Independence through Ventilation

– *Interview with Prof Douglas A McKim*

Conducted by Dr Chan Yeow

TREMENDOUS EPHEMERAL interest has been generated in amyotrophic lateral sclerosis (ALS) by the recent Ice Bucket Challenge, which was a global phenomenon. Coincidentally, Singapore's Ministry of Health had recently invited Prof Douglas A McKim to be the Healthcare Manpower Development Plan Visiting Expert for Home Ventilation in Financial Year 2014/2015. Hopefully, the fruits of his visit in January this year will be more durable and impactful.

Prof McKim, a respirologist from Ottawa, Canada, has dedicated his professional life to caring for patients with neuromuscular weakness (such as those with ALS), enabling them to live independently using breathing and cough assistance. Dr Chan Yeow, director of Tan Tock Seng Hospital's Home Ventilation and Respiratory Support Service, checks in with Prof McKim to find out more about his work.

Dr Chan Yeow – CY: Douglas, welcome to Singapore. Can you tell us what some of the things that have left a deep impression on you are?

Prof Douglas A McKim – DM: I was not quite expecting such a green city. I was expecting a dense concrete jungle, somewhat like Hong Kong. You have such towering green trees with beautiful ferns growing on their branches and orchids everywhere!

Working with patients requiring ventilation

CY: Tell us about a typical work week.

DM: I work in Ottawa Hospital Rehabilitation Centre as director of the CANVent (Canadian Alternatives in Non-Invasive Ventilation) programme within the Respiratory Rehabilitation programme. I look after patients with respiratory insufficiency due to neuromuscular and restrictive thoracic disorders, and have one other colleague who looks after COPD (chronic obstructive pulmonary disease) rehabilitation. I spend approximately one-and-a-half days weekly in clinic seeing new patients and reviewing follow-up patients. I also review patients in acute care and ICUs (intensive care units) within the hospital, and provide advice

on ventilator weaning, non-invasive airway clearance and tracheostomy decannulation. The other half of my time is spent as a sleep physician – I am director of the Sleep Laboratory, and I see patients, often with severe sleep disordered breathing, respiratory failure and review sleep studies.

CY: Long term ventilation is such a niche area. How did you get started?

DM: All of us encounter a moment that marks a turning point in our professional lives. In my case, as an internal medicine resident in respirology in 1987, I looked after a pregnant mother with severe chest wall restriction from kyphoscoliosis. She was in severe hypercapnic respiratory failure. My attending physician prescribed non-invasive ventilation (NIV) for her, and we managed to help her deliver a healthy daughter. I have been intrigued by NIV from that time onwards.

CY: In my case, it was looking after a middle-aged gentleman in ICU with high cervical fracture. How did the mother and daughter do, and are you still in touch with her?

DM: Her condition was stable after delivery so she gradually withdrew from NIV and follow-up. I have only recently (after 20 years) seen her again in clinic. She was again in respiratory failure due to severe restriction, so we have restarted her on NIV and also introduced LVR. Her daughter is now a beautiful young lady!

The value of sighs

CY: What is LVR?

DM: Lung volume recruitment. The vital capacity (how much air one can maximally exhale into a spirometer) is an index that tells us about the performance of the pulmonary system. It is affected by lung volume, lung stiffness, resistance in the small airways, chest wall stiffness (ribcage and abdomen), and condition of the breathing muscles.

You and I take between five to ten sighs each hour, and each sigh is double or triple our resting tidal volume. During exercise or straining, we may inhale to full lung capacity. Deep breaths have been shown to help stimulate surfactant production, which decreases alveolar surface tension. With muscle weakness, one is unable to inhale deeply, and over time, the intercostal muscles and sternocostal joints stiffen. The alveoli remain small, and may collapse from mucus plugging, and over time the load to the breathing muscles actually increases, which constitutes additional insult to the already weak breathing muscles.

CY: I don't think we dwelt so much on breathing muscles in medical school.

DM: This is an area of great importance. You would not imagine a cardiologist not considering the state of the myocardium. A student can tell you that a stiff, weak joint should be mobilised and yet we ignore a vital organ surrounded by joints! Why should a respirologist not be interested in the state of the breathing muscles and thorax?

Managing patients with different degrees of respiratory deterioration in neuromuscular disease

DM: In our practice, every patient with ALS or a myopathy is immediately referred to us. Initially, their breathing condition may be normal, and we consider them at-risk. We encourage infection prevention (flu and pneumococcal vaccination) and make the baseline lung function measurements.

As their condition progresses, they have difficulty taking deep breaths and coughing effectively. We introduce LVR (using a manual resuscitator bag and a one-way valve) and teach manually assisted cough. They may also benefit from mechanical cough assistance with "in-exsufflation".

In the next stage, they begin to have difficulty sleeping, having many arousals and dips in oxygen, particularly during rapid eye movement (REM) sleep. This is because our skeletal muscles (except the diaphragm and eye muscles) become paralysed during REM to prevent us acting out our dreams. At this stage, they may benefit from non-invasive mask ventilation during sleep.

By the time they need some ventilatory assistance in the day (more than 12 out of 24 hours), we typically prescribe mouthpiece ventilation if they are able to speak, safely swallow and protect their airway. This allows them to get additional breathing support as and when needed, and also allows them to breath-stack to recruit more normal lung volumes.

At any point along this trajectory, if they develop respiratory failure from infections, using these techniques offers them a better chance of not needing intubation, or if intubated, being extubated back to NIV and cough assistance.

CY: Isn't 24-hour ventilation a very expensive proposition to a healthcare system?

DM: On the contrary! Specifically, for 24-hour ventilated Duchenne patients, in more than 80 patient-years, we have only had four admissions attributable to respiratory causes. If you consider that each day in ICU costs thousands of dollars, this constitutes a tremendous cost savings, and since NIV is less complicated, quality of life may be better.

The province of Ontario operates a ventilator equipment pool for long term ventilator users. Through bulk purchase and recycling of used ventilators, it was able to decrease the equipment cost for a fully ventilated patient to less than CAD$1,000 (S$1,100) per patient per year (2005).

CY: What are your thoughts on tracheostomy?

Prof McKim with one of his long-time Duchenne muscular dystrophy patients, who has been using mouthpiece ventilation for over 12 years

DM: Often they are not necessary when the patient has adequate bulbar function, and the correct NIV and airway clearance therapies are applied. Patients with severe bulbar dysfunction will not do well on NIV and will die unless trached. Tracheostomy can be compatible with good quality of life, but is more costly and more care is required with more frequent hospitalisation. The patients also suffer complications that come with the surgery and an artificial airway.

Ventilation, independence and quality of life

CY: How is the quality of life of your patients on 24-hour ventilation?

DM: Pretty good. I have had students who have graduated from college, and are holding jobs. They can even attend ice hockey matches. One of my ALS patients was the deputy governor of the Bank of Canada, and he continued working even when he was on 24-hour ventilation.

Choosing to stop ventilatory treatment

CY: That sounds too good to be true. Do you have patients who suffer and want to stop it all?

DM: We initiate advance care planning discussions early, and frequently review their decisions. This is one of the most difficult but essential tasks in my work. Basically we have treatments that prolong survival, but the illness often takes its toll in disability and dignity. When I was a medical student, we were less clear in our understanding and used terms like "passive euthanasia". Now we recognise that giving up ventilation is equivalent to a cancer patient foregoing surgery or chemotherapy, or even a patient with ischaemic heart disease who refuses to take antiplatelet therapy. Basically, each patient has a right to decide what treatment to receive as long as they have the capacity to make this decision and are fully informed.

CY: In Asia, often the family makes a collective decision. Filial piety, Asian values…

DM: Yes, perhaps true, but in our view anyway, the patient has the final say. It is the patient who lives with the decision.

CY: Do patients suffer when ventilation is withdrawn?

DM: We need to make sure the terminal weaning is done carefully, with patient comfort ensured. Sufficient narcotics and sedatives should be administered to ensure absence of shortness of breath, pain or fear, but the patient should not "die because of the narcotic dosing". This is the principle of double effect, and is widely accepted.

Concluding thoughts

CY: What is your assessment of our care of patients with neuromuscular respiratory insufficiency?

DM: I have witnessed enthusiastic teams, with fantastic collegiality, in several of your hospitals. It is good that you have family physicians involved in your work. The system of co-payment is wise, will contribute to sustainability of the services and prevents abuse of healthcare funding.

CY: However, chronic respiratory insufficiency is not even recognised as an entity, and our patients have difficulty paying for their care.

DM: This is a shame. In Ontario, the provincial health plan recognises that these patients are more complex than the typical COPD/asthmatic patient, and therefore, not only recognised, there is a special, extra fee code for caring for them.

CY: What are your plans for the future?

DM: We are developing a CANVent NIVAM (Non-Invasive Ventilation and Airway Management) website, which will make educational materials and instructional videos available to patients and their families. We have also completed and published some retrospective data on the effects of LVR, and are currently performing two prospective randomised studies on these techniques in multiple sclerosis and Duchenne muscular dystrophy.

CY: Thank you for this interview and for all the things we have learnt from you. Wishing you, your family, your team, and patients all the very best!

DM: Thank you for the opportunity to visit this beautiful country and to learn from your approaches as well. All the best to you all too! ■