

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:

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- 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:

Sir Bert Massie (Chairman)	Chairman of the Disability Rights Commission
Dr Steve Sturman MBChB (hons), FRCP	Consultant in Neurology & Rehabilitation Medicine, City Hospital, Birmingham
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Dr Ros Sinclair	Chartered psychologist and British Polio Fellowship Trustee
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Post-Polio Clinics Directors Network
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Disclaimer: The following are unofficial notes which have not been read by or approved by the speaker.

- Purpose of this call is to tell about the British Polio Fellowship – why and how the definition was developed and queries to date from other countries.
- The British Polio Fellowship (BPF) was established 69 years ago. There are approximately 8,000 members; 20 on the staff. The Fellowship offers support, information, welfare, counsel for people's rights.
- The British Government said there are 120,000 people in the UK with polio. BPF believes the number ranges from 60,000 – 80,000 but uses the larger number.
- The number of people with polio who lead normal lives but are now being affected by PPS are coming back.
- A recent survey of BPF members shows that it takes an average of six years for them to get a diagnosis.
- An "expert" panel of health professionals, laymen with polio and other allied professionals was established. BPF hopes with a better definition, people could be diagnosed faster and get on to the right treatment.
- BPF gathered all the published definitions they could find. All had a lot in common but none stood out to be helpful for diagnosis for a general practitioner or a lay person who wanted to know if they had PPS or not.
- It could not be proved in a large number of cases that people actually had polio – some were never told they had polio or there were inadequate medical records.
- A matrix of diagnosis criteria was developed. Originally it had three columns – Clinically Definite, Clinically Possible and Clinically Probable.
- Due to a lot of resistance from medical professionals, the "Clinically Probable" was dropped.
- BPF did not set a number of years of stability because there is no universal agreement on that number.
- The definition is considered to be a work in progress. BPF would like feedback and see how to get further consensus.
- The definition was circulated widely in Europe. After it was published, they received some praise and some queries.

- Sweden does not like "Clinically Possible". Netherlands does not like "Clinically Possible". England (Dr. Spencer) thinks it should include dizziness, contrary to what the Netherlands thinks.
- Since agreeing to a definition it was placed on an internet site that general practitioners can access.
- Now developing medical education courses for healthcare professionals.
- The BPF would be grateful to receive verbal or written comments on the definition; would like to get as much consensus as possible.

Question: The elimination of the "probable" category, besides the objections wanting to have the black and white, yes or no, were there specifics as to why people objected to that?

- BPF wanted to be as inclusive as possible so patients can be put on Patient Pathways to be treated.

Question: The "Possible" would include people who did not have confirmed polio?

- Yes.

Question: What about the people who do have confirmed polio but you cannot rule out other factors?

- They do not want to put down "Probable" when they have to justify in court that someone has not received the right diagnosis.

Comment: After hearing the presentation, it is understood that the major concern of the BPF is trying to help the people in the UK get appropriate treatment. It fits the needs of the BPF, it's logical and makes sense.

- It's crucial for BPF to give general practitioners who know nothing about PPS the ability to look up a definition and move the people in the right direction.
- The BPF has not added anything scientific but hopes that their expert panel can strengthen the issues addressed here. Any biomarkers the group can help the BPF with to make the definition better would be appreciated.

Comment: One of the simplest things to do would be a pilot study among general practitioners and see what kind of consensus BPF can get.

- BPF did a survey of general practitioners and 17% said they might be able to diagnose.

Comment: It was suggested BPF show them the definition and ask again.

- This is the sort of thing BPF would do in the courses they want to devise.
- Good idea to give the definition and see if BPF could get consensus from that.

Comment: In the States we were trying to do something that would fit the ICD-9 codes. There is no way to differentiate reliably what are late effects or post polio. There wasn't really any reason for that unless you had a code. The other reason is we wanted a clinical definition so that everyone would agree that you have certain things. We want it for research studies.

Comment: When the label "PPS" is used, people stop looking for anything else. Under the BPF it is less of a concern because the end result of what the patient is looking for once they get that definition is entry into the specialist and hopefully the specialist can make the determination to rule out the other things.

Comment: One of the original discussions in the '80s was raised by Dalakas - PPMA Post Polio Muscle Weakness. However, it applied to too few people.

- The BPF is trying to come up with something general practitioners could relate to and have people move on

Question: Would this be helpful for Sweden, Spain and France? Spain doesn't acknowledge PPS.

- BPF circulated the definition hoping that they all could use it in some way. No feedback from Germany.

Dr. Maynard will forward the definition to Verner Reich and Mr. Ball will follow up.

Comment: Agree with "Clinically Definite" and "Clinically Possible". In a sense at least it throws out the other definition problem.

- BPF wants to be as inclusive as possible.

Comment: Concern was expressed about Point #7 – oftentimes the patients come to the clinic with medical, orthopedics, or conditions that may cause the pain. Some people may latch on to it; the Social Security Administration may latch on to that and that causes concern.

Question: Have noticed the issue which is in March of Dimes criteria – symptoms persist for at least a year is not included. What are your thoughts?

- BPF does not have an answer for that one. Will look at the research.
- BPF has seen figures less than 15 years – 8 was used.

Comment: 15 years was decided as a compromise. The disadvantage of putting the number is there is no basis – "several" is reasonable. If it is a year, it helps you make certain that it is not another disorder.

Comment: When patients from Asia or Africa are evaluated, complete with good history of their present problems in some cases they do not have the 15 years in yet.

Comment: Third World Country patients are usually very complex from the standpoint that they had a different experience with initial rehab.

Question: Does the BPF use the term "late effects" of polio?

- BPF sees it as an "or" late effects. It is mentioned in their literature.

Comment: When looking at topics at the conferences, it is really about late effects of polio. They call it PPS.

Comment: In Denmark they try to differentiate between late effects and PPS. When there is new evidence of weakness it is called PPS.

They differentiate between how they exercise the patient. If the patient has PPS they are not trained so hard. If they have late effects of polio and their muscles are stable they are trained hard.

In Denmark, they use a manual muscle test and usually see the patient over the years and listen to what the patient tells them (cannot do the stairs any more). Then they do not train them so hard and address lifestyle changes or suggest wearing a brace. Each patient is treated individually, listening to all their problems.