Breathing Problems of Polio Survivors

by Linda Bieniek , Judith Fischer, Joan Headley, and Edward Anthony Oppenheimer, MD .

Reprinted from Polio Network News, Spring 2001 Vol. 17, No. 2 with permission of Gazette International Networking Institute, 4207 Lindell Blvd., #110, St. Louis, MO 63108-2915.

Link to GINI Website

People with neuromuscular disease such as post-polio often need help with breathing, not because there is something wrong with the lungs, but because the respiratory muscles may be weakened or paralyzed and the chest wall is stiff and inelastic. If scoliosis is present, the work of breathing may be even harder. Other conditions such as chronic obstructive pulmonary disease (COPD) and sleep apnea (obstructive, central, or mixed) may also be occurring. And, overlaying all of these factors, is the simple fact of aging.

How did acute poliomyelitis affect breathing? What is bulbar polio? What is spinal polio?

The poliovirus affected, in many different patterns, the nerve cells in the lower brain (bulbar) and spinal cord that control the muscles of the body. Poliovirus does not damage the lung tissue or the nerves to the airway muscle.

When the bulbar nerves were destroyed (bulbar polio), the muscles of the throat were weakened. This resulted in choking during eating and a diminished ability to cough.

When the spinal nerves were affected (spinal polio), muscles of the arms and legs, and trunk muscles needed for breathing and for taking a deep breath for coughing were weakened. Polio survivors may have had some combination of bulbar and spinal polio, so there may be corresponding throat muscle and limb/respiratory muscle weakness. Involvement of the upper part of the spinal cord weakened the key breathing muscles - the diaphragm and chest musculature.

How does respiratory muscle weakness affect breathing?

The diaphragm is the key muscle for inspiration (breathing in). When it is weakened by polio, the work of breathing becomes harder, especially when a person is lying down. With each breath, the abdominal contents have to be pushed down, but when sitting upright, gravity assists the diaphragm by pulling the contents down. Polio survivors, especially those with scoliosis, compensate by breathing faster but more shallowly because they lack the muscle strength to stretch a stiff rib cage. They may also have smaller lung volumes that further reduce respiratory muscle efficiency and drastically increase the work of breathing. This can lead to underventilation and respiratory failure.

How does respiratory muscle weakness affect sleep?

Respiratory muscle weakness contributes to sleep-disordered breathing. During REM (rapid eye movement) sleep, relaxation of many voluntary muscles, including the shoulder, chest, and abdominal muscles, often occurs. And, these muscles are used to assist breathing when the diaphragm is weak. Consequently, a weak diaphragm has difficulty sustaining adequate breathing, especially when lying down. This leads to a decreased level of oxygen in the blood, or SaO_2 desaturation. SaO_2 desaturations can extend into nonREM sleep and contribute to arousals, inducing sleep fragmentation, and decreasing the amount of time in REM sleep. The quality of sleep deteriorates.

What are the signs of breathing problems?

A polio survivor experiencing a combination of any signs and symptoms in the following list should immediately seek a respiratory evaluation, preferably by a pulmonary physician (pulmonologist) with experience in neuromuscular disease.

- Shortness of breath on exertion (dyspnea)
- Need to sleep sitting up (orthopnea)
- Retention of carbon dioxide (CO_2)
    - morning headaches
    - poor concentration and impaired intellectual function
- Sleep disturbances
    - not feeling rested in the morning
    - sleepiness during the day
    - dreams of being smothered and/or nightmares
    - restless and/or interrupted sleep
    - fatigue or exhaustion from normal activities
    - snoring
- Claustrophobia and/or feeling that the air in the room is somehow bad
- Anxiety
- Difficulty in speaking for more than a short time
- Quiet speech with fewer words per breath
- Use of accessory muscles to breathe
- Weak cough with increased susceptibility to respiratory infections and pneumonias

Are all polio survivors at risk for breathing problems?

No. Individuals who used an iron lung, or barely escaped one, during the acute phase should be aware of potential problems. Those survivors who did not need ventilatory assistance during the acute phase, but who had high spinal polio resulting in upper body weakness and/or diaphragm weakness, and those with scoliosis (sometimes referred to as chest wall deformity) may also be at risk.

Other factors contributing to breathing problems are asthma, COPD, smoking, obesity, and sleep apnea: either central, obstructive, or mixed.

Another compounding factor is diminished vital capacity (VC), which happens to everyone as they age.

Why do these problems often go unnoticed?

The reasons are varied and can be complex. The onset of respiratory problems is insidious, and this gives an individual time to become accustomed to each decrease in function. Thus, one is not immediately aware that anything is wrong, and a treating physician may not recognize the signs or be familiar with the option of home mechanical ventilation. The person's spouse or family should be questioned about signs and symptoms, changes in activity levels and breathlessness, and sleeping patterns.

Additionally, facing breathing difficulties can be frightening, for both the survivor and their loved ones, and there can be a reluctance to address them. Sometimes this fear stems from inaccurate information about the problem and the solutions or from earlier polio-related experiences.

What is underventilation?

The alveoli in the lungs are tiny air sacs at the end of the respiratory tract where gas exchange with the blood occurs. In underventilation (medically known as chronic or global alveolar hypoventilation), the saturation of oxygen in the blood falls due to increased carbon dioxide (CO_2). Normally the alveoli should clear most of the CO_2 out with each breath. Instead, the CO_2 accumulates (called hypercapnia), and thus there is decreased room in the alveoli for oxygen.

Hypercapnia and decreased oxygen saturation are the hallmarks of underventilation or hypoventilation. The signs and symptoms of underventilation usually appear first during sleep.

Some people seem to suddenly experience life-threatening respiratory failure due to CO_2 accumulation (hypercapnia). They may not have been aware of gradually increasing symptoms and signs, particularly since they are often not physically active nor regularly monitored with simple pulmonary function tests. Polio survivors may think that they are breathing fine until an upper respiratory infection, which makes breathing in harder for everyone, causes serious problems, partially due to an ineffective cough and the inability to eliminate secretions.

Who is the most qualified to evaluate breathing problems?

Astute family physicians will order a referral to a pulmonologist, preferably one experienced in neuromuscular disease. Pulmonologists specialize in all breathing-related disorders, however, most focus on more acute problems such as intensive care. The pulmonologist who focuses on neuromuscular diseases understands that the problem is due more to respiratory muscle weakness and the restrictive nature of the disease rather than the lungs themselves.

The IVUN Resource Directory compiles a list of pulmonologists and respiratory health professionals who are knowledgeable about and committed to home care and long-term

mechanical ventilation. It is available at www.post-polio.org/ivun.html or in hard copy from International Polio Network.

What tests will a pulmonologist order?

Pulmonary function tests can be performed in a physician's office with a simple spirometer (an instrument for measuring the capacity of the lungs) or in a fully-equipped pulmonary function laboratory.

Pulmonary function tests should include:

- *Vital capacity (VC) both sitting and supine (lying down), FVC, FEV_1.* VC measures the total volume of air one can breathe out completely after inhaling a full breath. VC is usually done forced, as fast as possible, and is known as FVC. When this fast forced expiration is performed, the volume breathed out in the first second is known as FEV_1. VC is sometimes done slowly and is called SVC. These tests can be done while standing, sitting, or supine. A drop in VC over 25% in the supine position indicates significant diaphragm weakness. When VC declines to less than 1 L (liter), underventilation often occurs.
- *MIF and MEF.* Maximum inspiratory force (MIF) and maximum expiratory force (MEF) are measured by breathing in and out with maximal effort, through a closed mouth tube attached to a pressure measuring device. This measurement reflects inspiratory and expiratory muscle strength.
- *Peak cough flow.* In people who have had polio, cough is often not effective enough, due to respiratory muscle weakness. A weak cough can lead to poor secretion removal, increased respiratory infections, and pneumonia.

What is an ABG and should polio survivors who suspect breathing problems have one?

An arterial blood gas (ABG) should be ordered when VC falls or symptoms of underventilation develop. An ABG invasively measures levels of oxygen, carbon dioxide, and pH in the blood and assesses pulmonary gas exchange. A noninvasive measurement of oxygen saturation in the blood is pulse oximetry, but it is not as complete or sensitive as an ABG.

What is pulse oximetry?

The blood oxygen saturation can be measured noninvasively using a pulse oximeter. It is a probe placed usually on a highly oxygenated part of the body, such as the finger. Infrared light is released and analyzed by recording its changing absorption in the arterial blood. Nocturnal oximetry is becoming more useful in screening for abnormalities that often occur first during sleep. Some oximeters have a memory module to record 8-12 hours of oxygen and pulse rate data.

What is a sleep study and when is it necessary?

Sleep studies (formally known as polysomnography) are usually performed in a sleep laboratory over one or two nights to record multiple variables simultaneously, such as sleep stages, rapid eye movement (REM), snoring, airflow at the nose and mouth, arousals, heartbeat, chest wall breathing motion, and oxygen saturation, to assess sleep disorders (such as sleep apnea). These studies include EEG (brain wave), ECG (electrocardiogram), and often a video record of sleep movements.

Sleep studies are recommended for an individual exhibiting signs of nocturnal underventilation, but not daytime underventilation, or for asymptomatic individuals with a VC < 1 to 1.5 L.

Sleep studies are often recommended to detect sleep apneas.

What is sleep apnea?

Defined as the lack of breathing through the nose and mouth for at least ten seconds, sleep apnea can be obstructive or central or mixed. Obstructive sleep apnea (OSA) occurs when tissues in the throat collapse and block airflow in and out of the lungs during sleep, although efforts to breathe continue. Central apnea occurs when the brain fails to send appropriate signals to the body to initiate breathing. There is neither airflow nor chest wall movement.

In sleep apnea, breathing ceases, oxygen in the blood decreases, arousal occurs, sleep ends, and breathing resumes. The individual then drifts back to sleep and another apnea occurs, with this cycle continuing throughout the night, resulting in hundreds of arousals from sleep.

OSA at first occurs when individuals sleep on their backs, but eventually apneic episodes are present with any sleep position. A number of factors make snoring and apnea worse, such as obesity and nasal obstruction. Smoking causes the lining of the upper airway to swell, alcohol and sedative drugs cause the muscles in the back of the upper airway to relax, and excessive weight decreases the size of the upper airway.

When there are nighttime breathing problems in a person with neuromuscular disease, such as post-polio, they are more likely due to respiratory muscle weakness, rather than OSA. However, some individuals may have only OSA while others may have both respiratory muscle weakness and OSA.

What is the treatment for sleep apnea?

Sleep apnea is best treated with the use of a continuous positive pressure airway (CPAP) device to push the tongue out of the way and keep the airway open during sleep. If underventilation and sleep apnea occur simultaneously, a bilevel positive pressure device is recommended to help improve ventilation and also keep the airway open.

What is CPAP?

CPAP stands for Continuous Positive Airway Pressure. Air flows continuously into the airway via the nose with use of a nasal mask. CPAP keeps the airway open, but does not adequately assist respiratory muscle activity.

CPAP is primarily used to treat obstructive sleep apnea, and thus is normally used only at night during sleep. CPAP units are not ventilators. Higher pressures may make exhaling uncomfortable and difficult. Newer CPAP units, called Auto CPAPs, automatically provide varying levels of pressure based on the individual's needs during the night. Because OSA is prevalent among the general population, many companies offer CPAP units.

What is mechanical ventilation (MV)?

Mechanical ventilation is the use of machines to help people breathe when they are unable to breathe sufficiently on their own. It is most often used for a few days in a hospital setting, when people are recovering from surgery or during a serious illness. However, some people may be unable to breathe on their own after the acute illness is over and may require long-term MV. Other people may have a stable, chronic condition that prevents them from breathing on their own; they may need to use a ventilator only at night or both at night and during the day.

Assisted breathing through mechanical ventilation can help people sleep better and result in improved lung function during the day. MV can restore the gas exchange and prevent respiratory failure.

When is mechanical ventilation an option?

A general guideline, but not a rule, suggests that mechanical ventilation be initiated when there is a 50% decline in VC or a VC of under 1 L. Each symptomatic survivor needs a comprehensive evaluation by a knowledgeable physician to determine when to start MV. This is especially true of the survivors of bulbar poliomyelitis. Mechanical ventilation helps rest the respiratory muscles, resets the sensitivity to CO_2 of the controller in the brain, and improves pulmonary mechanics by providing more functional lung expansion.

Is exercise a treatment option?

Chest expansion exercises may help keep the chest wall more elastic and avoid loss of lung volume. There are four ways this can be done: with an Ambu-type resuscitation bag, using a volume ventilator for air stacking, using glossopharyngeal (frog) breathing, or using an IPPB (intermittent positive pressure breathing) machine.

If I need mechanical ventilation, what are the options?

The older technology of negative pressure ventilation, which developed during the polio epidemics to keep respiratory polio survivors alive, has given way to the newer technology of positive pressure ventilation.

Negative pressure ventilators apply intermittent negative pressure (like a vacuum) to the chest and abdomen by means of an iron lung or Porta-Lung, a chest shell (cuirass), or a form of body jacket or wrap. Some polio survivors still use the iron lung, Porta-Lung, or cuirass.

The pneumobelt is also still used. It is an inflatable corset around the abdomen which acts by pressing on the abdomen to augment exhalation. Inspiration is not assisted. The pneumobelt can only be used in the upright. seated position and does not work if one is either underweight or overweight.

Positive pressure ventilation uses a bilevel pressure support device or a volume ventilator to deliver air noninvasively via a face mask, nasal mask, nasal pillows, mouthpiece, lipseal, or oro-nasal combination, called noninvasive positive pressure ventilation (NPPV). Positive pressure can also be delivered invasively through a tracheostomy tube (TPPV).

What is the difference between a bilevel device and a volume ventilator?

Bilevel pressure devices continuously deliver air, but the inspiratory pressure can be adjusted separately from the expiratory pressure. Bilevel devices are also used with a face or nasal mask or nasal pillows, and, like CPAP, used mainly at night. However, bilevel devices can only deliver a certain amount of pressure that may not be enough for people with respiratory muscle weakness and underventilation. There are many bilevel pressure devices on the market, but the only one that can be truly called BiPAP® is from Respironics.

Compared with volume ventilators, bilevel devices are lightweight, less expensive, easier to use, and adjust better to leaks. However, disadvantages include not being well-suited for tracheostomy ventilation, having no internal battery, not as commonly used with an external battery, may be noisier, and the expiratory pressure is unnecessary for some people and may cause thoracic discomfort. Examples of bilevel devices include Respironics Bi-PAP® S/T and BiPAP® Synchrony™; Puritan Bennett KnightStar 330® and KnightStar 320®; ResMed VPAP® II ST; and Breas PV102.

Volume ventilators deliver a preset volume of air. These machines can deliver much more air than bilevel devices, and thus enable deeper breaths for improved coughing and air stacking. They may be necessary for people with poor lung elasticity, such as those with pulmonary fibrosis, and stiff chest walls, as with kyphoscoliosis, when bilevel is not enough. Volume ventilators, though larger and heavier and more expensive than bilevel units, are quieter, have more alarm features, overcome airway secretions and resistance, have an internal battery, work from battery power easier, can be used for 24 hours, and are wellsuited for tracheostomy use.

Examples of volume ventilators include the LP6 Plus, LP10, and Achieva® from Puritan Bennett, PLV®-100 and PLV®-102 from Respironics, TBird® Legacy from Thermo Respiratory Alternate Care, and PV501 from Breas.

A new generation of ventilator technology has produced the LTV™ series from Pulmonetic Systems, Inc. These new ventilators are compressorless and run by turbines. They are very small

- about the size of a laptop computer - and lightweight, about 13 lbs., but more expensive. They also have the capability to offer both volume and pressure support.

Who decides which equipment to use? How?

The answers to these questions focus on whether there is breathing muscle weakness or whether there are reasonably normal breathing muscles but obstructive sleep apnea (OSA).

If there is nighttime breathing abnormality due to muscle weakness, it is best treated with a machine that assists ventilation, such as the aforementioned bilevel pressure device or volume ventilator.

If the upper airway tends to close off during sleep, OSA occurs. This is often successfully treated by continuous positive airway pressure (CPAP) which delivers a constant flow of air to keep the airway open. It is certainly possible to have weakness in the throat/pharyngeal area that produces OSA, without weakness of the breathing muscles. If this is the case. CPAP is the appropriate treatment.

Whether to use a portable volume ventilator or bilevel pressure device is an important decision. Sometimes the choice reflects the experience and training of the pulmonologist or respiratory therapist. Often cost is a factor. In Europe, the costs of the bilevel pressure devices and the volume ventilators are not as disproportionate as they are in the United States.

The ventilator which is most comfortable for the user and fits his or her individual ventilatory needs best should be the overriding choice. The physician, respiratory therapist, and ventilator user should collaborate on determining the best system, although ventilator users are not often given the opportunity to try different ventilators and systems.

Where is the best location to start noninvasive positive pressure ventilation (NPPV)?

Generally, NPPV is started in the home by a respiratory therapist from the home health care agency or durable medical equipment (DME) provider, as ordered by the physician (who is not present). However, home health respiratory therapists have limited time, and the home health care agency does not receive reimbursement for the time needed. The therapist usually cannot demonstrate the whole range of equipment and interface options available. Followup is needed repeatedly in the first months to ensure that the equipment and interfaces are comfortable and working properly to achieve optimal benefit.

The use of NPPV can also be initiated as part of hospital discharge. where there should be more time to do it properly. However, many pulmonary critical care physicians are highly ICU-focused, and they and the respiratory therapy staff may not have the experience and interest needed to get optimal results.

When possible, the best approach may be for the individual to visit the home MV unit of a medical center for daytime assessment and for teaching the use of NPPV. The person can try

various equipment and interfaces to see which would work best under the guidance of an experienced physician and respiratory therapist. The day visits can be repeated as needed for setup and followup. This is in addition to the home health care/DME company's home visits after the system is selected and ordered.

What does a respiratory therapist do?

Respiratory therapists work under the direction of a physician, usually in hospital settings where they perform intensive care, critical care, and neonatal procedures. Polio survivors usually interact with a respiratory therapist in their physicians' offices and/or in their homes during visits from a therapist hired by a home health agency or a home medical equipment supply company (Apria, Lincare, etc.).

Respiratory therapists perform procedures that are both diagnostic and therapeutic. Some of these activities include: taking arterial blood gas (ABG) specimens and analyzing them to determine levels of oxygen, carbon dioxide, and other gases; measuring the capacity of the lungs to determine if there is impaired function; and studying disruptive sleep patterns. Respira tory therapists help set up and maintain the various equipment (CPAP and bilevel devices, venti lators) to assist breathing, adjust the settings, adjust the mask or other interface, instruct in their use, and monitor compliance.

What is an interface?

An interface connects the tubing from the volume ventilator, bilevel device, or CPAP to the person using it. Examples include nasal or facial masks, nasal pillows, lipseal, mouthpiece, or tracheostomy. Often the individual is handed only one mask to try by a respiratory therapist from a home health care company, but several different masks may need to be tried in order to find one that fits properly. Many people do best when they can choose at least two (nasal) interfaces that are comfortable and rotate between them. Individuals can use a mouthpiece if they have nasal congestion due to a cold.

Adapting to and making adjustments to an interface is critical and often requires patience and determination. Many users creatively adapt their masks to achieve the best fit.

What is a tracheostomy and when is it necessary?

A tracheostomy is an artificial airway created during a surgical operation called a tracheotomy. A tracheostomy may be necessary for polio survivors who cannot tolerate noninvasive ventilation, for those who can no longer be adequately ventilated noninvasively, and for those who need ventilation more than 20 hours per day. Other considerations include a severely impaired cough and inability to clear secretions, and significant swallowing problems. Some polio survivors who need 24-hour ventilation may prefer to use noninvasive ventilation.

The decision to undergo a tracheotomy is a serious one and should be made by a fully informed individual along with caregivers in consultation with the physician.

How can a polio survivor improve cough?

Retained secretions in people who have impaired coughing ability can turn a common cold into pneumonia. To improve cough, there are two forms of assisted coughing.

Manually assisted coughing involves another person administering a thrust to the chest and abdomen of the individual with neuromuscular disease immediately after that individual takes a big breath.

Mechanically assisted coughing can be performed with the CoughAssist.. by J.H. Emerson Company. The CoughAssist applies positive pressure to provide a deep breath in, then shifts rapidly to negative pressure to create a high flow out, as with a normal cough.

What is cor pulmonale?

Cor pulmonale (right-sided heart failure) can occur in post-polio and in other diagnoses, such as COPD. In both, the low oxygen level causes pulmonary hypertension, liver enlargement, and swollen feet. Cor pulmonale is detected by a careful physician examination, keeping respiratory muscle weakness in mind.

What about oxygen?

Tony Oppenheimer, MD, retired physician in pulmonary and critical care medicine from Southern California Permanente Medical Group, cautions, "Administering oxygen does not provide assistance to the weakening respiratory muscles, but gives both the patient and the doctor the false impression that appropriate treatment is being provided. Underventilation is mistaken for an oxygen transfer problem. Indeed, administering oxygen can mask the problem.

"Also there is a danger of causing respiratory depression by giving oxygen. It will improve the oxygen saturation, but not the underventilation. It may increase the danger of dying of sudden respiratory failure.

"Some situations may require administering oxygen, such as cor pulmonale or pneumonia due to infection or aspiration. If this occurs in people with respiratory muscle weakness and hypoventilation, then it is important to provide both assisted ventilation and supplemental oxygen, and use ABGs to monitor them."

A useful analogy from Lisa S. Krivickas, MD, Spaulding Rehabilitation Hospital, Boston, in regard to people with respiratory failure from neuromuscular disease is that the lungs are like deflated balloons that are not strong enough to inflate.

To inflate the balloon, mechanical assistance to force air into the balloon is needed. Blowing oxygen across the mouth of the balloon (the equivalent of using supplementary oxygen delivered by nasal cannula) will do nothing to inflate the balloon.

References

American College of Chest Physicians. (1998). *Mechanical ventilation beyond the intensive care unit: report of a consensus conference of the American College of Chest Physicians.* Chest, 113, Supplement, 2895-3445.

Bach, J.R. (1999). *Guide to the evaluation and management of neuromuscular disease.* Philadelphia, PA: Hanley& Belfus, Inc.

Bach, J.R. (1996). *Pulmonary rehabilitation: The obstructive and paralytic conditions.* Philadelphia, PA: Hanley & Belfus, Inc.

Faber, H.K. & Amoss, H.L. (1956). *Acute poliomyelitis*. In Tice's Practice of Medicine, 335359.

Fischer, J.R. & Headley, J.L. (1995). *Postpolio breathing and sleep problems.* IVUN News, 11 (4), 2.

Gay, P, & Edmonds, L. (1995). *Severe hypercapnia after low-flow oxygen therapy in patients with neuromuscular disease and diaphragmatic dysfunction.* Mayo Clinic Proceedings, 70 (4), 327-330.

Hill, N.S. (2001).*Long-term mechanical ventilation.* New York, NY: Marcel Dekker, Inc.

Hsu, A. & Staats, B. (1998).*"Postpolio" sequelae and sleep-related disordered breathing.* Mayo Clinic Proceedings, 73, 216-224.

Maynard, F. & Headley, J. (1999).*Handbook on the late effects of poliomyelitis for physicians and survivors.* Saint Louis, MO: Gazette International Networking Institute.

Oppenheimer, E.A. (2000).*Oxygen is NOT for hypoventilation in neuromuscular disease.* IVUN News, 14 (1), 4-5.

Romaker, A. (1995).*New breathing problems in aging polio survivors: respiratory muscle weakness.* Polio Network News, 11 (4),1, 7-8.