Late post-polio deterioration

Figure 1  Still from the March of Dimes film, The Daily Battle (March of Dimes Birth Defects Foundation) – used as the cover of Tony Gould’s book – A Summer Plague, 1995.
In the first half of the 20th century, polio was one of the most feared of conditions in the UK and USA. Few people were unaffected during the epidemics of the 1940s and 50s, which preceded the introduction of immunization with the Salk inactivated vaccine in 1956 and the Sabin live attenuated vaccine in 1962. Patients remember the anxiety and mass hysteria surrounding each new outbreak of poliomyelitis, and fear of polio was often greatest amongst medical personnel, many of whom were affected (Gould 1995; Mulder 1991; Halstead 1995). This fear is graphically illustrated on the cover of Tony Gould’s classic description of the history of poliomyelitis in the 20th century (Fig. 1) (Gould 1995).

There are striking accounts of the acute illness – patients recall bathing in affected areas and developing non-specific flu-like symptoms, often following periods of prolonged exertion (Russell 1949). When the diagnosis was suspected they were frequently admitted to large ‘isolation hospitals’ in the country outside major areas of population. Many were not allowed to see their family or friends, and all speak of their profound anxiety and helplessness. They were treated with strict bed rest and some required ventilation with an Iron Lung (an efficient form of ventilation particularly suited to polio because it allowed concurrent physiotherapy and postural drainage) (Fig. 2). Those with severe bulbar weakness required a tracheostomy to protect the airway, but few survived.

Early treatment and rehabilitation generally consisted of strict and total bed rest with pain relief, splinting and casting so many patients lay for weeks or months in a posterior plaster jacket or cast. This lead to disuse and atrophy of muscles not already affected by the disease (Mulder 1991). The early rehabilitation of polio was greatly influenced by the vision of Sister Kenny, an Australian nurse practising in the United States, who advocated the use of warm, moist heat, using wool packs, together with early mobilization of affected limbs. This work was given further impetus at Warm Springs, Georgia, a rehabilitation centre, supported by President Franklin D Roosevelt following his own acute polio contracted at the age of 39. Here techniques of hydrotherapy and early mobilization with splinting and passive movements, were developed to prevent contractures and joint ankylosis.

Early rehabilitation

In experienced centres, during the first few weeks of mobilization, the main thrust of management was to prevent deformity by stretching and splinting of affected limbs. Intensive physiotherapy aimed to retrain affected muscles to normal strength and function. Appropriate orthoses were provided to compensate for loss of function – callipers, crutches and wheelchairs. The aim was not only to facilitate mobility, but also to prevent undue wear and tear. Careful postural re-education and attention to activities

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of daily living were fundamental to successful early rehabilitation (Green 1949).

Later rehabilitation
 Much of the later rehabilitation involved multiple orthopaedic procedures and the provision of appropriate orthoses. The orthopaedic management was aimed at maintaining stable mobility. Multiple surgical procedures were often undertaken to lengthen or shorten tendons, transfer muscles in affected limbs, fuse unstable joints and correct other abnormalities.

THE ‘POST–POLIO SYNDROME’
 It gradually became clear that, after a period of prolonged stability, many patients with residual disability following earlier paralytic poliomyelitis developed late functional deterioration – impairment of activities of daily living, mobility, upper limb function and respiratory capacity (Campbell et al. 1969; Codd et al. 1985; Editorial, The Lancet, 1986; Howard et al. 1988; Halstead 1991; Kidd et al. 1996). In this context, much has been written about the illness of Franklin D. Roosevelt (Gallagher 1985). There is no doubt that after a prolonged period of stability, his disabilities increased with age and in his later years he even had difficulty lighting a cigarette, driving a car and holding a cup of coffee.

In 1962, Zilkha described 11 patients who developed new weakness and fasciculations 17–43 years after polio, in muscle groups apparently not involved clinically in the acute disease. This late functional deterioration has been referred to as the ‘post-polio syndrome’ and ‘late effects of polio’. I prefer to use the term ‘post-polio functional deterioration’ because this better and more clearly defines the clinical problem. All the definitions recognise a variety of new difficulties with daily living that have begun in later life following an earlier attack of acute poliomyelitis. Bradley (1987) defined a ‘post-polio syndrome’ consisting of progressive musculoskeletal deformity as a consequence of the original weakness, which may be associated with nerve entrapment. He recognized muscu-
loskeletal pain syndromes without progressive weakness and considered progressive post-polio muscular atrophy as a separate entity when there was a new history of muscle atrophy with or without decreased muscle strength in an asymmetrical distribution compatible with previous polio, and electrophysiological features of acute denervation superimposed on chronic denervation-reinnervation in the absence of another neuromuscular cause.

Dalakas and colleagues (Dalakas et al. 1986; Dalakas & Hallat 1998; Dalakas 1988) defined the ‘post-polio syndrome’ as new neuromuscular symptoms - 25 to 35 years after reaching maximum recovery from acute paralytic poliomyelitis - unrelated to any orthopaedic, neurological, psychiatric or systemic medical illness. This definition included complaints such as muscle and joint pain, reduced exercise tolerance, impairment of activities of daily living, limb atrophy, cramps and fatigue but specifically excluded musculoskeletal symptoms due to back injuries, radiculopathy, compression neuropathies and other medical, neurological, orthopaedic or psychiatric illnesses. These criteria are somewhat inconsistent because, in my experience, most patients with post-polio functional deterioration have musculoskeletal abnormalities which can be directly related to the consequences of the original illness including entrapment neuropathy, radiculopathy and orthopaedic problems.

We have reported a series of 209 consecutive patients (Howard et al. 1988) of whom 163 (78%) developed late functional deterioration due to the following factors:

- respiratory (99 cases)
- neurological (20 cases)
- orthopaedic (17 cases)
- general medical (27 cases).

Scoliosis developed in 94 patients and was invariable if paralytic poliomyelitis had occurred before the growth spurt. Virtually all the patients with late functional deterioration had been severely affected during the acute illness and all had significant residual weakness. 34 patients had worsening limb function associated with difficulty using callipers, cervical spondylisis, pharyngeal instability, osteoarthritis, osteoporosis, back pain and contractures. Other conditions were also relatively frequent in this population of severely disabled patients and the following factors contributed to progressive functional deterioration by increasing disability and levels of dependence:

- chronic urinary symptoms secondary to calculi;
- severe trophic changes of the legs;
- coincidental medical disorders such as hypertension, diabetes mellitus and peptic ulceration;
- depression;
- pregnancy.

In a subsequent consecutive series of 283 patients (Kidd et al. 1996), a change in the clinical pattern became apparent with most patients being referred because of orthopaedic, neuromuscular or neurological problems rather than the development of ventilatory insufficiency. Perhaps most of those who had developed respiratory muscle weakness during the acute illness had already presented and been treated.

Windebank et al. (1991) published a series of 50 patients who had paralytic polio in Olmsted County. They were closely followed up using the Mayo Clinic standard scoring system for all neurological examinations. Although 64% reported new symptoms, only 18% were found to have any functional deterioration. Also, in only four patients could new or worsening weakness be substantiated clinically, and there was a clear radiculopathy or nerve entrapment to explain their functional deterioration. The frequency of symptoms of late deterioration did not change over a 5-year prospective assessment (Windebank 1995).

Pathogenesis

The aetiology of post-polio functional deterioration is uncertain but a number of theories have been proposed. Firstly it may represent a process of neuronal attrition due to normal ageing. This is unlikely because the condition is more dependent on the latent period following the acute illness than age, and new symptoms may start well before the age of 60. Furthermore, anterior horn cell dropout is not normally seen below 70 years (Tomlinson & Irving 1977) and very seldom do muscle biopsies show angulated fibres (Dalakas 1988, 1991).

It has been suggested that there is a geographical relationship between past notification rates of polio and current mortality from motor neurone disease in the UK (Martyn et al. 1988; Martyn 1990). However, this has not been confirmed. Indeed Armon et al. (1990) noted the relative paucity of classical motor neurone disease developing in survivors of paralytic poliomyelitis, and Swingier et al. (1992) were
Unable to show any geographical association between past mortality from poliomyelitis and present morbidity and mortality from motor neurone disease in Scotland.

There is some evidence of mild inflammatory change on muscle biopsy, abnormal activated peripheral blood T lymphocyte subsets and ganglioside specific IgG and IgM, as well as active pathological changes in spinal cord and muscle (Drachmann et al. 1967; Dalakas 1988; Pezeshkpour & Dalakas 1988; Feve et al. 1992). The significance of these findings is unclear. There is undoubtedly an ongoing intrathecal antibody response to poliomyelitis – oligoclonal IgM bands specific to polio virus were detected in the CSF of 21/36 (58%) patients with post-polio syndrome but none of 67 controls (Sharief et al. 1991). Once again the significance of this finding is uncertain (Melchers et al. 1992; Muir et al. 1996).

It is possible that one mechanism contributing to post-polio functional deterioration is related to a continuous and widespread process of denervation and reinnervation occurring in all muscles clinically and subclinically affected by the original poliomyelitis. Thus, the remaining healthy motor neurones can no longer maintain unstable new units, with the eventual loss of single fibres. This is suggested by electrophysiological findings of active denervation, including spontaneous activity and increased fibre density with jitter and blocking on single fibre studies. However, similar changes are seen in both affected and unaffected muscle groups in surviving polio patients with and without ‘post-polio syndrome’, thus it is uncertain whether these electrophysiological changes are of any clinical significance. Furthermore, muscle biopsy shows features of ongoing denervation, including small angulated fibres, group atrophy and fibre type grouping, in both stable patients and those with late onset of deterioration (Weichers & Hubbell 1981; Bradley 1987; Cashman et al. 1987; Ravits et al. 1990; Dalakas et al. 1995).

### Factors contributing to post-polio functional deterioration

The condition usually occurs in patients with significant residual disability following the original polio. The extent of the original limb, trunk, respiratory and bulbar weakness is an important factor in predisposing to the development of late functional deterioration. Some patients are referred because of the development of disability even though the initial polio left no residual sequelae, or the original diagnosis was equivocal. In my experience, these patients always have an alternative explanation for their symptoms, often a myopathy or a fatigue syndrome.

Frequently there is progressive wasting and weakness in limbs already affected by poliomyelitis but this may also occur in the contralateral limb, i.e. secondary to weight bearing or distorted mechanics. Indeed, a compensatory hypertrophy may occur in the contralateral limb (Wilson et al. 2000). Progressive joint contractures secondary to the distorted mechanics lead to limb impairment such as knee hyperextension, hip arthrosis and scoliosis.

The effects of growth are often significant. Polio developing before the growth spurt usually leads to progressive scoliosis because of the eccentric development of the spinal musculature. Also, polio developing in the early years leads to limb shortening, which causes growth retardation during the adolescent growth spurt. The use of callipers, crutches and wheelchairs can lead to the development of compression neuropathies.

### What the patients notice

(See table 1.) Patients become aware of deterioration in their functional capacity after many years of stability – often weakness, pain and fatigue. Most patients notice an insidious onset of progressive impairment but sometimes functional deterioration occurs as a consequence of a precipitating event such as a fall or intercurrent illness. Patients describe lack of strength.

<table>
<thead>
<tr>
<th>What the patient notices</th>
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<tr>
<td>Prolonged plateau level of function</td>
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<tr>
<td>Late functional deterioration</td>
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<tr>
<td>Onset 10 + years after acute poliomyelitis</td>
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<tr>
<td>Weakness, pain and fatigue</td>
</tr>
<tr>
<td>Onset usually slow and steady, but may develop suddenly and may progress at irregular intervals</td>
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<tr>
<td>May follow intercurrent event (immobility and strain)</td>
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<tr>
<td>Lack of strength and endurance</td>
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<td>Inability to undertake ‘normal’ activities</td>
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<td>Prolonged fatigue following physical activities</td>
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<td>Difficulty in recovering from periods of immobility</td>
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<tr>
<td>Musculoskeletal complaints:</td>
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<tr>
<td>Increasing muscle weakness</td>
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<tr>
<td>Pain in muscles and joints</td>
</tr>
<tr>
<td>Unreliability of previously stable joints (often upper limb)</td>
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<tr>
<td>Changes in gait and a tendency to fall</td>
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<tr>
<td>Cramps and fasciculations</td>
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or endurance, which causes difficulty with normal activities such as walking to the shops or washing their hair. There may be prolonged fatigue following physical activity, or difficulty in recovering from periods of immobility. They often experience new musculoskeletal symptoms including cramps and fasciculations, increasing weakness of a limb or the trunk, unreliability in a previously stable joint (often in the upper limbs), pain in muscles and joints and changes in the joint (often with a tendency to trip or fall).

Respiratory complaints related to diaphragm and respiratory muscle weakness, and leading to progressive nocturnal hypoventilation, include breathlessness on exertion, orthopnoea and breathlessness when the trunk is in water. Nocturnal ventilatory impairment may lead to snoring, abnormal sleep movements, morning headache, daytime hypersomnolence, impaired intellectual function, irritability and depression.

CAUSES AND MANAGEMENT OF LATE FUNCTIONAL DETERIORATION

Orthopaedic complications
These are extremely common and reflect the prolonged abnormal stresses applied to joints due to muscle weakness - fixed flexion deformities, hyperextension or lateral instability of the knee or hip (Fig. 3). Other causes of deterioration include progressive instability of joints, fractures, osteoporosis and osteoarthritis. Scoliosis frequently worsens over many years (Fig. 4). Degenerative joint disease is most common in weight bearing joints and weakened limbs. Cervical spondylosis is extremely common and causes neck pain and variable sensory radicular symptoms. Cord compression occurs in some patients (Fig. 5) and the diagnosis may be difficult if limbs are already affected by polio. Obesity frequently contributes to orthopaedic deterioration.

Many aspects of the management of these patients require detailed and specialized orthopaedic assessment. A range of simple supports to knee, ankle and cervical spine, or correction of worn and damaged orthoses, may lead to considerable functional improvement. These include the provision of new callipers, braces, foot orthoses, knee and pelvic supports, shoe raises, collar, harness and seating. Hip and knee deformities may be helped with physiotherapy, hydrotherapy, night splints or foam supports.
Cervical decompression is usually indicated in the presence of severe established radiculopathy or myelopathy. If progressive scoliosis is contributing to respiratory insufficiency then spinal surgery may be undertaken, or bracing without fusion may be attempted but these are technically difficult procedures (Fig. 6). With severe bilateral genu recurvatum causing posterior knee pain, if orthotic support fails, bone block procedures using the patella have proved effective (Hong-Xue et al. 1991).

Respiratory insufficiency

Respiratory muscle weakness may lead to progressive nocturnal hypoventilation exacerbated by chest wall deformity, progressive scoliosis or other factors stressing critically compromised ventilation – respiratory tract infections, obstructive airways disease, obesity, pregnancy and tracheostomy complications (Lane et al. 1974; Howard et al. 1974).

The strategies and methods of artificial ventilation used in poliomyelitis are long-established but require special adaptation in the presence of scoliosis. Some patients who were ventilated during the acute illness continue to require long-term intermittent or continuous support. Others were weaned after the acute illness but subsequently have required intervention after developing ventilatory failure. In general, patients who received ventilation during the acute illness have residual respiratory muscle weakness and remain most at risk of later respiratory compromise. Diaphragmatic weakness is indicated by paradoxical abdominal movement on respiration and a fall in the forced vital capacity (FVC) of more than 25% from standing to supine. A continuing decline in the FVC may precede the development of symptoms. Nocturnal oximetry will begin to show the characteristic pattern of prolonged periods of nocturnal desaturation before the development of hypercarbia and hypoxia on daytime arterial blood gas measurement.

At this stage ventilatory support is necessary. The indications for positive and negative ventilation have been described in detail previously (Howard & Spencer 1993; Howard & Williams 1999). Negative pressure ventilation using an Iron Lung (Fig. 7) or cuirass shell (Fig. 8) was the mainstay of respiratory support during the polio epidemics. Pressure changes applied to the trunk, but not the head, cause air to pass in and out of the lungs. Its current use is confined to those patients in whom the Iron Lung has been...
their mainstay of assisted ventilation, having been introduced before the availability of other convenient systems.

The drawbacks to negative pressure ventilation include lack of portability along with a physiological effect of increasing obstructive apnoeas during sleep because negative pressure applied to the thorax leads to an increased tendency for the upper airway to collapse in the region of the velopharynx. In addition, negative pressure ventilators may not lend themselves to use for patients with major thoracic deformities.

A mechanical rocking bed was described in 1953 and the technique has remained popular and effective in patients whose respiratory weakness is predominantly diaphragmatic (Fig. 9). It is the only method of respiratory assistance that leaves the patient unconnected to any apparatus. Although diaphragmatic weakness commonly involves the crura, it is unusual for rocking bed users to experience oesophageal reflux. The disadvantages include nonportability and engineering problems in providing a smooth motion.
may develop worsening dysphagia, which is not usually associated with other evidence of bulbar weakness. This rarely progresses to aspiration, and symptomatic management by speech and language therapists is usually appropriate. New neurological disorders in the survivors of polio have included motor neurone disease, multiple sclerosis (Chroni et al. 1995), syringomyelia, epilepsy and meningioma but there is no evidence to suggest these associations are anything other than coincidental.

General medical disorders
General medical factors contributing to late deterioration have included chronic urinary disturbances such as renal or bladder calculi. Other important problems include the coincidental development of diabetes and hypertension.

Other aspects of management
The effective management of post-polio functional deterioration requires a closely-knit multidisciplinary approach, which involves both specific management of increasing impairment and a process of enabling the patient to cope with new disabilities. Polio survivors are a remarkable group of patients. They tend to be extremely motivated and driven; they have conquered their disability, often by ignoring it completely, and have the most amazing stories of achievement (Gallagher 1995). However, in the management of post-polio functional deterioration this approach can be counter-productive because many continue to believe that increasing disability can be overcome by more exercise. Thus, as late post-polio functional deterioration develops, many patients use intensive exercise regimes to regain muscle mass, strength and function. Although some forms of exercise are often helpful this needs to be carefully assessed by a multidisciplinary team. Immobility will lead to wasting and stiffness but it is essential to strike a balance between exercise regimes that can alleviate symptoms, without causing increasing weakness and fatigue in post-polio muscles. Non-swimming exercises in warm water are often helpful in conditioning to exercise, mobility and reducing pain. Regular graded exercise should be broken up by regularly spaced periods of rest. The aim of management is to prevent overuse and subsequent deterioration and the key is to find the correct balance between activity and rest with careful prioritizing, planning and pacing of activities, life-style modification.

During the 1940s outbreak of polio in Copenhagen, the demand for Iron Lungs outstripped the supply. Patients were kept alive by relays of medical and dental students using a type of ‘bel lows’ to provide ventilation. This led directly to the development of intermittent positive pressure ventilation and the birth of modern intensive care. Positive pressure ventilation is the most efficient form of assisted ventilation and was first applied in the polio epidemics. For continuous use, or, in practical terms, greater than 16 hours a day, a connection to the trachea via a cuffed endotracheal tube, or directly by tracheostomy, is required. Machines delivering a predetermined pressure are now more common. The advantage lies in the patient being able to vary tidal volume on a breath-to-breath basis. These ventilators are leak-compensating, which provides another advantage as intermittent speech is possible during the inspiratory stroke of the ventilator. They are particularly suitable for long-term use in neurological disease and are usually simple and reliably constructed, making them suitable for home use. It is these ventilators that have commonly been used to deliver positive pressure through nasal masks, so-called ‘Nasal Intermittent Positive Pressure Ventilation’.

Neurological problems
Skeletal deformity due to previous poliomyelitis contributes to the development of multiple peripheral nerve entrapments leading to functional deterioration. A small proportion of patients may develop worsening dysphagia, which is not usually associated with other evidence of bulbar weakness. This rarely progresses to aspiration, and symptomatic management by speech and language therapists is usually appropriate. New neurological disorders in the survivors of polio have included motor neurone disease, multiple sclerosis (Chroni et al. 1995), syringomyelia, epilepsy and meningioma but there is no evidence to suggest these associations are anything other than coincidental.
and the provision of assistive devices including orthoses, braces and corsets.

Physicians must recognize that patients may notice changes in function that are not revealed by increasing weakness on neurological examination. What appears to be a slight worsening of an overwhelming disability, may be devastating to patients but doctors often take the wrong approach by failing to understand the patient’s fear of increasing immobility. Patients often need to be reassured that the condition is not a form of motor neurone disease, and nor is it likely to progress to severe impairment, although new functional disabilities are possible and must be appropriately managed. It should be explained that post-polio muscles work harder and have reduced nerve supply, thus even apparently unaffected muscles may have had severe damage during the acute illness and be substantially weak, or at least vulnerable to the effects of apparently minor insults.

Many patients have impaired respiratory function. They must be made aware of the signs of a developing chest infection and should have a prophylactic supply of antibiotics, and receive influenza and pneumococcal immunization. They must also avoid smoking. It is essential that patients with known respiratory muscle weakness are monitored regularly with FVC measurements (erect and supine) and nocturnal oximetry (El-Kabir et al. 1998).

Sleep disturbance is common (Williams et al. 1998). Whilst respiratory insufficiency may cause obstructive sleep apnoea or hypoventilation, a variety of polysonmias, in particular periodic limb movement disorder and the restless legs syndrome, may cause considerable sleep disruption.

Excess weight contributes to impaired mobility, the development of osteoarthritis and respiratory insufficiency because of hypoventilation and obstructive sleep apnoea. Weight loss is often very difficult because of the reduced mobility and a dietician is an extremely important member of the multidisciplinary team. The most important intervention however, is the development of a graded exercise programme suited to the individual.

These patients are particularly prone to depression, and coexisting anxiety is also common. Whilst treatment with antidepressants and anxiolytics is often helpful, there is no doubt that cognitive behavioural therapy is rapidly emerging as an extremely valuable tool in management.

What appears to be a slight worsening of an overwhelming disability, may be devastating to patients but doctors often take the wrong approach by failing to understand the patient’s fear of increasing immobility

Pain management can be difficult because the pain is often generalized and not localized to a joint or limb. Simple physical measures such as warmth, cold, massage or passive stretching may be of great value. Transcutaneous electrical nerve stimulation and acupuncture are also extremely helpful in some situations. Analgesics, particularly non-steroidal anti-inflammatory drugs, are required by some patients, particularly where there is an inflammatory component. In other patients a more holistic approach to long-term pain management will be provided by a pain clinic.

Fatigue is particularly difficult to treat. Many factors contribute to its development, and exclusion of an underlying disorder is essential before effective management can be undertaken. The most important aspect of treating fatigue is the development of a graded exercise programme with clear and achievable goals. Medication is not usually helpful.

A considerable burden falls on the carers who have often been the only support for decades. They must be recognized, supported and involved in ongoing rehabilitation and management.

The British Polio Fellowship (http://www.britishpolio.org) (Fig. 10) provides a unique and invaluable resource for patients and their families. The Society is unfailingly supportive and knowledgeable about the condition and the provision of rehabilitation, self-management, welfare benefits, disability equipment, housing and holiday accommoda-
Using conventional definitions these patients cannot be considered to have the ‘post-polio syndrome’. There is no doubt that the severe physical stresses of post-polio disability lead to the development of progressive orthopaedic, respiratory, and general medical problems, often exacerbated by intercurrent events. These problems may present with atypical clinical features, because of the extent of underlying muscle atrophy and weakness, and so be difficult to diagnose. Moreover, many of these problems are potentially treatable and most patients can be helped to understand and manage increasing disability. It is therefore important to be very cautious before attributing functional deterioration to the nebulous diagnoses of ‘post-polio syndrome’ or ‘progressive post-polio muscular atrophy’. If increasing disability really cannot be attributed to progressive orthopaedic, respiratory or general medical problems, it is at least reassuring that it is extremely unusual for the impairments to progress to severe disability.

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