42. Post-polio

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Summary

The most common post-polio symptoms include reduced muscle strength, a longer period of recovery for previously affected muscles, general fatigue and painful joints and muscles. This has brought about a new concept called the post-polio syndrome where new or aggravated muscle weakness is present in addition to other symptoms. Possible consequences of these symptoms are limited daily activities at work or in leisure time with the added risk of inactivity and general health-related problems. There is also a risk of overloading because of too much activity. Consequently, a suitable individual level of activity and a carefully monitored exercise programme are recommended.

Examples of suitable forms of exercise are water gymnastics and Nordic pole walking. Traditional fitness and strength training could be recommended for some individuals. Exercise will provide added strength, improved aerobic fitness and reduced pain.

Polio and its residual effects

Causes and prevalence

Polio or poliomyelitis is a viral disease marked by a gastroenteric inflammation which, in a small number of people, leads to an inflammation in the spinal cord and sometimes also in the motor neurons of the brain stem. As a consequence, paralysis occurs to a lesser or greater extent. Whether immunologic changes occur in the later stages of the disease is currently the subject of numerous studies and discussions. Actively administered vaccination programmes have led to polio being practically eradicated throughout the world,

although a limited number of outbreaks have occurred in recent decades, including Europe. However, polio is still present in some parts of Africa and Asia.

It has been known for more than 100 years that individuals who have recovered from acute polio may experience aggravated or additional symptoms following a relatively stable period of time, sometimes several decades after the initial onset.

Yet, the concept of developing strategies for clinical assessment and treatment only gathered serious momentum around 20 years ago (1). The industrialised countries have a significant number of people with post-polio residual paralysis. In Scandinavia, the last epidemic occurred in the first half of the 1950s. Asia, Africa and South America have seen more recent cases of polio. As a consequence, younger people with post-polio syndrome have arrived in the Nordic countries. Even if not all polio patients suffer from additional or aggravated problems – around 80 percent as indicated by clinical evidence and around 50 percent as indicated by population-based evidence – most of them seek medical care. In Sweden, around 10,000–15,000 people are estimated to have been diagnosed with post-polio syndrome. The prevalence of post-polio syndrome is probably even higher than these figures suggest.

Symptoms and underlying mechanisms

The additional problems are of a varying nature (2). Many are associated with an increase in muscular weakness, which is one of the most common post-polio symptoms. The added weakness may affect muscles previously affected by the disease, as well as those muscles in which no earlier weakness has been perceived. Increased muscular fatigue and difficulty in regaining muscular strength following muscular exertion are not unusual. In the event of muscular over-exertion, the individual may experience muscle pain during or after the exertion.

If the respiratory muscles have been affected by polio, causing breathing problems, the respiratory problems may be further accentuated by additional ventilatory problems. A state of underventilation may arise or be aggravated, mainly at night. Although this is a fairly unusual problem, it does lead to anxiety before a correct diagnosis can be made and will have an effect on the individual's general functioning.

Another recurrent problem is the general fatigue that many individuals experience in addition to muscular fatigue. This is usually due to a lower level of fitness, i.e. a reduced circulatory capacity because the patient is out of shape, but also because of an inappropriate degree of exertion, not enough rest periods or the training being of too long duration.

Another type of problem is associated with overloading and consequential instability in joints surrounded by weakened muscles. This problem is not necessarily directly linked to an additional or increased weakness of the muscles. Symptoms due to overloading can also occur without the muscular structure being significantly weakened, but where the load on the "better" side of the body is greater. Other problems that are perhaps a little harder to explain include reduced sensitivity and enhanced intolerance to cold. Psychological symptoms such as apprehension, anxiety, depression, irritability and concentration difficulties are also frequently reported.

The different organ symptoms and general fatigue, the onset of which may occur very quickly, may also restrict daily life activities as well as both work and leisure time. However, a relatively high proportion of post-polio patients in an active age continue to work (3). The problems in managing work are, however, accentuated with the onset of additional symptoms at which point more than half of those in working life report difficulties at work. Many with post-polio symptoms find it difficult to carry on with their normal leisure-time activities and the majority find alternative activities (3). In spite of this, many of these individuals are satisfied with their leisure time activities and have consequently adapted well to a reduced function.

The term post-polio syndrome (PPS) was created at the beginning of the 1980s when a more systematic assessment and recording of additional symptoms began (1). The criteria for diagnosing PPS have since changed somewhat according to the literature and additional or aggravated muscle weakness is no longer a common symptom, but a mandatory criterion. According to Gawne and Halstead (2), the criteria for the post-polio syndrome (PPS) are:

1. A confirmed history of paralytic polio including neurogenic changes as indicated by electromyography scan (EMG).
2. A stable period of at least 20 years following the initial recovery.
3. Additional or aggravated muscle weakness in muscles affected by polio, often with additional symptoms.
4. No other medical reasons for additional symptoms.

Specific muscular changes have specific names such as post-polio muscle atrophy (PPMA) and post-polio muscular dysfunction (4). Not all persons with residual polio symptoms are diagnosed with the post-polio syndrome since the diagnosis requires additional symptoms with increased muscular weakness. Nor is the post-polio syndrome included in the ICD-10 diagnostic criteria. However, the diagnosis code B91 "Sequelae of poliomyelitis" is included.

Muscular function

In case of an anterior horn nerve cell loss as seen in poliomyelitis, compensatory mechanisms in the form of collateral innervation (sprouting) are activated. A reinnervation of denervated muscle fibres occurs through the regrowth of sinuvertebral nerve endings from surviving motor axons. As a result, the remaining motor units will contain a significantly increased amount of muscle fibres. Collateral innervation is an important mechanism for the improvement of muscle function in the early stages of polio. However, denervation and subsequent reinnervation has been noted in some patients several decades after the onset of polio (5). This is interpreted as a loss of anterior horn cells or certain motor units losing a part of the collaterally innervated muscle fibres. These are then taken over by other expanding motor units. There are many different theories as to the cause of the anterior horn nerve cell loss such as aging or a shorter life span owing to over-utilisation or partial nerve damage as a consequence of polio.

The motor unit of the muscles affected by polio is enlarged up to 11 times in size which together with a double transverse surface of individual muscle fibres amount to more than five times the muscle fibres in a normal motor unit. Very large motor units constitute a risk factor for additional weakness (6). Some retrospective studies indicate that patients with an initially severe paralysis that is followed by a stable phase of considerable improvement, reasonably good function and level of activity are at a greater risk of a late onset of aggravated or additional muscle weakness (7).

Another essential compensation for loss of motor units is the growth (hypertrophy) of the remaining muscle fibres. As mentioned above, an approximate doubling is measurable. However, the degree of muscle fibre hypertrophy varies significantly and is probably dependent on the relative load experienced by the muscle in question. An increase in both type 1 and 2 muscle fibre areas may occur. Conversely, no significant compensatory increase in muscle fibres is noticeable in patients with near to normal muscle strength. A long period of "muscle over-utilisation", as seen in the tibialis anterior muscle when walking, can lead to an increase in the proportion of type 1 muscle fibres (8) as a consequence of fibre type transformation.

The two compensatory mechanisms, i.e. reinnervation and muscle fibre hypertrophy, allows for muscles with a considerable loss of motor units to sustain normal or near to normal muscle strength. Neuromuscular transmission disorders are thought to be one of the reasons for muscle weakness and increased muscular fatigue (9), but are unlikely to be the only explanation for muscle weakness.

A number of mechanisms could cause the muscular fatigue and lack of muscle endurance experienced by patients. Another likely reason is that patients with post-polio syndrome and an increased muscle weakness continue with the same absolute muscle activation despite a lower maximal strength. As a result, the relative load increases, resulting in intensified and sudden tiredness.

Another possible reason for muscular fatigue is an inefficient restitution following muscle activity (10, 11).

Joint structure and pain

Orthopaedic problems in general are fairly common in individuals with post-polio symptoms. Those that walk often report problems with their lower extremities while those using a wheelchair or crutches often report problems with their upper extremities due to the load placed on their shoulders and/or wrists, respectively. The patients that seem to be suffering most from muscle and joint pain have a moderately reduced function and comparatively high level of activity (12). There is a substantial need for orthoses and other types of orthopaedic aids, particularly orthopaedic shoes and inserts.

Principles of assessment and treatment

It is important to distinguish between late effects of polio or other more nonspecific symptoms of weakness or pain. Other simultaneous diseases with similar symptoms occur fairly frequently in post-polio individuals (13). A clinical examination should be carried out by a medical consultant experienced in treating post-polio patients, preferably at a specialist clinic.

Apart from identifying symptoms and reduced functions, the examination should incorporate an EMG scan to verify the extent of the polio disease. The individual's vital capacity is also measured to exclude or confirm the presence of polio effects on respiratory muscles.

No specific pharmacological treatment for muscular weakness and fatigue was previously available. However, in the past few years, clinical studies have been able to identify an inflammatory reaction in the central nervous system in post-polio patients – not a renewed progression of the polio infection. Therefore, trial treatments with intravenous immune globulin have been carried out resulting in improved muscle strength, an enhanced feeling of well-being, reduced pain and increased physical activity (14). However, this type of treatment is still experimental and only available under the supervision of a specialist clinic. It is imperative that pain caused by a relatively high level of activity, specific loading on unstable joints or biomechanical conditions are alleviated. The patient should be given guidance on the appropriate level of activity and the use of mobility aids, and orthotic devices should be prescribed and adjusted. Pain-relieving measures may be necessary in the form of antiflogistic and analgetic preparations combined with heat treatment and/or transcutaneous stimulation of the nerves or acupuncture.

Effects of physical activity

As is evident from the above, muscle fibre hypertrophy is the consequence of a daily and relatively heavy load on specific muscles. Hence, there is a significant "spontaneous" adaptation to the demands placed on an individual's physical activity. Priority should be given to the development of strength rather than endurance (17). The anticipated effect of strength training is an improved neural activation. However, initially large muscle fibres are unlikely to expand any further. Because the muscles in an extremity may be subject to a varying degree of strength reduction, the weakest muscles could be restricting the activity while the '"stronger" muscles become relatively inactive. The effects of physical exercise on such muscles will then be similar to those on inactive muscles unaffected by polio. There have been numerous discussions concerning the damaging effects of too much physical activity. It is not unlikely that an inappropriate activity intensity and duration may lead to increased weakness and fatigue that will last for several days. However, if discovered early, such possible "overactivity" should be reversible and an adjustment of the physical activity and exercise should be made.

The level of aerobic fitness is often reduced and worsened by reduced muscle strength, pain and inactivity. If possible, find a mode of physical activity where the muscle weakness is less restrictive than the circulatory capacity. Aerobic fitness will most likely improve

together with the endurance capacity of the muscles. The initial effect of physical activity is almost certainly peripheral with an improved muscle adaptation to aerobic exercise and enhanced efficiency. However, it is important to modify the exercise and physical activity programme to suit the individual in question.

Indications

Physical activity and exercise are only used for the purpose of secondary prevention. However, it is still not known whether a modified physical training of post-polio individuals prevents the development of new symptoms.

The symptoms of post-polio confers an increased risk of inactivity with the development of health problems. It is crucial to inform patients that inactivity can lead to aggravated symptoms such as increased weakness, pain and fatigue in addition to other conditions such diabetes, cardiovascular disease, osteoporosis and obesity. Hence, great importance should be given to promote physical exercise in the prevention of such conditions. It is also recommended to adapt the physical activity with the aim of sustaining and improving biomechanical conditions as well as maintaining the optimal level of aerobic fitness.

Prescription

Strength and muscular endurance training

Because residual effects of polio have a great symptomatic variation, particularly regarding the degree of reduced muscle function, it is important that physical exercise programmes are adapted to the individual patient. Clinical and functional targets should be clearly specified. The exercise programme should not just include the weakest muscles or muscles groups. Try different methods of optimal muscle training.

According to the documentation available, it is possible to exercise and strengthen moderately weakened and polio-affected muscles measuring > 3 on a scale of 0–5 (15). A number of studies indicate that weight training increases the strength of individual muscles. The load of the body itself, together with low-intensity training, has proven to benefit muscular function (16).

Exercise programmes should also include intervals of endurance training as spontaneous adaptations appear to prioritise strength before endurance (17).

Table 1. Recommendations for physical activity considering polio status and reduced strength (18).

Polio status	Muscle strength	Training
Stable	Normal	Without restrictions
Stable	Reduced	Short period of strength training (4–6 weeks)
Unstable	Reduced	Sub-maximal training
Unstable	Significantly reduced	Low-intensity training
Serious atrophy	Serious atrophy	No training

Stable polio does not refer to a subjective perception of progressive muscle weakness. In contrast, unstable polio does refer to a progressive muscle weakness. In the case of unstable polio, it is essential to determine whether there is over-utilisation or inactivity.

It is possible to exercise respiratory muscles affected by polio. Improved endurance is obtained by exercising the respiratory muscles once a day for a period of 10 weeks using an apparatus that provides different levels of inhalation resistance. Before and after each exercise, the patient uses his/her own ventilator for a minimum period of 30 minutes (19).

General rules: The patient's response acts as a training guideline. Initially, the training should be carefully monitored with shorter than normal training sessions. Several short sessions of training are better than one session of a long duration. A long duration of pain or tiredness (24 hours or more) after training is an indication that the muscle load needs to be reduced. A perceived exertion scale (20) can be used to stop the patient from exceeding a certain level during training.

People affected by polio have a longer post-exercise muscle recovery period than people with a normal muscular system. Consequently, a training frequency of more than twice a week is not recommended.

Eccentric strength training is not recommended as it involves a greater load compared to concentric strength training, which confers an increased risk of overloading.

Aerobic fitness training

Fitness training is possible for patients with sufficient strength in larger muscle groups. The basic principle of applied training involves a load of 60–80 percent of the maximal oxygen uptake/heart rate for a minimum duration of 30 minutes, usually twice a week.

If the larger muscle groups lack sufficient strength, a similar effect can be obtained through peripheral muscle strength training, i.e. low-intensity training with many repetitions.

Recommended forms of exercise

A number of examples of suitable forms of training are provided below with references to available scientific evidence. Table 2 lists documented scientific studies.

Table 2. Forms of exercise with documented benefits.

Form of training	Activity	Intensity	Frequency (times/week)	Length of training period	References
Aerobic fitness training	Ergometer bicycle training/legs	70% of max HR*	3	16 weeks	22
Aerobic fitness training	Ergometer cycling/arms	70–75% of max HR for 20 min.	3	16 weeks	23
Strength training	Dynamometer	Max. isokinetic 12 × 8 sec. Max. isometric 12 × 4 sec.	3	6 weeks	15
Strength training	Weights	10 RM** × 3	Every other day	2 years	24
Strength training	Weights	Dynamically controlled level of exertion gradually increased to extreme exertion	4	12 weeks	25
Combination programme (pneumatic resistance/light aerobic fitness training)	75% of 3 RM × 3	Sub-max.	3 2	10 weeks	26, 16
Water exercises		Sub-max.	2	5 months	21

* Max HR = maximal heart rate.

** RM = Repetition maximum. 1 RM is the maximum amount of weight one can lift in a single repetition for a given exercise.

- **Dynamic water exercise:** Training should take place in a heated swimming pool, preferably in a group session. The unloading of body weight in water reduces the load on muscles and joints. The water gently resists movement with many opportunities to vary and modify the training intensity. Various muscle groups are exercised and the water facilitates individual exercise programmes. Research indicates that sub-maximum exertion alleviates pain and reduces heart rate (21).

- **Using a bicycle ergometer:** Best suited for patients with good muscle strength in the lower extremities. The fitness benefits have been established (22). Outdoor cycling is only recommended for patients with very good muscle function. A good balance is vital and there is an increased risk of falling when mounting and dismounting the bicycle due to a reduced muscle function. Cycling uphill is not recommended.

- **Light aerobic fitness training:** Group exercises incorporating modified movement techniques in sitting or lying positions. According to one study, the result is an improved aerobic capacity and increased strength in specific muscle groups (16).
- **Swimming:** Exercising in a normally heated swimming pool is suitable for patients with good muscle function as well as competent swimmers. There are no documented studies on this, however.

- **Nordic pole walking:** Walking with poles unloads the lower extremities which is advantageous. Nordic pole walking is a safer way of walking, but requires good function of the upper extremities to avoid injuries due to overloading. There are no documented studies on this.

- **Weight lifting:** Provided that the exercises have been adapted and are carefully monitored, there is no reason not to do weight training. This type of training can work well as optional home training. Studies indicate improved strength without any muscle damage (15, 24, 25).

Advice and recommendations on physical activity

Experience has shown that those with post-polio symptoms have a fairly high level of physical activity (12). Consequently, the patient's physical activity routines should be reviewed and discussed when constructing a physical exercise programme. It is not usually necessary to change the daily physical activities carried out, although the degree of intensity may need to be reduced and the activities spread all throughout the day.

Appropriate physical activities should be recommended, also based on the patient's personal objectives. It is a question of the patient saving his/her strength for what is important in life and finding a balance between physical activity and rest. It is also important to recommend mobility aids and energy-saving measures for both work and leisure-time.

Functional mechanisms

Because compensatory mechanisms lead to an enlargement of the muscle fibres in muscles affected by polio, the increased strength gained through physical exercise is believed to be the effect of increased neural activity.

The improvement of aerobic fitness follows the same principles that apply to well-trained healthy individuals. In individuals with a significantly reduced muscle function, the peripheral adaptation of muscles with an increased oxygenation is most important.

Participation in a dynamic water exercise group means regular physical activity of a 40-minute duration which partly explains the pain relief experienced (21). It is also evident that taking part in a physical activity in itself has an alleviating effect on pain during and after the activity.

Functional tests

If deemed necessary, a standard health examination should be carried out in order to exclude cardiovascular disease or any other relevant diseases.

- **Functional tests** for the assessment of muscle function, e.g. various movements and standing up motions from different heights of sitting.

- **Muscle strength test**. A rough assessment of muscle function is possible using a scale from 0 to 5 when measuring muscle weakness (3 or below). A myometer can be used in the clinical settings if a more objective measurement is required. If a dynamometer is available for the measurement of isometric and isokinetic muscle strength, this should be used at the start of exercise and at some point later.

- **30 m walking test** at a self-chosen maximum speed. The duration of the session and the number of steps are recorded. If lower extremity muscle function is significantly reduced and the walking velocity slow, i.e. < 1.5 m per second at maximum speed, there is a risk of the walking velocity quickly slowing down if muscle function is further reduced.

- **Provided that the patient has sufficient muscle function, a standardised maximal or submaximal exercise test** should be carried out together with an electrocardiogram (ECG) if a cardiovascular disease exists.

- **Spirometry** for measuring respiratory function. Respiratory muscles may be affected by polio without the patient being aware of it.

- **Pain diagram and VAS** (Visual Analogue Scale) to identify the type of pain and its extent and intensity. The pain diagram provides a useful indicator of potentially overloaded muscles.

- **Different questionnaires** for the assessment of perceived quality of life and health. No disease-related questionnaire exists, but the Life Satisfaction Scale (27) and Nottingham Health Profile (NHP) (28) are both used. The latter produces a clear physical profile and is therefore suitable for this patient category.

Contraindications and risks

There are no known contraindications to correctly prescribed physical activity. However, it is important to have sufficient knowledge of the propagation and gravity of polio and to be able to recognize clinical indications of muscle overload. Physical activity should never lead to increased or additional pain. There is an insignificant and undocumented risk of a permanently reduced function because of muscle overload. The onset of osteoporosis is possible in which case the risk of falling must be taken into consideration when recommending physical activity. In case of a substantially reduced respiratory capacity, respiratory muscle fatigue may set in following exertion. As a number of patients with residual effects of polio have reached a mature age with the added risk of cardiovascular disease and high blood pressure, the physical exercise programme should be planned taking this into consideration.

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