

Myopathies of the trunk

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Disclosures

- None

Learning objectives

- The symptomatology associated with truncal (axial) muscle weakness
- The associated signs
- The major disorders in which truncal weakness is a common component
- The major disorders that may present with truncal weakness
- Management options

Key message

- In patients with isolated truncal weakness a specific underlying cause may not be found

Myopathies of the trunk

Introduction

Weakness of the paraspinal muscles may predominantly affect the anterior or the posterior muscles

Anterior muscle weakness

Difficulty raising head when lying

Posterior muscles

Head drop

Bent spine (camptocormia)

Myopathies of the trunk

- Causes of “bent spine”
 - Idiopathic scoliosis
 - Non-neuromuscular
 - Spinal disease
 - Parkinsonism and related disorders
 - Non-myopathic
 - Myasthenia gravis
 - ALS/MND
 - CIDPN

Myopathies of the trunk

- Weakness of neck flexion
 - Common in
 - inflammatory myopathies
 - Myasthenia
 - Myotonic dystrophy
 - Absent in many dystrophies

Myopathies reported to cause bent spine syndrome/dropped head syndrome

- IIM
- IBM
- FSHMD
- RYR1
- Myofibrillar
- Metabolic
- Primary amyloidosis
- FHL1
- Late-onset nemaline myopathy

Diagnostic issues

- Additional clinical features paramount
- Muscle biopsy occasionally helpful
- EMG rarely of diagnostic value

Management

- Difficult, and essentially only supportive measures available

References

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Camptocormia as presenting manifestation of a spectrum of myopathic disorders

Muscle & Nerve 2015 (In press)

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Sporadic inclusion body myositis presenting with severe camptocormia

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