

# Muscle disorders in the elderly

Myopathy is the term used to describe diseases of the muscle that are not attributable to nerve dysfunction. It is an umbrella term that encompasses myopathies that are inherited and those that are acquired. Here, we consider the common myopathies that can occur in later life. We discuss the clinical manifestations, investigation and management of drug-induced, toxic and endocrine myopathies as well as the inflammatory muscle disorders. We also consider the inherited muscle disorders that can present in older patients, including myotonic and oculopharyngeal dystrophy, and facioscapulohumeral. We briefly discuss the neuromuscular disorder myasthenia gravis, which whilst not a primary disorder of muscle, has many overlapping features of a myopathy.

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The diagnosis of muscle disorders in the elderly presents a clinical challenge. This is not only because many of these conditions are complex and overlapping, but also due to a misguided acceptance among many clinicians that deterioration in function and muscle strength among the elderly is usually a result of the “ageing” process rather than a pathological disease entity.

The importance of reaching the correct diagnosis is apparent for a number of reasons. Many muscular disorders presenting in the elderly are acquired<sup>1</sup> and therefore potentially reversible. The complications of such disorders, including recurrent falls and aspiration, have significant morbidity and mortality and in the case of certain inherited disorders presenting in the elderly, the potential implications for future generations in establishing a correct diagnosis are vitally important.<sup>2</sup> In the following article, we present a brief overview of muscle diseases affecting the elderly, highlighting typical presenting features and diagnostic

investigations. We will also discuss some rare neuromuscular problems presenting for the first time in later life.

## Muscle disorders

### Myopathy

Symptoms suggestive of a myopathy presenting in an older person are similar to those presenting at any age and weakness is usually the predominant feature. This weakness is commonly proximal and presents as difficulty rising from a chair or climbing the stairs. This may be due to the increased eccentric contractions of proximal muscles as part of their antigravity role. These eccentric contractions have been shown to be more greatly affected when the muscle becomes weaker. However, distal weakness is also recognised (often in inherited conditions) and in this case, the patient may present with problems when writing, or with hand grip or grasp.<sup>2</sup>

Patients also complain of malaise and may experience a fever. Discolouration of the

urine can occur, and represents rhabdomyolysis.<sup>3</sup> Sensory symptoms are not associated with pure myopathies and reflexes are preserved until very late in the disease process, although in older patients with multiple comorbidities such distinctions are often difficult to demonstrate clinically. Muscle atrophy is also generally a late sign.<sup>4</sup>

A clue to the aetiology of a new onset myopathy may be indicated from the time course of the illness. Weakness over a period of hours may suggest a toxic cause whereas that presenting over days is more likely to represent an acute dermatomyositis or rhabdomyolysis. Symptoms developing over weeks or months may be a result of an endocrine disorder, polymyositis or drugs, and that occurring over months to years may indicate a hereditary illness.<sup>5</sup>

The investigations needed to confirm a diagnosis and identify a likely cause for an individual myopathy will depend on the clinical picture. These will include assessments of full blood count, renal and liver function, and thyroid function as well as calcium,

magnesium and phosphate, creatine kinase (CK) and erythrocyte sedimentation rate (ESR).

Urine dipstick is also vital in every case to look for myoglobinuria. Autoantibodies may also be requested. Electrophysiology can be helpful. Electromyography (EMG), which can localise the pathology to the muscle rather than the nerve or anterior horn cell, is widely used to aid diagnosis. EMG findings will depend on the cause of the myopathy, but hallmark changes include low amplitude, polyphasic, brief duration potentials with voluntary contraction. Muscle biopsy is often used if an inflammatory disorder is postulated and MRI can also be helpful in this regard. Genetic testing may be offered if a muscular dystrophy is a possibility. Mild cases progress slowly and the patient may have a normal life span. In such cases, the patient may go undiagnosed until later life.

### Drug-induced myopathy

A number of common and widely-prescribed drugs have been identified as causing features of myopathy which can either be fleeting or progressive, painful or painless and, in severe cases, can cause rhabdomyolysis and renal failure.<sup>3</sup> It is well known that statin therapy can cause such effects,<sup>6</sup> but other commonly used drugs, including steroids, beta-blockers, and a host of other cardiac drugs, can cause myopathy.<sup>4</sup> Such drugs used in combination can increase the risk of such effects. Hypokalaemia, a common side-effect of many diuretics, can also produce a myopathy.

Patients presenting with a drug-induced myopathy tend to have proximal weakness, which

in severe cases can progress to muscle atrophy. CK is usually elevated in statin-induced as well as other painful myopathies but may be normal. Treatment involves removal of the offending drug, which usually results in good recovery.<sup>4</sup>

### Toxic myopathy

Toxic myopathy encompasses not only alcohol-induced disease but also myopathy associated with drugs of abuse. Although drug abuse is rare amongst the elderly, it is worth remembering that heroin, methadone and cocaine can all induce symptoms of muscle weakness and rhabdomyolysis.<sup>4</sup> Alcohol abuse among the elderly, however, is not rare and this can cause both an acute as well as a chronic myopathy. Acute myopathy may follow an alcoholic binge<sup>4</sup> or can be secondary to hypokalaemia associated with vomiting and can usually be reversed by removing the toxin or by potassium replacement.<sup>4</sup> In the chronic form associated with long-standing alcoholism, where there is proximal weakness and wasting of the shoulder and pelvic girdle, effects are often difficult to reverse.<sup>3</sup> CK levels may or may not be elevated and patients may not offer an alcohol history. In such situations, a collateral history may be useful and other clues should be sought including macrocytosis and deranged LFT's.

### Endocrine myopathies

Endocrine dysfunction is common in later life and a number of these disorders can present with, or have been linked to, muscle dysfunction.

Both hyper- and hypothyroidism can present with features of proximal muscle weakness and myopathy, the latter

being more common and more severe in hyperthyroidism. In hypothyroidism, the CK is often elevated and, if there is clinical suspicion, other signs suggestive of the diagnosis should be sought. In the case of thyrotoxicosis, CK is often normal. In both conditions, symptoms usually resolve on treating the underlying disorder.<sup>4</sup>

Patients with Cushing's syndrome commonly have symptomatic myopathy at presentation. Weakness is proximal and symmetrical, and there is often muscle pain. Other features suggestive of the diagnosis should be sought including truncal obesity, purple striae, moon face and supraclavicular fat pads.<sup>10</sup> Patients with a steroid-induced Cushing's syndrome are most at risk of developing muscular symptoms. In these patients, CK levels may be normal and EMG studies are usually non-specific. A muscle biopsy may help in distinguishing a steroid-induced myopathy from an inflammatory illness and treatment involves removal of the offending drug.<sup>4</sup>

Disease processes involving the utilisation and metabolism of vitamin D and calcium are also common amongst the elderly and can similarly affect muscle strength.

In patients with osteomalacia, there will be difficulty rising from a chair due to proximal muscle weakness. Patients have waddling gait and often bone pain. Patients with osteomalacia are likely to have hypocalcaemia and hypophosphataemia but CK levels may be normal and EMG studies are of little benefit. Recovery of muscle strength may take some weeks or months following treatment with vitamin D3.

Conversely patients with



hypercalcaemia as a result of primary hyperparathyroidism can also present with myopathies although these will often be late in the illness. The mechanism of weakness is not known, although CK is usually normal and treatment of the disease process will provide symptomatic improvement. Other endocrine causes of muscle weakness in the elderly include Addison's disease as well as a small number of other disorders of carbohydrate metabolism presenting in late adulthood, such as maltase deficiency, although these are exceedingly rare.

### Inflammatory muscle disorders

The idiopathic inflammatory muscle disorders occurring most commonly

in older people are inclusion body myositis, dermatomyositis and polymyositis.

#### Inclusion body myositis

Inclusion body myositis (IBM) is a relatively rare disorder with a prevalence of 51.3 per million population in those over 50 years of age.<sup>7</sup> However it is a disease predominantly affecting the elderly. IBM presents with a combination of proximal and distal weakness characteristically affecting the quadriceps and long flexors of the fingers, and a profound weakness of the forearms can occur. The weakness is usually asymmetrical and can also affect flexion of the neck. Dysphagia associated with the condition is the main cause of morbidity and mortality in IBM.<sup>8</sup>

It is not clear what causes IBM

but there is evidence that a T-cell mediated response may be involved along with accelerated degeneration in the muscle.<sup>2</sup> Diagnosis is by means of nerve conduction studies, measurement of CK and muscle biopsy, which is often diagnostic. Steroids do not alter the progress of this condition and loss of mobility is usual within 10 years of diagnosis.<sup>8</sup>

#### Dermatomyositis

Dermatomyositis is an autoimmune disorder which has two peak incidences: one in childhood and a second in later life. It presents with a symmetrical proximal muscle weakness, pain, dysphagia and often characteristic skin rashes. These include a scaly eruption over the metacarpophalangeal joints known as Gottrons papules and a purple or 'heliotrope' rash over the eye lids. There is often a 'V'

shaped photosensitive rash over the anterior chest wall.

Dermatomyositis may be associated with internal malignancy in as many as 20% of cases and this is particularly common in the elderly.<sup>4</sup> Therefore, it is vital to examine the patient thoroughly, and consideration should be given to further investigation of specific malignancies depending on the history given.

The “gold standard” investigation is muscle biopsy although CK can help with disease monitoring.

High-dose steroids, as well as steroid-sparing agents, are employed in the treatment of this disorder. Immunoglobulins and monoclonal antibodies have also been used successfully.<sup>9</sup>

### Polymyositis

Polymyositis holds many similarities with dermatomyositis, and these may be considered overlapping disorders. Polymyositis is also thought to be an autoimmune disorder which presents with proximal muscle weakness but unlike dermatomyositis, no link with malignancy has been identified. It is thought the disorder may be triggered by certain infections, including human T-cell leukaemia virus 1 (HTLV1) and human immunodeficiency virus (HIV) or by certain drugs such as D penicillamine.<sup>10</sup> Polymyositis causes weakness in the pelvic and shoulder girdles and may limit functionality, particularly for elderly people. Dysphagia is common, occurring in up to one third of patients and patients may have signs of associated connective tissue disorders such as Sjogren’s syndrome.<sup>4</sup>

Diagnosis and treatment is as for dermatomyositis, discussed above.

## Inherited disorders of the muscle

### Myotonic dystrophy

Myotonic dystrophy is a common autosomal dominant disorder which presents most often in late adolescence and middle age.<sup>11</sup> Despite this, milder forms of the illness have been recognised in more elderly patients.

There are a number of characteristic features on clinical examination which help to identify the disorder, namely ptosis and frontal balding. There is a generalised weakness of the facial muscles which may progress to involve other muscle groups and there is a risk of respiratory muscle weakness. Patients are unable to quickly open and close their hands and thus their grip is commonly affected. Particularly in the case of older patients, there is a risk of cardiomyopathy and associated cardiac arrhythmias, which can lead to sudden death.<sup>2</sup> Other systemic complications include diabetes and cataracts as well as hepatic and gastrointestinal complications. Diagnosis is based on isolation of the DM1 gene in families with a known history of the disorder. Where there is no documented history, identification of this disorder relies on recognition of the clinical features. CK is often normal. There is no treatment that is known to alter the progression of the illness but appropriate treatment of cardiac and other complications is likely to increase longevity. It is

important to establish a diagnosis even in an elderly person who may be only mildly affected. This is because the disease demonstrates anticipation and therefore future generations are likely to be more severely affected at an earlier age.<sup>3</sup>

### Oculopharyngeal dystrophy

Oculopharyngeal dystrophy is a late-onset disorder commonly affecting adults in their fifties or older. Presentation is with ptosis or dysphagia and there may be mild weakness of the proximal muscles of the shoulder and pelvic girdle. There is usually a positive family history. The disorder is autosomal dominant and identification of the PABPN1 gene confirms the diagnosis in affected individuals.<sup>2</sup> Life expectancy is normal although the patient may eventually be wheelchair bound. Treatment is aimed at relieving problematic dysphagia and, where appropriate, surgery for ptosis.

### Facioscapulohumeral muscular dystrophy

Patients with facioscapulohumeral dystrophy occasionally present in old age; however, more commonly the disease presents in early adult life. The disorder affects the muscles of the face, scapula and the upper arm. CK levels may be increased or normal and diagnosis is based on identification of a deletion on the FSHMD1A gene.<sup>2</sup> Treatment is supportive.

### Becker’s muscular dystrophy

Duchenne muscular dystrophy is a widely recognised disorder in the paediatric population and death occurs in late teens. However,

a milder form of the disorder—namely Becker’s muscular dystrophy—presents in adolescence with muscle weakness of the limb girdle. Calf pseudohypertrophy is a common finding on examination and there may be contractures as well as significant muscle wasting.<sup>4</sup> In some cases where females have been affected more mildly, symptoms may not present until middle age.<sup>12</sup> In these cases the heart is often affected and a cardiomyopathy can occur. In patients presenting in late middle age, CK levels may be normal although in younger patients CK is usually elevated. EMG, muscle biopsy and genetic testing will help to confirm the diagnosis and treatment is supportive.

### Myasthenia gravis

This disorder is a neuromuscular condition rather than a muscular disorder; it is relatively uncommon with a prevalence of one in 17,000.<sup>13</sup> As well as its classical presentation affecting young women under the age of 40, there is a second peak in incidence in men over 60 years. The disorder is characterised by fatigable muscle weakness commonly affecting the ocular and bulbar muscles, although the proximal limb girdle and respiratory muscles are also often involved.

Diagnosis is based on clinical demonstration of muscle fatigability combined with a history of diplopia, ptosis, dysphagia, speech disturbance or weakness affecting other muscle groups. A history of other associated autoimmune conditions particularly hypothyroidism may also be present in the patient or relatives. There is a recognised familial predisposition in about 5% of cases.<sup>13</sup>

Anti-acetylcholine receptor antibodies should be measured in all patients suspected of having the disorder and where the result is negative, antibodies against muscle specific kinase (MuSK) should be requested.<sup>2</sup> A tensilon test should be performed to help aid the diagnosis and repetitive nerve stimulation/electromyogram (EMG) studies should also be performed.

It is also vital to obtain a computed tomography (CT) or magnetic resonance imaging (MRI) scan of the neck to look for thymomas, which are commonly found in association with myasthenia gravis. Where thymomas are identified, surgery should be offered to the patient.<sup>2</sup>

Treatment of the disorder is lifelong, particularly in older patients, and will include cholinesterase inhibitors in combination with immunosuppressant. In a myasthenic crisis, intravenous immunoglobulin and plasma exchange have been shown to produce rapid clinical improvement. Patients should be regularly followed-up to allow early identification of possible deterioration and intervention.

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