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Axial myopathies: an elderly disorder

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The term axial myopathy is controversial. For some (1), the disorder is caused by a myopathic condition with generalized involvement of the axial musculature, although, clinically, weakness is predominant at either the cervical or thoracic level. For others, bent spine syndrome and dropped head syndrome are very separate diseases (2).

Even if the names of the two syndromes are different, bent spine is often quoted as *camptocormia* (from the Greek *camptos* meaning bent and *cormos* meaning tree trunk) or reducible kyphosis or proclinospine, or para-vertebral myopathy. On the other hand, dropped head syndrome corresponds to floppy head syndrome (3) or to isolated neck extensor myopathy (4). Some recent reviews have focused on *camptocormia* and head drop (5, 6).

It, therefore, seemed worthwhile to take a closer look at these quite unusual syndromes, especially bent spine syndrome: Is trunk extensor muscle weakness an elderly disorder? Are there specific clinical signs? Are ancillary examinations useful? What muscles are involved? Pathophysiology? Primary *vs*. secondary syndromes? Are dropped head and bent spine the same disease? Are they familial cases?

1. Bent spine syndrome is undoubtedly a late disease: mean age at onset 62.08 years, mean age at first examination 66.9 years. Male sex is affected in 60% of cases.

2. Semiology of bent spine syndrome is characteristic. An erect position is not possible. An abnormal stooping posture is found, often progressive during examination. *Camptocormia* is exaggerated on walking and, in some patients, begins upon exercise. On the other hand, the forward attitude disappears in a supine position that is different from simple dorsal kyphosis. Extension of the neck is always possible and the neck muscles are normal. Patients complain of transient low back pain but extensor *spinae* muscles are not painful. "Fold sign" is usual, due to atrophy of the para-spinal muscles A dorsal vertical fold appears between the scapulae during the erect position, then disappears.

3. Complementary methods. Muscle computed tomography (CT) scan and magnetic resonance imaging (MRI) are usually abnormal and informative (7). Para-spinal muscle biopsy is technically not easy and the interpretation is sometimes difficult: the muscle specimen is often too small. Serum creatine kinase is normal or moderately elevated. Para-spinal muscle electromyography shows non-specific or slight abnormalities suggesting a myopathic, rather than a neurogenic disorder. The appearance of abnormal spinal muscle on CT scan is characteristic: empty muscle with normal outlines, sometimes slightly flattened. Hypodensity of the muscle is constant. There is a loss of substance, the muscle seems to be washed out, but the volume is usually preserved. Coronal MRI is significant: vertical para-spinal muscles become thinner and disappear with a striking rarefaction of muscle *fasciculi* particularly in comparison to controls. The most important change in para-spinal muscle biopsy is fibrosis and adiposis. Also found are necrosis and regeneration, variations in fibre size, type II fibres atrophy. The pattern of muscle biopsy is rather myogenic. Inflammatory lesions are not unusual but do not imply focal myositis. Likewise, mitochondrial abnormalities – probably related to age – are frequent: ragged red fibres and complex I and III deficiency. Therefore, primary *camptocormia* is a para-vertebral myopathy (8).

4. The muscles involved are divided into two groups. Their function is different. The deep layer is comprised of *multifidi*, short oblique muscle extensors, but also rotators. The superficial layer includes long muscles: *longissimus dorsalis*, *ilio-dorsalis lumborum*, *spinalis thoracis*. These are extensor *spinae*. These muscle masses occupy the space between the transverse and spinous processes on both sides of the vertebral column. Electromyography shows that no activity is required during standing erect for *erector spinae* except for forced extension. *Erector spinae* become active during forward flexion, except for very rapid flexion, similar to a fall. The *erector spinae* come into action when the trunk assumes an erect position. However, while standing erect, normal subjects require very slight activity, sometimes reflex of some intrinsic muscles of the back, probably intermittent during a shift of the centre of gravity. At the same time, *longissimus dorsalis* is the origin of continuous monitoring. Finally, activity is controlled due to force of gravity between sets

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of muscles: the short deep muscles (*multifidi*) stabilize the individual inter-vertebral joints, *longissimus* and the abdominal muscles stabilize the spine as a whole.

5. Pathophysiology. There is a destabilization in the complex interaction of spinae muscles. Several parameters are involved (9).

Muscular parameters: There is a weakness of the *erector spinae* muscles, prime movers. Electromyographic activity normally when body bending is reduced. *Multifidi,* essentially stabilizers and rotators, are also weak. Likewise, for abdominal muscle stabilizers, but also *flexores* and *rotatores* of the spine (obliquous *externus* and *internus*, transverse *abdominis*, *rectus abdominis*). Neck extensor muscles, on the other hand, are normal.

Postural parameters: The permanent stabilization of the trunk is impaired. The resistance to gravity, continuously monitored by *longissimus* is modified, likewise the intermittent activity of other muscles when there is a shift in the centre of gravity. There is an increased instability of stance when leaning forward in addition to dyssynergy due to elderly age: decrease in stride length and walking velocity, increase in stride duration and double-support duration (10).

Inter-limb coordination parameters: Relationships between normal head movements and trunk weakness are decreased. Likewise, diagonal backward displacement of body mass centre, after hand and leg force, is impaired.

6. Primary *vs*. secondary bent spine syndrome. Primary syndromes are typical in their semiology. A not unusual form appears at the onset of exercise and disappears at rest. This exertional *camptocormia* has the same features upon examination. The loss of para-vertebral muscles is less obvious and the fold size is sometimes absent. In some primary syndromes, the pathology is slightly modified. In addition to fibrosis, adiposis and necrosis, there are either inflammation or ragged red fibres. Myoadenylate deficiency is not infrequent. The main secondary syndromes are observed in hysteria (world war neurosis), in Parkinson's disease, in neonatal hypotonia, in phosphorylase or carnitine deficiency, and in neuromuscular diseases (facioscapulohumeral dystrophy, inclusion body myositis, motor neuron disease, in particular the Vulpian type. An interesting and under-diagnosed form is proximal myotonic myopathy (PROMM) (11) with progressive painful para-spinal muscle weakness exaggerated by exercise, slight myotonia, cataract, high gamma GT level, angular fibres, type II atrophy on muscle biopsy.

7. Dropped head and bent spine syndrome are separate entities. According to some Authors (1), these two diseases are different manifestations of axial myopathy and have a similar aetiology. Both syndromes are late manifestations of a non-specific myopathic disease, restricted to para-spinal musculature. Although clinical involvement is more or less limited to one level, it seems, to these Authors, that it is a myopathy affecting mainly axial musculature. In fact, many clues favour the notion of two separate diseases (Table I). Muscles involved in dropped head are different with three layers: First: splenius, trapezius; Second: *semispinalis capitis*, *semispinalis cervicis*, *longissimus dorsalis*; Third: *rectus posterior major, rectus posterior minor, obliquus superior*. Pathophysiologically phasic muscles are more involved in dropped head, the course is more chronic. The main secondary forms are spasmodic antecolis, dermatomyositis, myotonic dystrophy, amyotrophic lateral sclerosis. Familial cases seem exceptional.

8. Familial cases. These are not infrequent in bent spine syndrome (10 cases). Inheritance is autosomal dominant. Onset is late. Low back pain is common. Serum creatine kinase levels are normal. Electromyography is myopathic. There are no specific changes in muscle bi-

Table 1. Differences between dropped head and bent spine syndrome.

	Dropped Head	Bent spine
Primary	Rare	Not infrequent
Muscles	Neck extensors	Erector ***spinae***
Pathophysiology	Phasic > tonic	Phasic + tonic
Muscle CT Scan	Hypodensities	Myopathic
Coronal MRS	?	Loss of paraspinal muscles
Muscle biopsy	Inflammatory, IBM	Rather myogenic
Onset	Subacute	Insidious
Outcome	Unchanged or Improved	No improvement
Handicap	Moderate	Stick or wheelchair
Secondary	Systemic and Multisystemic disease	NMD
Familial case	Exceptional	Some but no informative

opsy. The course is slowly progressive. Unfortunately, a genetic study was not possible because of the late onset and the elderly age of some family members.

In conclusion, primary bent spine syndrome is a primary progressive axial muscle disease, sporadic or familial late onset myopathy or related to aging. It is a relentless disease with no response to treatment-drugs, rehabilitation, surgery. It is different from neck muscle weakness.

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