

Living and Breathing with Weak Respiratory Muscles

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Network member Dr John Colebatch was born in 1928 at Northam, Western Australia. He entered first year medicine at the University of WA, completing the course and graduating in medicine from Adelaide University in 1951.

In April 1953 while working at the Infectious Diseases Branch of the Royal Perth Hospital, he contracted poliomyelitis, becoming totally paralysed and requiring treatment in an iron lung. After a year recovering in hospital he rejoined the medical staff of the Royal Perth Hospital with responsibility for infectious diseases and rehabilitation.

In 1958 he commenced research on lung disorders in the Department of Medicine, University of Sydney. He continued his research at the University of California, San Francisco and then at the University of Oxford. In 1964, he was appointed to the academic staff of the University of New South Wales as Senior Lecturer in Medicine and joined the Department of Respiratory Medicine, becoming Associate Professor in 1970 and Chairman of the Department in 1985. In 1970-71 he was a Senior Fulbright-Hayes Research Scholar and a Visiting Associate Professor of Physiology at the Harvard University School of Public Health. From 1968 to 1992, he was a Senior Specialist in Respiration to the Royal Australian Navy. In 1989, he received the inaugural award of the Fisons medal of the Thoracic Society of Australia and New Zealand. In 1992, in recognition of his research on the lungs, he was made a Member of the Order of Australia.

Because of increasing muscular weakness – the post-polio syndrome – Dr Colebatch retired at the end of 1992 and was appointed a Visiting Professor of Medicine, continuing part time work until 1996. Muscular weakness led to the use of a ventilator at night and an electric wheel chair to maintain mobility.

We are grateful to John for writing this article especially for Network News. He will be pleased to respond to any questions on this subject – just send them to the Editor.

The return of muscle weakness 20 or more years after the initial paralysis and its relentless progression is well known to all who have experienced the post-polio syndrome. In this account, which is based on my own experience, I will be primarily concerned with the recurrence of respiratory muscle weakness in those patients who during the acute stage of their illness suffered respiratory muscle paralysis. I will assume that the lungs are normal and that chronic respiratory failure is the result of respiratory muscle weakness alone.

Breathing, when it fails, is a whole of life issue; it affects all activities during the day, as well as the quality of sleep. The aim of management is to maintain the best possible activity during waking hours. To understand how this can be achieved it is necessary to consider two important issues – first, how breathing is regulated and second, how muscles produce movement and enlarge the chest.

Background

In April 1953, while working in an Infectious Diseases Hospital, I became completely paralysed as a result of poliomyelitis and was placed in an “Iron Lung”. It was 74 days before I was freed from this coffin-like machine. About three months after my initial paralysis when I stood up for the first time I was unable to breathe at all, owing to the effect of gravity increasing lung volume. Since then I have always worn an abdominal support and/or a tight belt around the lower abdomen before standing up. It was one year before I was able to leave hospital, walking with the aid of crutches. I maintained a reasonably active life, including working overseas, but after 25 years it became clear that muscle

weakness was returning and gradually its severity increased until once again I was unable to walk and required assistance for showering and dressing. The relentless progress to dependence has been a deeply distressing experience.

In 1991, after a "Sleep Study" I was advised to use a machine to produce continuous positive airway pressure (CPAP) during the night. I persevered with this treatment, but was able to sleep using it on one night only. Subsequently I realized that CPAP was not the appropriate treatment for anyone with weak inspiratory muscles and discontinued my use of this machine.

In September 1992, after another "Sleep Study", I started using a fixed volume ventilator at night. Initially, there was some improvement in sleep quality, but this did not persist. Because it prevented spontaneous breathing, I found this ventilator intolerably uncomfortable to use. As a result of its use with an excessive level of ventilation, breathlessness during the day became severe and interfered with activity. After three weeks of nocturnal ventilation, I became troubled with irregular heart beats. By decreasing the amount of ventilation during the night, the irregular heart beats were abolished and breathlessness decreased.

In November 1992, I started using a BiPAP Ventilator which was intended to allow me to initiate breathing and be comfortable to use. However, this early model was unduly sensitive and was triggered into inspiration because the heart beat caused a small flow of air (about 10 ml) into the lungs. As a result, my breathing was not normally regulated, but was abnormally increased because it was initiated by the heart beat rather than by my own inspiratory efforts. I was soon troubled again with severe daytime breathlessness and irregular heart beats and, in addition, a feeling of impending death. These problems were overcome by the use of a modified BiPAP which was not triggered by the heart beat.

Recognition of Failing Breathing Muscles

Difficulty with breathing is likely to develop in post-polio subjects who suffered respiratory muscle paralysis during the acute stage of their illness. Initially there is increased breathlessness and fatigue and, later, difficulty sleeping. To assess this problem it is essential to measure:

- breathing capacity, and
- arterial blood gases.

Breathing capacity is usually assessed by measurements of vital capacity (VC) and the maximum volume expelled in 1 second (FEV1). Before there is a serious problem with respiratory failure, both VC and FEV1 decrease to less than 50% of the expected value. But a more significant measurement is inspiratory capacity (IC) or the volume which can be inspired from the resting position. It is this volume which represents how much the subject can breathe. As lung volume increases, inspiratory muscles are shortened and IC decreases, that is, the ability to inspire decreases. That is why it is more difficult to breathe when sitting or standing compared with lying down. A tight belt around the lower abdomen will decrease lung volume when standing and increase IC and therefore the ability to inspire.

Arterial blood gases require collection of arterial blood which is normally done in a hospital in close relation to the laboratory where the measurements are made. An increased arterial CO₂ level defines the presence of chronic respiratory failure. By the time this becomes a problem, arterial CO₂ will usually have risen from a normal level of 40-45 mm Hg to around 55 mm Hg. If arterial CO₂ remains in the normal range, assisted ventilation is unlikely to be required. In the presence of normal lungs, oxygen saturation is well maintained, so that oximetry is unreliable and of little value in assessing the presence of chronic respiratory failure.

The respiratory muscles and chest wall together make a pump which moves air into and out of the lungs and is referred to as “ventilation”. The pump is driven by the nervous system. When the pump cannot provide adequate ventilation there is a feeling of breathlessness and the arterial CO₂ increases until the quantity produced is expired in the smaller volume of ventilation. This situation is referred to as “chronic respiratory failure”.

When weakness of breathing muscles has caused chronic respiratory failure nocturnal ventilation is required. This ensures adequate ventilation during the night and allows recovery of fatigued muscles. The aim of this regimen is to maintain activity and independent breathing during the day. To minimise the increase in lung volume with gravity, when sitting or standing, it is essential to provide abdominal support. This will help to maintain the ability to inspire.

Managing Nocturnal Ventilation

When breathing is failing, one of the symptoms is difficulty sleeping. Sleep studies may be undertaken to help define the breathing problem. However, these studies are complex and in my view may lead to errors in management. Notwithstanding evidence of obstructive sleep apnoea, which is commonly observed, treatment with CPAP is not appropriate for anyone with weak respiratory muscles. This is because an increase in airway pressure increases lung volume and shortens respiratory muscles making it more difficult to further increase lung volume and maintain breathing. It is as if the subject were sitting up all night instead of lying down and instead of resting the inspiratory muscles it makes their work more difficult.

Breathing is driven by the acidity of fluids around the brain which in turn reflect metabolic activity. The best type of breathing assistance is one which allows this regulation to continue in its natural state. That is, the subject’s brain should drive the ventilator as part of the chest pump, the ventilator should not determine the amount of ventilation independently of the subject’s metabolism. It follows that the subject initiates inspiration to which the machine responds and supports – a demand-driven machine. A pressure support ventilator fits this criterion and such machines are in common use. My preference is for the BiPAP Ventilator operated in demand mode. I have used this ventilator every night for more than eleven years.

Fixed-volume ventilators are uncomfortable to use at night for anyone who retains some ability to breathe independently. They also do not permit the subject to regulate his breathing. A similar disadvantage applies to a pressure-assist ventilator set to a fixed time, which excludes the subject from initiating inspiration. If there is concern about the occurrence of apnoea during sleep, this possibility can be overcome by setting the ventilator to a slow back-up rate, say 8 per minute, which will maintain sufficient ventilation if needed, but not interfere with the subject’s own breathing.

Improving Daytime Activity

One of the objects of nocturnal ventilation is to improve daytime activity. For this to be achieved the level of ventilation during the night must not exceed what the subject can maintain when breathing independently during the day. This is best achieved by using a ventilator in demand mode with the subject retaining regulation of breathing.

There should be no attempt to decrease to “normal” an increased arterial CO₂. The level of CO₂ is set by the ability of the chest pump to respond to the demands for breathing during daytime activity. To lower CO₂ during the night, in these circumstances by excessive ventilation, will increase daytime breathlessness. It may also be dangerous by causing irregularity of the heart beat. Normally CO₂ is a little higher during the night than during the day and this situation should be allowed to continue. If excessive nocturnal ventilation is

suspected it may be worthwhile to measure the electrolytes. The risk is an abnormal loss of potassium resulting in irregularity of the heart beat. This disorder may be fatal. Potassium supplements will correct this problem.

Weakness of post-polio muscles is an ongoing problem which cannot be overcome by exercise. It is sufficient to try to maintain the activities of daily living. To do this it is essential to have adequate periods set aside for rest during the day. (Exercise programs are for an earlier stage of life.) Post-polio paralysis is not a disease of muscle, rather it reflects a loss of those motor nerve cells and fibres on which initial recovery depended. The surviving muscles with their remaining nerve supply are normal. It follows that nutritional supplements or anabolic steroids aimed at improving muscle function are unlikely to have any lasting effect. On the other hand an adequate intake of the vitamin B group may be helpful for their essential role in the nervous system.

Besides exercise, metabolism and with it the demand for breathing is increased by intake of food. Breathlessness during the day, while at rest, is distracting and can be decreased by limiting intake of food to the smallest amount that can be tolerated. This regimen has the additional benefit of discouraging an increase in body weight when any increase is a serious disadvantage for continuing activity.

In situations where exertion cannot be avoided such as toileting it is an advantage to have the use of a ventilator. This can make the difference between dependence and independence.

The slowly progressive nature of the post polio syndrome means that new challenges to activity and independence arise year by year and even each few months. To meet the particularly difficult challenge of respiratory failure and yet maintain some independent activity it is essential to consider carefully all possible adjustments. Simple things such as limiting daytime intake of food and ensuring rest periods can help to reduce discomfort and make the days a little more enjoyable.



An iron lung on display in the Prince Henry Hospital Museum. John Colebatch and Ian Neering who share their experiences of respiratory failure in this *Network News* both spent considerable time in a similar “coffin-like” box when they contracted polio in the 1950s.

Sleep Apnoea Revisited

Ian Neering PhD MSc

Long-time Network member Ian Neering was Associate Professor of Physiology and Pharmacology at the University of NSW and now, as is appropriate with his degree of decrepitude (Ed. his description, not mine!), runs a drug consultancy from his home office.

Ian's article below provides further insights into the subject of sleep apnoea, and reinforces the message of Dr Colebatch's article – it is imperative for polio survivors to have their particular causes of sleep apnoea correctly diagnosed and appropriately treated.

Your articles in the last PPN *Network News* have prompted me to put down my experiences with sleep apnoea as they might be of interest to some of your readers. Let me stress that these are my personal experiences and people should not necessarily extrapolate from them to form any sort of generalisation.

By way of background, I should say that I contracted polio in 1950 when I was around 4 years of age. I was completely paralysed and spent a good deal of time in an iron lung. Over the years, with the sort of therapies most of your readers will know only too well, I made a pretty good recovery and was able to lead quite an active life without any prostheses of any kind. I have quite a marked kyphoscoliosis with significant deformity and associated low functional lung volumes. At around the age of 40, I started to notice the signs of PPS and my general condition started to deteriorate. I took to using a walking stick and was forced to modify my life style significantly. When I had respiratory infections, they lasted longer and were increasingly troublesome. I now need crutches to get around.

In 1993, a dose of flu saw me admitted to hospital with life-threatening respiratory failure which basically means that my lungs could not function sufficiently well to provide me with an adequate supply of oxygen. I was on a ventilator for a few days but was fortunate enough to make a good recovery. In the aftermath of my period in intensive care, sleep studies were performed and sleep apnoea was diagnosed. The respiratory specialist theorised that cumulative fatigue of my respiratory muscles, partially as a consequence of the sleep apnoea, was responsible for the respiratory failure and a CPAP device was prescribed.

Let me tell you, I struggled with that machine! Let alone the fact that no mask seemed to fit around my nose properly and jets of air burbled and squeaked from the poor seal around my face unless I lay "just so" in bed and didn't move a muscle. Not conducive to a good night as you can imagine. I felt no improvement in my overall well being after weeks of persevering and follow up visits to the doctors.

I'm afraid I have a general antipathy to the medical profession not uncommon amongst old polios and a general mistrust of "blanket treatments". I, like many polios, have become rebellious towards doctors' prescriptions and prefer to find my own bumbling way to the solutions that best suit me. At my wit's end, I decided to take matters into my own hands and here I have an advantage over most in that I am a physiologist by training.

I need to digress for a moment here. Your excellent articles on sleep apnoea didn't say very much about the sleep study used to definitively diagnose the condition. When you go in for an overnight study, you are fitted up with a number of sensors. Cables are attached to your scalp for an EEG which records your brain waves and allows the investigators to assess your level of sleep. A finger probe records oxygen saturation of the blood which is the critical measure telling us we are not receiving sufficient oxygen as a result of the sleep apnoea. A more sensitive measure of ventilation is that of carbon dioxide but not all sleep studies include this. Wires are also attached to our chest so that the activity of the respiratory muscles can be recorded. This is known as an EMG. It's important to understand that EMG records muscle activity only. A reduced EMG activity recording could be caused by a reduced drive from the motor nerves controlling the muscle or from a weakened muscle, unresponsive to nerve stimulation. There may be other sensors attached to measure airflow, leg movements and so

on. A good sleep study will look at all of these parameters; an inadequate one will not, and may lead to a misdiagnosis.

Now, your article referred quite correctly, to two main types of sleep apnoea: obstructive sleep apnoea and central sleep apnoea. In the former, the problem is mechanical in that respiratory muscles may be working well but actual airflow into the lungs is restricted for any of a variety of reasons. In the latter, there is reduced respiratory drive from the brain to the muscles of respiration resulting in a reduced or absent inspiratory effort. Weakness of respiratory muscles will exacerbate both forms of apnoea. Dr Joyner, in her classification, quite reasonably, has delineated a category just for polios which contains aspects of both forms of apnoea though, strictly speaking, does not add a third category to the two groups already defined.

A sleep study, by looking at the temporal relationship between the activity in the respiratory muscles, air flow and oxygen saturation of the blood, can determine what form of sleep apnoea you suffer from. The problem however is that getting to sleep with all those wires attached to you, in what for many of us is an unpleasant environment with unfortunate associations is not all that easy! The consequence is that we may not actually reach the deepest form of sleep during which REM or dream sleep occurs. The investigating physician therefore will not have a complete picture of what your breathing does during sleep.

Enough digression. I was lucky enough to be able to borrow (with appropriate bribing and inducements) some bits of equipment and set up to monitor my sleeping at home. I got hold of an oximeter to measure blood oxygen saturation. Diaphragm EMG was measured using a little battery operated portable device used by sports people. I measured airflow by taping a thermistor (which measures temperature of the air) under my nose. Measuring EEG was beyond the range of my ability to equip. It wasn't really necessary. The hardest part was logging all the data. After all, I needed to be able to record oxygen saturation, EMG and airflow over a period of at least 6 hours. After fiddling with chart recorders and ending up almost buried in paper upon waking, I ended up recording all the data continuously on my computer. My partner was not particularly impressed that I had turned our bedroom into a laboratory but I soothed her with a vision of a well-rested husband who would not be snappy and grumpy if he had a decent night's sleep. I can't adequately describe my satisfaction to you when I finally saw the definitive result. Obstructive apnoea is characterised by an **increased** inspiratory effort coincident with the decrease in airflow. In other words, when the airway is obstructed, we try a whole lot harder to breath in, at least initially, and you can see this in an increased response in the tracing of the EMG from the muscles that help us inspire. In my tracing, respiratory effort just faded away coincident with airflow, breathing stopped because the muscles were no longer being asked to contract, a sign of a centrally mediated sleep apnoea. My problem I now knew was that during sleep, the drive to my inspiratory muscles, the messages from my brain telling my muscles to breath (respiratory drive), was reduced. In fact it would cease all together, causing blood oxygen content to fall until all those alarm mechanisms that the body has to detect such a dangerous malfunction caused me to wake up.

It is known that respiratory drive decreases during sleep even in normal individuals. Many polios have weakened respiratory muscles. It seems not unreasonable to suppose that for some polios the combined reduction in respiratory drive from the brain and the weaker muscles could result in the sleep apnoea. EMG recordings would be unable to dissect out this scenario. Remember, the EMG cannot distinguish between a reduction in nerve impulses from the brain to the muscle, or a weakly contracting muscle.

How did this new knowledge make a difference? Well, it meant that the CPAP machine was quite inappropriate for my needs. CPAP is great for the obstructive types of apnoeas where the positive air pressure from the machine "splints" the airways in an open position and prevents their collapse during the respiratory cycle. It's a bit like breathing in a gale. CPAP won't help you if you simply stop breathing because its flow of air is continuous. There's no cycling. Armed with my new data, I was able to show my respiratory physician that a different form of therapy was required. He promptly prescribed a BIPAP machine which is a bit like a ventilator in that it cycles through an inspiratory and an expiratory phase. There is an obstructive component to

my sleep apnoea and the BIPAP machine will help against this also. That machine is now more than 10 years old and has become an essential chattel of my life.

Since using sleep support, I have had fewer respiratory infections and those I have had, have had a more benign outcome.

Why have I felt the need to put all of this down on paper? My main motivation is a fear that some of you may go through the same misdiagnosis that I did. My message is: **for those of us who had respiratory paralysis during the acute phase of polio infection (and possibly for others), CPAP may NOT be the treatment of choice** either because of muscle weakness, reduction of central drive or a combination of both.

I still have trouble with masks. Having spent much time and effort making plaster casts of my face and attempting to mould masks, cast masks, and experiment with various materials, I've found that the "disposable" resuscitation masks sold by the St John's Ambulance people work as well as any. Use of these with a fabric "gasket" and my partner's design of head harness has enabled me to sleep relatively comfortably even allowing me to toss and turn as the dictates of comfort demand.

One other fascinating aspect of these machines that I've not seen mentioned anywhere else is the tendency, for me at least, to cause psychological dependence. This manifests as an inability to sleep at all without the machine. On occasion when I've been camping and my car battery fails, in a power outage, or when I have had to travel in an emergency without my machine, I simply can not sleep without it. On one occasion I had no sleep for two consecutive nights. On the third night, I was so exhausted I did manage to sleep fitfully. I wonder if others have noticed this effect?



Vale – Alicia Lee and Joan Clarke

It saddens us to report the passing in recent weeks of two long-time Network members.

Dr Alicia Lee contracted polio as a child in 1937. Alicia served on the Network's Management Committee for several years in the 1990s and her input was much valued. If she saw injustice of any kind, she was not one of the silent majority but did something about it. For example, as Convenor of the *Callan Park Action Group*, Alicia was a vocal opponent of the State Government's several attempts to sell off the site. Alicia was also an inveterate letter writer, to politicians and newspapers alike, on a wide range of subjects. She insisted that language describing people with disabilities be positive and enabling. As a staunch advocate of access for people with disabilities, Alicia saw the need to not focus solely on the requirements of those using wheelchairs but urged others to recognise the different needs of those with walking difficulties. Alicia's willingness "to get out there and do it" will be missed.

Joan Clarke passed away peacefully on 1 June. Joan contracted polio at the age of three in 1923, while she was in a train travelling between Sydney and Goulburn. She went on to become an author, editor and researcher. Two of her books are of interest to polio survivors: *Dr Max Herz, Surgeon Extraordinary : The human price of civil and medical bigotry*, published in 1976, and Joan's autobiography *All on One Good Dancing Leg* which was published in 1994. Joan contributed to the Network over many years by convening a Telephone Support Group covering the Manly Peninsula area, and by writing articles, letters and poems for *Network News*. Readers might remember her humorous piece *In Memory of Oscar*, written about her faithful scooter.

Her son Jon is holding a Wake in celebration of "*Joan's Glorious Life*" on Friday 9 July from 12 noon at North Sydney Leagues Club, 12 Abbott Street, Cammeray 2062. Everyone who knew Joan is welcome to come along. Enquiries to Jon Clarke on (02) 6684 1710 or by email to <jonc@nrhs.nsw.gov.au> or mail to 15 Hottentot Crescent, Mullumbimby 2482.