia is reported in the literature. These include PD,^{16,17} myopathy and muscular dystrophy,^{18–20} multiple system atrophy,⁹ and other neurodegenerative disorders like Alzheimer disease and dementia with Lewy bodies.^{11,21} Relatively rare cases are associated with drug side effect,²² radiation-induced myopathy,²³ tetanus,²⁴ and calpainopathy.¹⁸ Our study replicated prior reports and identified several etiologies not previously reported: progressive supranuclear palsy, central core myopathy, dystonia-myoelonus syndrome, and postpolio syndrome. We found the most common causes of camptocormia to be idiopathic axial myopathy, PD, and degenerative spine disease. Other large case series in the literature examine the prevalence and causes of camptocormia in subspecialty neurology clinics where this presentation is mostly seen, i.e., movement disorders and neuromuscular clinics. A series of 63 cases (73% female) reported the most common etiology was paraspinal myopathy.¹⁴ Of 52 patients presenting with camptocormia at the neuromuscular clinic at our institution, a specific diagnosis was reached in 54%, the most common causes being facioscapulohumeral muscular dystrophy and inclusion body myositis.²⁵ In another study of 275 patients with PD, 6.9% had camptocormia. This was associated with more severe disease, longer disease duration, dementia, and higher levodopa doses.¹⁶ The pathophysiology of camptocormia in PD is not known, but speculated mechanisms include myopathy, extra-pyramidal system dysfunction, or dystonia.¹⁷ The wide range of possible etiologies make it essential for the investigating physician to carry out a thorough evaluation and objective measurement of forward flexion when possible.

It is important to distinguish degenerative joint disease (without fixed deformity) from isolated idiopathic axial

Table 4 Treatment modalities

Treatment modality	Patients receiving therapy, n (%)	Most common targeted etiology	Response to treatment
Physical therapy	199 (72.1)	Various causes	Mild/none
Levodopa	85 (30.8)	PD	Mild/variable
Surgical spine fixation	15 (5.4)	PD/degenerative spine disease	Moderate/good
Prednisone	14 (5.1)	Myopathy/autoimmune	Mild/none
Immunosuppression	13 (4.7)	Inflammatory myopathy	Mild/none
Botulinum toxin	10 (3.6)	Dystonia	Mild/moderate
IVIg for immunosuppression	9 (3.3)	MG	Moderate
Subthalamic nucleus deep brain stimulation	2 (0.7)	PD	Mild improvement in 1 case
Pallidal deep brain stimulation	2 (0.7)	PD	None
Spinal stimulation/lidocaine injection/ECT	None	—	—

Abbreviations: ECT = electroconvulsive therapy; IVIg = IV immunoglobulin; PD = Parkinson disease. Commonly used therapies in our cohort and response based on patient and physician judgement.

myopathy. In our cohort, patients were assigned the diagnosis of degenerative spine disease based on CT and MRI spine findings of severe osteoarthritis, and the opinion of the evaluating orthopedic or spine specialist. The EMG revealed primarily neurogenic changes; however, some cases had mixed findings of both neurogenic and myopathic features. Differences between the pattern of paraspinal muscle fiber degeneration seen in spinal osteoarthritis vs paraspinal myopathy have been reported previously.^{6,17} The exact pathogenesis is unknown, and potentially some degree of overlap exists between the 2 conditions. A careful evaluation and exclusion of other treatable pathologies is strongly recommended in these cases.

Diagnostic testing should be guided by history and examination, and a diagnostic algorithm is proposed in the

figure. Commonly performed tests include MRI or CT imaging of the spine, needle EMG, and CK. Individualized tests include muscle biopsy, surface EMG, and specific serology for genetic and autoimmune myopathies. Spine imaging commonly reveals nonspecific muscle atrophy and fatty infiltration and may vary based on chronicity of camptocormia.²⁶ As shown by prior series,^{14,25} our cohort confirms that CK elevation in these patients is usually mild and nondiagnostic. A specific diagnosis is most likely to be achieved where a targeted test is performed based on high clinical index of suspicion; for example, gene testing for facioscapulohumeral muscular dystrophy, muscle biopsy for inflammatory myopathy and myositis, serology for autoimmune myopathy, or surface EMG to confirm dystonia, which could lead to efficacious therapy.

Figure Suggested algorithm to evaluate camptocormia based on clinical presentation

Treatment for camptocormia appeared ineffective in most cases. However, when therapy could be targeted to a specific treatable disease, a better response was seen.

There are cases of dystonia successfully treated with pallidal and subthalamic deep brain stimulation reported in the literature. Srivannichapoom and Hallett¹⁷ reviewed 56 cases of camptocormia in PD, 51 of which underwent subthalamic stimulation and 5 pallidal stimulation. Of these, 61% had improvement with stimulation. However, most cases have limited follow-up and modest improvement. Complete resolution of camptocormia, as described in case reports,^{27,28} was not replicated in our patients with PD with camptocormia. This variation may reflect complex pathophysiology and an unclear neural substrate underlying camptocormia, which most likely represents a heterogeneous condition. Spinal stimulation may provide short-lived benefit,²⁹ but was not performed in our cohort. Botulinum toxin can be efficacious in dystonic camptocormia. Prior reports have targeted the iliopsoas and external oblique muscles with some benefit.³⁰⁻³² Levodopa leads to unpredictable partial benefit in some patients with PD. The mechanism of this response is not well-understood and is particularly interesting in view of higher doses of levodopa being associated with camptocormia; albeit this may be a surrogate of more severe disease.⁶ In our cohort, there was moderate response to immunotherapy in inflammatory myopathy and myasthenia gravis. The most commonly used methods of physical therapy and extension bracing have limited benefit in the short term and may serve to slow down paraspinal muscle deconditioning.³³ Lidocaine injections of the external oblique muscles of patients with PD were reportedly efficacious in a few cases,³⁴ but were not used in our patient population. Overall, response to treatment in camptocormia is partial at best, with higher likelihood of improvement where therapy can directly target the underlying etiology. Long-term follow-up along with objective measurement of camptocormia will increase our knowledge of this heterogeneous condition.

This study has several strengths. We describe the largest cohort of camptocormia patients evaluated at a single center, without limiting our selection to a subspecialty clinic; hence all causes of camptocormia were captured. The broad array of diagnostic and treatment options noted in this patient cohort allowed us to put forward a diagnostic and treatment algorithm. We highlight important clinical points to be considered when evaluating camptocormia and provide useful groundwork for future studies. A limitation was the lack of objective measurement of the degree of forward spinal flexion. This made it difficult to accurately capture severity of camptocormia and response to treatment. Since this was a retrospective study, diagnostic workup performed for each patient varied based on the evaluating physician. Diagnosis assigned was based on best physician judgement at the end of diagnostic workup, and due to the retrospective nature, some diagnoses may have evolved and changed over time. Being

TAKE-HOME POINTS

- Camptocormia is a 45° or greater forward flexion of the spine caused by a variety of neurologic and musculoskeletal disorders.
- Thorough investigation should be guided by history and physical as a diagnosis can be made in most cases.
- Treatment options are limited. Better prognosis may be expected when a treatable etiology is discovered.
- Objective measurement of the extent of spinal flexion is important for diagnosing and assessing treatment response.

a tertiary referral center, several patients elected to follow-up locally after completion of diagnostic evaluation, hence data on response to treatment and long-term course of the condition were limited.

Camptocormia is a progressively disabling condition. Pathophysiology is diverse and incompletely understood. The underlying etiology should be sought in all cases, as it may lead to effective treatment in some patients. Objective documentation of the extent of camptocormia will help quantify deformity and response to treatment. We propose a diagnostic and treatment approach based on a large single-center cohort of camptocormia.

Author contributions

F. Ali: data collection, data analysis, manuscript preparation. J.Y. Matsumoto: manuscript editing and supervision. A. Hassan: manuscript editing and supervision.

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Disclosure

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References

1. Brodie BC. Pathological and Surgical Observations on the Diseases of the Joints. London: Longman; 1818:276.
2. Rosanoff S. La camptocormie. *Rev Neurol* 1915;22:937-939.
3. Sandler SA. Camptocormia; a functional condition of the back in neurotic soldiers. *Arch Neurol Psychiatry* 1946;55:158-160.

4. Sandler SA. Camptocormia, or the functional bent back. *Psychosom Med* 1947;9:197–204.
5. Sutro CJ, Hulbert B. Hysterical flexion deformity of the vertebral column; camptocormia. *Bull US Army Med Dep* 1946;5:570–574.
6. Laroche M, Delisle MB, Aziza R, Lagarrigue J, Mazieres B. Is camptocormia a primary muscular disease? *Spine* 1995;20:1011–1016.
7. Friedman JH. Episodic camptocormia in PD. *Mov Disord* 2001;16:1201.
8. Van Gerpen JA. Camptocormia secondary to early amyotrophic lateral sclerosis. *Mov Disord* 2001;16:358–360.
9. Skidmore F, Mikolenko I, Weiss H, Weiner W. Camptocormia in a patient with multiple system atrophy. *Mov Disord* 2005;20:1063–1064.
10. Gomez-Puerta JA, Peris P, Grau JM, Martinez MA, Guanabens N. Camptocormia as a clinical manifestation of mitochondrial myopathy. *Clin Rheumatol* 2007;26:1017–1019.
11. Brucki S, Nitrini R. Camptocormia in Alzheimer's disease: an association? *Mov Disord* 2008;23:156–157.
12. Terashima M, Kataoka H, Sugie K, Horikawa H, Ueno S. Coexistence of chronic inflammatory demyelinating polyneuropathy and camptocormia. *J Neurol Neurosurg Psychiatry* 2009;80:1296–1297.
13. Jordan B, Eger K, Koesling S, Zierz S. Camptocormia phenotype of FSHD: a clinical and MRI study on six patients. *J Neurol* 2011;258:866–873.
14. Laroche M, Cintas P. Bent spine syndrome (camptocormia): a retrospective study of 63 patients. *Joint Bone Spine* 2010;77:593–596.
15. Barrett E, McCreesh K, Lewis J. Reliability and Validity of Non-Radiographic Methods of Thoracic Kyphosis Measurement: A Systematic Review. *Manual Therapy*, vol 19. Philadelphia: Elsevier; 2014:10–17.
16. Tiple D, Fabbri G, Colosimo C, et al. Camptocormia in Parkinson disease: an epidemiological and clinical study. *J Neurol Neurosurg Psychiatry* 2009;80:145–148.
17. Srivannitchapoom P, Hallett M. Camptocormia in Parkinson's disease: definition, epidemiology, pathogenesis and treatment modalities. *J Neurol Neurosurg Psychiatry* 2016;87:75–85.
18. Liewluck T, Goodman BP. Late-onset axial myopathy and camptocormia in a calpainopathy carrier. *J Clin Neuromuscul Dis* 2012;13:209–213.
19. Zenone T, Streichenberger N, Puget M. Camptocormia as a clinical manifestation of polymyositis/systemic sclerosis overlap myositis associated with anti-Ku. *Rheumatol Int* 2013;33:2411–2415.
20. Goodman BP, Liewluck T, Crum BA, Spinner RJ. Camptocormia due to inclusion body myositis. *J Clin Neuromuscul Dis* 2012;14:78–81.
21. Gavrylova N, Limousin N, Belin J, de Toffol B, Praline J. Camptocormia as presenting sign in dementia with Lewy bodies. *Clin Neurol Neurosurg* 2013;115:2397–2398.
22. Kiuru S, Iivanainen M. Camptocormia, a new side effect of sodium valproate. *Epilepsy Res* 1987;1:254–257.
23. Seidel C, Kuhnt T, Kortmann RD, Hering K. Radiation-induced camptocormia and dropped head syndrome: review and case report of radiation-induced movement disorders. *Strahlenther Onkol* 2015;191:765–770.
24. Kaji S, Shibayama H, Sato S, Fukutake T. Acute camptocormia as the presenting symptom of tetanus. *J Neurol Neurosurg Psychiatry* 2014;85:936–937.
25. Ghosh PS, Milone M. Camptocormia as presenting manifestation of a spectrum of myopathic disorders. *Muscle Nerve* 2015;52:1008–1012.
26. Margraf NG, Rohr A, Granert O, Hampel J, Drews A, Deuschl G. MRI of lumbar trunk muscles in patients with Parkinson's disease and camptocormia. *J Neurol* 2015;262:1655–1664.
27. Nandi D, Parkin S, Scott R, et al. Camptocormia treated with bilateral pallidal stimulation. *J Neurosurg* 2002;97:461–466.
28. Hellmann MA, Djaldetti R, Israel Z, Melamed E. Effect of deep brain subthalamic stimulation on camptocormia and postural abnormalities in idiopathic Parkinson's disease. *Mov Disord* 2006;21:2008–2010.
29. Arii Y, Sawada Y, Kawamura K, et al. Immediate effect of spinal magnetic stimulation on camptocormia in Parkinson's disease. *J Neurol Neurosurg Psychiatry* 2014;85:1221–1226.
30. Wijemanne S, Jimenez-Shahed J. Improvement in dystonic camptocormia following botulinum toxin injection to the external oblique muscle. *Parkinsonism Relat Disord* 2014;20:1106–1117.
31. Colosimo C, Salvatori FM. Injection of the iliopsoas muscle with botulinum toxin in camptocormia. *Mov Disord* 2009;24:316–317.
32. Azher SN, Jankovic J. Camptocormia: pathogenesis, classification, and response to therapy. *Neurology* 2005;65:355–359.
33. Ye BK, Kim HS, Kim YW. Correction of camptocormia using a cruciform anterior spinal hyperextension brace and back extensor strengthening exercise in a patient with Parkinson disease. *Ann Rehabil Med* 2015;39:128–132.
34. Furusawa Y, Mukai Y, Kawazoe T, et al. Long-term effect of repeated lidocaine injections into the external oblique for upper camptocormia in Parkinson's disease. *Parkinsonism Relat Disord* 2013;19:350–354.

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