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Vivo 65 – Case report

Vivo 65 improves long-term non-invasive ventilation in a 17-year old patient with Rett syndrome.

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Introduction

Rett syndrome is a rare genetic neurological disorder that predominantly affects girls. It is characterized by a slow growth of the brain causing a progressive loss of motor skills and speech. Over time children with Rett syndrome experience increasing problems with the use of muscles that control movement, coordination and communication. Uncoordinated breathing and seizures are associated with the syndrome. Problems with breathing include breath-holding, abnormally rapid breathing called hyperventilation, forceful exhalation of air and swallowing air. These problems tend to occur during waking hours, but other breathing disturbances such as shallow breathing or periodic breathing can occur during sleep. There is no cure for Rett syndrome, all treatments are directed toward relieving symptoms and providing support.

Case presentation

A 17-year-old female has been followed in the Pediatric Pulmonary Clinic. The patient was diagnosed with Rett's syndrome at age of two and initially exhibited a wide variety of symptoms including physical and mental impairments. Muscle rigidity and severely compromised mobility gradually progressed and resulted in restrictive lung disease with chronic respiratory problems. The diagnosis of chronic respiratory insufficiency was followed by non-invasive ventilation treatment for symptom relief. Non-invasive ventilation is an established treatment modality to improve ventilation by providing ventilatory support through the upper airways, i.e. by providing the patient with a volume of air through a tightly fitted facial or nasal mask.

Children diagnosed with Rett syndrome are often easily irritable and the patient had frequent crying spells where she would cry or scream for extended time periods. The crying spells made it very difficult to provide effective ventilation. The main limitation of the non-invasive ventilation was related to substantial air leaks during the crying spells, which made it difficult to set an adequate trigger for the delivery of breaths.

The patient received nocturnal, non-invasive ventilation with a full-face mask (Trilogy 100, Philips, Netherlands). The ventilator operated in Average Volume Assured Pressure Support (AVAPS). Over the last year, the ventilator settings were increased multiple times based on an elevated carbon dioxide blood level and a worsening respiratory status. For final settings see table 1 below.

Table 1. Trilogy settings	
Ventilation Mode	AVAPS
Tidal Volume	400 mL
Breath Rate	15 BPM
IPAP max	35 cmH_2O
IPAP min	25 cmH_2O
EPAP	5 cmH_2O
Inspiratory Trigger	1L/min

Despite escalation of the ventilator settings the CO_2 blood level remained elevated with an average of 62.5 mmHg (range: 56-69 mmHg) over the past year, see Figure 1 below. Due to progression of disease the patient required a more efficient non-invasive ventilation. The clinical team made the decision to transition the patient to a Vivo 65 ventilator (Breas, Mölnlycke, Sweden) with the goal to provide more comfortable and better synchronized ventilation. The following ventilator settings were prescribed, see table 2 below.

Table 2. Vivo 65 settings	
Ventilation Mode	PSV(TgV)
Target Volume	400 mL
Breath Rate	15 BPM
Pressure max	40 cmH_2O*
Pressure min	30 cmH_2O*
PEEP	5 cmH_2O
Inspiratory Trigger	3

* Including Positive End Expiratory Pressure (PEEP)

After transitioning the patient from the Trilogy 100 to the Vivo 65 ventilator, a reduction in CO_2 level was seen. The graph below depicts the changes in CO_2 levels during the recent year, see Figure 1.

"Within a few weeks this remarkable ventilator has improved the patient's quality of life."

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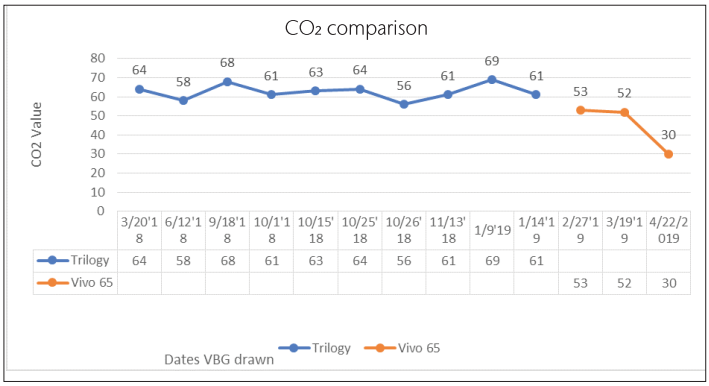

Figure 1. Comparison of CO_2 levels after treatment with Trilogy (blue) and Vivo 65 (orange)

In addition, the downloaded treatment report from the Vivo 65 indicated improvements in overall ventilation, magnitude of leaks and work of breathing when compared to the Trilogy 100 report, see table 3 and 4 below. The patient's family reported improvement in quality of life including, better airway clearance and less frequent alarms during the nights.

Table 3. Trilogy, download report	
Session	Average values*
Volume Vte (ml)	138
Leakage (L/min)	41.5
Total breath rate (BPM)	19.6
Spontaneously Triggered Breaths	69.4

* from treatment period 12/6/2018 to 01/03/2019

Table 4. Vivo 65, download report	
Session	Average values*
Volume Vte (ml)	497
Leakage (L/min)	30.7
Total breath rate (BPM)	16.1
Spontaneously Triggered Breaths (eSync)	27.1

* from treatment period 02/19/2019 to 03/14/2019

Discussion

Carbon dioxide (CO_2) is produced as a normal by-product of metabolic processes in cells and the gas normally diffuses into the bloodstream to be exhaled from the lungs. Respiratory failure occurs when the lungs cannot properly remove CO_2 from the blood. Hypercarbia, i.e. high blood level of CO_2, is a dangerous condition that if left untreated may cause damage to vital organs. It is therefore imperative to provide effective ventilation to keep CO_2 level within a normal physiological range. Normal CO_2-values are 35 to 45 mmHg or 41 to 51 mmHg when measured in arterial or venous blood, respectively.

Despite repeated efforts to improve ventilation during the last 12-month period venous CO_2 levels remained elevated. It was believed that the most significant reason for suboptimal ventilation was the inability to achieve adequate ventilator/patient synchronization secondary to substantial air leaks during frequent crying spells. An alternative mechanical ventilator with a more sensitive breath triggering mechanism was suggested.

The Vivo 65 ventilator offers a unique patented trigger technology called "eSync". Information from the flow sensor detects the start of patient effort or diaphragmatic contraction, see Figure 2. The Vivo 65 does not require a leak measurement as part of its highly responsive trigger algorithm and therefore is "leak independent". The trigger technology is designed to be sensitive enough to detect very small efforts.

Figure 2. Vivo 65. The eSync triggering algorithm calculates the patient diaphragmatic flow during inspiration using Joules of energy. The minimal trigger of the Vivo 65 is 0.2 L/sec^2 compared to 1-2 L/min for the average home ventilator.

The patient's ventilatory status was optimized by the highly responsive eSync trigger technology. Spontaneously triggered breaths decreased from an average of 69% to 27% and the back up rate setting was reduced from 20 breaths per minute (BPM) to 16 BPM. The Vivo's eSync technology detects the beginning of the patient effort and continues to deliver the precise flow and volume required to maintain the desired settings independent of leaks and without requiring the patient to excessively increase their work of breathing. It is believed that the improved patient/ventilator synchrony contributed to the reduction of CO_2 venous blood levels below normal values (i.e. 30mmHg) after 2 months.

Although the short-term outcome suggests that treatment with Vivo 65 will reduce the clinical and physiological symptoms of chronic respiratory failure in a young patient diagnosed with Rett Syndrome, continued monitoring of the changes in CO_2-levels and oxygen saturation is warranted to ensure long-term success in this difficult case. The end-tidal CO_2 monitoring and pulse oximetry functions of the Vivo 65 will facilitate the day-to-day monitoring in the home environment. With the aim to reduce the number of invasive procedures, the non-invasive end-tidal measurement may serve as a complement to the periodic arterial or venous blood sampling.

Conclusion

Overall, the patient's ventilatory status was optimized by the highly responsive eSync system of the Vivo 65 ventilator. The ventilator improved the patient's overall ventilation and airway clearance while decreasing the CO_2 blood levels, leaks, patient work of breathing and alarm activation during nighttime hours.
Within a few weeks this remarkable ventilator has improved the patient's quality of life. The patient's family is truly grateful.

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