

Respiratory issues in the Polio and Post-Polio Syndrome population; causes, symptoms, testing and management.

Introduction

This factsheet is for anyone who had polio or has Post-Polio Syndrome (PPS) who would like to learn more about respiratory complications due to polio or PPS and how they may be managed.

Post-Polio symptoms

People who had polio years ago may now be experiencing new symptoms. These can include new or increased muscle weakness and fatigue with or without other symptoms like muscle and joint pain, muscle atrophy or wasting, breathing or swallowing difficulties or cold intolerance.

There is more information on PPS in other British Polio Fellowship factsheets.

Respiratory complications and polio

Not everyone with a history of polio will have respiratory issues. However, with the onset of PPS or even just with ageing, breathing issues may either become more apparent or develop, so it is worth considering if you think these might be affecting you. This can be the case even if you did not need support with your breathing during the original polio episode.

Polio survivors may have weak respiratory muscles and/or a restricted chest cavity which reduces how much air can be moved in and out of the lungs (ventilation). In the blood stream, this can lead to low oxygen levels and/or high levels of carbon dioxide (a waste product of breathing). If sufficiently serious, this can lead to respiratory failure; type I due to low oxygen levels, and type II due to high carbon dioxide levels, often also with low oxygen.

Weak muscles can also make it difficult to cough strongly enough to clear the lungs which can lead to chest infections or further carbon dioxide being trapped in the lungs by phlegm.

These changes can come on slowly and be hard to recognise.

A small proportion of people will have had on-going respiratory issues since the original polio. Weakness of the breathing muscles meant they continued to require breathing support through machines that supported ventilation either at night or throughout the 24h period. These people will be managed under a medical service and require regular and ongoing checks to make sure the mechanical support remains at the right level as it may need to be increased over time.

For those who have previously not been diagnosed with breathing issues, other risk factors such as smoking will also influence the likelihood of additional respiratory-related disorders. Breathing problems can also be signs of other problems such as issues with the heart; any new breathing problems should always be checked with your GP.

Symptoms to look out for that suggest you might have a respiratory issue include:

- Shortness of breath
- Feeling like you can't take a big breath
- Morning headaches
- During the daytime feeling very sleepy
- Sleep being disturbed, possibly waking with a gasp
- Bed partners reporting snoring or stopping breathing in your sleep
- Difficulty clearing phlegm with or without a chest infection

Signs that you're breathing ability has become affected:

- Faster breathing rate brought on by low levels of physical effort
- Your stomach and chest movements become uncoordinated when you breath in and out
- Your chest is not expanding well
- Using other muscles such as those of the neck or needing to support your chest by pushing down through your arms to get a big breath

Potential new respiratory issues related to polio

There are four possible respiratory issues related to having had polio.

- a) Breathing becoming disturbed when asleep (sleep apnoea and hypopnea)**
- b) Chest muscle and abdominal muscle weakness**
- c) Restriction of the chest wall**
- d) A reduced ability to clear secretions**

It would be expected that long term issues with breathing become more apparent at night before any issues are noticed with breathing during the day. This is due to the reduced number of muscles working when you are asleep, as well as the position of being flat, making it more difficult to breathe. However, because the changes can occur slowly over time you may not notice breathing is becoming more difficult. Therefore, people whose breathing is challenged by their restrictive chest wall or muscular weakness may not be picked up until a crisis arises in their breathing, generally a chest infection.

a) Breathing is disturbed when asleep (sleep apnoea and hypopnea)

Obstructive sleep apnoea (OSA) is a complete break in breathing while sleeping due to restriction of the upper airways, lasting 10 seconds or more and can occur repeatedly through the night. Hypopneas are reductions in the flow of air by at least 30% lasting 10 seconds or more. Both are characterised by being sleepy in the day, fatigue, snoring and sleep not being refreshing. For people with PPS, poor sleep quality can contribute to fatigue symptoms and it requires assessment to see if your fatigue could be partly due to sleep related breathing disturbances. One tool often used to measure this is the Epworth Sleepiness Scale.

If you share the bedroom with a partner they might see and hear that you snore and hold your breath, then gasp to start breathing again. Some changes to breathing overnight is normal but when it happens frequently enough, it causes you to partially wake up and then disturbs the quality of sleep.

OSA and hypopnea syndrome are found relatively frequently in the general population with a higher number of cases in men than women. There is some evidence it is more frequent in a PPS population than in the general population.

It is highly linked to being overweight and having a larger neck. Any extra bulk around the neck can make this problem worse as the muscles relax during sleep and the weight of additional neck mass makes it harder to breathe. The position of the lower jaw can also impact, if it is set further back it is more likely to put pressure on the airway when relaxed. The apnoeas/hypopnoeas don't happen during all of sleep but just the deepest parts of sleep known as Rapid Eye Movement sleep (REM). During REM sleep, the only muscle functioning to maintain your breathing is the diaphragm under the ribs, as the other muscles switch off during this sleep period. The muscles and other tissues of the neck become floppy and narrow the space for breathing, so obstructing breathing completely every few breaths. Therefore, any additional upper airway resistance or mass and any diaphragm weakness will have a greater effect in REM than during other periods of sleep.

In order to diagnose breathing issues at night, a sleep study or polysomnography is carried out. Usually this involves wearing a probe on a finger overnight to monitor your oxygen saturation levels and also heart rate.

If the level of disturbance to the sleep is high, treatment will be considered by the medical team. Recommendations to lose weight might be one line of treatment. A breathing machine called Continuous Positive Airway Pressure, usually shortened to CPAP, delivered via a mask over the mouth and nose might be indicated. If this is the case you would be seen in clinic to trial the machine, learn about it and get used to the mask. For people that can't manage the CPAP, a device worn in the mouth to change the jaw position 'a mandibular advancement device' might be helpful.

There are some reasons why your brain might disturb your breathing at night, termed central sleep apnoea (CSA), but this is rare and no more common in those with polio compared to a population without polio.

b) Muscle weakness leading to problems breathing

Most people are aware that inadequate breathing (underventilation or hypoventilation) can lead to low oxygen levels. As with other neuromuscular conditions, PPS related weakness can also lead to too much carbon dioxide being retained in the lungs and high levels in the bloodstream (hypercapnia). This usually begins at night when the muscles do not work as effectively. Several of

the symptoms are similar to obstructive sleep apnoea which may lead to misdiagnosis.

If your breathing muscles are becoming weaker you might notice that it is harder to breathe in some situations. This could include while lying flat, going up stairs, when asleep, after putting on weight or during a chest infection. Pregnancy is another situation when there is more stress on the breathing system. Muscle weakness can lead to respiratory failure if the impact on the breathing system is great enough. However it is relatively uncommon in those with a polio history that respiratory failure progresses to needing requiring ventilator support during the day.

How much your breathing is affected will depend on the level and pattern of muscle weakness. The weakness might happen on its own but is often in combination with the chest wall deformity described in section c) Restriction of the chest wall.

If there is any indication your muscles might be getting too weak and can't adequately support your breathing (such as the signs and symptoms listed above), the medical team could use a number of tests to check for this. See table 1 for a list of tests that might be used to investigate respiratory issues. The tests can be used to check oxygen levels, carbon dioxide levels or muscle strength.

Table 1. Tests that can be used to help diagnose respiratory issues

Test	Oxygen level	Carbon dioxide level	Muscle strength
Oxygen Saturation finger probe	yes		
Capillary or arterial blood gas	yes	yes	Can be implied from results
Lung function tests		Can be implied in some cases	Can be implied in some cases
Respiratory Muscle testing		Can be implied in some cases	yes
Overnight oximetry	yes		
Capnography or transcutaneous CO2		yes	

Treatment is again linked to the severity of the effect on your ability to adequately manage gas exchange during breathing. If it is indicated, non-invasive mechanical ventilation (NIV) delivered via a mask will assist gas exchange. Ventilation may well only be required at night time and can be a single level (CPAP) or Bi-levels of pressure (BiPAP) to support breathing depending on the type of respiratory failure you have. With BiPAP ventilators, inspiratory and expiratory pressures can be adjusted separately according to the person's breathing needs. People with weak diaphragms may have difficulty breathing in, so may need the inspiratory pressure to be set at a higher level than the expiratory pressure, to help them take in as much air as possible without making it harder to exhale. The symptoms of hypoventilation can be improved using BiPAP ventilators. Unless you also have other lung diseases, oxygen is not normally indicated either with the machines or alone.

c) Restriction of the chest wall

The ribs should move freely in what is described as a bucket handle action with each breath. They should open out and lift up from the trunk creating more space in the lungs. For some people, polio affected the nerves related to the muscles of the trunk. As children grew, any imbalance in muscle strength could affect the growth and shape of the bones leading to unusual spinal curves and twists known as kyphosis, scoliosis or a combination of both. The resulting altered spine shape reduces the ability for the rib cage to produce a 'bucket handle' action on breathing in. This, combined with any weakness, leads to restriction of the chest wall movement, i.e. a reduction in the size of deep breath. Over time, the chest wall can get very stiff and even more restricted. This means the lungs don't open as well, the amount of air moved with each breath is reduced and there can be areas of lung that remain collapsed and never take part in gas exchange during breathing.

Again, the person may be able to breathe well enough under normal circumstances but struggle during times of increased demand, such as a chest infection or an increased load to move against such as constipation. Reduced movement and an increased load can lead to respiratory failure. As people struggle to get in big breaths, rapid shallow breathing might be seen. This can indicate type I respiratory failure. When severe this can quickly lead to

exhaustion and on to type II respiratory failure. Signs of any impact on carbon dioxide in blood gases will usually trigger a trial of NIV.

d) Reduced ability to clear secretions

Polio is not associated with an increased production of phlegm but removal of secretions during a chest infection can be an issue if your cough strength is poor. As with breathing muscles, muscles involved in coughing may be adequate during normal circumstances but may not be strong enough when faced with the additional burden of a chest infection. An effective cough needs not only adequate volumes of air taken in (phase 1) but coordination in the throat to close the glottis (phase 2) and abdominal strength to build up the pressure against the glottis that gets released quickly as a cough (phase 3). This generates a large enough force to clear secretions to the upper airways and out of the mouth. A reduction in muscle strength in the trunk and abdomen could affect the 1st or 3rd phase while a reduction in control of the glottis could affect the 2nd phase.

An inability to clear secretions puts people at risk of chest infections and a build-up of secretions can add to any chronic areas of lung collapse and eventually impact on the ability to breathe.

If you do find you are producing phlegm and struggle to clear it, this does need assessment. Physiotherapists can do this and give advice on secretion removal techniques which might include the 'active cycle of breathing', a combination of breathing in, holding and sharp breaths out, gravity assisted secretion drainage, or finally huffing, a type of breath out. It will need exploring with you to make sure you can still use the prescribed technique even when you are at your most tired.

Other techniques that have been shown to be useful for some people with polio and problems clearing secretions include:

- Being taught how to breath stack (either independently using glossopharyngeal breathing or with a device called a lung volume recruitment bag) to get a bigger breath in
- Manual assisted coughing (MAC) to aid expiration. It should be considered that MAC normally requires assistance from another person

and can be uncomfortable as you have to apply pressure to the stomach or lower chest, timed with the cough to get the effect.

- For patients with very thick secretions, a type of medication to break secretions down, mucolytics could be considered. However, this will need to be monitored to assess if this truly makes clearance easier.
- The use of mechanical devices such as the mechanical insufflator/exsufflator (MI:E) to assist your cough. This not only pushes air in via a face mask, to give a bigger breath, but also sucks the air out to help the secretion removal.

All of these techniques require assessment from a health professional trained in their use, usually a physiotherapist. Advice will then be given on the most effective technique for you and will need to be monitored over time.

How to get help

If you notice any of the symptoms listed previously, particularly if they persist for more than a couple of weeks, keep a record of your symptoms and talk them over with your GP (including details of your polio history and any neuromuscular weaknesses). Your GP may order some tests or prescribe medication, for example antibiotics if infection is suspected, before perhaps making a referral to a physiotherapist or respiratory specialist.

Not all new breathing difficulties will be due to previous polio or PPS and should always be investigated thoroughly.

There are some things you can do for yourself, such as positive habits to improve sleep, good nutrition, improving posture, avoiding smoking or smoky areas, avoiding alcohol especially before bed and losing weight.

To summarise

There are a number of reasons why people with polio may suffer from breathing issues. These include the breathing becoming disturbed when asleep, muscle weakness, restriction of the chest wall or a reduced ability to clear secretions.

These breathing issues may not have been present previously and not everyone who has had polio will develop them.

Generally new symptoms will come on slowly, but you may not notice the impact until you have an additional burden on your respiratory system, such as a chest infection.

Any new signs or symptoms should be investigated bearing in mind the background of polio and potential for muscle weakness.

Medical disclaimer

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Glossary of terms

- Respiratory failure type 1- Inadequate gas exchange leading to low oxygen levels in the blood
- Respiratory failure type 2- Inadequate gas exchange leading to high carbon dioxide levels in the blood
- Obstructive sleep apnoea (OSA)- a complete break in breathing while sleeping due to restriction of the upper airways.
- Hypopneas- reductions in the flow of air by at least 30%
- Rapid Eye Movement (REM)sleep – periods sleep marked by eye movement, faster heart and breathing rates and more brain activity
- Continuous Positive Airway Pressure (CPAP) – ventilator delivering a single level of pressure to support breathing
- Bi-level Positive Airway Pressure (BiPAP) - ventilator delivering two levels of pressure to support breathing with the in-breath and the out-breath
- Central sleep apnoea (CSA)– pauses in breathing due to an issue in the central nervous system
- Non-invasive mechanical ventilation (NIV) – collective term for all ventilation delivered via an external device like a mask rather than a tube directly inside the body
- Manual assisted coughing (MAC) – pressure delivered to the abdomen in time with a cough to aid flow of air and secretions out of the body
- Mechanical insufflator/exsufflator (MI:E) – a device that can deliver positive pressure to assist a breath in as well as negative pressure to assist a cough out

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