Improving long-term non-invasive ventilation in a patient with Duchenne Muscular Dystrophy

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Duchenne Muscular Dystrophy is a genetic condition that causes progressive muscle weakness. Muscle weakness occurs due to a lack of the dystrophin protein, muscle fibres break down and are replaced by fibrous and or fatty tissue causing the muscle to weaken gradually. Not only are the peripheral muscles weakened but so are the respiratory muscles. As the disease progresses so does the decline in lung function and the requirement for ventilatory support. Non invasive ventilatory support increases survival in these patients [1,2], To improve adherence [3] and survival [4] it is important to ensure optimal ventilation and decrease patient ventilator asyncrony [3,4]. We report a patient with Duchenne muscular dystrophy who experienced patient ventilator asynchrony which improved with changing his ventilator.

Case

Our patient is now a 32-year male with Duchenne muscular dystrophy who was followed up in the Pediatric Pulmonary Clinic in Chicago. He was diagnosed with muscular dystrophy at the age of four and initially exhibited a wide variety of symptoms (muscle rigidity and severely compromised mobility gradually progressed and resulted in restrictive lung disease with chronic respiratory problems). He became a wheelchair user at 10 years of age. He had not received any oral steroids; however, he did receive and continues to use Albuterol and corticosteroids for wheezing. At 20 years old a gastrostomy was inserted for weight gain. His disease had progressed and now he was experiencing bulbar impairment. When he was 21 years old, (2011), he received his first BiPAP machine after having a sleep study, electrocardiogram (ECG) and pulmonary function tests (PFT). PFT showed a forced expiratory volume in one second (FEV1) of 0.58L (13% pred), forced vital capacity (FVC) of 0.66L (12% pred), maximum inspiratory pressure (MIP) 29 cmH_2O and maximum expiratory pressure (MEP) 9 cmH_2O. His ECG showed typical sinus tachycardia at 104 bpm. His main symptom was restlessness and loss of energy. His arterial blood gases (ABG's) had also deteriorated. Initially he did well on a respiratory assist device (RAD) (passive circuit with an oronasal mask) and was using it on average 10 hours per day. In 2014 at the age of 23 years old his blood gases worsened, and he complained of being short of breath during the day and despite enteral feeding he had weight loss. A subsequent sleep study was performed, and he was transferred from a RAD device to a life support non-invasive ventilator (NIV), with the option to use mouthpiece ventilation (MPV). He used a Trilogy 100 ventilator (Philips, Murrysville, USA) for 22 to 24 hours a day. However, despite having a near 24/7 ventilatory requirement, he preferred not to use the MPV mode.

Between 2014 to 2018 (4 years), his total volume was increased from 400 to 600 ml. There was not significant weight gain between 2014 and 2018 but, an increased volume was required to maintain his ABG's, and to help with his air hunger sensation. His ABG's in 2018 on a Trilogy 100 averaged a pH of 7.37 and $PaCO_2$ of 48 mmHg. In March 2019 the patient was transitioned to the Vivo 65. 4 – 6 weeks after the transition to the Vivo 65 ventilator (Breas, Mölnlycke, Sweden), the patient's blood gas values averaged a pH of 7.40 and a $PaCO_2$ of 45mmHg. Table 1 shows a direct comparison of the patient's ventilator settings and ABG's.

Table 1; Ventilatory parameters

Ventilator Settings	Trilogy 100	Vivo 65
Ventilation mode	AVAPS	PSV-TgV
Tidal volume/ target volume (ml)	600	450
Breath Rate (BPM)	20	20
IPAP min (cmH_2O)	35	25
IPAP max (cmH_2O)	45	35
PEEP/EPAP (cmH_2O)	5	5
Inspiratory trigger setting	1.0 l/m	1
Ventilator Download Report		
Treatment Period	12/2018 to 4/2019	04/2019 to 05/2019
Volume Vte (ml)	436	527
Leakage (L/min)	46.5	30
Total breath rate (bpm)	25	24
Spontaneously Triggered Breaths (%)	64.3	44
Alarms/night over one month	Circuit Disconnect Alarm 10 nights out of 30 nights	0
Blood Gases		
pH	7.37	7.40
pCO_2 (mmHg)	*48*	*45*

Discussion

The Vivo 65 delivered a greater target tidal volume at lower pressures compared to the Trilogy 100. This meant we were able to reduce the tidal volume to weight ratios from 12ml/kg to 8 ml/kg. Volume weight ratios higher than 10 ml/kg have been shown association with increased risk of pulmonary barotrauma and should be avoided [5]. High tidal volume delivery has also been associated with decreased venous return, as well as reduced cardiac output. Importantly recent studies have shown decreased mortality with the use of lower tidal volume in patients requiring invasive and NIV [6,7]. We were able to ventilate our patient with optimal tidal volumes ranging from 6 – 8 ml/kg [8].

The Trilogy 100 recorded an expiratory tidal volume (Vte) average of 436 ml for a tidal volume setting of 600 ml. One explanation for this is asynchrony caused by leak induced double triggering [9]. This is where the Vte is lower than the set volume, and a second breath delivered in rapid succession whilst the patient's breathing out [4]. Evidence of this is seen in Figure 1. In contrast to the Trilogy 100, minimal auto-triggering was not seen when the patient used the Vivo 65. One explanation for this is that on this device the trigger is independent of leaks, this nearly eliminates the leak induced auto/double-triggering. Therefore, in our patient the Vivo 65 provided improved patient synchrony as a result of no auto or double triggers in the presence of leaks.

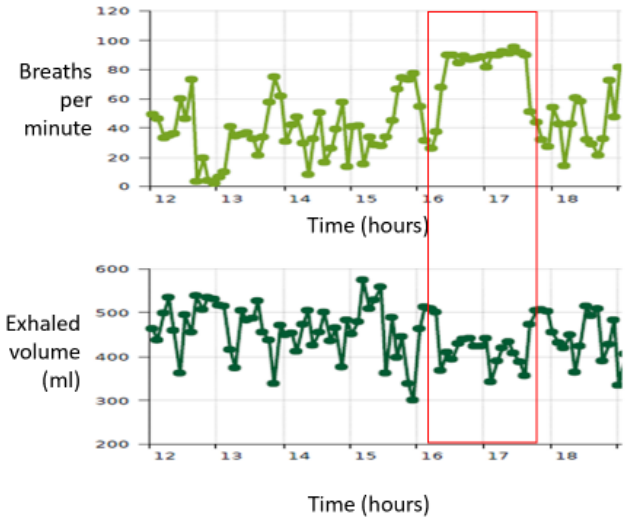

Figure 1 shows the breaths per minute and exhaled volume of the patient from his downloaded ventilator information in January 2019. He had a spontaneous respiratory rate of 64.3%. Highlighted in red where the patient triggered breath rate is 80 – 90 bpm with an associated exhaled tidal volume range of 350 to 450 ml. During these periods leak induced double triggering was noted.

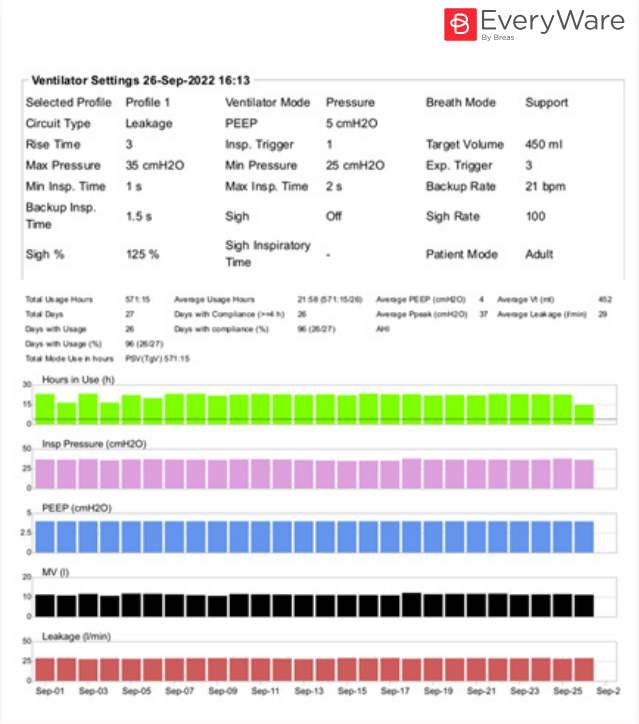

Figure 2 shows the most recent download from the Vivo 65. There is excellent adherence and stable pressures and minute ventilation (MV) with minimal leakage, the patient spontaneous respiratory rate is 44%.

Conclusion

For this patient the Vivo 65 provided superior ventilation, with less patient ventilator asynchrony. The patient had improvements in his carbon dioxide level with a lower target volume, lower pressure support and fewer patient reported alarms at night. The patient reported "increased comfort, a better night's sleep and more energy in the day".

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