POLIOMYELITIS

New Problems From An Old Infection

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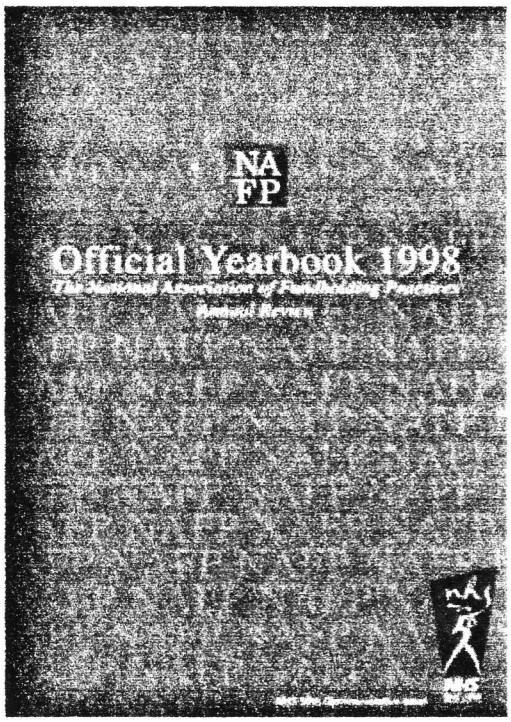

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There are currently around 30,000 post-poliomyelitis survivors in the UK, although the last major epidemic was over 40 years ago. Around half of these will develop the post-polio syndrome which is a recently recognised disorder characterised by late onset of weakness in muscles affected during the acute illness. Indirect effects of polio, especially degenerative disorders of the joints and soft tissues also contribute to the increasing disability that these patients are developing. It is essential to accurately analyse the cause of any new symptoms in polio survivors so that specific treatments can be applied, or modifications to their pattern of activities recommended.

Is poliomyelitis a disease of the past? It is now over forty years since the last major epidemic took place in the UK and there can hardly be a doctor still practising who has much first-hand experience of acute poliomyelitis occurring in this country. The general feeling is that once the patient recovered from the acute illness, his or her condition remains stable, and is irrelevant to any new problems. From the patient's point of view, however, there is a feeling of isolation and frustration. Isolation because of lack of contact with other people who have had polio or with any organisation that can offer help. Frustration because they feel that the medical profession and other health care workers have little expertise or interest in the problems that result from their polio; new problems as well as old.

The numbers of post-polio patients is considerable. In the USA it is estimated that there are over 600,000 survivors who have had this infection and in the UK the number is thought to be around 30,000. The new interest in polio is not however only due to the potentially large patient population, but mainly to the recent recognition of the post-poliomyelitis syndrome (PPS). There has been debate about the importance, or even the existence, of this condition over the last 20 years, but much of this confusion is related to semantic difficulties. PPS is best regarded as the development of new weakness in muscles which were previously affected by the acute infection. The confusion has arisen because postpolio patients also develop muscle weakness and other new symptoms as indirect late consequences of polio, due to, for instance, degenerative soft tissue and joint conditions. The wider term 'late effects of polio' is best used to describe these indirect consequences as well as PPS, which should be reserved for the specific late onset neuromuscular disorder that polio causes.

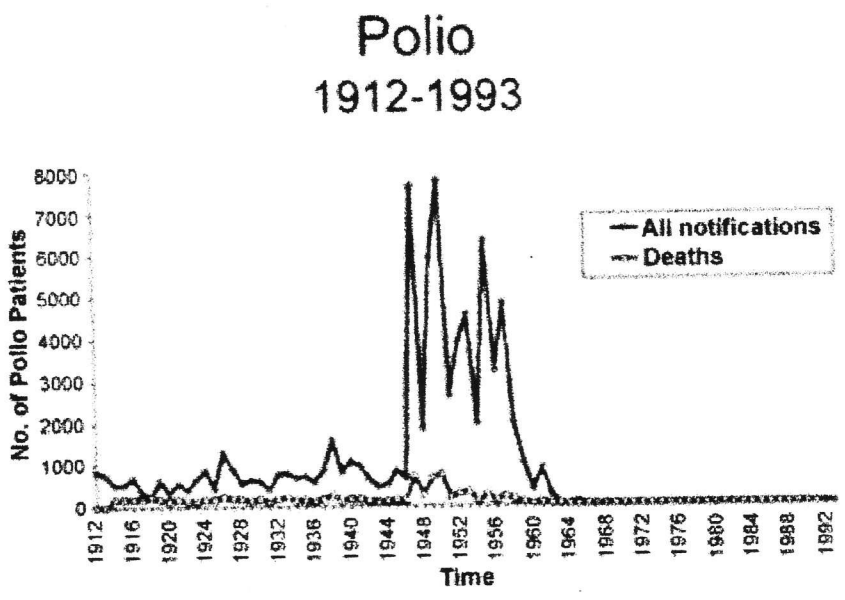

Figure 2: Notifications of Polio in UK 1912 - 1993

Causes of PPS

To understand PPS one has to be aware of what happens during and after the acute infection. The polio virus is an enterovirus which in most subjects only causes a non-specific influenza-like illness with or without diarrhoea. The majority of people do not develop classical paralytic poliomyelitis. When this does occur this is due to damage to lower motor neurons which causes the peripheral nerve fibres or axons supplying the muscle fibres to die back. This leaves the muscles fibres 'orphaned' and in the healing process the remaining axons sprout new terminals to innervate these muscle fibres. New motor units, which are the group of muscle fibres supplied by any one axon, may increase to as much as 10-15 times the normal size. The formation of these 'giant' motor units contributes to the recovery of muscle strength in the weeks, months and even the first couple of years after the acute illness. For many years they appear to function satisfactorily, but in some subjects the axons eventually begin to die back. The muscle fibres cannot then function and begin to atrophy.

The cause of this late degeneration of the axons and the death of their cell bodies in the spinal cord and brain stem is uncertain. There is no evidence for any reactivation of the polio virus or for any abnormal immunological response that might cause cell death. Ageing itself is unlikely to be responsible since PPS is not age-related. Cell and axon death may be related to an inability to sustain the higher metabolic demands required by the giant motor units, particularly if the muscles have been much used. Active muscles have a higher stimulation frequency of their muscle fibres than little used muscles and this places a greater metabolic load on the peripheral nerve cells.

Diagnosis of PPS

Recognition of PPS is based on identifying which patients are at risk and on recognising relevant new symptoms. PPS is commoner in patients who were more severely affected by the acute illness. Those who were hospitalised or ventilator-dependent during their initial infection or in whom all four limbs were paralysed are at high-risk. It is more common in those who contracted polio after the age of 10, possibly because the healing process occurred less satisfactorily with more extensive sprouting of new terminals, and also because the infection is usually more severe in those who contracted polio at a later age. It is unusual for PPS to be detectable within thirty years of the acute illness. The mean onset is around 35 years, but it can occur at any time, even as long as 75 years after the original infection. PPS is detectable in around 50% of patients who have had polio, but in 80% of these (40% of the total) it is only slowly progressive. It is the remaining 20% (10% of the total) in whom it advances rapidly. The bulk of the patients who are now developing PPS were those who developed polio in the major epidemics between 1946 and 1957 (figure1).

Electromyographic (EMG) and peripheral nerve conduction studies may exclude other disorders but have proved to be of little help in diagnosing PPS, despite its being due to peripheral nerve fibre degeneration. This is because even in patients without clinical features of PPS there is often some detectable peripheral nerve fibre degeneration. This suggests that many polio patients may show subclinical features of PPS after a certain interval, but in only some is it clinically apparent.

The cardinal symptom of PPS is the appearance of new weakness in muscles which were previously affected by the acute illness. Patients who had polio often develop trick movements and other strategies to carry out activities despite muscle weakness and only a slight loss of strength may prevent these from continuing to be performed. The patient may simply be aware of a deterioration in the function of a part of the body, such as a hand or arm, or there may be difficulty in walking or an increasing frequency of falls, perhaps due to instability of a lower limb joint due to new muscle weakness. Polio causes a patchy muscle weakness and PPS can be equally patchy in the muscles that it affects (figure 2).

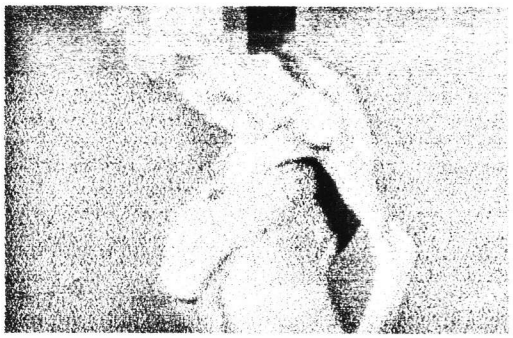

Figure 2: Polio patient showing extensive but patchy muscle atrophy

A second important symptom is the development of muscle fatigue. This is the loss of strength of a muscle with continued or repetitive activity. It initially affects muscles that are weakened, since at any given level of exertion they are exercising at a relatively greater fraction of their capacity than a normal muscle. Patients may find that they become progressively weaker as they walk further and have to stop in order to regain their strength. Muscles which are over-exercised may become painful, either during or more commonly some hours after the exercise. This pain may have a burning character to it and muscle cramps are also common. After overexercising patients may feel generally physically tired for as long as one or two days.

While it is commonest for PPS to affect the limb muscles it can also involve those that are responsible for speech, swallowing and respiration. The patient's voice may become weaker, dysphagia may become a problem and aspiration of food, drink or saliva into the tracheo-bronchial tree may occur. Patients may present with weight loss through avoiding eating because of the difficulties that it causes and this may exacerbate any muscle weakness. If the dilator muscles of the upper airway are involved, particularly those in the pharynx and those controlling the larynx, obstructive sleep apnoeas may be seen. These are characterised by snoring or stridor-like noises during sleep associated with an irregular respiratory pattern, a feeling of waking up un-refreshed and increasing daytime sleepiness. This sleepiness should be distinguished from the feeling of physical weariness due to overuse and fatigue of the limb muscles.

A separate respiratory problem arises if the chest wall muscles weaken as part of the PPS. In this situation they are unable to maintain an adequate level of alveolar ventilation during sleep and both hyrpercapnia and hypoxia develop. This may cause frequent awakenings from sleep, un-refreshing sleep and daytime sleepiness. This type of respiratory failure is commonest in patients who were initially treated in an iron lung as a form of ventilatory support during their acute illness. Their vital capacity, measured with a spirometer, is usually less than around 1 litre. More sophisticated tests of their inspiratory muscles usually reveals profound weakness and many of these patients also have a weak cough due to expiratory muscle weakness or laryngeal abnormalities.

Other late effects of polio

These features which indicate the appearance of PPS should be distinguished from the indirect effects of polio. The most important of these are soft tissue and joint degenerative conditions. The tendons and ligaments may become inflamed or weakened, osteoarthritis is common and joints become unstable. Peripheral nerve entrapment is also common and this often causes, for instance, shoulder pains if the cervical spine is involved or sciatica from lumbar spine disorders.

Assessment of these symptoms is often complicated by the presence of depression and anxiety. These may be the result of years of social isolation due to the physical effects of the original infection, but very often depression worsens when symptoms of PPS or the other late effects of polio appear. Uncertainty about the future, in particular about the ability to continue with the previous way of life and to remain independent, becomes a major problem. The appearance of new symptoms often brings back thoughts of

the original illness as a child and all the implications of this. Some patients deny that they are developing new difficulties, but more commonly the fear of what the future will hold and difficulties in coping with a loss of physical function precipitate a consultation with a general practitioner.

Management of PPS

The management of PPS and the other indirect effects of this infection requires an accurate assessment of the cause of the current symptoms and in particular a recognition of what is and what is not due to PPS. It is important to establish whether or not the original illness really was polio or a different neurological or infective condition. The severity and extent of muscle, joint and soft tissue abnormalities should be assessed. If the respiratory muscles are thought to be involved, estimation of the vital capacity is essential. A sleep study is often needed to assess whether or not the patient is developing obstructive sleep apnoeas or hypoventilating during sleep. The evaluation of the patient's problems may require a specialist opinion, especially if weakness of the respiratory muscles is suspected.

The principle of treatment for PPS is that excessive use of affected muscles should be avoided. This contrasts with the 'use it or lose it' theory of rehabilitation which was in vogue for polio 'victims' in the 1940's and 50's. At that time it was thought best to retrain the muscles as much as possible and to continue with exercise in order to keep them strong. In retrospect this may have contributed to overuse of the muscles and have increased the current prevalence of PPS. Advice about pacing activities, building in time for rest between these, reducing the intensity of exercise, and stopping before fatigue or pain develop, is important. Patients may benefit from relaxation treatment, passive stretching exercises and hydrotherapy or swimming, in which the weight of the body is supported by the water while the joints can be kept mobile. It is essential to try to enable the patient to continue with his or her activities, but to prioritise these and to carry them out in different ways. For instance, if the lawn has to be cut in the summer, this should be done in two or more stages with adequate rest periods rather than all at once. Activities around the home such as washing up should be done while sitting on a high kitchen stool rather than standing if the leg muscles are affected. The height of shelves may need to be altered particularly in the kitchen and broad-handled kitchen utensils used so that they can be gripped with less muscle strength than the conventional types.

Muscle activity can also be reduced by attention to posture and gait. Exercises to improve the mobility of the spine may help. Simple appliances such as a heel lift in a shoe can equalise the length of the two legs, reduce pelvic obliquity, and improve back ache and gait. Calipers for the limbs together with, for instance, lumbar spine supports or a walking stick may enable patients to stand or walk for longer intervals.

Patients who have had polio are often cardiovascularly unfit, particularly if their legs were involved. Individually assessed exercise programmes may improve this. Occasionally malnutrition leads to generalised muscle atrophy which can be reversed following nutritional advice. More commonly however weight loss is required. Obesity increases the energy requirements, may contribute to obstructive sleep apnoeas, and increases the load on damaged or weakened weight-bearing joints.

Mechanical aids or assistive devices may be used to reduce muscle activity. This may be as obvious as a wheelchair which enables the patient to become mobile without having to walk, or a stairlift which enables the patient to use the upstairs level of the home without having to climb the stairs. Carefully positioned handrails may perform a similar function and a walking stick may also be of help with mobility. If the upper airway or chest wall muscles become weak the external aid that is required may be a nasal continuous positive airway pressure (CPAP) system or a ventilator to support respiration. Nowadays this can usually be provided in a non-invasive form either as a nasal mask with a portable ventilator or occasionally as a negative pressure system such as cuirass. These are simpler than the traditional iron lung which was the only non-invasive system available during most of the polio

epidemics. Patients may need to be reassured that the need for ventilatory support does not imply the need for an iron lung again. Some patients who have a weak cough or recurrent aspiration may require cough-assistance techniques or even a tracheostomy to protect the airway and to enable ventilatory support to be given. Advice about the consistency of food, head position and tricks to facilitate the pharyngeal phase of swallowing helps to prevent aspiration, and to maintain an adequate nutritional state.

A variety of drugs have been tried in order to attempt to improve muscle function in PPS. These include pyridostigmine, an anticholinesterase inhibitor, which has been promoted in view of its ability to improve neuromuscular junction transmission. Studies of its use have shown that it is no better than placebo and it may even worsen PPS in the long term. Other drugs such as amantadine and selegeline which increase the availability of dopamine in the central nervous system have also been proposed, but have not been effective. Myotrophin, which influences nerve terminal sprouting has also been evaluated. None of these drugs can be recommended in the present state of knowledge. Oral steroids should be avoided in PPS if possible, because of the myopathy that they can cause.

Management of other late effects of polio

The indirect effects of the polio infection such as degenerative changes in the joints and soft tissues may require treatment with non-steroidal anti-inflammatory drugs and occasionally a surgical approach, particularly if weight-bearing joints become unstable. This is most commonly needed for the knee joints. Joint replacements may be required and occasionally even amputation of a useless limb, usually an arm, but it is unusual for a scoliosis in PPS to require a surgical approach. Many patients who had polio develop osteoporosis and preventative treatment should be considered for this. It is also important to assess the psychosocial implications of these late problems following polio, and for other members of the family or carers to understand the nature of the patient's problems, what measures need to be undertaken and what the outlook is likely to be. Depression may respond to tricyclic or SSRI antidepressants, but betablockers, benzodiazepines and other central nervous system depressants should be avoided.

Conclusion

With this broad-ranging approach the symptoms of the late effects of polio and in particular PPS often improve. Anxiety can be relieved and the previous way of life restored and maintained. The British Polio Fellowship, Ground Floor, Unit A, Eagle Office Centre, The Runway, South Ruislip, Middlesex, HA4 6SE, tel 0181 842 189, acts as a central organisation for patients throughout the UK who have had polio, and provides information and support which most patients find useful. It has local branches and has recently produced a video about the late effects of polio which is intended not only for patients, but also for those involved in their care. Hopefully this will increase the awareness of the new problems that are arising in patients who have had polio and will lead to better provision of care for them across the country.

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