

Finding causes of and managing fatigue in PPS

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Summary

Fatigue is the most frequently mentioned complaint of people with post-polio syndrome (PPS). And fatigue is often severe. However fatigue is not very specific and a prominent complaint in many neuromuscular disorders, in many chronic diseases such as multiple sclerosis, in oncology and even on itself in chronic fatigue syndrome. How to understand fatigue in PPS and how to deal with it is the aim of the two fatigue sessions.

In Session I the focus will be on the magnitude of the problem of fatigue in PPS, the different forms of fatigue and the factors playing a role in fatigue in PPS. In Session II the focus will be on the assessment and treatment of fatigue in clinical practice and what people with PPS can do to reduce fatigue.

Contents

Session I on Friday 11.15 - 12.30 AM covers the following topics:

- the problem of fatigue in PPS; prevalence and severity
- types and definitions of fatigue in PPS
- the mechanisms and factors contributing to fatigue in PPS

Session II on Friday 2.45 - 4.00 PM covers the following topics:

- how to assess fatigue in PPS
- how to treat fatigue in PPS
- energy conservation techniques

Causes of fatigue

Fatigue is mentioned by up to 80% of people with post-polio syndrome and the scores on fatigue questionnaires are often high, indicating severe fatigue. But, what is fatigue? Fatigue can be defined as 'a persistent, subjective sense of tiredness that interferes with usual functioning'. This refers to the general feeling of fatigue, however local muscle fatigue is also often present and is among the symptoms to define post-polio syndrome: 'new muscle weakness or abnormal muscle fatigability'. In post-polio syndrome fatigue is most frequently related to physical factors.

Local muscle fatigue

Local muscle fatigue is the decline in the ability of the muscle to generate force. Several factors have been identified that may play a role in this:

- I - Failure to drive muscles from the central nervous system due to alterations in the central nervous system to activate the nerve cells due to polio - the precise mechanism is not understood;
- II - Transmission failure from the nerves to the muscle fibers due to the fact that nerve connections with muscle fibers that were formed in the recovery phase after the acute polio are of less quality and therefore less able to sustain the transfer of the signal from the nerve to the muscle;
- III - Decreasing capacity of the muscles due to post-polio syndrome to meet the physical requirements needed to execute daily life activities. The muscles that slowly decline in strength have to work at an

increasing level of their maximal capacity and this will be inversely related to the duration physical activities can be maintained.

IV - A decrease in endurance properties of muscles. Muscles that are chronically used at a certain load, especially leg muscles, change their properties towards endurance, however not fully. Shortages of relevant enzymes have been reported.

V - Especially less and not affected muscles may be chronically under loaded in daily life and suffer from disuse. As a consequence they are less loadable.

General fatigue

General fatigue, the feeling of being tired, may have several causes.

I - People with post-polio syndrome may feel fatigued due to the fact that they are constantly acting above or in the upper range of their physical capacities. This may result in a chronic state of exhaustion. It is important to realize that movement efficiency is often reduced. This implies that walking may cost twice (or even more) the energy of normal walking in case of two affected legs.

II - Brain alterations due to polio virus damage have been suggested as a possible cause of general fatigue.

III - Recent studies have demonstrated signs of chronic inflammation in the cerebrospinal fluid in PPS. This may also play a role in fatigue.

IV - Deconditioning of the cardio respiratory system. Persons with post-polio syndrome have been found to be deconditioned, or to have a condition comparable to a sedentary life style.

V - Psychological factors, such as 'giving up the fight', social factors related to the persons life situation, and sleep problems may all contribute to fatigue. However, these are not the main causes of fatigue in post-polio syndrome.

Other causes of fatigue

It is very important to rule out other causes of fatigue. Of course the list of potential causes is very long, but a few common causes such as anemia, hypothyroidism, depression need to be mentioned.

Factors associated with fatigue in PPS

In a recent study, so far unpublished data, several of the above mentioned factors were found to be associated with fatigue. Lower physical functioning, more pain, sleep problems, lower well being and an active coping style were found to be associated with fatigue. Of course, in this study not all potential factors were included.

Pharmacological treatment

No pharmaceuticals have been proven effective in reducing fatigue. Randomized controlled trials (RCT's) in which drugs are tested against a placebo and both investigators and patients are blinded for the intervention are the gold standard to prove effectiveness of interventions. Results from such studies have so far been disappointing in that no drug was found to be effective.

Modafinil, a drug used in narcolepsy, was recently demonstrated as not effective in reducing fatigue in a study by Vasconcelos OM, *Neurology* 2008 confirming the negative results reported earlier by Chan KM in *Muscle and Nerve* in 2006.

Intravenous Immunoglobulines (IvIg) have been studied in two trials by Borg K, in *Lancet Neurology* in 2006 and by Farbu E, *European Journal of Neurology* in 2007. The study by Borg found effect for muscle strength and for 'vitality'. The study by Farbu found an effect for pain. Both studies however found no effect for fatigue.

Pyridostigmine, a drug that improves neuromuscular transmission was demonstrated not effective in two studies, one by Trojan DA in *Neurology* in 1999 and one study by Horemans HL in *Journal of Neurology Neurosurgery and Psychiatry* in 2003.

Other drugs were investigated in only one study involving limited numbers of patients. Negative results were found by Dinsmore S for high-dose *prednisone*, and by Stein DP, for *amantadine*, an anti-inflammatory drug, both published in *Annals of New York Academy of Sciences* in 1995. A study in 2005 by On AY, demonstrated a significant effect of *lamotrigine*, an antiepileptic drug, supposed to have neuroprotective properties, on fatigue. So far confirmative studies have not been published.

Finally, a recent pilot study by Skough K, in 2008, found no effects for coenzyme Q10.

Assessment of and Managing fatigue in individuals with PPS

Assessment

I Medical evaluation

It is very important to start with a thorough medical evaluation to exclude other pathologies as mentioned earlier.

II What is meant by fatigue?

The next thing is to go into a full consideration of the complaint of fatigue. Is it local or general, related to activity and which activities, does it increase over the day, does it respond to rest.

III Are other contributing factors present?

Consideration has to be given to sleep quality, mood disorders and coping styles.

IV What is the activity pattern?

The activities of daily life have to be inventoried. What is the activity level of a person, what kind of work does someone do, what are social and home activities, how is mobility outdoors. Are there any aids being used for walking, mobility in and outdoors and so on.

V What is the social system?

How is the person's social environment, does he or she have sufficient support and understanding at home and work.

VI What are the own perceptions?

How does someone value his or her complaints of fatigue (and other complaints, post-polio syndrome, polio residuals and so on).

VII What are the physical capacities?

What is somebody able to do given the polio residuals and co-morbidities. What is the physical burden of activities such as standing, walking, transfers, stair climbing, and the individual's various activities.

VIII Conclusion

Finally a conclusion can be made on which factors cause or sustain fatigue. These are to be targeted in interventions.

Assessment tools

Tools that may be of value in the assessment of fatigue are validated questionnaires to assess fatigue severity, pain, coping styles, mood, and physical functioning.

Diaries to inventory daily life activity are extremely useful to gain insight in what someone's life looks like, in what a person actually does over the days. A common finding is that people with PPS appear to be much more active than they spontaneously report.

Clinical tests may be included to determine the extent of the polio residuals, and capacity tests to assess physical abilities.

Management

The first important thing is that the person obtains insight in the factors contributing to fatigue. The next and crucial thing is readiness to change. Many factors contributing to fatigue are related to behavior and to cognitions. For instance, if someone is constantly overusing oneself, but considers that as normal, it will be impossible to obtain any change in behavior. It is well known that many polio survivors are so-called 'over achievers' who are not easily prepared to reduce their activities.

Occasionally, the reverse is also seen that some polio individuals may avoid physical activity, for instance if they think that this may damage muscles, and they may very well be mainly fatigued due to the vicious circle of fatigue, inactivity, physical deconditioning and so on.

To diminish fatigue energy conservation skills are often to be learned. This may be done individual or in group therapy programs. On the other hand regular physical activity is advised to maintain physical functioning. This implies an individual non-fatiguing exercise program that can be easily done at home, or in an (adapted and accessible) fitness setting. Environmental adaptations at home or work, transportation aids, braces and assistive devices may all be needed tailored to the individual's needs. Rehabilitation therapy is therefore usually multidisciplinary organized and may involve physical and

occupational therapists, social workers, psychologists, orthotists, shoe technicians and adaptation technicians.

Preferably, the effect of multidisciplinary interventions to reduce fatigue should evaluate the achievements obtained after the program and during follow-up.