Unusual Cases: Postpolio syndrome

Patients who thought they had long ago conquered the weakness and disability of polio are now being visited by an unwelcome reminder of their childhood illness.

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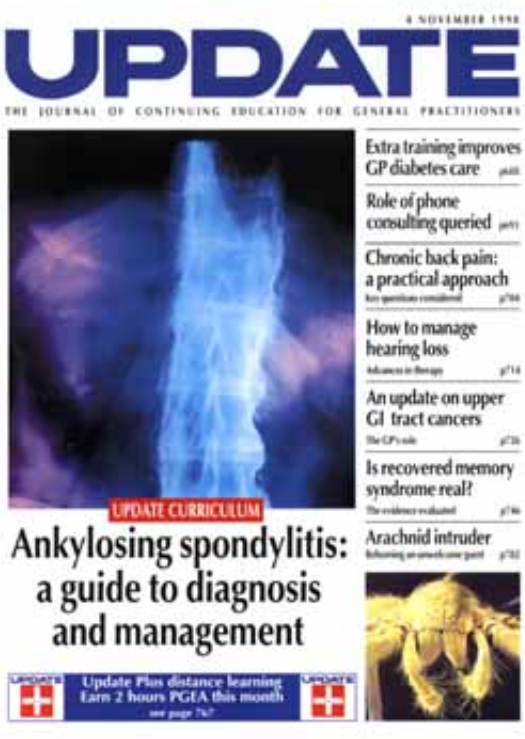

UPDATE
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The Case

A 55-year-old man asks for a sick note. He claims to have had increasing difficulty driving his lorry recently, and says the task has now become impossible. Weakness in his right leg, which was affected by polio in childhood, is stopping him from operating the pedals, although this has never been a problem before.

Examination reveals profound weakness and wasting of the calf musculature. Despite considerable scepticism, the GP issues a sick note.

A flurry of letters follow from the patient's insurers, all asking why he has only just developed this problem when the polio attack occurred 30 years ago. It is only after recourse to the literature that the question can be answered. The patient is suffering from postpolio syndrome.

Of the estimated 30,000 Britons with residual limb weakness from old poliomyelitis, about a quarter go on to develop postpolio syndrome (PPS) many years after the original illness. The weakness and muscle pain they endured as children revisits them after 30-40 years of stability, only this time in a more chronic, insidious and deceptive fashion.

PPS comes as a devastating blow to people who have spent a lifetime trying to conquer the functional deficits left by the original illness.

The syndrome involves a constellation of symptoms -- new muscle weakness, usually in a limb with previous polio weakness, muscle wasting, muscle pain at rest and with use, arthralgia, fatigue and psychological problems.

GPs face a daily stream of patients with musculoskeletal symptoms, myalgia, arthralgia and fatigue. Most can safely have their symptoms ascribed to degenerative joint disease, soft tissue injury, depression or nonspecific self-limiting causes.

PPS is often misdiagnosed because its symptoms closely resemble and often coincide with these common complaints. However, as a typical group practice is likely to include three or four polio survivors and one patient with PPS, most GPs are likely to see this condition.

DIAGNOSIS

PPS was fully described as recently as 1985.*[1-3]* Its identification followed numerous case reports in the 1970s and 1980s.

The diagnosis is based on a number of criteria:*[4]*

- history of documented paralytic poliomyelitis in childhood;
- partial recovery of motor function and functional stability for at least 15 years;
- residual asymmetric muscle atrophy with weakness (Figure 1);
- development of new pain and weakness;
- no other illness to account for the symptoms.

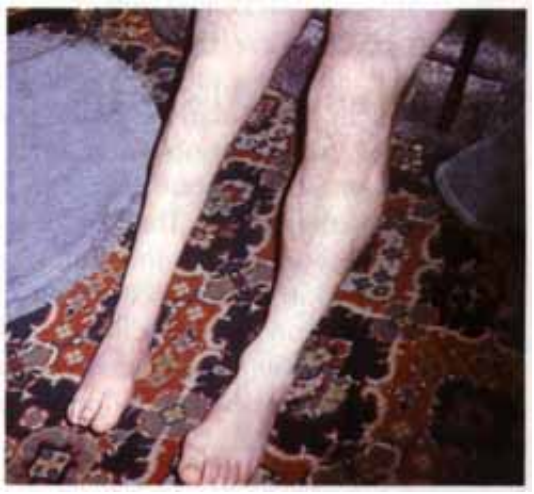

Figure 1. Atrophy of the calf musculature in postpolio syndrome

Common symptoms

PPS is typically characterised by the development of increasing weakness in a limb previously affected by polio. Some 90% of patients experience pain in the muscle with use, and 40% have muscle cramps at rest. Fatigue, both generalised and specifically on performing physical tasks and activities, is reported by 65% (Box 1).*[5]*

These symptoms lead to a range of functional problems. New weakness in a leg may result in difficulty with walking or stair climbing. Other activities of daily living such as bathing, dressing and using the toilet may become difficult, depending on the muscle groups involved. Many patients adapt by developing new techniques that preferentially use the unaffected muscles. The onset of functional deficits often has major lifestyle implications -- affecting the patient's ability to work, for example.

Box 1. Major symptoms.

- Increasing weakness in the limb previously affected by polio.
- Pain in the muscle with use.
- Muscle cramps at rest.
- Generalised fatigue.
- Fatigue on performing physical activities.

Less-common symptoms

As well as the major symptoms, a host of others occur with less frequency (Box 2). Some patients report new weakness in a limb or muscle group they had previously considered normal. Usually, this is because the muscle group had a minimal weakness unrecognised by the patient, or because the original polio affected less than 50% of the anterior horn cells supplying that limb and no weakness resulted at the time.

The onset of PPS unmasks this subclinical polio, and the limb becomes weak.

A small number of patients may develop dysphagia because of new weakness in the bulbar musculature. Detailed barium studies have shown that many more have a subclinical involvement of the swallowing mechanism.*[6]*

Patients who suffered involvement of the respiratory musculature in the original polio attack, for example those who used an iron lung or developed scoliosis, are candidates for further weakness of the respiratory musculature. This is likely to manifest itself in three ways:

- Chronic hypoventilation especially while asleep, leading to nocturnal hypoxia and hypercapnia, which in turn lead to poor sleep, daytime headaches and drowsiness. Some of these patients are treated with assisted ventilation devices.
- More frequent chest infections, because clearance of secretions is reduced.
- Reduced exercise tolerance, which exacerbates any mobility problems.*[7]*

Genu recurvatum -- hyperextended knee -- may occur for the first time in PPS (Figure 2a). The muscle tone around the knee becomes unbalanced through weakness of one group, allowing the deformity to develop. When the knee is hyperextended the leg muscles have to work harder.

Treating the deformity with a leg brace (Figure 2b) which prevents hyperextension reduces the work required to walk a given distance. It also reduces pain in the joint arising from the deformity.

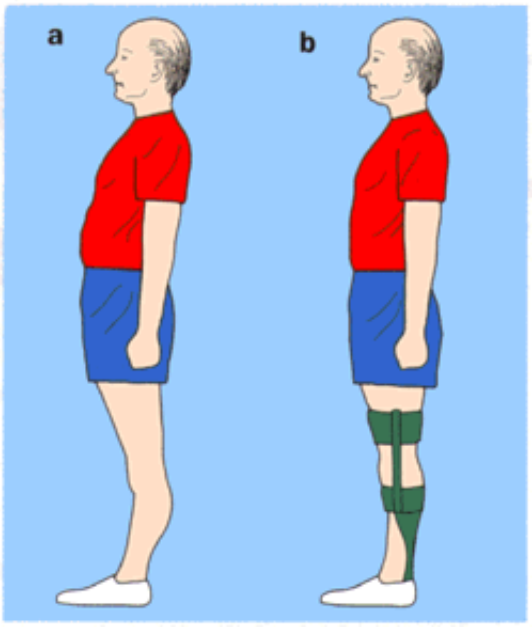

Figure 2. a) Hyperextension of the knee (back knee). b) Knee-ankle-foot orthosis.

Box 2. Uncommon symptoms.

- New weakness in a muscle group previously considered normal.
- Dysphagia due to weakness in the bulbar muscles.
- Weakness of the respiratory muscles in patients whose respiratory musculature was affected in the original polio attack.
- Hyperextended knee.

Psychological effects

As well as the physical symptoms, many patients with PPS have to cope with psychological and cognitive sequelae. These encompass fatigue after mental tasks, problems with attention, concentration and memory, and sleep abnormalities.

One study has shown changes in the reticular activating system on MRI scanning. The researchers postulate that the polio virus damages these neurones at the time of primary infection, and hence cognitive changes can occur with the onset of PPS.*[8]*

Box 3. Psychological sequelae.

- Fatigue after mental tasks.
- Attention problems.
- Problems concentrating.
- Memory problems.
- Sleep abnormaiities.

EXAMINATION

Clinical examination is likely to reveal lower motor neurone type wasting and weakness in the involved muscle groups. Fasciculation may be present and be reported by the patient.

Examination of all four limbs is worthwhile, as there may be weakness in a limb that is regarded as normal by the patient.

The most severely affected limbs, particularly the legs, are likely to be cold, and there may be trophic changes of the type found in peripheral vascular disease.

There are no diagnostic tests and the diagnosis is based on the clinical findings.

WHO IS MOST AT RISK?

A well designed study has shown that 28% of all cases of paralytic polio develop PPS 20-40 years later, with a peak incidence at 30-34 years after the childhood illness.*[5]*

The same study has shown that the risk of PPS is higher if:

- the patient is female;
- the original attack was severe, with marked residual disability;
- if it occurred in late rather than early childhood.

Recent weight gain is strongly associated with the development of PPS.

Once a patient develops PPS, a slow step-wise deterioration is likely, with periods of relative stability interspersed with periods of further increasing weakness. Overall muscle power is lost at a rate of 1% a year.*[2]*

WHAT CAUSES PPS?

Theories abound, but the jury is still out on the definitive pathogenesis of PPS. The favoured theory

suggests that after an attack of polio the number of lower motor neurones supplying a muscle is depleted by over 50% because of death of the anterior horn cells. The remaining lower motor neurones then attempt to compensate by forming terminal axon branches (sprouts), which innervate neighbouring muscle units that have lost their innervation. However, these new giant neurones with multiple terminal branches are inherently unstable and there is continuous reinnervation and denervation of adjacent muscular units.

A point is reached at which the rate of denervation exceeds the rate of reinnervation, possibly due to metabolic exhaustion of the motor neurone. At this stage, muscle strength is lost, and PPS is believed to start.*[9,10]*

MANAGEMENT

Treatment is largely supportive, with the GP playing a central role.

Correct and early diagnosis is probably the most important contribution the GP can make. Delay in diagnosis can lead to anxiety, and compound the patient's difficulties.

The onset of PPS may mean enforced early retirement, and have a major bearing on social security and insurance entitlements.

Drug treatment has been ineffective. Trials with several agents, including corticosteroids and anticholinesterases, have failed to show any convincing benefit.*[11,12]*

Interventions that may help include the use of orthotic devices and advice on exercise.

Orthotic devices

These are devices that restrict the movement of a joint. Foot drop is a common deformity in PPS (Figure 3a), due to weakness of the dorsiflexor muscle group. If uncorrected, it can lead to a permanent deformity through shortening of the Achilles tendon.

An orthotic device that maintains the foot in dorsiflexion prevents the deformity, and reduces the effort required in walking (Figures 3b and 4).

A knee brace is another example of an orthotic device that may be useful in PPS.

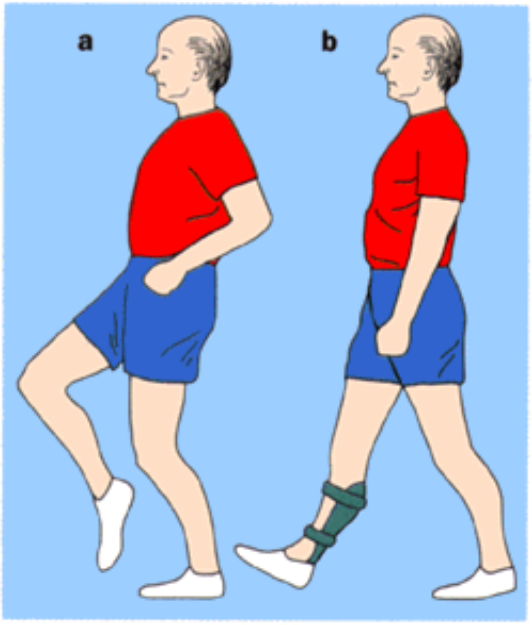

Figure 3. a) Weakness in ankle dorsiflexion (foot drop). b) Ankle-foot orthosis.

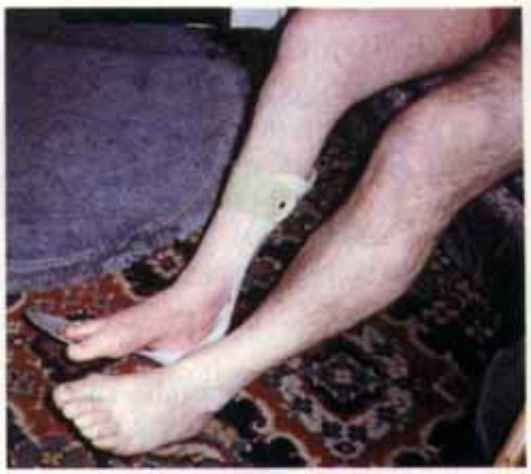

Figure 4. An orthosis for foot drop.

Exercise

PPS patients are likely to be unfit because their disabilities preclude vigorous exercise. However, an exercise programme forms an important part of any treatment package.

It has been shown that a graded programme, in which the affected muscle groups are gradually subjected to increasing isotonic exercise over a period of time, can produce increased strength and improved function. Patients should be advised to avoid exercising to the point of muscle pain or exhaustion.

An occupational therapist or physiotherapist can often tailor an exercise programme to suit the individual patient, based on muscle strength in each limb.

REFERRAL

Referral to a rehabilitation specialist is often beneficial. The consultant can then co-ordinate further referrals for physiotherapy, exercise, orthotic devices and other interventions.*[13,14]*

There are also several support groups, where PPS sufferers can seek additional help (see useful addresses).

The Outcome

The patient's weakness continued to increase, and later he went on to develop a troublesome foot drop. This was treated with an orthotic device to maintain the foot in dorsiflexion.

The patient also complained of increasingly troublesome rest pain, muscle spasms and cold in the affected limb. A combination of conventional analgesia and a small dose of amitriptyline provided only partial symptomatic relief.

He has been referred to a specialist in rehabilitation, and is slowly coming to terms with this latest phase of a chronic disease.

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USEFUL ADDRESSES

- The British Polio Fellowship, Ground Floor, Unit A, Eagle Office Centre, South Ruislip, Middx HA4 6SE, tel: 0181 842 4999; fax: 0181 842 0555.

  Provides helpful advice sheets, literature and other support for patients. Most of its work today is concerned with PPS.

- Lincolnshire Postpolio Network, PO Box 954, Lincoln LN5 5ER, tel: 01522 888601, web site: http://www.lincolnshirepostpolio.org.uk/.

  Informal support network run by PPS patients.

- Other web sites with useful links:
  http://members.aol.com/DEMP12/pps_help.html
  http://home.earthlink.net/~polioinfo/
  Many PPS groups in the USA issue information on the internet

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