Original Research

Pulmonary Dysfunction and Sleep Disordered Breathing as Post-Polio Sequelae: Evaluation and Management

John R. Bach, MD* and Augusta S. Alba, MD**

Lincolnshire Post-Polio Library copy by kind permission of Dr. Bach.

From the *New Jersey Medical School of the University of Medicine and Dentistry of New Jersey, and the Department of Physical Medicine and Rehabilitation, University Hospital, Newark, NJ, and the **Department of Physical Rehabilitation Medicine, New York University and Godwater Memorial Hospital, New York, NY.

Reprint requests [Original Publication]: John R. Bach, MD, Dept of Rehabilitation Medicine, University Hospital B-239, The New Jersey Medical School-UMDNJ, 150 Bergen St., Newark, NJ 07103.

Post-polio sequelae can include sleep disordered breathing and chronic alveolar hypoventilation (CAH). Both conditions develop insidiously and can render the post-polio survivor susceptible to cardiopulmonary morbidity and mortality when not treated in a timely and appropriate manner. These conditions can be diagnosed by a combination of spirometry, noninvasive blood gas monitoring, and ambulatory polysomnography. Sleep disordered breathing is most frequently managed by nasal continuous positive airway pressure, while tracheostomy ventilation is the most common treatment for ventilatory failure. We report the more effective and comfortable techniques recently made available for managing sleep disordered breathing and the use of noninvasive treatment options for CAH, respiratory failure, and impaired airway clearance mechanisms. One hundred forty-three cases are reviewed.

It is difficult to estimate the number of post-poliomyelitis individuals with ventilatory insufficiency in the United States. There were over 500 000 people afflicted with poliomyelitis in this country over the 35-year period from 1928, the year that the iron lung was perfected, to 1962.[1,2] This includes about 300 000 from 1948 to 1955 alone. In a study of 1540 poliomyelitis patients admitted to one hospital, 1290 (84%) had acute paralytic polio; 186 (14.5%) required ventilatory support and survived for at least 3 months.[3] In the 1952 polio epidemic of Denmark, 345 of 2300 patients (15%) had ventilatory failure and/or impairment of swallowing. The mortality was 94% for those with respiratory paralysis with bulbar involvement, and 28% for those with respiratory paralysis without bulbar involvement.[4] Twenty-five of the 200 surviving ventilator-assisted individuals (12.5%) remained so.

By 1952, Danish mortality figures for ventilator-assisted individuals had decreased from 80% to 41% overall, partly because of more frequent use of tracheostomy.[4] Specialized centers in the United States also reported significant decreases in mortality by "individualizing" patient care. From 1948 to 1952, 3500

patients were treated at Los Angeles General Hospital. Fifteen percent to 20% required ventilatory support. General mortality decreased from 12% to 15% in 1948 to 2% in 1952 without the use of tracheostomy for ventilatory support.[5] Although many patients at Los Angeles General Hospital, particularly those with bulbar polio, had tracheostomies placed for management of secretions while they were ventilated by body ventilators (BVs), in other centers where few tracheostomies were performed mortality also decreased to about 2%.[5] The previously high fatality rate was not because of the inability of BVs to ventilate patients, but because of swallowing insufficiency and aspiration of secretions.[5] Better nursing care and attention to managing airway secretions, including the use of exsufflation devices in some centers, were factors in decreasing mortality rates.[6-11] These latter devices continue to be important for ventilator-assisted individuals with access to them.[12-16]

In a national survey of the late effects of poliomyelitis, Halstead et al[17] found that 42% of the respondents reported new breathing problems. Some post-poliomyelitis individuals who never before required ventilatory assistance were using ventilators.[18,19] The first report of postpoliomyelitis late onset ventilatory insufficiency was in 1970.[20] More recently, the majority of ventilator-assisted individuals have been described as late onset.[12,18] In 1987, Bach et al[13] studied 75 ventilator-assisted individuals, of whom 31 were late onset an average of 18 years post-polio. Six subsequently died, but 21 additional post-poliomyelitis individuals had become ventilator users by 1990.[12] We are currently managing 112 ventilator-assisted individuals, 64 (58%) of whom have late-onset ventilatory insufficiency.

Pathophysiology of Late-Onset Pulmonary Dysfunction

For post-poliomyelitis individuals, acute respiratory failure most frequently results from complications of chronic alveolar hypoventilation (CAH). This is due to a restrictive pulmonary syndrome secondary to post-polio respiratory muscle weakness and often scoliosis. Post poliomyelitis individuals also have a predilection for sleep-disordered breathing, which can lead to CAH.

Loss of vital capacity is directly related to respiratory muscle weakness.[21] Vital capacity, which normally plateaus at age 19 years and decreases about 1.0% to 1.2% per year thereafter, decreases at a greater rate for many postpoliomyelitis individuals. Fischer[18] reported a 1.7% loss of forced vital capacity per year in 20 post-poliomyelitis individuals, while Bach et al[13] reported a 1.9% ± 1.2% loss of vital capacity per year. Vital capacity loss in post poliomyelitis individuals occurs because of the effects of aging, fatigue, and/or accelerated loss of remaining anterior horn cell collaterals on residual respiratory muscle strength, pulmonary compliance, and function.

The loss of pulmonary volumes leads to insidious ventilatory insufficiency. Hypercapnia is likely when the vital capacity falls below 55% of predicted normal.[21] Symptoms[14] may be minimal for nonambulatory patients as gradual resetting of respiratory control centers accommodates CAH. Hypoxia, hypercapnia, and loss in vital capacity are exacerbated when intrinsic lung disease, kyphoscoliosis, sleep disordered breathing, or obesity complicate respiratory muscle weakness. The rapid shallow breathing pattern with loss of the ability to take occasional deep inspirations (sighs) appears to contribute to loss of compliance, increased stiffness of the rib cage, and chronic microatelectasis.[22,23] Decreased pulmonary compliance increases the work of breathing.

Patients with expiratory muscle weakness also can have difficulty clearing secretions, especially during upper respiratory tract infections (URIs). Chronic mucus plugging can lead to ventilation/perfusion imbalance, worsening atelectasis, pneumonias, pulmonary scarring, further loss of lung compliance, *cor pulmonale*, and eventually cardiopulmonary arrest. Risk is heightened in the presence of weak oropharyngeal musculature. Mucus plugs can also cause sudden hypoxia and respiratory failure.

Sleep-disordered breathing, which is the occurrence of apneas and hypopneas, either obstructive or central, occurs in 37.5% of the general population over 62 years of age.[24] A symptomatic individual with a mean of 10 apneas plus hypopneas per hour is said to have the obstructive sleep apnea syndrome (OSAS).[25,26]

There appears to be a greater incidence of sleep disordered breathing in post-poliomyelitis individuals due to paresis of intrinsic muscles of the larynx[5] and damage to respiratory control centers from the encephalitic process of the primary viral infection.[5,27-29] Guilleminault and Motta[30] noted a mixed obstructive-central apnea picture in five post-poliomyelitis individuals. Steljes et al[31] demonstrated obstructive or mixed apneas in seven of eight post-poliomyelitis individuals with complaints of fatigue, muscle weakness, muscle or joint pain, sleep problems, and breathing difficulties. Sleep disordered breathing alone can result in CAH, hypoxia, right ventricular strain, and when severe, acute cardiopulmonary failure.

Evaluation of CAH and Sleep Disordered Breathing

Evaluation and management of a restrictive pulmonary syndrome and CAH should not await hospitalization for acute respiratory failure, nor should patients be labeled as having post-polio syndrome without an adequate evaluation of pulmonary function, including sleep studies when indicated. While post-polio sequelae symptoms and signs include increasing fatigue, headache, sleep disturbances, decreased muscle strength, muscle aches, dyspnea, cyanosis, irritability, anxiety, and depression,[32] these symptoms are also characteristic of CAH or OSAS.[14] Other symptoms and signs of CAH or OSAS include hypersomnolence, poor concentration, impaired intellectual function and memory, nightmares, decreased libido, and recent changes in body weight. Patients with impaired oropharyngeal muscle strength can also complain of increased difficulty controlling upper airway secretions.[14]

Since most post-poliomyelitis individuals are now middle aged or elderly, obstructive airway disease may also be a factor. Therefore, besides a focused history and physical examination, pulmonary evaluation of post-poliomyelitis individuals should often include general pulmonary function studies. Significant intrinsic lung disease is present when the pO_2 is chronically less than 60 mm Hg in the absence of hypercapnia and reversible factors. This indicates the need for oxygen therapy. If the pulmonary dysfunction is primarily restrictive, regular measurement of vital capacity with a hand-held spirometer is sufficient for monitoring patient progress and response to treatment. The vital capacity should be measured in the sitting position, supine position, with other positional changes, and while using thoraco-lumbar orthoses when appropriate.

The artificial inspiratory capacity (AIC) is useful for determining the potential for assisting ventilation by noninvasive means as opposed to tracheostomy.[33] The AIC is a measure of the maximum volume of air that can be held with a closed glottis. It is obtained by the air stacking of mechanical insufflations or by glossopharyngeal breathing (GPB).[16] The AIC is a function of pulmonary compliance and the strength of bulbar musculature. For post-poliomyelitis individuals, an AIC of at least 1000 ml is needed to achieve adequate peak expiratory flows for effective coughing and therefore permit optimal continued use of noninvasive aids without resort to tracheostomy or mechanical exsufflation.

Any patient with less than 50% of predicted normal supine vital capacity should undergo oxygen saturation (SaO_2) monitoring and pCO_2 monitoring during sleep. The capnograph, which can be used to measure end-tidal pCO_2, and pulse oximeter must be capable of summarizing and printing out the data. [12,14] These studies are most conveniently performed on an ambulatory basis. The perhaps more accurate transcutaneous CO_2 sleep studies can be done on an inpatient basis with CO_2 sensing electrodes.

The diagnosis of CAH is established by the presence of nocturnal pCO_2 greater than 50 mm Hg. Without nocturnal pCO_2 data, continuous oxyhemoglobin desaturation with a mean less than 95% for 1 hour or more during sleep in a symptomatic patient with vital capacity less than 50% of predicted is sufficient to establish the diagnosis and initiate treatment.

With CAH, nocturnal hypoventilation is often more severe than daytime hypoventilation.[15] Without a significant obstructive apnea component, nocturnal SaO_2 monitoring typically reveals a relatively smooth drop from the baseline waking SaO_2. If left untreated, insidiously progressive hypercapnia can occur in these patients and a compensatory metabolic alkalosis results in elevated central nervous system bicarbonate levels which contribute to depression of the ventilatory response to hypoxia and hypercapnia, and exacerbate CAH.

For symptomatic patients with vital capacities greater than 50% of predicted normal and with inconclusive nocturnal SaO_2 and CO_2 studies, the symptomatology cannot be explained on the basis of a severe paralytic/restrictive pulmonary syndrome alone. These patients may undergo ambulatory polysomnography to evaluate for the presence of sleepdisordered breathing.[34]

Pulmonary Managements

Both sleep disordered breathing and CAH can be reversed, respiratory control mechanisms normalized, and life quality and longevity increased with treatment. The primary goals for the appropriate patient should be to maintain normal alveolar ventilation around the clock, to provide clearance of airway secretions, to maintain or improve pulmonary compliance, and to address the factors which cause sleep-disordered breathing.

Sleep-disordered breathing. Any treatable conditions associated with OSAS should be identified and appropriately managed.[35,36] Otherwise, continuous positive airway pressure (CPAP) is effective for the majority of patients for whom no treatable etiologic condition can be found. CPAP works as a pneumatic splint to maintain an open airway. CPAP of 5 cm to 15 cm H_2O is usually adequate. Independently varying the inspiratory and expiratory pressures improves effectiveness and comfort. The Bi-PAP ventilator (Respironics Inc, Monroeville, Pa) has been designed to achieve this. For many patients with a combined paralytic/restrictive ventilatory insufficiency and sleep-disordered breathing, both CPAP and Bi-PAP may be ineffective. The use of noninvasive methods of assisted ventilation by intermittent positive airway pressure (NV-PAP) can be delivered via oral, nasal, or oral-nasal patient-ventilator hose interfaces. NV-PAP is usually delivered at higher than typical CPAP or Bi-PAP pressures. This can assist or support the patient's ventilation and maintain upper airway patency [12-14,30,37-41]

Even when effective, CPAP, Bi-PAP, and NV-PAP may not be tolerated due to discomfort or air leakage from an inadequately fitting CPAP mask.[42] For these patients, a commercially available kit is available for molding a custom nasal interface (SEFAM mask, Lifecare Inc [now Respironics Inc], Lafayette, Colo). Such interfaces are comfortable and effective at higher pressures. However, they are expensive ($785), delicate, and require refabrication every 6 weeks to 5 months. A transparent, durable, custom-molded, low profile nasal interface is also available.[43] It is comfortable and cosmetic and costs $500. Various other orthotic and surgical options are available when positive airway pressure cannot be tolerated.[44-48]

Chronic alveolar hypoventilation. There are post-poliomyelitis individuals for whom neither sleep-disordered breathing nor the restrictive pulmonary syndrome are severe enough to cause CAH, but who with both conditions require aid to maintain adequate ventilation.[12,13] The symptoms of CAH are often ignored or misdiagnosed, and the individual is often suboptimally managed even when the diagnosis is

made accurately. Untreated post-poliomyelitis individuals with CAH may present with recurrent bouts of pneumonia and acute respiratory failure from otherwise benign URIs. Such patients are hospitalized, and receive intermittent positive pressure breathing (IPPB) treatments for the delivery of bronchodilators and other frequently ineffective medications for patients with purely restrictive pulmonary conditions.

Xanthine derivatives are commonly used despite the lack of good evidence of their efficacy in increasing diaphragm contractility in this population and their potentially serious side effects.[49] Likewise, oxygen therapy is frequently unnecessarily prescribed both before and after discharge. The use of long-term oxygen therapy instead of emphasis on providing maximal insufflations, optimal ventilation, and assisted coughing can worsen hypercapnia, diminish respiratory drive, and increase microatelectasis which in turn decreases pulmonary compliance. When carbon dioxide retention is severe, it can lead to acute respiratory arrest.[14,18,20,50] Even low-flow oxygen therapy may cause anemia by inhibiting erythropoetin synthesis, and thereby hamper respiratory exchange. Finally, since few physicians are familiar with noninvasive techniques of assisted ventilation, reluctant patients are often coerced into accepting a tracheostomy with its potential complications and inconveniences.

Ventilatory assistance or support can be provided by BVs, devices which act directly on the body, or by NV-PAP. Negative-pressure BVs create negative pressure on the chest and/or abdomen, causing air to flow into the lungs through the nose and mouth.[51] They include the rocking bed,[52] iron lung,[53] Porta-Lung,[51] the cuirass,[54,55] and negative pressure wrap ventilators.[51] These devices have been used for nocturnal ventilatory support for decades by hundreds of ventilator-assisted individuals. [13,16,56] They are inconvenient and not always effective. Travel is difficult and sleeping intimately with a significant other may be impossible. In a small study of post-poliomyelitis individuals using BVs, Steljes et al[31] observed poor sleep quality, a high apnea hypopnea index characteristic of OSAS, hypoxia, and hypercapnia. Use of BVs is associated with obstructive sleep apneas and often severe oxyhemoglobin desaturations in the majority of individuals who use them.[57] Despite the severity of the desaturations, there is little evidence that they are life-threatening. The usual presence of heavy snoring and observed apneas often seen in patients with classic OSAS may not be present for patients using BVs. [57]

The intermittent abdominal pressure ventilator (IAPV) consists of an inflatable bladder in an abdominal belt. The bladder is cyclically inflated by a positive pressure ventilator. The alternating pressure on the abdominal contents indirectly moves the diaphragm and ventilates the patient. The IAPV generally augments the patient's tidal volume by over 300 ml.[58] It is most effective in the sitting position at 75° to 85°. It is the method of choice for daytime ventilatory support for most individuals with less than 1 hour of free time off the ventilator because it is cosmetic, practical, and ideal for concurrent GPB and wheelchair use.[58]

Recently, NV-PAP has been recognized as an effective alternative to tracheostomy ventilation and BV use for nocturnal supported ventilation as well as for daytime aid. Intermittent positive pressure ventilation (IPPV) via nasal access is generally simplest and should be tried first.[12,14,39-41] The use of custom interfaces may be tried.[12,14,37] If nocturnal blood gas monitoring does not demonstrate sufficient normalization of ventilation with this method, mouth IPPV with a Bennett lip seal (Puritan-Bennett, Boulder, Colo) should be tried.[13,15] A complete seal can always be obtained even if one must plug the nose with cotton pledgets and modify the strap system for additional support. Ventilator volumes of 1000 ml to greater than 2000 ml may be necessary depending on the ongoing insufflation air leakage. For ventilator-assisted individuals living alone who are unable to manage a strap retention system, a strapless oral-nasal interface can be constructed.[37,43] For all three methods, air can be delivered via custom molded interfaces. Mouth IPPV is also an excellent alternative to pressure support ventilation or

interspersing periods of tracheostomy IPPV with CPAP as a means of ventilator weaning.

Avoiding tracheostomy permits the patient to more readily learn GPB. GPB should be taught to those with less than 1000 ml of vital capacity and adequate oropharyngeal muscle strength for functional swallowing and speech.[16,59,60] GPB is the use of the tongue and pharyngeal muscles to add to a maximal inspiratory effort by projecting boluses of air past the vocal cords into the lungs. Effective GPB permits a patient to sustain ventilation for up to hours despite having little or no vital capacity. It also normalizes speech production and provides deeper breaths for shouting and coughing. Progress with GPB should be monitored by regularly measuring the volume of air per gulp and the number of gulps per breath.

Although NV-PAP methods are generally preferable to BVs or tracheostomy for long-term ventilatory support, BVs continue to be useful for temporary support for some patients during URIs.[15,16] They can also be used during tracheostomy site closure,[61] and for patients who prefer them to NV-PAP.

One hundred forty-three ventilator-assisted individuals, including those reported in two previous studies, have been reviewed.[12,13] They had a mean recent vital capacity of 734 ± 573 ml in the sitting position and all were mechanically ventilated by noninvasive methods. Five of the patients had vital capacities that were at least 20% (150 ml) less in the sitting position than when supine. Thirty-five patients had vital capacities that were at least 20% (150 ml) less in the supine position than when sitting.

Seventy-one patients were late-onset ventilator-assisted individuals beginning aid at age 41.9 ± 14.2 years (range: 9 to 78), a mean of 29 ± 12.1 years (range: 3 to 59) post-polio. These 71 ventilator-assisted individuals have benefited from ventilatory assistance a mean of 1 1.55 -t 9. 1 years. Three of the 71 required only daytime ventilatory assistance. Seventeen of the 71 used ventilatory aids overnight only. Ten others used them overnight and up to 8 hours during the daytime. The other 41 late-onset ventilator-assisted individuals progressed to require ventilatory assistance about 24 hours a day. All but two of these ventilator-assisted individuals required ventilatory assistance temporarily at onset of polio. Ten of the 71 ventilator-assisted individuals died after 12.2 ± 9.9 years of ventilatory assistance. Four ventilator-assisted individuals died overnight while using mouth IPPV without a Bennett lip seal. One man died suddenly at age 52. One woman died from a meningioma, one from a cerebrovascular accident, and one from cardiac failure associated with *cor pulmonale*. One man died from pneumonia after he underwent a tracheostomy.

Seventy-two patients have been ventilator-assisted since onset of polio. They have benefited from aid for 37.5 ± 3.6 years. Twenty ventilator-assisted individuals died after 24.5 ± 5.8 years of ventilatory support. Their deaths were associated with the following: alcohol and drug abuse, 4; sleep without using a lip seal, 3; cardiovascular disease, 2; unknown, 2; pneumonia, 2; and 1 each from emphysema, inadequate nocturnal ventilatory support using a rocking bed, complications of tracheostomy, an acute abdomen, a motor vehicle accident, cancer, and renal failure.

Thirty-five of the 143 ventilator-assisted individuals had tracheostomies placed for management of acute medical or surgical conditions. Eleven ventilator-assisted individuals retained the tracheostomy for continued ventilatory assistance. Five of these 11 ventilator-assisted individuals died, all within 4 years of tracheostomy placement, from pulmonary disease associated with mucus plugging and/or substance abuse in four cases and *cor pulmonale* in the other case. The other 24 ventilator-assisted individuals had the tracheostomy sites closed and returned to noninvasive ventilatory support once the acute conditions had been resolved. Several patients had been tracheostomized as many as three times, but each time returned to noninvasive support. Of these 24 ventilator-assisted individuals who have been ventilator assisted for 25.5 ± 13.7 years, only one has died thus far. Her death was associated with substance abuse.

The avoidance of tracheostomy in this group as a whole has permitted 59 of these individuals to master

GPB sufficiently to use it for time free of mechanical ventilation. The 59 ventilator-assisted individuals with an average vital capacity in the sitting position of 481 ml had an average glossopharyngeal maximum single breath capacity[16] of 2133 ml. Their average free time without using GPB was 189 minutes sitting, 150 minutes supine, but 307 minutes sitting when using GPB. Twenty-four ventilator-assisted individuals with 5 minutes or less free time without GPB had 2 hours or more free time using it.

Assisted coughing and oximetry biofeedback. Manual and/or mechanical assisted coughing should be used as necessary. This may be every 10 to 15 minutes around the clock during URIs.[62,63] When peak expiratory flows are inadequate by manual techniques, mechanically assisted expulsion of airway secretions, which is both less labor intensive and more effective, can be achieved with mechanical exsufflation devices.[6-8,14,15,61-63]

Oximetry can be used for biofeedback to assist the patient with CAH in maintaining more normal daytime ventilation as well as for monitoring the efficacy of assisted ventilation.[63] The importance of this versatile and convenient technique was first described for poliomyelitis individuals in 1948, but only recently have oximeters become widely available.[64] For biofeedback, the patient should be instructed to keep his SaO_2 at 95% or greater throughout daytime hours and supplement his breathing if necessary with periods of IPPV via the mouth.[13,15,16] The oximeter can effectively gauge ventilation provided that supplemental oxygen therapy is not used. Continuous SaO_2 monitoring is also especially useful during URIs when continuous desaturation is a sign of atelectasis or pneumonia and sudden desaturation an indication of acute mucus plugging.

Tracheostomy and its indications. Fischer reported converting 27 patients from ineffective BV use to tracheostomy ventilation.[18] Bach et al[12,13] averted tracheostomy by converting over 40 patients from inadequate BV aid to NV-PAP. Intubation and tracheostomy also can be avoided for some ventilator-assisted individuals in the intensive care setting.[61] A survey of ventilator-assisted individuals who have used both tracheostomy ventilation and noninvasive methods indicated the patients' clear preference for the latter. The great majority of patients stated that they ate, spoke, and looked better using noninvasive aids. Most also felt healthier and slept better.

The use of an endotracheal tube can cause a sore throat, laryngitis, glottic edema, mucosal ulceration, laryngeal or tracheal stenosis, tracheal dilatation, tracheal-innominate artery fistula, atelectasis, and pulmonary infections.[65-68] Long-term tracheostomy is often complicated by chronic bacterial colonization,[66,67] granulation tissue formation and bleeding, chronic mucus plugging, tracheal stenosis in 17% to 65% of patients, and many other less frequent but potentially serious complications including accidental tracheostomy disconnection and respiratory arrest without possible resort to GPB.[69-72] Speech and swallowing also can be hampered by the presence of an indwelling tracheostomy.[73]

In 1955, an International Consensus Symposium defined the indications for tracheostomy as the combination of respiratory insufficiency with swallowing insufficiency and disturbance in consciousness or vascular disturbances.[5] "If a patient is going to be left a respirator cripple with a very low vital capacity, a tracheotomy may be a great disadvantage. It is very difficult to get rid of a tracheotomy tube when the vital capacity is only 500 cc or 600 cc and there is no power of coughing, whereas, as we all know, a patient who has been treated in a respirator (Iron Lung) from the first can survive and get out of all mechanical devices with a vital capacity of that figure."[5] Although the indications for tracheostomy established in 1955 are equally appropriate today[5] a tracheostomy is also indicated when: there is a strong history of substance abuse; the patient is uncooperative; the patient has severe intrinsic lung disease with ongoing difficulties in managing respiratory tract secretions for which a mechanical exsufflation device (for mechanically assisted coughing) may be contraindicated or unavailable and access to effective

assisted coughing is inadequate; there are uncontrolled seizures; or there are orthopedic conditions which interfere with the use of noninvasive alternatives, ie, facial fractures, etc.

Basic rehabilitation. Counseling is critical to explain: the importance of the therapeutic goals and how to attain them; the need to avoid unnecessary oxygen therapy; the need to avoid sedatives, obesity, heavy meals, malnutrition, extremes of temperature, humidity, excessive fatigue, crowded areas, or exposure to respiratory tract pathogens; the need for appropriate flu and bacterial vaccinations and early medical attention during URIs; how to help the patient optimize his level of functioning and daily activities; and finally, how to prepare for any elective surgical procedures. Basic rehabilitation techniques including the use of lung expansion techniques by air stacking of mechanical insufflations[74] or GPB[16,59,60] to approach the maximum inspiratory capacity should be performed at least twice a day. The earlier and more aggressively this is introduced, the better may be the ultimate effect on maintaining or possibly improving pulmonary compliance, decreasing microatelectasis, alveolar surface tension, and the work of breathing. A positive pressure blower (Zephyr, Lifecare Inc [now Respironics Inc], Lafayette, Cole), intermittent positive pressure breathing (IPPB) machine, or portable ventilator is useful for delivering the mechanical insufflations.

Although there is little in the medical literature concerning the use of a combination of lung expansion techniques and respiratory muscle exercise for post-poliomyelitis individuals, this combination has been shown to increase pulmonary volumes, respiratory muscle strength, and endurance for spinal cord injured patients with restrictive pulmonary syndrome.[75] Likewise, inspiratory muscle training exercise has been shown to increase respiratory muscle endurance for patients with progressive neuromuscular disease provided that the vital capacity at onset of training was at least 30% of predicted normal.[76,77] These techniques may, thus, be of value for post-poliomyelitis individuals; however, it must be noted that ventilator-assisted individuals have their greatest risk of complications when the vital capacity is 30% or less. Thus, the ventilator-assisted individuals who need help the most accomplish the least by inspiratory muscle training.

Physical therapists can train the postpoliomyelitis individual and caregiver in chest physical therapy techniques, postural drainage, and manual assisted coughing. For some patients, strengthening of the accessory muscles of respiration may improve the vital capacity in the sitting position and increase expiratory flow for more effective coughing.

Preparing for Surgical Intervention

Post-poliomyelitis individuals require surgical intervention more frequently than the general population. It is important to prepare early for post-surgical respiratory management. To be prepared for any emergency surgical procedure, a hospital of the patient's choice should be made aware of the patient's equipment and any special needs before such intervention is realized. For the majority of hospitals who do not own portable ventilatory equipment, this might require 1 month or so of advanced notice to the hospital's risk and claims services, because the use of non-hospital equipment is usually prohibited. Nursing and respiratory therapy in-services may be required and waivers signed assigning liability for mechanical failure to the ventilator providers rather than to the hospital. Ventilator-assisted individuals should be requested to have their ventilatory equipment available throughout their hospitalizations.

Pulmonary function testing should be done on all post-poliomyelitis individuals prior to any elective surgical intervention. Although either restrictive or obstructive pulmonary impairment significantly increases the risk of respiratory complications, the presence of both conditions greatly augments this risk. Except for markedly obese individuals or those with severe sleep-disordered breathing,[27-31] it is unlikely that any patient with greater than 65% of predicted normal vital capacity has significant risk of

respiratory morbidity resulting from surgery and general anesthesia than a physically intact individual.

The lower the vital capacity below 65% of predicted normal, the greater the likelihood of complications. Such patients or those with CAH of any etiology should be trained in receiving maximum insufflations with an IPPB machine or portable ventilator and be taught to use mouth and nasal ventilation in the event of postoperative ventilator weaning difficulties. These techniques should be used for short periods of time daily for 1 to 2 weeks prior to surgery. The patient and the caregivers also should be introduced to both manually assisted and, where available, mechanically assisted coughing as described previously.

Since few centers have expertise in managing ventilator-assisted individuals who use noninvasive methods, it is especially important for a knowledgeable physiatrist or pulmonologist to be involved in any inpatient management. General anesthesia should be avoided in favor of local or regional anesthesia whenever possible and nonessential elective procedures avoided. Simple inhalation anesthetic techniques should be used whenever possible. The use of opioids and muscle relaxants should be used sparingly or avoided. For any surgical procedure requiring intubation, the patient should be extubated postoperatively as soon as full consciousness is regained and noninvasive ventilatory aid immediately provided as necessary. Ventilator-assisted individuals should be returned to their equipment as soon as possible. Besides the differences in flow characteristics, the expiratory volume alarms of the usual hospital ventilators can make it impossible to resume the 24 hour noninvasive ventilatory support techniques to which the patient may be accustomed. Ventilator-assisted individuals with less than 2 hours of free time should also have their back-up ventilator available.

Hypercapnia should be corrected by assisted ventilation. Low-flow oxygen therapy should only be used with caution in the presence of hypoxia (pO_2<60mm Hg) combined with normocapnia or hypocapnia (pCO_2<40mm Hg). Likewise, bronchodilators and methylxanthines should be used when specifically indicated by the presence of reversible bronchospasm. The importance of maximal insufflations and assisted coughing can also not be overemphasized. Mechanical-assisted coughing is especially useful post-abdominal surgery, because these devices require no abdominal muscle effort and abdominal pressures are increased significantly less than during unassisted coughing.[11] Transient microatelectasis, which might cause hypoxia (pO_2 15 mm Hg or more less than predicted for age), can be reversed by frequent maximal insufflations and aggressive assisted coughing. Chest percussion and postural drainage also may be somewhat useful for this purpose.

References

1. Historical Statistics of the United States. *Colonial Times to 1970,* Bicentennial Edition, Part 1. US Department of Commerce, Bureau of the Census, 1975; pp 8,77.
2. *Statistical Abstracts of the United States,* 110th ed. US Department of Commerce, Bureau of the Census, 1990; p 116.
3. Kaufert FL, Kaufert JM. Methodological and conceptual issues in measuring the long term impact of disability: the experience of poliomyelitis patients in Manitoba. *Soc Sci Med.* 1984; 19:609-618. [PubMed Abstract]
4. Lassen HCA. The epidemic of poliomyelitis in Copenhagen, 1952. *Proc R Soc Med.* 1953; 47:67-71.
5. Hodes HL. Treatment of respiratory difficulty in poliomyelitis. In: *Poliomyelitis: Papers and Discussions Presented at the Third International Poliomyelitis Conference.* Philadelphia, Pa: JB Lippincott; 1955:91-113.
6. The OEM Cof-flator Portable Cough Machine. St Louis, Mo: Shampaine Industries.
7. Barach AL, Beck GJ, Bickerman HA, Smith W. Physical methods simulating mechanisms of the

human cough. *J Appl Physiol.* 1952; 5:85-91.

8. Barach AL, Beck GJ, Smith RH. Mechanical production of expiratory flow rates surpassing the capacity of human coughing. *Am J Med Sci.* 1953; 226:241-248.

9. Bickerman HA, Itkin S. Exsufflation with negative pressure. *Arch Intern Med.* 1954; 93:698-704.

10. Barach AL, Beck GJ. Exsufflation with negative pressure: physiologic and clinical studies in poliomyelitis, bronchial asthma, pulmonary emphysema and bronchiectasis. *Arch Intern Med.* 1954; 93:825-841.

11. Beck GJ, Scarrone LA. Physiological effects of exsufflation with negative pressure. *Diseases of the Chest.* 1956; 29:1-16.

12. Bach JR, Alba AS, Shin D. Noninvasive airway pressure assisted ventilation in the management of respiratory insufficiency due to poliomyelitis. *Am J Phys Med Rehabil.* 1989; 68:264-271.

13. Bach JR, Alba AS, Bohatiuk G, Saporito L, Lee M. Mouth intermittent positive pressure ventilation in the management of postpolio respiratory insufficiency. *Chest.* 1987; 91:859-864. [PubMed Abstract]

14. Bach JR, Alba AS. Management of chronic alveolar hypoventilation by nasal ventilation. *Chest.* 1990; 97:52-57. [PubMed Abstract]

15. Bach JR, O'Brien J, Krotenberg R, Alba AS. Management of end stage respiratory failure in Duchenne muscular dystrophy. *Muscle Nerve.* 1987; 10:177-182. [PubMed Abstract]

16. Bach JR, Alba AS, Bodofsky E, Curran FJ, Schultheiss M. Glossopharyngeal breathing and non-invasive aids in the management of post-polio respiratory insufficiency. *Birth Defects.* 1987; 23:99-113. [PubMed Abstract]

17. Halstead LS, Wiechers DO, Rossi CD. Late effects of poliomyelitis: a national survey, In: Halstead LS, Wiechers DO, eds. *Late Effects of Poliomyelitis.* Miami, Fla: Symposia Foundation; 1985:11.

18. Fischer DA. Poliomyelitis: late respiratory complications and management. *Orthopedics.* 1985; 8:891-894. [PubMed Abstract]

19. Howard RS, Wiles CM, Spencer GT. The late sequelae of poliomyelitis. *Q J Med.* 1988; 66:219-232. [PubMed Abstract]

20. Hamilton EA, Nichols PJR, Tait GBW. Late onset of respiratory insufficiency after poliomyelitis. *Ann Phys Med.* 1970; 10:223-229. [PubMed Abstract]

21. Braun NMT, Arora NS, Rochester DF. Respiratory muscle and pulmonary function in polymyositis and other proximal myopathies. *Thorax.* 1983; 38:616-623. [PubMed Abstract]

22. De Troyer A, Deisser P. The effects of intermittent positive pressure breathing on patients with respiratory muscle weakness. *Am Rev Respir Dis.* 1981; 124:132-137. [PubMed Abstract]

23. Estenne M, De Troyer A. The effects of tetraplegia on chest wall statics. *Am Rev Respir Dis.* 1986; 134:121-124. [PubMed Abstract]

24. Carskadon M, Dement W. Respiration during sleep in the aged human. *J Gerontol.* 1981; 36:420-425. [PubMed Abstract]

25. George CF, Millar TW, Kryger MH. Identification and quantification of apneas by computer-based analysis of oxygen saturation. *Am Rev Respir Dis.* 1988; 137:1238-1240. [PubMed Abstract]

26. He J, Kryger MH, Zorick FJ, Conway W, Roth T. Mortality and apnea index in obstructive sleep apnea. *Chest.* 1988; 94:9-14. [PubMed Abstract]

27. Hill R, Robbins AW, Messing R, Arora NS. Sleep apnea syndrome after poliomyelitis. *Am Rev Respir Dis.* 1983; 127:129-131. [PubMed Abstract]

28. Petrén K, Ehrenberg L. Etudes cliniques sur la poliomyélite aigue. *Nouv Inconog Salpetriere.* 1909; 22:373,546,661.

29. Plum F, Swanson AG. Abnormalities in central regulation of respiration in acute and convalescent poliomyelitis. *Arch Neurol Psych.* 1958; 80:267-285.

30. Guilleminault C, Motta J. Sleep apnea syndrome as a long-term sequelae of poliomyelitis. In:

Guilleminault C, ed. *Sleep Apnea Syndromes*. New York, NY: KROC Foundation; 1978:309-315.

31. Steljes DG, Kryger MH, Kirk BW, Millar TW. Sleep in postpolio syndrome. *Chest.* 1990; 98:133-140. [PubMed Abstract]

32. Jubelt B, Cashman NR. Neurological manifestations of the post-polio syndrome. *CRC Critical Reviews in Neurobiology.* 1987; 3:199-220. [PubMed Abstract]

33. Bach JR. Pulmonary rehabilitation. In: Mitchell CW, ed. *Rehabilitation of the Aging and Older Patient.* Baltimore, Md: Urban & Schwarzenberg Inc (in press).

34. Williams A, Santiago S, Stein M. Screening for sleep apnea? *Chest.* 1989; 96:451-453. [PubMed Abstract]

35. Lombard R Jr, Zwillich CW. Medical therapy of obstructive sleep apnea. *Med Clin North Am.* 1985; 69:1317-1335. [PubMed Abstract]

36. Smolley LA. Obstructive sleep apnea: avoiding diagnostic pitfalls. *Journal of Respiratory Disease.* 1990; 11:547-552.

37. Bach JR, McDermott I. Strapless oral-nasal interfaces for positive pressure ventilation. *Arch Phys Med Rehabil.* 1990; 71:908-911. [PubMed Abstract]

38. Curran FJ, Colbert AP. Ventilator management in Duchenne muscular dystrophy and postpoliomyelitis syndrome: 12 years experience. *Arch Phys Med Rehabil.* 1989; 70:180-185. [PubMed Abstract]

39. Carroll N, Branthwaite MA. Control of nocturnal hypoventilation by nasal intermittent positive pressure ventilation. *Thorax.* 1988; 43:349-353. [PubMed Abstract]

40. Leger P, Jennequin J, Gerard M, Robert D. Home positive pressure ventilation via nasal mask for patients with neuromuscular weakness or restrictive lung or chest-wall disease. *Respiratory Care.* 1989; 34:73-79.

41. Waldhorn RE, Herrick TW, Nguyen MC, O'Donnell AE, Sodero J, Potolicchio SJ. Long-term compliance with nasal continuous positive airway pressure therapy of obstructive sleep apnea. *Chest.* 1990; 97:33-38. [PubMed Abstract]

42. McDermott I, Bach JR, Parker C, Sorter S. Custom fabricated interfaces for non-invasive intermittent positive pressure ventilation. *International Journal of Prosthodontics.* 1989; 2:224-233. [PubMed Abstract]

43. Bonham PE, Currier GF, Orr WC, Othman J, Nanda RS. The effect of a modified functional appliance on obstructive sleep apnea. *Am J Orthod Dentofacial Orthop.* 1988; 94:384-392. [PubMed Abstract]

44. Clark GT, Nakano M. Dental appliances for the treatment of obstructive sleep apnea. *J Am Dent Assoc.* 1989; 118:611-619. [PubMed Abstract]

45. Katsantonis GP, Walsh JK, Schweitzer PK, Friedman WH. Further evaluation of uvulopalatopharyngoplasty in the treatment of obstructive sleep apnea syndrome. *Otolaryngol Head Neck Surg.* 1985; 93:244-250. [PubMed Abstract]

46. Thawley SE. Surgical treatment of obstructive sleep apnea. *Med Clin North Am.* 1985; 69: 1337-1358. [PubMed Abstract]

47. Riley RW, Powell NB, Guilleminault C, Mino-Murcia G. Maxillary, mandibular, and hyoid advancement: an alternative to tracheostomy in obstructive sleep apnea syndrome. *Otolaryngol Head Neck Surg.* 1986; 94:584-588. [PubMed Abstract]

48. Yokoyama A, Yamashita N, Mizushima Y, Yano S. Inhibition of natural killer cell activity by oral administration of theophylline. *Chest.* 1990; 98:924-927. [PubMed Abstract]

49. Bergofsky EH. *Cor pulmonale* in the syndrome of alveolar hypoventilation. *Prog Cardiovasc Dis.* 1967; 9:414-437. [PubMed Abstract]

50. Bach JR, Beltrame F. Alternative methods of ventilatory support. In: Rothkopf MM, Askanazi J, eds. *Home Intensive Care.* Baltimore, Md: Williams & Wilkins (in press).

51. Goldstein RS, Molotiu N, Skrastins R, Long S, Contreras M. Assisting ventilation in respiratory failure by negative pressure ventilation and by rocking bed. *Chest.* 1987; 92:470-474. [PubMed Abstract]

52. Drinker PA, McKhann CF The iron lung. *JAMA.* 1986; 255:1476-1480. [PubMed Abstract]

53. Kinnear W, Petch M, Taylor G, Shneerson J. Assisted ventilation using cuirass respirators. *Eur Respir J.* 1988; 1:198-302. [PubMed Abstract]

54. Splaingard ML, Frates RC, Jefferson LS, Rosen CL, Hanison GM. Home negative pressure ventilation: report of 20 years of experience in patients with neuromuscular disease. *Arch Phys Med Rehabil.* 1985; 66:239-242. [PubMed Abstract]

55. Bach JR. Psychosocial aspects of home mechanical ventilation. In: Kutscher AH, ed. *The Ventilator: Psychosocial and Medical Aspects; Muscular Dystrophy, Amyotrophic Lateral Sclerosis, and Other Diseases.* New York, NY: Foundation of Thanatology (in press).

56. Bach JR, Penek J. Obstructive sleep apnea complicating negative pressure ventilatory support in patients with chronic paralytic/restrictive ventilatory dysfunction. *Chest.* 1991; 99:1386-1393. [PubMed Abstract]

57. Bach JR, Alba AS. Total ventilatory support by the intermittent abdominal pressure ventilator. *Chest.* 1991; 99:630-636. [PubMed Abstract]

58. Dail C, Rodgers M, Guess V, Adkins HV. *Glossopharyngeal Breathing Manual.* Downey, Calif: Professional Staff Association of Rancho Los Amigos Hospital, Inc; 1979.

59. Dail CW, Affeldt JE. *Glossophalyngeal Breathing* (video). Los Angeles, Calif: Department of Visual Education, College of Medical Evangelists; 1954.

60. Bach JR, Alba AS. Noninvasive options for ventilatory support of the traumatic high level quadriplegic. *Chest.* 1990; 98:613-619. [PubMed Abstract]

61. Sorter S, McKenzie M. *Toward Independence: Assisted Cough* (video). Dallas, Tex: BioScience Communications of Dallas, Inc; 1986.

62. Bach JR. Mechanical exsufflation, noninvasive ventilation and new strategies for pulmonary rehabilitation and sleep-disordered breathing. *Bull NY Acad Med* (in press). [PubMed Abstract]

63. Elam JO, Hemingway A, Gullickson G, Visscher MB. Impairment of pulmonary function in poliomyelitis: oximetric studies in patients with the spinal and bulbar types. *Arch Intern Med.* 1948; 81:649-665.

64. Bain JA. Late complications of tracheostomy and prolonged endotracheal intubation. *Int Anesthesiol Clin.* 1972; 10:225-244. [PubMed Abstract]

65. Niederman MS, Ferranti RD, Zeigler A, Merrill W, Reynolds HY. Respiratory infection complicating long-term tracheostomy: the implication of persistent gram-negative tracheobronchial colonization. *Chest.* 1984; 85:39-44. [PubMed Abstract]

66. Johanson WG, Pierce AK, Sanford JP, Thomas GD. Nosocomial respiratory infections with gram-negative bacilli: the significance of colonization of the respiratory tract. *Ann Intern Med.* 1972; 77:701-706. [PubMed Abstract]

67. Johanson WG, Seidenfeld JJ, Gomez P, De Los Santos R, Coalson JJ. Bacteriologic diagnosis of nosocomial pneumonia following prolonged mechanical ventilation. *Am Rev Respir Dis.* 1988; 137:259-264. [PubMed Abstract]

68. Moar JJ, Lello GE, Miller SD. Stomal sepsis and fatal hemorrhage following tracheostomy. *Int J Oral Maxillofac Surg.* 1986; 15:339-341. [PubMed Abstract]

69. Deutschman CS, Wilton P, Sinow J, Dibbell D, Konstantinides FN, Cerra FB. Paranasal sinusitis associated with nasotracheal intubation: a frequently unrecognized and treatable source of sepsis. *Crit Care Med.* 1986; 14:111-114. [PubMed Abstract]

70. Pingleton SK. Complications of acute respiratory failure. *Am Rev Respir Dis.* 1988; 137:1463-1493. [PubMed Abstract]

71. Fishburn MJ, Marino RJ, Ditunno JF Jr. Atelectasis and pneumonia in acute spinal cord injury. *Arch Phys Med Rehabil.* 1990; 71:197-200. [PubMed Abstract]
72. Bonanno P. Swallowing dysfunction after tracheostomy. *Ann Surg.* 1971; 174:29-33. [PubMed Abstract]
73. O'Donohue W. Maximum volume IPPB for the management of pulmonary atelectasis. *Chest.* 1976; 76:683-687. [PubMed Abstract]
74. Huldtgren AC, Fugl-Meyer AR, Jonasson E, Bake B. Ventilatory dysfunction and respiratory rehabilitation in post-traumatic quadriplegia. *Eur J Respir Dis.* 1980; 61:347-356. [PubMed Abstract]
75. DiMarco AF, Kelling JS, DiMarco MS, Jacobs I, Shields R, Altose MD. The effects of inspiratory resistive training on respiratory muscle function in patients with muscular dystrophy. *Muscle Nerve.* 1985; 8:284-290.
76. Martin AJ, Stern L, Yeates J, Lepp D, Little J. Respiratory muscle training in Duchenne muscular dystrophy *Dev Med Child Neurol.* 1986; 28:314-318. [PubMed Abstract]
77. Milner-Brown HS, Miller RG. Muscle strengthening through high-resistance weight training in patients with neuromuscular disorders. *Arch Phys Med Rehabil.* 1988; 69: 14-19. [PubMed Abstract]

▲ Top of Article ▲

Content Catalogue   **Source Catalogue**

Reception   **Library**   **Networking**   **Directory**   **What's New?**

The Lincolnshire Post-Polio Network
Registered Charity No. 1064177
An Information Service for Polio Survivors and Medical Professionals

All enquiries, book requests, medical article requests, membership fees, items for newsletters and donations to
The Secretary, Lincolnshire Post-Polio Network
PO Box 954, Lincoln, Lincolnshire, LN5 5ER United Kingdom
Telephone: +44 (0)1522 888601
Facsimile: +44 (0)870 1600840
Email: info@lincolnshirepostpolio.org.uk
Web Site: www.lincolnshirepostpolio.org.uk

redistribution of any articles via any media, with the exception of the aforementioned, requires permission from The Lincolnshire Post-Polio Network and where applicable, the original author(s).

Document preparation: Chris Salter, Original Think-tank, Cornwall, United Kingdom.
Created: 1st March 1999
Last modification: 21st January 2010.