

BRITISH POLIO FELLOWSHIP EXPERT PANEL – DEFINITION OF POST POLIO SYNDROME

Summary Level Definition

“Post Polio Syndrome (PPS) is a neurological condition that can occur in people who have had polio. After an interval of several years of stability, people may then develop increasing weakness, stamina problems, fatigue and pain. PPS may respond to a range of therapies which might prevent further deterioration”.

Symptoms include the onset of new weakness or abnormal fatigue in previously affected or unaffected muscles; a general reduction in stamina; muscle and/or joint pain; muscle atrophy; breathing, sleeping and/or swallowing problems; or cold intolerance. Symptoms may lead to loss of endurance or function.

A diagnosis of PPS assumes the absence of any other conditions that could explain the above symptoms. Such conditions should be considered and excluded by appropriate investigations”.

Operational Definition

The following operational definition of Post Polio Syndrome supports and expands upon the summary definition above. It is derived from consensus statements based on the extensive review of various medical and scientific literature, studies and previous definitions.

As there is no diagnostic test for Post Polio Syndrome it is commonly defined by a **symptom complex** that includes new muscle weakness, decreased endurance, pain and fatigue.

The onset of PPS may be gradual or it can occur suddenly. It occurs irrespective of ageing. Symptoms may sometimes appear to be triggered by various events like surgery, falls or immobility.

The following are the main criteria that need to be considered in making a diagnosis of “Clinically Definite PPS” or “Clinically Possible PPS” and are also shown graphically in the table below:

▪ **Clinically Definite PPS**

“Based on various consensus statements, a diagnosis of “Clinically Definite PPS” comprises a confirmed history and/or physical evidence (however slight) of polio, a period of functional recovery and stability, new muscle weakness, or abnormal muscle fatigue, with evidence of neurogenic change, and the exclusion of any other possible conditions”.

▪ **Clinically Possible PPS**

It is recognised that there are patients who have PPS symptoms, but there is less diagnostic certainty. This would include patients with the following characteristics - a possible history of polio where there may be no previous physical manifestation, new muscle weakness, or abnormal muscle fatigue, (with no evidence of neurogenic change), a complex of symptoms that are generally recognised to be those for PPS, and the exclusion of any other possible conditions.

Diagnosis criteria (See below for more detail on each of criteria)	Clinically Definite PPS	Clinically Possible PPS
1. Definite history / physical evidence of polio	√	
2. History of possible polio		√
3. Period of recovery and stability	√	√
4. New muscle weakness – with evidence of neurogenic change	√	
5. New muscle weakness – no evidence of neurogenic change		√
6. Appropriate complex of symptoms	√	√
7. No other disorder / medical explanation	√	√

1. Definite History of Polio / Physical Evidence

The patient's original medical records, history and/or physical evidence provide a confirmed diagnosis and history of the original polio illness.

2. History of Possible Polio

Some people may not have confirmation of prior polio or a physical manifestation of the illness, but do have some history and/or current symptoms, which taken together, indicate a **possibility** of polio.

For example, this could apply to a patient in whose family, or circle of friends, there was an incidence of polio, or who is known to have come from an area where there was a polio epidemic or outbreak, and/or who suffered an illness that at the time was not diagnosed as polio.

3. Period of Recovery and Stability

Partial or fairly complete neurological and functional recovery after the original polio illness followed by a period of neurological and functional stability. As guidance only, the period of stability will generally be 15 years or more.

4. New Muscle Weakness – with evidence of Neurogenic change

Muscle weakness may be confirmed clinically by the presence of clear lower motor neuron features. When and if available / appropriate, EMG testing may confirm this and establish a baseline for repetitive testing or offer alternative diagnosis. Other tests that may be useful are nerve conduction tests to assess nerve damage, Manual Muscle Testing (MMT), reflex and exercise testing for endurance. Other causes of neuromuscular weakness will also need to be excluded by appropriate tests.

Although it is accepted that electro diagnostic testing has limitations in confirming neurogenic weakness, and will not provide a definitive diagnosis of new weakness, it may help to exclude some of the other common causes of neurogenic weakness as well as other, more rare, conditions.

5. New Muscle Weakness – No evidence of Neurogenic change

A patient who has a possible history of polio may be experiencing new weakness that testing cannot confirm to be neurogenic (see Point 4) but is consistent with the symptoms of PPS.

6. Appropriate complex of symptoms

These may include two or more of the following health problems occurring after the stable period: extensive general fatigue, abnormal muscle fatigue, decreased endurance, muscle pain, joint pain, new weakness in muscles previously affected or unaffected, new muscle atrophy, functional loss, breathing or swallowing problems, cold intolerance.

7. No Other Disorder / Medical Explanation

Exclusion of medical, orthopaedic, and known neurological conditions that might cause the health problems listed in Point 6 above, although these other conditions may coexist with PPS. Depending on symptoms, the most obvious possible causes to rule out are orthopaedic problems related to the original polio; injuries, breathing problems, other neuromuscular diseases, and other diseases that commonly cause fatigue, such as thyroid problems, diabetes or heart disease.

EMG testing and other relevant tests (see Point 4 above) may be used as a means to exclude other known neurological conditions that may present similar symptoms.

Main symptoms of PPS

With reference to Point 6 in the criteria for diagnosis above, the following are considered to be the most common health problems resulting from PPS.

▪ Weakness

New weakness is often seen as the most recognisable symptom of PPS.

It can sometimes be difficult to separate weakness from muscle fatigue, which could be thought of as 'weakness that develops over time' or through usage.

Recent definitions of PPS usually include both weakness and muscle fatigue (also known as peripheral fatigue, or stamina or endurance problems) as essential symptoms. Both symptoms can occur not only in muscles previously known to be affected by polio, but also in muscles where no damage has been apparent until now.

Studies have varied in their findings but have shown for example: weakness in around 2/3 people with polio, weakness in previously affected muscles in up to 85% of people with polio, and weakness in previously unaffected muscles in up to 58% of people with polio.

▪ Fatigue

Post Polio fatigue is often experienced as two separate types of fatigue: a general, sometimes overwhelming, exhaustion (which may include mental fatigue), and localised muscle fatigue, often described as increasing physical weakness, loss of strength and endurance during exercise, and a heavy sensation in the muscles.

Muscle fatigue after even minimal exercise can lead both to increasing weakness and the aching that is common in PPS. It may lead to general exhaustion or fatigue at the end of the day or it may even last for several days. Muscle fatigue can be a result of the muscle overuse that is thought to play a large part in PPS.

General fatigue is an overwhelming feeling of exhaustion and weakness. Sometimes mental fatigue can be the result of muscle fatigue and muscle overuse. Fatigue can

also result from sleep disturbance, sleep apnoea (stopping breathing for intervals) or breathing problems.

Rest usually relieves fatigue, unless it has been building up for days or more, when it may take longer to feel better.

General fatigue has been found in around half of people with polio¹⁴, increasing to 80% for fatigue during exercise.

There is also a theory that any brain stem damage during the initial polio illness can cause increased lack of energy and alertness, sleepiness and concentration problems later on.

▪ **Muscle pain**

Muscle pain is very common and is usually described as aching, especially after activity, or felt as burning, spasms or cramps. This pain may be the result of muscle overuse and may occur with twitching or fasciculations, especially later in the day or at night.

Research has shown pain can relate to activity levels, and particularly the intensity of activities, which is usually higher in people who had polio as they are using their muscles at their maximum much of the time.

▪ **Joint pain**

Where joints are no longer held in place by strong muscles, they may become unstable, resulting in joint pain. It may also be caused by injuries to the tendons or ligaments due to overuse of unstable joints. Weakness and injuries around joints may also lead to the pain of compressed nerves.

▪ **Muscle loss**

Also known as muscle atrophy, this loss of muscle bulk has been found in 20-30% of people with PPS.

▪ **Sleep disturbance**

Sleep disturbances are common and may relate to sleep apnoea, breathing difficulties, pain, muscle twitching or general overtiredness. Sleep apnoea (lack of oxygen because of not breathing well or often enough) can be due to repeated shutting of the airway from throat weakness, sometimes made worse by weight gain. It can also occur when there are brief interruptions in the brain reflex that triggers breathing.

▪ **Breathing problems**

Breathing problems are more common in people who needed help with their breathing when they had polio, but can occur later on in anyone with polio. They are caused by muscle weakness of the diaphragm and chest muscles, scoliosis or sleep apnoea.

Breathing problems may develop very slowly. Early signs may be frequent waking from sleep, sometimes with choking or gasping; nightmares, snoring, morning headaches, coughing, daytime sleepiness, difficulty speaking at length, lack of concentration and breathlessness with exercise.

It is extremely important for both doctors and people with polio to understand there may be breathing problems later on, and these need thorough assessment.

▪ **Swallowing problems**

Weakness in the muscles used for chewing and swallowing may lead to frequent choking, gagging, or food becoming stuck in the throat. There may also be voice and speech changes, such as hoarseness or a nasal sounding voice, especially after speaking for a while or when tired.

Research into swallowing problems has shown slowly progressing new weakness even in some people who were not aware of any change and had not originally had bulbar polio. Swallowing muscles do not usually weaken over time², so this convinced many doctors that PPS occurs irrespective of ageing.

Most often, swallowing problems stay mild and progress only very slowly. Advice from a speech and language therapist may prove helpful.

▪ **Cold intolerance**

Sensitivity to cold, like many PPS symptoms, may be felt in one area of the body for example in an arm or leg which may have a poor blood supply, or may be felt as a general cold intolerance, or intolerance to any large temperature change.

▪ **Other symptoms**

As there has not been enough research into PPS, there are many other symptoms that may or may not be related to polio. When other possible causes are ruled out, polio is sometimes thought to be the cause. These symptoms may include balance or dizziness problems, face or eye muscle weakness, digestive or urinary problems relating to polio damage.

If the autonomic nervous system (the part of the nervous system controlling muscles people do not have to think about moving, like the heart, stomach or bladder) has been affected by polio, then generalised cold intolerance, blood pressure and heart rate problems may occur.

Further sources of information

As well as the clinical publications, studies and papers on the subject of Post Polio Syndrome, further information can be obtained from The British Polio Fellowship, FREEPHONE 0800 018 0586; Website: www.britishpolio.org.uk or email: infobenefits@britishpolio.org.uk.

References

References include:

- Farbu E, Gilhus N E, Barnes M P et al. EFNS guidelines on diagnosis and management of post-polio syndrome. Report of an EFNS task force. European Journal of Neurology 2006.
- Halstead L S. Assessment and Differential Diagnosis for Post-Polio Syndrome. Orthopedics 1991.
- Hughes R, Brainin M, Gilhus N E. European Handbook of Neurological Management 2006.
- Jubelt and Agre. Characteristics and Management of Postpolio Syndrome. JAMA 2000.
- Ramaraj R. Post-poliomyelitis syndrome: clinical features and management. British Journal of Hospital Medicine, Dec 2007.
- The British Polio Fellowship. Post Polio Syndrome. Guidelines – summarising clinical guidelines for primary care 2007.

Expert Panel Members

The Expert Panel members who contributed to the above definition are:

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