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Presentation, Etiology, Diagnosis, and Management of Camptocormia

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Key Words
Movement disorder · Neuromuscular disease · Spinal deformity · Muscle disease · Parkinson · Dystonia

Abstract
Camptocormia (bent spine syndrome, cyphose hystérique) is an abnormality characterized by severe forward flexion of the thoracolumbar spine which typically increases during walking or standing and completely disappears in supine position. Camptocormia can be due to central nervous system diseases, such as Parkinson's disease, dystonia, multisystem atrophy, or Alzheimer's disease, due to peripheral nervous system diseases, such as primary myopathy, secondary myopathy, motor neuron disease, myasthenia, or chronic inflammatory demyelinating polyneuropathy, due to side effects of drug treatment, due to disc herniation, arthritis or spinal trauma, or due to paraneoplasia. Only rarely may camptocormia be attributable to psychiatric disease. The diagnosis is based on clinical findings, imaging of the cerebrum or spine, needle electromyography, or muscle biopsy. Treatment options are limited and frequently futile and rely on conservative measures, such as psychotherapy, physiotherapy, use of orthoses, drugs, injection of botulinum toxin, withdrawal of causative drugs, electroconvulsive therapy, or invasive measures, such as surgical correction or deep brain stimulation. The outcome is generally fair. Some patients profit from therapy whereas others do not respond to treatment and become progressively immobile.

Introduction

Camptocormia, also known as bent spine syndrome, or cyphose hystérique, is a major disabling, non-fluctuating, acquired postural abnormality due to various different causes, characterized by an involuntary forward-flexed posture of the thoracolumbar spine [1, 2]. The term camptocormia derives from the Greek words 'kamptos' (to bend) and 'cormos' (trunk) [3]. The abnormal posture leads to lumbar kyphosis and is increased or only visible when the patient stands up or walks [3, 4]. Camptocormia is most frequently due to disorders of the striated muscles of the spinal column, due to central nervous system (CNS) disorders, or due to some rare conditions (table 1). Camptocormia has to be delineated from Pisa syndrome (pleurothotonus) characterized by a tonic flexion of trunk and head to one side along with scoliosis. This review aims to summarize and discuss recent findings and future perspectives concerning the clinical presentation, etiology, diagnosis, and treatment of camptocormia.

History

Camptocormia was first documented in the 17th century by the Spanish painter Francisco de Zurbaran. 'Functionally bent spine' was first described by Brodie in 1818 [5]. The next description under the term 'functionally bent back' was published by the same author 19 years later [6, 7]. The term camptocormia was first coined in 1914

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Table 1. Conditions associated with camptocormia

Conditions	References
CNS disorders	
Organic	
Parkinson's disease	19, 27, 33, 60
Dystonias	2, 14, 28, 38
Abdominal segmental dystonia	37
Multisystem atrophy	34, 35, 65
Alzheimer's disease	36
Basal ganglia disorders	14
Essential tremor	14
Viljuisk encephalomyelitis	26
Psychiatric	
Gilles de la Tourette syndrome	28
Conversion disorder	41
Psychogenic	65
Oppositional defiant disorder	41
Neurosis	14
PNS disorders	
Primary myopathies	
Myotonic dystrophy type 1	16, 21
Myotonic dystrophy type 2	39
Axial myopathy	7, 84
Dysferlinopathy	24
Nemaline myopathy	20, 26
Mitochondrial myopathy	40, 47
Congenital myopathy	40
XMPMA	85
Secondary myopathies	
Hypothyroid myopathy	43
Isolated thoracic extensor myopathy	48
Inflammatory myopathy (PM)	18, 40, 44
Dermatomyositis	40, 44
Chronic axial myositis	41
Focal myositis[1]	45, 46
Inclusion body myositis	3, 26
Myasthenia gravis	47
Myopathy with nemaline rods	20
Facioscapulohumeral muscular dystrophy	26
Neurogenic	
Amyotrophic lateral sclerosis	26, 48, 50, 75
CIDP	51
Drugs	
Olanzapine	57
Donepezil	59
Valproate	60, 86
Systemic steroids	27
Varia	
Lumbar disc herniation	52
Dystonia from neck trauma	53
Arthritis	61
Trauma	61
Malignancies	54
Idiopathic	26

PM = Polymyositis; CIDP = chronic inflammatory demyelinating polyneuropathy; XMPMA = X-linked myopathy with postural muscle atrophy. [1] Camptocormia in Parkinson's disease and multisystem atrophy.

by the French neurologists Souques and Rosanoff-Saloff who described the abnormality in World War 1 soldiers traumatized by shell shocks [8, 9]. The first described soldier was wounded by a bullet which entered along the axillary border of the scapula and emerged near the spine, resulting in a trunk being bent almost at a right angle [9]. To the 'poilu' (nickname of the French WW1 soldier) this condition was known as 'cintrage' (aching). The man was 'cured' by application of plaster corsets [9]. In 1919, two further cases were described by Roussy and Lhermitte [10]. The first was an infantryman, blown into the air by a bursting shell, who experienced violent pain and remained stooped to the right after regaining consciousness [10]. He was also cured by a plaster corset. The second case was a 'chasseur' who experienced respiratory distress, mutism and camptocormia after being buried in an explosion. He recovered after a single séance of electrical treatment [10]. Though these cases were classified as hysterical, the traumatic injury alone may sufficiently explain camptocormia [11]. The association between camptocormia and Parkinson's disease was first described by Djaldetti et al. [12] in 1999. Camptocormia due to a genetic disease was first described in a patient with myotonic dystrophy type 2 carrying a ZNF9 gene mutation [13]. The first patient with multisystem atrophy (MSA) and camptocormia was reported by Reichel et al. [14] in 2001.

Frequency

There are few data about the frequency of camptocormia available. In a single-center epidemiological study on 275 consecutive outpatients with Parkinson's disease, the prevalence of camptocormia was 6.9% [15]. The occurrence of camptocormia in this study was related to the severity of Parkinson's disease [15].

Clinical Presentation

Camptocormia is clinically characterized by an excessive involuntary trunk flexion due to progressive weakness of the extensor vertebral muscles [16]. Camptocormia is enhanced during standing and walking [17] and relieved in recumbent or supine position [18–20]. In quite a number of cases, camptocormia is associated with lower back pain [1, 3, 7, 21–24] but in others it is painless [19]. Camptocormia may be associated with concomitant weakness of the gluteus maximus and hip and genuflexion [3]. Camptocormia is associated with dropped head

syndrome only in single cases [25, 26], such as in patients with myotonic dystrophy type 1 [16], MSA, or postencephalitic Parkinson syndrome [26]. Nearly all patients with camptocormia have spondylarthrosis, rendering it a risk factor for developing camptocormia [27]. In a study of 16 patients, mean age at onset of camptocormia was 65 years and mean age at onset of neurological abnormalities was 52 years [28]. Almost 69% of the patients had Parkinson's disease, 25% had dystonia, and one Gilles de la Tourette syndrome [28]. The family history may be positive for muscle disease in up to 50% of the cases [23].

Etiology

To explain the etiology and pathomechanism underlying camptocormia, two schools of thought have emerged. The first considers camptocormia to be a CNS disorder, resulting in focal action dystonia of the spine [27]. According to this hypothesis the CNS structure supposed to be affected is the striatum and its projections to the reticulospinal tract or the thalamus [27]. Arguments in favor of this hypothesis are the beneficial effect of deep brain stimulation on camptocormia in single cases [27] and the reduced midbrain and pons volume in patients with Parkinson's disease and camptocormia. The second hypothesis attributes camptocormia to peripheral nervous system (PNS) involvement, in particular myopathy of the antigravity muscles associated with trunk extension [27]. This hypothesis is supported by myopathic electromyographies (EMGs), hypodensities on muscle MRI, myopathic changes on muscle biopsy and occasional improvement upon steroids [27]. Camptocormia is most frequently due to organic disorders and only rarely a manifestation of a psychiatric disorder [28, 29].

Organic Disorders

CNS Disorders

The CNS disorder most frequently reported in association with camptocormia is Parkinson's disease [1, 12]. Clinical features of camptocormia in Parkinson's disease include old age, predominantly male sex, long disease duration, and early axial involvement [3]. Camptocormia usually develops after onset of Parkinson symptoms. Only in single cases camptocormia may be the initial manifestation of Parkinson's disease [30]. Camptocormia in Parkinson's disease starts to develop between the age of 60 and 70 years [3] and the latency with which camptocormia occurs after onset of Parkinson's disease is 5–10 years [1, 31]. Camptocormia is most frequently found in patients with more severe Parkinson's disease. Camptocormia in Parkinson's disease is attributed to either a CNS or PNS origin. Arguments for a CNS cause are that camptocormia occurs together with CNS disorders, that the severity of camptocormia in Parkinson's disease is associated with reduced midbrain and pons volume [32], that half of the Parkinson patients with camptocormia develop restless leg syndrome [32], that Parkinson patients with camptocormia present with enhanced muscle activity on polysomnography, suggesting affection of the central control of movements during sleep [32], that parkinsonism in patients with camptocormia is frequently associated with dystonia [28], that there may be additional, non-dopaminergic neuronal dysfunction in the basal ganglia or the brainstem in Parkinson patients with camptocormia [3], and that some patients with camptocormia respond to deep brain stimulation. Arguments for a PNS origin of camptocormia in Parkinson's disease are that axial muscles may show myopathic features on EMG or muscle biopsy [1] and that MRI of the axial muscles may show atrophy and fatty replacement of the thoracolumbar muscles [33]. CNS disorders associated with camptocormia in addition to Parkinson's disease include MSA [2, 34, 35], Alzheimer's disease [36], basal ganglia disorders [14], and abdominal segmental dystonia [37]. Because of the frequent association of camptocormia with Parkinson's disease [3, 19] or dystonias [28], some authors regard camptocormia even a segmental form of dystonia [14, 38].

PNS Disorders

PNS conditions associated with camptocormia include primary myopathies, such as myotonic dystrophy type 1 [16, 21] and type 2 [39], dysferlinopathy [24], nemaline myopathy [20], axial myopathy [7, 40, 41], or mitochondrial myopathy (table 1) [42]. Secondary myopathies with camptocormia include hypothyroid myopathy [43], dermatomyositis, polymyositis [43, 44], focal or segmental myositis [33, 45, 46], inclusion body myositis [3], or myasthenia gravis (table 1) [47]. The frequently observed myopathic abnormalities on EMG or muscle biopsy in Parkinson patients with camptocormia are attributed rather to chronic contractions of the antigravity muscles involved in compensatory trunk extension than a primary myopathy [48]. Why an overactive muscle, however, develops myopathic changes remains speculative. Because of the frequent association of camptocormia with myopathy, some authors generally regard camptocormia as a primary girdle myopathy with subclinical involvement of the pelvic and shoulder girdle muscles [49].

PNS disorders other than myopathy associated with camptocormia include amyotrophic lateral sclerosis [50], chronic inflammatory demyelinating polyneuropathy [51], or lumbar disc herniation [52]. Genes so far found mutated in patients with camptocormia are the RYR1 gene in axial myopathy [53], DMPK gene in myotonic dystrophy 1 [16, 21], ZFP9 in myotonic dystrophy type 2 [39], the dysferlin gene in dysferlinopathy and the parkin gene in Parkinson's disease [54].

Drugs

Rare causes of camptocormia are side effects of therapeutic drugs. Among these, neuroleptic drugs and anti-Parkinson drugs are the most important (table 1). Among the neuroleptic drugs, camptocormia was most frequently induced by olanzapine [55]. L-DOPA induced or worsened camptocormia in single cases [12] but may also have a beneficial effect in cases where it is due to Parkinson's disease [19, 56] or MSA (see below). In a single patient, camptocormia was induced by donepezil [57]. In several patients, camptocormia was induced by an overdose of valproic acid [58]. There is also information that systemic corticosteroids may cause camptocormia [pers. commun.]. If causative drugs are withdrawn, camptocormia may improve.

Miscellaneous

In individual patients camptocormia was associated with trauma [59, 60], arthritis [60], or malignancies (table 1) [61]. In the latter case, camptocormia was interpreted as a manifestation of a paraneoplastic syndrome.

Psychiatric Disorders

Although camptocormia was initially described as a conversion disorder in military personal [29, 62, 63], psychogenic camptocormia is rare [29, 64]. Particularly in the early description of the abnormality it was interpreted as a form of hysteria occurring in individuals with low self-esteem and confusion of identity, sadomasochistic behavior toward military authorities, and impotence [11]. Camptocormia has also been described in association with Gilles de la Tourette syndrome [28], manic-depressive disorder [65], or a psychogenic state [29].

Diagnosis

Since the etiology of camptocormia is quite heterogeneous, investigations in different directions have to be carried out at the beginning of the diagnostic work-up. Generally, the diagnosis may be established upon clinical findings, blood chemical investigations, imaging of the cerebrum, EMG, or muscle biopsy.

Biology

There are a number of blood chemical parameters which are useful in the diagnostic work-up of camptocormia. Among these are the blood sedimentation rate, C-reactive protein, electrolytes, such as calcium and phosphorus, creatine kinase, aldolase, or vitamin D. For the diagnosis of metabolic myopathies, determination of lactate and pyruvate during standardized exercise can be of additional help.

Imaging

Cerebral CT scans may show atrophy, basal ganglia calcification, basal ganglia lacunas, lenticular lesions, or reduced volume of the midbrain or pons [32]. Cranial MRI may show signal abnormalities of the basal ganglia in a small number of patients with camptocormia and Parkinson's disease [22]. MRI of the vertebral muscles may show features of a circumscribed myopathy, such as variable degrees of atrophy and fatty replacement of the thoracolumbar paraspinal muscles [66]. These alterations are similar to those seen in muscular dystrophy [67]. Localized changes from edema with contrast enhancement are considered to be an early sign, whereas atrophy or fatty degeneration are considered as late changes [1, 3, 68]. Some authors interpret these changes rather as secondary than the cause of camptocormia [52]. CT scans of the spinal muscles may show atrophy and hypodensity of the muscles being interpreted as fatty involution [69].

Needle Electromyography

Depending on the underlying cause, EMG may be normal, neurogenic or myogenic. A myogenic pattern may be recorded even in patients with Parkinson's disease [1]. Needle EMG of the paravertebral muscles may also reveal abundant fibrillations, positive sharp waves, or bizarre high-frequency discharges [33].

Muscle Biopsy

Muscle biopsy may be normal or may show mild myopathic features, inflammatory features suggesting focal inflammatory myopathy (focal myositis), or dystrophic features [67]. There may also be extensive diffuse or lobulated fibrosis as the only variant finding in camptocormia patients as compared to controls [68]. Muscle biopsy in Parkinson's disease patients with camptocormia may be divided into three groups, i.e. necrotizing myopathy, inflammatory myopathy, or mitochondrial myopathy [70].

Myopathic changes in patients with Parkinson's disease include abnormal fiber size variation, increase of internal nuclei, increase of connective tissue, or myofiber disarray, or fatty degeneration [1, 31]. In patients with advanced Parkinson's disease and camptocormia muscle biopsy may show end-stage myopathy with autophagic vacuoles, chronic inflammatory myopathy, non-specific myopathic changes, or mitochondrial myopathy [33]. Single cases may also show amyloid deposition and ragged red fibers [pers. commun.].

Gait Analysis

Kinematic, kinetic and biomechanical analysis may reveal exaggerated anterior pelvic tilt during terminal stance when walking in an upright posture. In a forward-bent posture, however, the anterior pelvic tilt may be significantly less [71]. Some authors assume that the extreme forward-bent posture is a compensatory mechanism to reduce the excessive pelvic tilt [71].

Table 2. Treatment of camptocormia

Treatment	Reference
Non-invasive	
Psychotherapy	52
Physiotherapy	4
Orthoses	4
Backpack treatment	87
Antidystonia medication	53
L-DOPA	58, 74, 76
Steroids	26, 69
Protirelin tartrate	35
Botulinum toxin	38, 77
Immunoglobulins	40
Cyclosporine	40
Invasive	
Subthalamic nucleus stimulation	80, 88
Posterior thoracolumbar fixation	83
Anterior interbody fusion	83

Treatment

Treatment options for camptocormia are limited and frequently futile [72]. Generally, treatment options may be classified as conservative or invasive (table 2). Conservative measures include psychotherapy, physiotherapy, application of drugs, injection of botulinum toxin, withdrawal of causative drugs, or electroconvulsive therapy. Invasive therapeutic measures include surgical methods or deep brain stimulation. Treatment of choice is the therapy of the underlying disorder and in case no disease-modifying agents are available orthoses, physiotherapy, and eventually analgesics are the only choice [4].

Psychotherapy

There are a number of psychotherapeutic techniques which can be applied to patients with camptocormia with a psychogenic origin. These include psycho-education regarding secondary gain, suggestions to improve posture, positive reinforcement, or behavioral therapy [56]. Persuasive re-education was particularly applied in WW1 cases but this psychological therapy was rather additive than persuasive [11].

Physiotherapy, Orthoses

Classical orthoses and physiotherapy often provide little correction, are often poorly tolerated [4], and are quickly abandoned [73]. Application of a thoracopelvic anterior distraction orthosis, however, results in a quality of life increase by 90% [4]. In single cases, physiotherapy and orthoses may relieve lower back pain [7].

L-DOPA

In the majority of cases with advanced Parkinson's disease, camptocormia is L-DOPA-resistant [1, 74]. However, in single cases, camptocormia associated with Parkinson's disease, dystonia, or MSA, administration of L-DOPA has been shown to be beneficial [62, 74, 75]. Depending on the investigated cohort, up to 20% of the Parkinson patients with camptocormia profit from L-DOPA therapy [3]. In a patient with MSA with predominant parkinsonism, camptocormia and Parkinson's disease markedly improved under L-DOPA [74]. In a patient with Parkinson's disease, adjustment of dopaminergic therapy by carbidopa-levodopa and entacarpone resulted not only in improvement of Parkinson's disease but also of camptocormia [76].

Immunoglobulins

Little data have been published demonstrating a beneficial effect of immunoglobulins (IVIG) in camptocormia [25]. An 81-year-old male with confirmed inflammatory myopathy of the paraspinal muscles experienced dramatic improvement to treatment with IVIG [25]. IVIG seem to be effective only in cases with inflammatory myopathy.

Botulinum Toxin

Injection of botulinum toxin into the rectus abdominis muscles has been shown to be beneficial in single cas-

es in which camptocormia was due to focal dystonia [28, 38]. Injections of botulinum toxin into the iliopsoas muscles may also relieve camptocormia [34]. In other patients with Parkinson-associated camptocormia, however, injection of botulinum toxin into the iliopsoas muscle was ineffective [72]. Botulinum toxin may not only be effective in patients with focal dystonia but also in patients with Parkinson's disease [77].

Electroconvulsive Therapy

In a single patient with camptocormia induced by olanzapine, discontinuation of the drug and application of L-DOPA was hardly effective, but electroconvulsive therapy was tried with success [61].

Miscellaneous

Patients with drug-induced camptocormia usually respond to withdrawal of antipsychotics or reduction of the daily doses. Single cases with inflammatory myopathy of the paraspinal muscles may profit from administration of steroids [27]. Steroids for camptocormia in Parkinson's disease, on the contrary, failed to show a beneficial effect [17]. Application of anticholinergics, amantadine, dopamine agonists, muscle relaxants or tetrabenazine is usually ineffective [52].

Deep Brain Stimulation

In single cases in which camptocormia is associated with Parkinson's disease or segmental dystonia, bilateral pallidal high-frequency deep brain stimulation [62, 78, 79] or bilateral subthalamic nucleus stimulation [80, 81] may have a beneficial effect. The therapeutic effect of deep brain stimulation suggests that, at least in single cases, camptocormia is indeed a CNS disease due to affection of the striatum and its reticulospinal and thalamic projections [27]. In a patient with long-standing crippling Parkinson's disease, camptocormia improved dramatically after bilateral subthalamic deep brain stimulation [79]. For pallidal stimulation deep brain stimulation electrodes are stereotactically implanted to target the internal globus pallidus [82]. Long-term pallidal stimulation results in significant functional improvement in the absence of any treatment-related adverse effects [82].

Surgery

In cases where conservative measures are unsuccessful, patients may profit from posterior thoracolumbar fixation, which may need to be augmented with anterior interbody fusion [83].

Conclusions

Camptocormia in the vast majority of cases is an organic disorder, either a manifestation of CNS disorders or due to affection of the peripheral nerves or the skeletal muscle. Only rarely is camptocormia caused by a psychiatric disorder. Drugs, trauma, or orthopedic problems may have a contributing effect. Since camptocormia is due to a number of various different disorders, the initial step in the management of camptocormia is detection of the underlying cause. Treatment should generally be directed towards the underlying etiology and pathomechanism. General measures, such as physiotherapy, orthoses, or botulinum toxin may be helpful in single cases. Only if the underlying cause is effectively treated can a substantial therapeutic effect be expected. If treatment of camptocormia is ineffective, patients sooner or later require walking devices and lastly a wheelchair. Since the ability to characterize the pathophysiology of camptocormia in Parkinson's disease with the available technologies is limited, an animal model of abnormal posturing is required to fully understand the postural phenomena and to develop effective treatment.

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