**CLINICAL VIGNETTE**

# Advanced non-invasive ventilation for post-polio syndrome chronic respiratory failure

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***Case presentation***

The patient is a 66-year-old male with hypertension, diabetes, and obesity who presents with exertional dyspnea. His past medical history is significant for polio at age 5 with bulbar symptoms and respiratory failure. The patient reports that he was in an "iron lung" and that the severe symptoms lasted for about three months. He spent approximately one year in the hospital and made significant recovery with the exception of moderate right shoulder weakness and very occasional coughing while eating. These symptoms were stable for the next 40-45 years.

At age 50, the patient noted shortness of breath during heavy exertion, fatigue, and progressive bilateral upper extremity weakness. Cardiopulmonary findings included an elevated right hemidiaphragm on CXR, restriction on PFTs, changes and hypercapnia ($PCO_2$ 58mm Hg) and hypoxemia ($PaO_2$ 40 mm Hg). He was assessed by neurology, and his progressive weakness was ultimately attributed to post-polio syndrome.

The patient's wife also noted that he was having episodes of shallow breathing, snoring, and choking at night. He was started empirically on continuous positive airway pressure (CPAP) by his outpatient physician for the treatment of possible sleep apnea without improvement of symptoms. He continued to be severely fatigued and dyspneic with exertion.

Polysomnography (PSG) with end tidal $CO_2$ monitoring ***demonstrated no obstructive apneas, central apneas, or hypopneas.*** Hypoventilation was demonstrated with baseline transcutaneous $CO_2$ ($TcCO_2$) 51-52 mm Hg and baseline oxygen saturations $SaO_2$ 93-95% on 1 LPM. Bi-level positive airway pressure (BPAP) titration improved $TcCO_2$ to 41-43 mm Hg at a setting of 16/8 cm $H_2O$.

The patient remained symptomatic with daytime fatigue and shortness of breath despite BPAP treatment and was referred for pulmonary/sleep evaluation. At the initial visit, the PAP machine was interrogated and was found to be at a sub-therapeutic setting of 9/5 cm $H_2O$. The setting was adjusted to 16/8 cm $H_2O$ with some improvement in his daytime fatigue. During the treatment process, the patient's new insurance carrier required repeat sleep study for ongoing coverage of the BPAP. Repeat polysomnogram with average volume-assured pressure support (AVAPS) titration provided the results in Table 1.

**Table 1.**

| | **Baseline PSG 3 months ago** | **Baseline awake at time of AVAPS titration** | **AVAPS titration** |
|---|---|---|---|
| AVAPS (cmH2O) | 0 | 0 | Max IPAP 25; Min IPAP 18; EPAP 6; Tidal Volume 640ml; BPM 20 |
| Position | Lateral | Lateral | Lateral |
| Sleep State | REM, NREM | N/A | REM, NREM |
| Baseline SaO2 Level | 93-95% | 86-88% | 95-97% |
| Supplemental O2 | 1 LPM | 0 LPM | 1 LPM |
| Minimum O2 Saturation | 90% | 86% | 92% |
| Recorded Sleep Time | 1.9 hours | N/A | 7.1 Hours |
| TcCO2 baseline (mmHg) | 51-52 | 60-62 mmHg | 43-46 mmHg |
| TcCO2 reading at higher than 50 mmHg | 100% | 100% | 0% |
| AHI | 0 | 0 | <1/hr |
| AHI during REM sleep | 0 | 0 | <1/hr |

Hypoventilation improved with an AVAPS setting of Maximum IPAP 25; Minimum IPAP 18; EPAP 6; Tidal Volume 640 mL with 20 breaths per minute $TcCO_2$ baseline improved to 43-46 mmHg from 60-62 mmHg. On the new AVAPS treatment settings, the patient reported significant improvement of the daytime symptoms.

***Discussion***

Post-polio syndrome (PPS) describes late manifestations of poliomyelitis, which may occur many years after the acute infection. The average time until onset is about 35 years with an incidence of up to 62%.[1]

The four criteria for diagnosis are: history of poliomyelitis with residual motor neuron loss, neurologic stability for many years post-recovery, gradual onset of new weakness or fatigue, and exclusion of other conditions that could cause similar manifestations.[2]

The muscular weakness is typically progressive, and asymmetric, and can be proximal or distal. Muscle fatigue often occurs with exertion. The weakness can also present as respiratory insufficiency, sleep apnea, and bulbar dysfunction.[3]

The patient's PSG demonstrated that his symptoms were caused not by obstructive sleep apnea but rather by hypoventilation secondary to neuromuscular weakness and partial diaphragmatic paralysis (suggested by imaging).

Respiratory insufficiency from post-polio syndrome can often be managed with non-invasive ventilation (NIV, also known as non-invasive positive pressure ventilation - NIPPV) with only a small minority requiring tracheostomy and invasive mechanical ventilation.[4] In patients with sleep disorders related to post-polio syndrome, use of attended, or in-lab, PSG is important to distinguish amongst obstructive sleep apnea, central sleep apnea, and hypoventilation, which are managed distinctly.

With this patient, PSG with AVAPS titration was chosen as opposed to standard BPAP titration study. AVAPS is an automatically titrating form of BPAP, where a set expiratory tidal volume is targeted by automatic adjustment of pressure support. This is thought to compensate for the variation in respiratory drive and muscle load with different sleep stages in chronic respiratory failure, or as in this patient's case, with progressive neuromuscular weakness.

AVAPS has generally demonstrated improvements in both daytime and nighttime hypercapnia in patients with chronic hypoventilation in small studies. A small, randomized crossover study demonstrated greater reduction in nocturnal carbon dioxide and bicarbonate levels over 6 weeks with AVAPS compared to CPAP or fixed BPAP.[5] AVAPS demonstrated improved control of nocturnal hypoventilation in 12 patients with obesity hypoventilation syndrome compared to BPAP, though with slight reduction in perceived comfort.[6] However, this has not been consistently shown. In a single-blinded RCT of 46 patients with obesity hypoventilation, $PCO_2$ and health-related quality of life were improved in both AVAPS and fixed BPAP compared to baseline without significant difference between the two groups.[7] Though more studies are needed at this time to establish superiority of AVAPS to fixed BPAP, the conceptual automatic adjustment with progressive weakness was a deciding factor in selecting this mode of non-invasive ventilation in this patient. Current groups that may potentially benefit from AVAPS are patients with chronic respiratory failure secondary to COPD, obesity hypoventilation syndrome, and respiratory neuromuscular weakness.

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