Bent spine syndrome: Presentation of four cases and literature review

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ABSTRACT

Objectives: Bent spine syndrome (BSS) or camptocormia is a cause of unstructured sagittal imbalance of difficult medical and surgical management. The purpose of this paper is to describe the causes of BSS and how to approach its treatment. **Materials and Methods:** Retrospective analysis of 4 cases of BSS treated at our center and review of the literature. **Results:** The 4 patients were women between 60 and 82 years of age. In 3 of them, BSS was due to an isolated atrophy of the paravertebral erector spinae muscles, and in 1 of them, it was due to an inflammatory myopathy. **Conclusions:** The prognosis of BSS is poor in all cases, so symptoms usually progress to an irreversible difficulty to stand upright.

Keywords: Camptocormia; erector spinae muscle atrophy; bent spine syndrome.
Level of Evidence: IV

Síndrome de la espalda inclinada. Presentación de cuatro casos y revisión de la bibliografía

RESUMEN

El síndrome de la espalda inclinada o camptocormia es una causa de desequilibrio sagital del tronco no estructurada de difícil manejo médico y quirúrgico. Puede ser secundario a enfermedades del sistema nervioso central o periférico, o de origen primario muscular, como la atrofia aislada de la musculatura paravertebral espinal. El diagnóstico se basa en la evaluación clínica, los estudios por imágenes, la electromiografía y la biopsia muscular. El síndrome de la espalda inclinada, cualquiera fuera su causa, tiene un pronóstico pobre, los síntomas suelen progresar hasta afectar la bipedestación de manera irreversible. Presentamos un análisis retrospectivo de cuatro casos de síndrome de la espalda inclinada tratados en nuestro centro y una revisión de la bibliografía.

Palabras clave: Camptocormia; atrofia muscular erectora espinal; síndrome de la espalda inclinada.
Nivel de Evidencia: IV

INTRODUCTION

The Bent Spine Syndrome (BSS) also known as camptocormia (from the Greek words: *kamptos* [to bend] and *kormos* [trunk]) is characterized by an involuntary flexion of the trunk in the standing position but not in the recumbent position. The BSS is an acquired postural abnormality and its causes are different from those of structural kyphosis. The BSS is also different from the Pisa Syndrome (pleurothotonus), which is usually characterized by a lateral flexion of the trunk secondary to a tardive dystonia related to the continued use of antipsychotics.[1]

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The BSS was described by Henry Earle (1815) and Brodie (1818), and reported by James Parkinson in one of his cases (1817).[2] The term "camptocormia" was coined by the French neurologist Souques and Rosanoff (1816) to describe an abnormal curvature of the trunk related to a "hysterical flexion" in the World War I soldiers who had sustained a shock during battle.[3] For a century, camptocormia was considered a psychiatric condition until Kiuru and Laroche[4] first associated it with organic diseases. The current causes of BSS are numerous, even secondary and genetic anomalies, as is the case of the myotonic dystrophy type 2 with a mutation of the *ZNF9* gene[5] or of the multiple-system atrophy.[6]

Our objective is to describe BSS causes and different treatment approachs.

We present 4 BSS clinical cases of muscular origin: 3 due to isolated atrophy of the paraspinal muscles and 1 due to inflammatory myopathy, which patient had been intervened three times and still her BSS symptoms had worsened.

Additionally to BSS symptoms, all patients had lumbar pain for which they had been treated by radiofrequency, facet joint infiltration, and physical therapy with little improvement. For diagnostic purposes, all patients had full-body MRI scans to search for impairment in other muscles, laboratory tests to detect inflammatory or autoimmune conditions, and a muscle biopsy (Table 1).

Table 1. Cases of BSS of muscular origin

	Sex	Age	History	Lumbar pain	Diagnosis	Previous interventions
1	F	62	Autoimmune Hepatitis, high-blood pressure, atenolol	Yes	Myositis	Lumbar arthrodesis
2	F	82	High-blood pressure, Breast Cancer	Yes	Isolated paraspinal muscle atrophy	Radiofrequency, epidural infiltration, facet joint infiltration
3	F	66	Uterus cancer, statins	Yes	Isolated paraspinal muscle atrophy	Radiofrequency, facet joint infiltration
4	F	60	Diabetes, high-blood pressure, bisoprolol, amlodipine	Yes	Isolated paraspinal muscle atrophy	No

F = female; M = male.

CASE 1

The patient was a 63-year-old woman who came to consult for a failed spine surgery. She had undergone 3 three lumbar spine surgeries, in 3 occasions during the previous 2 years, with no symptom resolution: lumbar pain and forward-bent trunk. She presented reducible kyphosis affecting the dorso-lumbar region and was unable to walk upright (Figure 1). She is currently without lumbar hardware (last lumbar hardware removal) (Figure 2). She walks supported by a walking frame to keep an upright position, and pushing on her thighs (similarly to the Gowers maneuver of patients with Duchenne muscular dystrophy). The MRI taken before the first surgery shows a healthy lumbar spine, with subtle signs of spondylopathy, but lumbar paraspinal muscles with fatty replacement (Figure 3). Full-body MRI scan showed symmetrical impairment in several muscles. Biopsies of the paraspinal and gastrocnemius muscles showed necrotic fibers within muscle fibers and interstitial inflammation. A main finding in the immunohistochemical analysis was the overexpression of MHC class I in the muscle cell membranes with abundant macrophages in perimysium and endomysium. Her diagnosis was myositis. A few months later, she suffered from autoimmune hepatitis and received prednisone and azathioprine therapy. The hepatic enzymes' values improved but the BSS did not since the fatty replacement changes in the paraspinal muscles were already present.

Figure 1. Patient unable to maintain an up-straight position while standing, a condition which subsided with recumbent positioning, proving a non-structural sagittal imbalance.

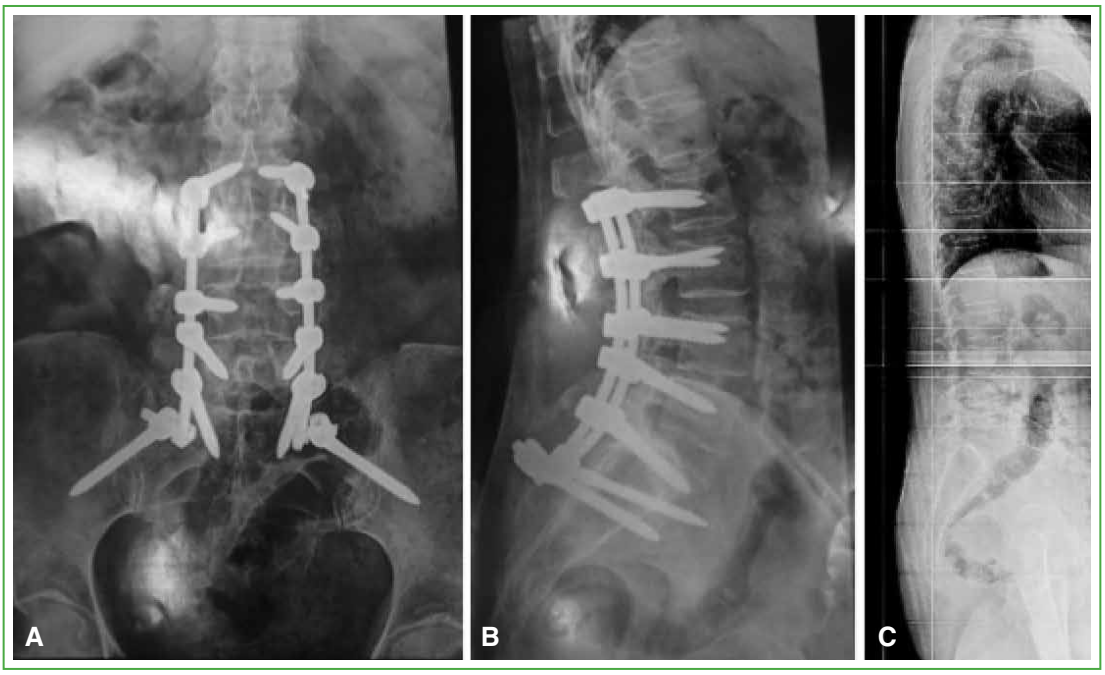

Figure 2. A and B. Radiographs showing hardware at L2-pelvis that worsen the BSS and lumbar pain. **C.** Radiograph after hardware removal.

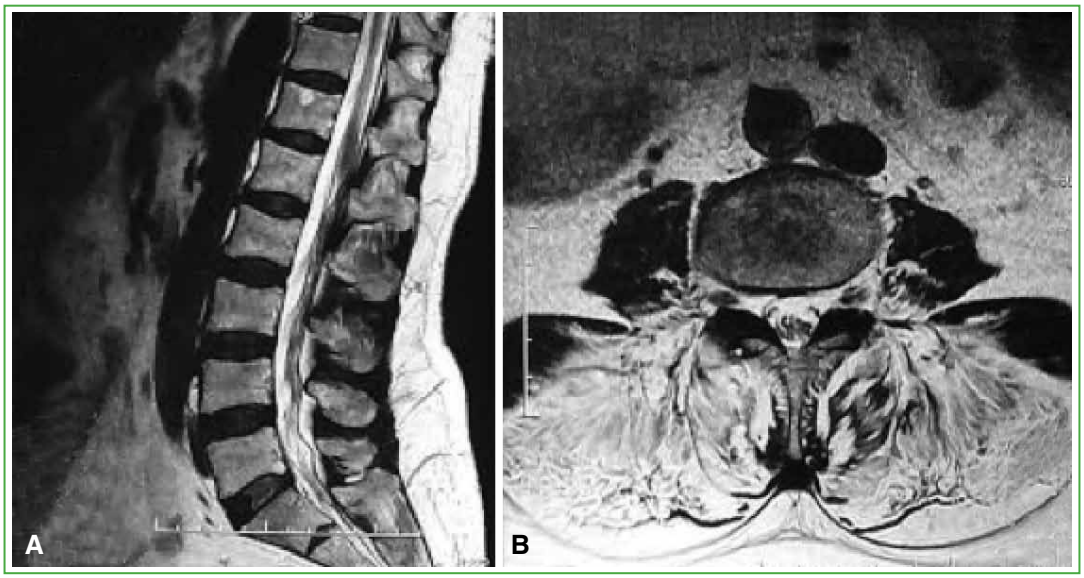

Figure 3. Preoperative MRI. **A.** In the sagittal section, no structural condition can be seen. **B.** Transverse section of T2 showing significant fatty replacement of the paraspinal erector muscles.

CASES 2, 3 AND 4

The three patients consulted for progressive lumbar pain while maintaining an upright position. They all had different forward-bent trunk angles, which subsided in recumbent position, and no spine structural conditions (Figures 4 and 5). None of them had history of myopathies or Parkinson's disease. Laboratory tests, full-body MRI and EMGs were performed to the three patients, and they were diagnosed with isolated spinal muscular atrophy. Table 1 shows the patient's prior conditions which may have been related to the development of BSS, although no direct link was able to be found (paraneoplastic syndromes, use of statins, diabetes mellitus). They were all advised not to pursue surgery and were treated with dorso-lumbar or lumbo-sacral orthoses, physical therapy and the complementary use of walkers as assistive devices.

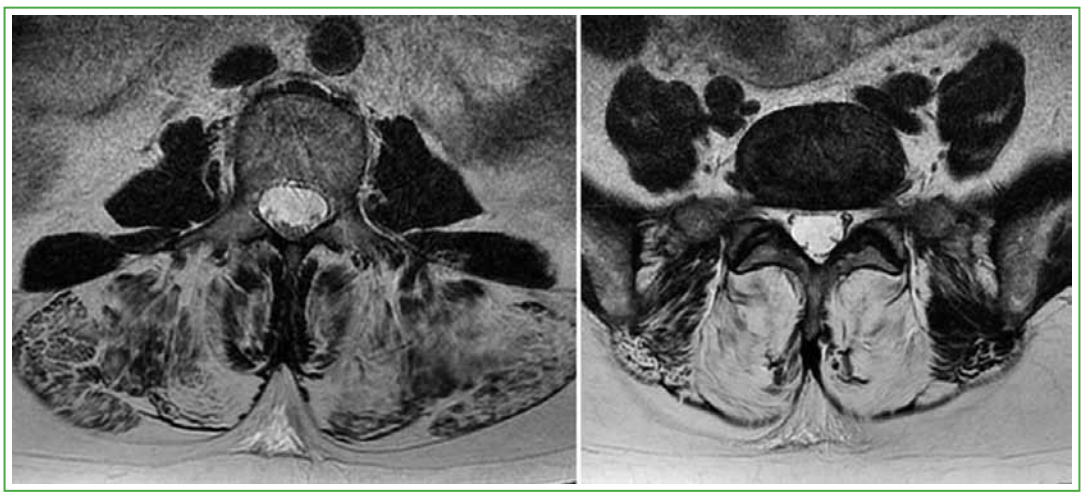

Figure 4. MRI transverse section of T2 of a 60-year-old woman with type 2 Diabetes history who presented progressive weakness to maintain an up-straight position. At L3 and L5 the scan shows a massive fatty replacement of the erector muscles with no volume decrease of the muscle compartment.

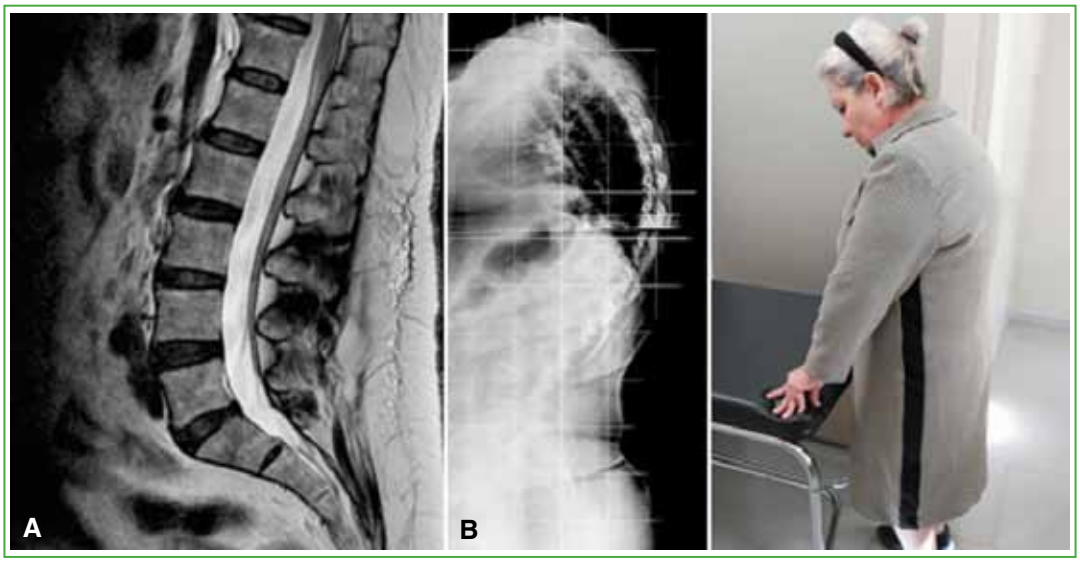

Figure 5. Figure 4 patient. **A.** MRI showing a healthy spine with no structural condition. **B.** Radiograph showing marked kyphosis and lordosis while standing due to muscle weakness. Full-body muscle MRI was performed. The atrophy was restricted to the lumbar erector muscles

DISCUSSION

Clinical presentation

There are currently no well-defined guidelines to characterize BSS. Most studies use a 15° to 45° angle of forward bending as principal criterion due to a progressive weakness of the paraspinal extensor muscles.[7-9] The BSS progressively manifests while standing or walking, and subsides with recumbent positioning.[10] In some cases, its presentation is painless; however, it is commonly associated with axial lumbar pain, especially when it is associated with Parkinson's disease.[11-13] The BSS is normally misconstrued as the Dropped Head Syndrome and their concurrent presentation is rare, although both conditions share many of their etiologic causes.[14] It is estimated that the average clinical presentation age for BSS is 65 years, and that 69% of the patients have history of Parkinson's disease and 25% of dystonia. Up to a 50% have family history of myopathy.[7,12] The BSS currently presents, most commonly, due to organic diseases and, only rarely, secondary to a mental disorder.[15] The general incidence of BSS is unknown. The only available published data is on its incidence concerning movement disorders. The relative frequency of primary or secondary myopathies is highly variable. In a BSS cohort from a rheumatology center, 65% of the subjects suffered from muscle dystrophy, 17% had Parkinson's disease, and 13% had myositis.[16] In a BSS cohort from a neuromuscular clinic, 25% had scapulohumeral dystrophy and 18% had inclusion body myositis.[14] There is evidence that the scapulohumeral dystrophy is underestimated in the BSS.[15]

Etiology

There is no consensus or standard classification on BSS. On the one hand, neurologist prefer to divide its causes according to whether they are disorders related to the central or the peripheral nervous system; on the other hand, we prefer to divide them into: muscular origin and neurological origin, according to the response to the local or systemic therapy.[7]

BSS of muscular origin

In this subgroup of patients, the BSS derives from weakness of the paraspinal erector muscles. The insufficiency of these muscles results in a postural imbalance that makes the trunk bend forwards. As it is not a structural deformity, posture posture corrects itself spontaneously with recumbent positioning.

Weakness of the extensor muscles may by a primary and idiopathic condition or secondary to several diseases that produce pathological changes to all the muscles responsible for the extension of the trunk (Table 2). BSS of muscular origin may be detected through changes in the EMG, muscle hypodensity in the MRI, and myopathic changes in the biopsy.

Table 2. Causes of muscular conditions that may result in BSS[14,19,22-24,38]

Myopathie causes	Characteristics	Diagnosis
Dystrophies		***Genetic***
Limb-girdle dystrophies	2 subtypes: LGMD1-2	Genetic: different chromosomes
Facioscapulohumeral Muscular Dystrophy		Chromosome 4 (4q35) genetic defect
Myotonic dystrophies	2 subtypes: autosomal dominant MD1 and MD2	Genetic
Duchenne dystrophy	X-chromosome recessive disorder Dystrophin abnormality	PCR. Genetic test: Xp21.2 deletion
Inflammatory		***Muscle biopsy***
Dermatomyositis	Calcification of connective tissue. Local compromise.	Clinical, biopsy
Polymyositis	Systemic compromise	Biopsy, anti-Jo AB (65%)
Nonspecific focal myositis	Unknown cause	Biosy: rule out sarcoma, infection
Inclusion Body Myositis	Unknown cause. Rule out polymyositis	Biopsy: vacuolar cytoplasmic degeneration, abnormal protein deposits
Chronic inflammatory polyneuropathy	Autoimmune. Similar to Guillain-Barré syndrome	Cerebrospinal fluid tap test, biopsy
Paraneoplastic Myositis	Reactive autoimmune	Biopsy
Myasthenia gravis	IgG against acetylcholine receptors on motor end plate	Tensilon test, acetylcholine anti-receptor antibodies
Metabolic-endocrine		
Hypothyroidism	Different etiologies	TSH
Osteomalacia	Vitamin D deficiency	Calcium and Vitamin D
Steroid	External intake	
Amyloidosis	Primary, secondary, familial	Amyloid deposits
Mitochondrial myopathies		
Carnitine palmitoyltransferase II deficiency	Autosomal recessive. Rhabdomyolysis episodes triggered by exercise or fasting	Genetic. Mutations in the CPT2 gene
Drugs	*External intake*	
Levodopa/carbidopa		
Olanzapine	Symptom improvement may take weeks after suspension	
Valproic acid		
Atomoxetine		
Donepezil		
Rivastigmine		

Primary muscle BSS

- Primary idiopathic isolated myopathy of the paraspinal muscles

This cause is one of the most common types in clinical practice. It manifests itself in adults with no history of neurological or autoimmune diseases, consulting for difficulties maintaining an upright position and a progressive fatigue during the course of the day. The main findings are fatty infiltration affecting the lumbar paraspinal muscles detected through imaging and biopsy.[16,17] In 1991, Laroche *et al*. published, for the first time, a series of 16 patients (mean age: 66 years) which is characterized by the late onset of a myopathy limited to the paraspinal muscles. All patients had in common an anterior flexion of the trunk which subsided with recumbent positioning, a fatty infiltration affecting the paraspinal muscles (according to CT scan), a shared pattern in the biopsy and family history. The authors concluded that the acquired lumbar kyphosis could be due to a delayed-onset primary myopathy, localized exclusively in the paraspinal extensor muscles. Since this first report, this initial hypothesis has been supported by other researchers[17-19] and by subsequent studies of the same research group.[16] Other authors confirm its idiopathic etiology, that it is prevalent in women and has cases of family inheritance.[17,19,20] The muscle weakness is strictly limited to the paraspinal extensor muscles, without any other clinically detectable muscle deficits. The BSS is responsible for functional disability and lumbar pain which worsen over and have little response to rehabilitation or medical treatment.[20] The most useful diagnostic tools are the imaging studies (CT or MRI) of the lumbar and dorsal sections of the paraspinal muscles.[17] The classic image is that of the paraspinal muscles with normal volume, but complete fatty replacement (Figures 4 and 5); this differentiates them from images of BSS neurogenic atrophy etiology, where muscle volume decreases, but muscle density is normal.[21] Ricq and Laroch conducted a study where 23 patients' CT scans were repeated after a 3.5-year follow-up, and they reported that some showed the same fatty infiltration pattern in the middle gluteal and the supraspinatus muscles, thighs and calves.[20] The EMG may detect myogenic patterns in the middle gluteal muscles and in the deltoid,[22] even if these muscles show no clinical compromise. In all the considered studies, the anatomical pathology of the muscle biopsies revealed variation in fibrosis and replacement of muscle fibers by adipose tissue.[17,19] The BSS of primary idiopathic origin may be considered in the group of progressive muscle dystrophies with late-onset. Although several research studies have found an inherited component, there are no genetic and molecular studies to identify the method of inheritance; it may be a congenital disease.[23] This data enables to speculate that this late-onset myopathy which is limited to axial muscles might be associated with one of the main muscle dystrophies, possibly to one of the limb-girdle muscular dystrophies

Secondary muscle BSS

The BSS may present as a symptom of several systemic muscle diseases. Dystrophies are hereditary diseases. The Facioscapulohumeral Muscular Dystrophy is a condition that causes progressive muscle weakness and that mainly affects the face, shoulder, and upper arm muscles. Its genetic defect is at the tip of the long arm of human chromosome 4 (4q35). The Facioscapulohumeral Muscular Dystrophy is one of many types of muscular dystrophies and probably the one most associated with BSS[23], and , in some cases, remains as an unrecognized cause.[15] Other dystrophies responsible for BSS are Limb-girdle Dystrophy and Myotonic Dystrophy type 1.[23] The BSS may be secondary to myositis, such as Dermatomyositis and Polymyositis, or focal myositis or inclusion body myositis. Diagnosis may be obtained through EMG, elevated CPK levels and muscle biopsy, characteristically revealing perivascular and inflammatory endomysial infiltrations with muscle fiber necrosis.[24] Certain metabolic or endocrine disorders, such as Hypothyroidism and Osteomalacia or Corticosteroid Myopathy, may cause secondary BSS.[18] Certain drugs associated with BSS, which are also considered a precipitating cause of the Pisa Syndrome, are the valproic acid, anticholinergic agents and dopamine agonists.[25] Finally, a study has reported two rare BSS causes: mitochondrial myopathy and amyloid myopathy.[26]

BSS of neurological origin

Among the neurodegenerative diseases, the BSS is most common in Parkinson's patients.[7] Within this population, BSS prevalence is 3-11.6% (according to several authors[11,26]) and studies on larger series report a prevalence of 10% prevalence.[27] BSS predisposing factors in Parkinson's patients are: old age, male sex, early axial muscle disorders (axial dystonia), Parkinson's severity, and a latency with an onset >5 years from Parkinson's diagnosis.[28,29]

Recent studies demonstrate that most neurological BSS result from lesions in the basal ganglia, responsible for the coordination of the postural reflexes in flexion/extension movements that enable keeping an erect posture.[30]

This functions is regulated by dopamine, which partially originates in the substantia nigra. The role of dopamine is essential for the function of the basal ganglia, its alteration results in disorders, such as Parkinson's Disease and Parkinson-Plus Syndromes, which include Multi-System Atrophy and Supranuclear Palsy.[27]

One feature of Parkinson's patients while standing is a slight anterior flexion of the trunk associated with Parkinson's axial dystonia.[31] In BSS patients, there is a severe anterior flexion which is sometimes associated with scoliosis. The BSS in Parkinson's patients was defined as an anterior flexion of the trunk >45°.[7,32] It is a common condition in a particular subgroup of Parkinson's patients, where axial dystonia is the core symptom, and there is excessive abdominal muscle activation. Patients with BSS are unresponsive to levodopa; however, it reduces akinesia, tremor and rigidity.[7,23,32] Levodopa-unresponsive effect in BSS's patients is likely to represent a specific form of Parkinson's disease where other non-dopaminergic dysfunctions in the basal ganglia may be responsible.[23]

For years axial dystonia was considered the cause of BSS in Parkinson's patients.[33-35] Lately, EMG, CT, MRI and biopsy studies have described focal myopathic changes in the paraspinal muscles of Parkinson's patients; however, its prevalence is not well-documented and the exact role of theses changes and BSS development are still unknown.[32,36] A new theory on the dysregulation of proprioception ability would explain the BSS mechanism in Parkinson's patients. The myopathic changes equal to those of a muscle that had tenotomy: the muscle tension is too low and the muscle receives a stimulus of hypercontraction. As opposed to a tenotomy, where the myopathological changes are due to a dysregulation of the polysynaptic reflex arch at the tendons level, the myopathological changes in Parkinson-related BSS could be the result of a dysregulation of proprioception at the level of the central nervous system. This pathophysiological concept is supported by the observation that lenticular lesions may cause BSS and that neurostimulation of the subthalamic nucleus may relieve BSS in Parkinson's patients, as it partially restores the proprioceptive ability. In the progression of the BSS, when muscle is replaced by chronic changes of fibrosis and fatty degeneration, the neurostimulation of the subthalamic nucleus will lose its effect on the muscle and, consequently, on the trunk inclination.[37]

Besides Parkinson's disease, the BSS may also occur in patients suffering from Multiple-System Atrophy[37,38] and Lewy Body Dementia,[39] and Amyotrophic Lateral Sclerosis, especially when the onset affects the respiratory muscles.[40] With regard to other neurodegenerative disorders, there have been individual reports of BSS in Alzheimer's patients.

Diagnosis

Due to the heterogeneous etiologies of BSS, we propose a diagnostic algorithm based on personal and family data, blood tests, imaging (CT, MRI), EMG, muscle biopsy and, eventually, genetic studies.

Clinical examination

Ask them about their disease history concerning conditions of a genetic or hereditary component, such as muscle dystrophies, Parkinson's disease (families with a high incidence of mutations in SPARK genes) or metabolic or autoimmune diseases.

At first, focus on primary signs of myopathies or movement disorders, such as Parkinson's disease or dystonias. In the case of myositis, the affected muscle may be asymmetrical and have inflammation signs. The angles of the trunk inclination must be measured with a goniometer (since inclination may vary with time) so as to verify the inclination changes.[41] The BSS progress may be useful in finding its etiology: inflammatory BSS has a faster evolution (weeks) while myopathic-primary BSS develops slowly (months or years). Some features should be recorded: spine conditions, drug intake (antipsychotics), lumbar pain, and the use of assistive walking devices (cane, walker, grocery cart, etc.).

Blood test

Some of the biochemical parameters that may be useful in finding its etiology are: acute phase reactants (C-reactive protein, erythrocyte sedimentation) and CPK for the inflammatory or autoimmune BSS, calcium and phosphate metabolism, vitamin D, thyroid hormones, lactate and pyruvate for the endocrine-metabolic BSS.[7,12,23]

Neurophysiological study

EMG is a useful diagnostic tool in BSS. Depending on the BSS etiology, the EMG may be normal or show myopathic or neurogenic changes. The choice of muscles to be examined should be determined by the physical examination. These may include dorsal and ventral trunk muscles (mainly paravertebral and rectus abdominis muscles) in patients with movement disorders. When myopathy is suspected a distal and proximal limb muscle evaluation should be added.

In the elderly, neurogenic pattern and spontaneous activity are not necessarily pathological.[42] Motor units of the paravertebral muscles differ from those of the limb muscles in that they have a smaller amplitude and a shorter duration.[19] A myogenic pattern may be recorded in Parkinson's patients.[8]

Imaging

Brain CT or MRI may show signs of atrophy, basal ganglia calcifications, lenticular lesions or trunk abnormalities.[11] In cases of Dystrophy or Idiopathic Restrictive Myopathy, MRI of the paraspinal muscles may show features of a circumscribed myopathy, such as variable degrees of atrophy and fatty replacement of the erector muscles (Figure 4).[43] The acute muscular changes that result in edema may be detected on MRI STIR sequence, as it is the case of myositis. In Parkinson's patients, findings may vary from edema and swelling in the paraspinal, quadratus lumborum and psoas muscles during the first 31 months,[36,44] up to the most common chronic late findings of fatty degeneration and replacement.[18,45,46] A disadvantage of the muscle MRI is its lack of specificity. Findings of muscle STIR hyperintensities should be followed by a muscle biopsy to differentiate myositis from a trauma, denervation or other possible causes of muscle diseases.[45]

Muscle biopsy

The most common indication for a muscle biopsy in BBS aims at identifying or excluding myositis. The biopsy of a paraspinal muscle cannot be replaced by another muscle biopsy, therefore prior imaging guidance (MRI, CT, ultrasound) should be considered. Biopsy sample must not be taken from the same site where the EMG was performed, because needle tracks may mimic inflammatory processes. In Table 2, we list the conditions that should be considered.

In Parkinson's patients, muscle biopsy shows consistent lesion pattern composed of myopathic changes with type-1 fiber hypertrophy, loss of type-2 fibers, loss of oxidative enzyme activity, and a thin granular layer of acid phosphatase reactivity of lesions.[44] They also show endomysial fibrosis variable in size and fatty degeneration that seems to correlate with the duration of BBS.[32]

Treatment

Treatment of BSS depends mainly on the etiology.

BSS treatment in Parkinson's patients

In the majority of patients, levodopa treatment provides no improvement on BSS symptoms,[7,32] with the exception of patients with an off-dystonia those with end of dose fluctuations.[46] Optimizing the Parkinson's disease therapy may improve BSS, as was described with continuous infusions of apomorphine to five Parkinson's patients with a three-year follow-up.[47]

Patients with abdominal dystonia with may partially benefit from a treatment with botulinum toxin injections in the rectus abdominis, external oblique and iliopsoas muscles.[7,48,49]

Bilateral neurostimulation of the subthalamic nuclei has been shown to improve core symptoms of Parkinson's disease.[46,50] However, neurostimulation has proven to be effective in treating BSS secondary to Parkinson's disease if instituted during the first 1.5 years after symptom onset of trunk inclination. Between 17 months and 3 years, results are inconclusive. After a 40-month period, no patient has improved, possibly as atrophic chronic changes and fatty replacement had already settled into the muscles.[37,51]

Surgical BSS treatment in Parkinson's patients

Most reported cases of BSS with surgical treatment were composed by Parkinson's patients. The high complication rate in these patients seems to be connected with their high susceptibility for Osteoporosis.[33] Parkinson's patients constitute a high-risk population for surgery, having a medical complication rate of 30%, a surgical

complication rate of 52%, and a revision rate of 35%.[34] Surgical spine procedures in Parkinson's patients increases the risk of BSS development. Minor surgical interventions have even a greater risk of resulting in instability, especially at the adjacent segment level.[9]

Treatment of dystonic BSS

In some published cases, patients have improved under treatment with either levodopa alone[50] or in combination with tetrabenazine, pimozide or anticholinergic agents.[51] Patients with Isolated Abdominal Dystonia may partially benefit from a treatment with botulinum toxin injections in the rectus abdominis and external oblique muscles.[49] As a last alternative, deep brain stimulation of the internal globus pallidus proved effective in this subgroup of patients diagnosed with primary dystonia.[52]

BSS treatment in patients with primary myopathies and Myositis

In cases of Myositis and Demyelinating Polyradiculoneuropathy, the choice of immunosuppressant treatment depends on the disease-specific state and its stage.

For patients with Focal Myositis, IV steroid therapy suffices.[45,53] In cases secondary to Polymyositis or Dermatomyositis, the combination of steroids and immunosuppressants (methotrexate, azathioprine, cyclosporine) depends on the disease own evolution.[10] Inclusion Body Myositis constitutes an exception since the immunosuppressant therapy and immunoglobulins have been proven ineffective. Therefore, the therapeutic approach should be focused on physical therapy.[54]

BSS orthosis treatment

Regardless of its etiology, BBS patients may benefit from orthosis to straighten trunk inclination and reduce back pain. In a prospective study, de Séze *et al*. evaluated the use of thoraco-pelvic anterior distraction orthosis in BBC patients who then experienced a significantly clinical and radiological improvement.[55] Wheel walkers and using a grocery cart for support may be an effective assistive walking device to improve the ability to walk and to abate lumbar pain.[56,57]

CONCLUSION

BSS prognosis, regardless of the etiology, is poor, symptoms usually progress to an irreversible difficulty to maintain an upright posture.

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REFERENCES

1. Yeh YW, Chen YC, Chen CK, Feng HM, Wang TY, Chen CL. Chronic Pisa syndrome associated with switching antipsychotics from olanzapine to ziprasidone. *Prog Neuropsychopharmacol Biol Psychiatry* 2009;33(1):162-3. https://doi.org/10.1016/j.pnpbp.2008.11.001
2. Parkinson J. An essay on the shaking palsy. 1817. *J Neuropsychiatry Clin Neurosci* 2002;14(2):223-36. https://doi.org/10.1176/jnp.14.2.223
3. Souques A. Réforme, incapacité, gratifications dans la camptocormie. *Rev Neurol* 1916;23:757-8.
4. Laroche M, Rousseau H, Mazieres B, Bonafe A, Joffre F, Arlet J. Value of x-ray computed tomography in muscular pathology. Personal cases and review of the literature. *Rev Rhum Mal Osteoartic* 1889;56:433-9.

5. Serratrice G. Clinical semiology of neuromuscular diseases. Bent spine myopathy or syndrome. *Acta Myol* 2007;26:1-4. https://www.ncbi.nlm.nih.gov/pmc/articles/PMC2949321/
6. Reichel G, Kirchhöfer U, Stenner A. Camptocormia-segmental dystonia. Proposal of a new definition for an old disease. *Nervenarzt* 2001;72:281-5. https://doi.org/10.1007/s001150050751
7. Azher SN, Jankovic J. Camptocormia: pathogenesis, classification and response to therapy. *Neurology* 2005;65:355-9. https://doi.org/10.1212/01.wnl.0000171857.09079.9f
8. Margraf NG, Wrede A, Rohr A, Schulz-Shaeffer WJ, Raethjen J, Eymess A, et al. Camptocormia in idiopathic Parkinson´s disease: a focal myopathy of the paravertebral muscles. *Mov Disord* 2010;25(5):542-51. https://doi.org/10.1002/mds.22780
9. Nakane S, Yoshioka M, Oda N, Tani T, Cida K, Suzuki M, et al. The characteristics of camptocormia in patients with Parkinson´s disease: a large cross-sectional multicenter study in Japan. *J Neurol Sci* 2015;358(1-2):299-303. https://doi.org/10.1016/j.jns.2015.09.015
10. Kuo SH, Vullanganti M, Jimenez-Shahed J, Kwan JY. Camptocormia as a presentation of generalized inflammatory myopathy. *Mucle Nerve* 2009;40(6):1059-63. https://doi.org/10.1002/mus.21403
11. Lepoutre AC, Devos D, Blanchard-Dauphin A, Pardessus V, Maurage CA, Ferriby D, et al. A specific clinical pattern of camptocornia in Parkinson´s disease. *J Neurol Neurosurg Psychiatry* 2006;77(11):1229-34. https://doi.org/10.1136/jnnp.2005.083998
12. Ricq G, Laroche M. Acquired lumbar kyphosis caused in adults by primary paraspinal myopathy. Epidemiology, computed tomography findings and outcomes in a cohort of 23 patients. *Joint Bone Spine* 2000;67(6):528-32. https://doi.org/10.1016/S1297-319X(00)00203-7
13. Margraf NG, Granert O, Hampel J, Wrede A, Schulz-Schaeffer WJ, Deuschl G. Clinical definition of camptocormia in Parkinson's disease. *Mov Disord Clin Pract* 2016;4(3):349-57. https://doi.org/10.1002/mdc3.12437
14. Ghosh PS, Milone M. Camptocormia as presenting manifestation of a spectrum of myopathic disorders. *Muscle Nerve* 2015;52(6):1008-12. https://doi.org/10.1002/mus.24689
15. Skidmore F, Anderson K, Fram D, Weiner W. Psychogenic camptocormia. *Mov Disord* 2007;22(13):1974-5. https://doi.org/10.1002/mds.21708
16. Laroche M, Cintas P. Bent spine syndrome (camptocornia): a retrospective study of 63 patients. *Join Bone Spine* 2010;77(6):593-6. https://doi.org/10.1016/j.jbspin.2010.05.012
17. Doherty KM, Noyce AJ, Silveira-Moriyama L, Nisbet A, Quinn N, Lees AJ. Familiar camptocornia: from dystonia to myopathy. *J Neurol Neurosurg Psychiatry* 2012;83(3):350-1. https://doi.org/10.1136/jnnp.2011.246561
18. Laroche M, Ricq G, Delise MB. Bent spine syndrome: computed tomography study and isokinetic evaluation. *Muscle Nerve* 2002;25(2):189-93. https://doi.org/10.1002/mus.10016
19. Mahjneh I, Marconi G, Paetau A, Saarinen A, Salmi T, Somer H. Axial myopathy: an unrecognised entity. *J Neurol* 2002;249(6):730-4. https://doi.org/10.1007/s00415-002-0701-9
20. Ricq G, Laroche M. Cyphose lombaire acquise de l ádulte par myopathie primitive des muscles paravertébraux. Aspects épidémiologiques, tomodensitométriques et évolutifs, à propos d´une cohorte de 23 patients. *Rev Rhum* 2000;67(10):908-13. https://doi.org/10.1016/S1169-8330(00)00034-X
21. Bulke JA, Crolla D, Termote JL, et al. Computed tomography of muscle. *Muscle Nerve* 1981;4(1):67-72. https://doi.org/10.1002/mus.880040112
22. Laroche M, Delisle MB, Aziza R, Lagarrigue J, Mazieres B. Is camptocormia a primary muscular disease? *Spine* 1995;20(9):1011-6. https://doi.org/10.1097/00007632-199505000-00007
23. Emery AE. The muscular dystrophies. *Lancet* 2002;359(9307): 687-95. https://doi.org/10.1016/S0140-6736(02)07815-7
24. Hund E, Heckl R, Goebel HH, Meinck HM. Inclusion body miositis presenting with isolated erected spinae paresis. *Neurology* 1995;45(5):993-4. https://doi.org/10.1212/wnl.45.5.993
25. Suzuki T, Matsuzaka H. Drug-induced Pisa syndrome (pleurothotonus). *Mol Diag Ther* 2002;16(3):165. https://doi.org/10.2165/00023210-200216030-00003
26. Delcey V, Hachulla E, Michon-Pasturel U, Queyrel V, Hatron PY, Boutry N, et al. La camptocormie: un signe de myopathie axiale. À propos de sept observationes. *Rev Med Interne* 2002;23(2):144-54. https://doi.org/10.1016/S0248-8663(01)00530-6

27. Yorikata A, Shimo Y, Takanashi M, Fukae J, Hatano T, Nakahara T, et al. Motor and non-motor symptoms of 1453 patients with Parkinson's disease: prevalence and risks. *Parkinsonism Relat Disord* 2013;19(8):725-31. https://doi.org/10.1016/j.parkreldis.2013.04.001
28. Spuler S, Krug H, Klein C, Medialdea IC, Jacob W, Ebersbach G, et al. Myopathy causing camptocormia in idiopathic Parkinson's disease: a multidisciplinary approach. *Mov Disord* 2010;25(5):552-9. https://doi.org/10.1002/mds.22913
29. Tiple D, Fabbrini G, Colosimo C, Ottaviani D, Camerota F, Defazio G, et al. Camptocormia in Parkinson disease. *J Neurol Neurosurg Psychiatry* 2009;80(2):145-8. https://doi.org/10.1136/jnnp.2008.150011
30. Dietz V, Zijlstra W, Assaiante Ch, Trippel M, Berger W. Balance control in Parkinson´s disease. *Gait Posture* 1993;1(2):77-84. https://doi.org/10.1016/0966-6362(93)90018-V
31. Jankovic J, Tintner R. Dystonia and parkinsonism. *Parkinson Dis Relat Disord* 2001;8(2):109-21. https://doi.org/10.1016/S1353-8020(01)00025-6
32. Bloch F, Houeto JL, Tezena du Montcel S, Bonneville F, Etchepare F, Welter ML, et al. Parkinson´s disease with camptocormia. *J Neurol Neurosurg Psychiatry* 2006;77(11):1223-8. https://doi.org/10.1136/jnnp.2006.087908
33. Babat L, McLain R, Bingaman W, Kalfas I, Young P, Rufo Smith C. Spinal surgery in patients with Parkinson's disease: construct failure and progressive deformity. *Spine (Phila Pa 1976)* 2004;29(18):2006-12. https://doi.org/10.1097/01.brs.0000138306.02425.21
34. Koller H, Acosta F, Zenner J, Ferraris L, Hitzl W, Meier O, et al. Spinal surgery in patients with Parkinson's disease: experiences with the challenges posed by sagittal imbalance and the Parkinson´s spine. *Eur Spine J* 2010;19(20):1785-94. https://doi.org/10.1007/s00586-010-1405-y
35. Micheli F, Pardal M. Dopa-responsive dystonic camptocormia. *Neurology* 2007;68(18):1543. https://doi.org/10.1212/01.wnl.0000265316.61721.63
36. Wunderlich S, Csoti I, Reiners K, Günthner-Lengsfeld T, Schneider C, Becker G, et al. Camptcormia in Parkinson's disease mimicked by focal myositis of the paraspinal muscles. *Mov Disord* 2002;17(3):598-600. https://doi.org/10.1002/mds.10110
37. Schulz-Schaeffer W, Margraf N, Munser S, Wrede A, Buhmann C, Deuschl G, Oehlwein C. Effect of neurostimulation on camptocormia in Parkinson's disease depends on symptoms duration. *Mov Disord* 2015;30(3):368-72. https://doi.org/10.1002/mds.26081
38. Skidmore F, Mikolenko I, Weiss H, Weiner W. Camptocormia in patients with multiple system atrophy. *Mov Disord* 2005;20(8):1063-4. https://doi.org/10.1002/mds.20521
39. Gavrylova N, Limousin N, Belin J, Praline J. Camptocormia as presenting sign in dementia with Lewy bodies. *Clin Neurol Neurosurg* 2013;115(11):2397-8. https://doi.org/10.1016/j.clineuro.2013.09.005
40. Gautier G, Verschueren A, Monnier A, Attarian S, Salort-Campana E, Pouget J. ALS with respiratory onset: clinical features and onset of non-invasive ventilation on the prognosis. *Amyotroph Lateral Scler* 2010;11(4):379-82. https://doi.org/10.3109/17482960903426543
41. Melamed E, Djaldetti R. Camptocormia in Parkinson´s disease. *J Neurol* 2006;253(Suppl 7):VII14-6. https://doi.org/10.1007/s00415-006-7004-5
42. Barkhaus PE, Periquet MI, Nandedkar SD. Quantitative motor unit action potential analysis in paraspinal muscles. *Muscle Nerve* 1997;20(3):373-5. https://doi.org/10.1002/(SICI)1097-4598(199703)20:3<373::AID-MUS19>3.0.CO;2-1
43. Haig AJ, Tong HC, Kendall R. The bent spine syndrome: myopathy + biomechanics = symptoms. *Spine J* 2006;6(2):190-4. https://doi.org/10.1016/j.spinee.2005.08.002
44. Margraf NG, Rohr A, Granert O, Hampel J, Drews A, Deuschl G. MRI of lumbar trunk muscles in patients with parkinson´s disease and camptocornia. *J Neurol* 2015;262(7):1655-64. https://doi.org/10.1007/s00415-015-7726-3
45. Ohana M, Durand MC, Marty C, Lazareth JP, Maisonobe T, Mompoint D, et al. Whole body muscular RMN to detect myopathies in non-extrapyramidal bent spine syndrome. *Skeletal Radiol* 2014;43:1113-22. https://doi.org/10.1007/s00256-014-1909-3
46. Yamada K, Shinojima N, Hamasaki T, Kuratsu J. Subthalamic nucleous stimulation improves Parkinson's disease-associated camptocormia in parallel to its preoperative levodopa responsiveness. *J Neurol Neurosurg Psychiatry* 2016;87(7):703-9. https://doi.org/10.1136/jnnp-2015-310926

47. Mensikova K, Kaiserova M, Vastik M, Kurkova S, Kanovsky P. Treatment of camptocormia with continuous subcutaneous infusions of apomorphine: 1-year prospective pilot study. *J Neural Transm (Vienna)* 2015;122(6):835-9. https://doi.org/10.1007/s00702-014-1297-9. **Erratum:** *J Neural Transm (Vienna)* 2015;122(9):1353. https://doi.org/10.1007/s00702-015-1385-5
48. Kataoka H, Tonomura Y, Eura N, Terashima M, Kawahara M, Ueno S. Painful abdominal contractions in patients with Parkinson disease. *J Clin Neurosci* 2012;19(4):624-7. https://doi.org/10.1016/j.jocn.2011.06.026
49. Bertram KL, Stirpe P, Colosimo C. Treatment of camptocormia with botulinum toxin. *Toxicon* 2015;107(Pt A):148-53. https://doi.org/10.1016/j.toxicon.2015.06.004
50. Deuschl G, Schade-Brittinger C, Krack P, Volkmann J, Schafer H, Botzel K, et al. A randomized trial of deep-brain stimulation for Parkinson´s disease. *N Engl J Med* 2006;355(9):896-908. https://doi.org/10.1056/NEJMoa060281
51. Bathia K, Quinn N, Mardsen C. Clinical features and natural history of axial predominant adult onset primary dystonia. *J Neurol Neurosurg Psychiatry* 1997;63(6):778-91. https://doi.org/10.1136/jnnp.63.6.788
52. O'Riordan S, Paluzzi A, Liu X, Aziz T, Nandi D, Bain P. Response to bilateral globus pallidus internal stimulation in three patients. *Mov Disord* 2009;24(Suppl. 1):1-653 p489. https://doi.org/10.1002/mds.22628
53. Dominik J, Sheenan G, Schleimer J, Wixom C. Response to the dropped hear/bent spine syndrome to treatment with intravenous immunoglobulin. *Muscle Nerve* 2006;33(6):824-6. https://doi.org/10.1002/mus.20506
54. Benveniste O. Inclusion-body myositis. *Rev Med Interne* 2014;35(7):472-9. https://doi.org/10.1016/j.revmed.2013.09.001
55. de Sèze M, Creuze A, Mazaux J. An orthosis and physiotherapy programme for camptocormia: a prospective case study. *J Rehabil Med* 2008;40(9):761-5. https://doi.org/10.2340/16501977-0252
56. Capecci M, Serpicelli C, Fiorentini L, Censi G, Ferretti M, Orni C, et al. Postural rehabilitation and Kinesio taping for axial postural disorders in Parkinson´s disease. *Arch Phys Med Rehabil* 2014;95(6):1067-75. https://doi.org/10.1016/j.apmr.2014.01.020
57. Upadhyaya C, Starr P, Mummaneni P. Spinal deformity and Parkinson disease: a treatment algorithm. *Neurosurg Focus* 2010;28(3):e5. https://doi.org/10.3171/2010.1.FOCUS09288