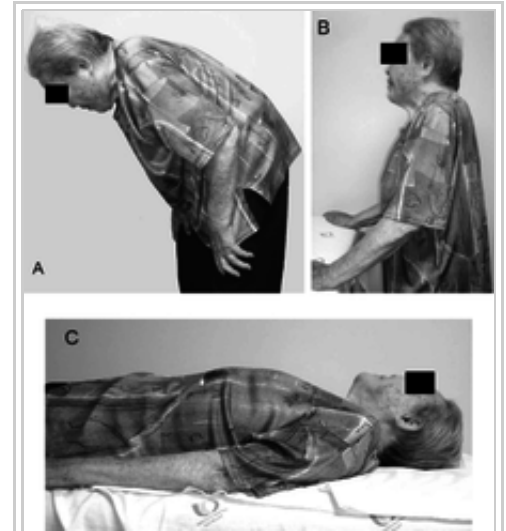


Camptocormia

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Camptocormia, also known as bent spine syndrome (BSS), is a symptom of a multitude of diseases that is most commonly seen in the elderly. It is identified by an abnormal thoracolumbar spinal flexion, which is a bending of the lower muscles of the back, occurring in a standing position. In order to be classified as BSS, the anterior flexion (the lower back bending) must be of 45 degrees anteriorly. This classification differentiates it from a similar syndrome known as kyphosis.^[2] Although camptocormia is a symptom of many diseases, there are two common origins: neurological and muscular. Camptocormia is treated by alleviating the underlying condition causing it through therapeutic measures or lifestyle changes.



A clinical photograph showing a patient with the flexed posture. It can be abated when laying down.^[1]

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History and society

Camptocormia comes from two Greek words meaning ‘bent’ (Camptos) and “trunk” (Kormos) and was coined by Alexandre-Achille Souques and B. Rosanoff-Saloff. These two men also created the definition of the disease that is widely accepted today.^[2]

When the disorder was first clinically studied around the time of First World War, it was believed to be a psychogenic conversion disorder that resulted from the server trauma of war. Souques et al. treated patients with psychological treatment and early versions of electrotherapy. Samuel A. Sandler used a similar approach to treat soldiers during the Second World War.^[2] The view of BSS as a conversion disorder led to a lack of awareness about the conditions and few diagnoses by physicians.

As time progressed and advances were made in our knowledge of neuroscience and physiology, biological mechanisms behind the irregular bending were identified. The current medically preferred term for the condition is Bent Spine Syndrome due to the psychological origin associated with camptocormia.^[2]

Camptocormia in the elderly

Bent Spine Syndrome is not limited to the elderly, but has an average age of onset of 66 years^[2] and is more common amongst men. This late age of onset is largely due to the increased preponderance of the conditions causing the symptom in older individuals- such as muscular weakness and neurological disorders like Parkinson's Disease. While BSS doesn't have any negative stigma in and of itself, those affected by it may be perceived differently due to the condition. For example, an elderly individual afflicted with the condition is viewed as very physically weak due to the severe bending of the back caused by the condition.

Characteristics and symptoms

The primary symptom of camptocormia is abnormal forward bending of the torso. This bending becomes worse while walking but does not appear when the affected individual is laying down in a horizontal position. This alleviation of the condition indicates that it is a manifestation of another disease or ailment and is not due to a spine that is actually bent.^[2] This is somewhat ironic due to the fact that the medically accepted name for the condition is Bent Spine Syndrome.

In an afflicted individual, the abnormal bending consists of an anterior flexion greater than 45 degrees.^[3] Due to this bending and the physical limitations caused by the conditions associated with the disease, it is usually impossible for an afflicted person to achieve a fully erect position. In addition, patients suffering from camptocormia often experience lower backpain due to the condition. BSS often appears in individuals afflicted with Parkinson's disease, muscular

dystrophies, endocrine disorders inflammatory conditions (myositis), or mitochondrial myopathies.^[1] As previously mentioned, the disease is more common in older individuals.

Pathology

When initially identified, Camptocormia was classified as a psychogenic disease. Although the condition is sometimes a psychogenic manifestation, camptocormia typically originates from either muscular or neurological diseases. However due to the wide variety of pathologies resulting in camptocormia, there is no singular cause that is most influential for the condition.

Muscular origin

Myopathic origin BSS can be secondary to various muscular disorders or occur as a primary idiopathy.^[2] These etiologies are termed secondary and primary BSS respectively. Idiopathic primary BSS is a late onset myopathy with progressive muscular weakness that is detected on the spinal extensor muscles in the elderly patients and is



Alexandre Achille Souques was one of the first major researchers of Camptocormia and created the definition and name used for the condition to this day

more predominant in females.^[2] The pathogenesis of primary BSS is typically related to fibrosis and fatty infiltration of muscular tissues and mitochondrial changes due to the aging process.^[2] Specifically, weakening of muscles occurs in the paravertebral muscles of patients. These paravertebral muscles have a great influence over the walking stance and gait of a patient, so fatty infiltration and degradation of these muscle lead to the characteristics that easily define BSS such as the anterior flexion of the back, but being able to keep themselves upright with any kind of support (e.g. holding onto a table).^[3]

Secondary BSS can have a multitude of causes making it hard to pinpoint to a specific muscular disorder. Some examples of diseases that have secondary BSS as a symptom being myopathy caused by muscular dystrophies, neuromuscular disorders, inflammatory muscle diseases, metabolic or endocrine disorders, and mitochondrial myopathies.^[2] A muscle biopsy would be able to clearly demonstrate whether Primary BSS or Secondary BSS is afflicting the patient due to primary BSS being much more identifiable.

Neurological origin

A multitude of neurological disorders cause BSS, including motor neuron disease, CNS disorders, and early amyotrophic lateral sclerosis.^[2] Usually, the bent spine is caused by dysfunctioning extensor spinal muscles with a neurological cause.

Neurological origin BSS may also result from damage to the basal ganglia nuclei that are a part of the cerebral cortex which play a major role in bodily positioning. Damage to this part of the brain can inhibit proper flexion and extension in the muscles necessary for maintaining an upright position. Additionally, the neurotransmitter dopamine plays a key role in the operation of basal ganglia. Abnormally low dopamine concentrations, such as that associated with Parkinson's disease, cause dysfunction in the basal ganglia and its associated muscle groups, leading to BSS.^[2] Studies have estimated the prevalence of BSS in people affected by Parkinson to be between 3–18%.^[1]

Gene mutations

Several gene mutations have been identified in patients with camptocormia. These including RYR1 gene in axial myopathy, DMPK gene in myotonic dystrophy, and genes related to dysferlinopathy and Parkinson's disease. These genes could serve as targets for gene therapy to treat the condition in the years to come.^[4]

Diagnosis

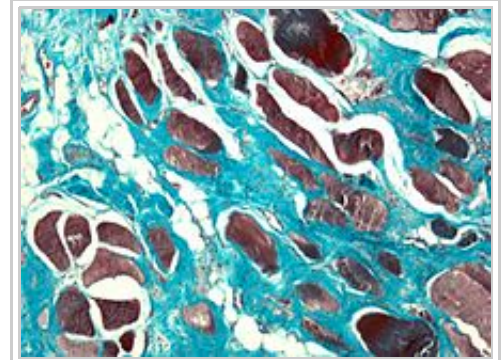
In order to qualify a patient's condition as BSS, the bending angle must be greater than 45 degrees. While the presence of the condition is very easy to note, the cause of the condition is much more difficult to discern. Conditions not considered to be BSS include vertebrae fractures, previously preexisting conditions, and ankylosing spondylitis. Lower back CT scans and MRIs can typically be used to visualize the cause of the disease.^[4] Further identification^[4] of the cause can be done by histochemical or cellular analysis of muscle biopsy.

Camptocormia is becoming progressively found in patients with Parkinson's disease.^[3] The diagnosis of Parkinson's associated camptocormia includes the use of imaging of the brain and the spinal cord, along with electromyography or muscle biopsies. As well as this, due to the nature of Parkinson's disease and the

symptoms it creates.

Muscle biopsies are also a useful tool to diagnose camptocormia. Muscle biopsies found to have variable muscle fiber sizes and even endomysial fibrosis may be markers of bent spine syndrome. In addition, disorganized internal architecture and little necrosis or regeneration is a marker of camptocormia.

Patients with camptocormia present with reduced strength and stooped posture when standing due to weakened paraspinal muscles. Clinically, limb muscles showed fatigue with repetitive movements.^[4] Paraspinal muscles, muscle parallel to the spine, underwent fat infiltration, which is the accumulation of substances within the tissue and cells in excess amounts. Electromyography may be used as well in diagnosis. On average, the paraspinal muscles of affected individuals were found to be seventy-five percent myopathic while limb muscles were to be fifty percent myopathic.^[4] Creatine Kinase activity levels in skeletal muscle can also be used as a diagnostic indicator that can be identifiable through blood tests.



Spinal muscle biopsy showing intense endomysial deposit of fibrosis (green) and fatty infiltration. We can also observe irregular distribution of involuted muscle fibers. (Trichrome, original magnification 9100)^[2]

Treatment and prognosis

Treatment and management

Due to the wide range of causes of Camptocormia, there is no one treatment that suits all patients. In addition, there is no specific pharmacological treatment for primary BSS. The use of analgesic drugs depend entirely on the intensity of the back pain. Muscular origin BSS can be alleviated by positive lifestyle changes including physical activity, walking with a cane, a nutritious diet, and weight loss.

Worsening of symptoms is possible but rare in occurrence.^[2]

Treatment of the underlying cause of the disease can alleviate the condition in some individuals with secondary BSS. Other treatment options include drugs, injections of botulinum toxin, electroconvulsive therapy, deep brain stimulation, and surgical correction.^[5]

Unfortunately, many of the elderly individuals affected by the BSS are not treated surgically due to age related physical ailments and the long postoperative recovery period.^[3]

Outcome and prognosis

This disease can lead to excess pressure on the spine, causing pain and discomfort.^[4] If the spine is bent too far, a patient may have difficulties breathing in the future because of the pressure of the spine pressed against the lungs. Camptocormia may also lead to aging, muscle weakness in the upper back, arthritis and other bone degeneration diseases.^[4] Due to loss of bone strength, injury to the spine and slipped discs become increasingly



Thymatron IV used for electroconvulsive therapy

significant. Camptocormia can lead to infection, tumors, diseases of the endocrine system and connective tissues. The success of the treatment method is largely dependent on the patient, but response to therapeutic methods is generally low.

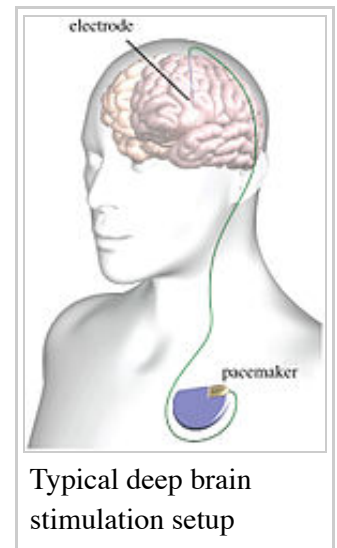
Research directions

Clinical studies reveal the heredity of camptocormia; however, the inheritance mechanism remains unclear.^[2] Current areas of research include molecular and genetic studies aimed at elucidating a possible inheritance model along with molecular pathological mechanisms, and proteins responsible for BSS. This research will help will facilitate improvement in the classification, diagnosis, and treatment of the condition. In addition, new technologies and animal models of postural abnormalities are being developed to understand camptocormia and design more effective treatment methods.^[4]



Deep brain stimulation

One treatment methodology that is very promising for the treatment of Camptocormia is Deep Brain Stimulation. Previously, deep brain stimulation, along with bilateral stimulation of the subthalamic nucleus and/or globes pallidus internus have been

used to treat patients with Parkinson's disease.^[5] Studies have shown that these similar treatments could be used on patients with severe Camptocormia. Using the Burke-Fahn-Marsden dystonia rating scale before and after treatment was used on patients of camptocormia; it was found that both patients found significant functional improvement and the ability to walk.^[5]



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