Use of high frequency chest wall oscillation in a child with Spinal Muscular Atrophy type 1: A case report

Lisa Lucyk [a*], Maria Earley [b], Nicola McClelland [c]
[a] Advanced Paediatric Physiotherapist
[b] Senior Physiotherapist
[c] Advanced Paediatric Physiotherapist
Staffordshire Children's Hospital at Royal Stoke, Newcastle Road, Stoke on Trent ST4 6QG

**Corresponding author*: *lisa.lucyk@uhnm.nhs.uk*

ABSTRACT

Case description: Spinal muscular atrophy (SMA) is an autosomal recessive neuromuscular disorder characterised by progressive muscular atrophy and weakness (Lunn et al., 2008). There are numerous clinical types according to age of onset of clinical signs and symptoms. Infants with SMA Type 1 are at an increased risk of developing sputum retention problems. High Frequency Chest Wall Oscillation (HFCWO) is increasingly being used to treat sputum retention and atelectasis in older children and adults. This case study details its successful use in a 14-month-old infant with SMA type 1 who had persistent left sided collapse and consolidation.

Discussion: The addition of HFCWO to the patient's usual chest clearance regime of mechanical insufflation-exsufflation (MIE), manual techniques and oropharyngeal suction contributed to the resolution of acute lung collapse and promoted successful weaning of respiratory support to pre-admission levels.
Conclusion: Despite not being licensed for use in children under two years of age this case demonstrates that HFCWO was used safely and successfully with a younger child with the support of the multi-disciplinary team and parental consent.

Key Points:
- Children with SMA 1 are at increased risk of lower respiratory tract infections and atelectasis as a result of poor airway clearance.
- High frequency chest wall oscillation is an emerging adjunct to aid chest clearance.
- High frequency chest wall oscillation has been shown in this case to be safe and effective in a child under the age of 2 with SMA.

Introduction

In 2016 Nusinersen (Biogen, 225 Binney Street, Cambridge, MA 02142) became the first drug therapy to be approved for the treatment of spinal muscular atrophy, a rare genetic neuromuscular condition that causes progressive muscle weakness and loss of movement. Evidence has shown that this new therapy, which is administered intrathecally at regular time periods, has shown measurable improvements in motor (Audic, F.et al.,2020) and respiratory function (Robinson et al., 2018) in patients with SMA. Despite this, children with SMA remain at high risk of lower respiratory tract infections and atelectasis due to an impaired cough and poor airway secretion clearance (Keating et al., 2011).

At Staffordshire Children's Hospital, we manage a growing cohort of patients with SMA Type 1, who are treated with intrathecal Nusinersen and cared for by our respiratory physiotherapy service. Due to their impaired respiratory function and inability to clear secretions independently, the majority of our patients perform daily chest clearance at home but remain prone to hospital admissions due to acute respiratory deterioration.

There are a wide range of adjuncts available to aid secretion clearance and improve lung volume, each with its own merits and limitations. High Frequency Chest Wall Oscillation (HFCWO) is one such adjunct, where extra-thoracic oscillations are applied to the chest wall usually by an inflated garment wrapped around the patient's chest and attached to an air pulse generator. This generator delivers rapid and repeating pulses of positive pressure causing an oscillatory or 'squeeze and release' effect on the chest wall, thereby creating a shearing effect to detach secretions from the bronchial wall, making secretions less viscous and as a result is an effective way of treating secretion retention and atelectasis (Mantellini et al., 2012). There are a number of different devices which provide this therapy including the Hill-Rom 'The Vest' Model 105 (Clinitron House,

Ashby Park, Ashby de la Zouch, Leicester, LE65 1 JG). The use of HFCWO via the Hill-Rom Vest is currently only licensed for patients over the age of two, unless specifically supported by the treating clinician. This is because there is concern that the pressure changes may cause airway collapse in younger children who have more compliant chest walls. This may also pose a greater risk in children with neuromuscular conditions such as SMA where chest wall compliance may be further increased. These risks may be reduced by using HFCWO in conjunction with non-invasive positive pressure ventilation. We report the use of HFCWO to treat secretion retention and collapse in a 14-month-old child with SMA.

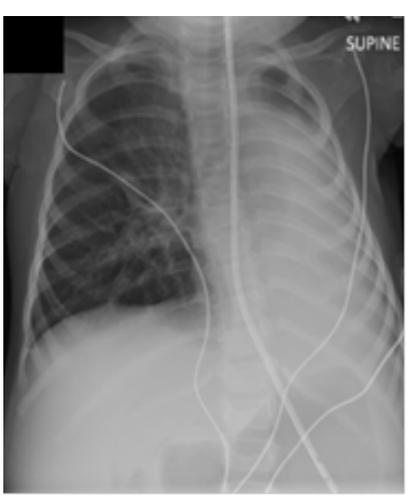
Case Presentation

A 14-month-old girl with SMA type 1 was admitted to our institution for elective administration of intrathecal Nusinersen. This is usually a day case procedure performed with adequate pain relief tailored to the individual patient (in this case Ketamine was used) and patients are typically discharged home later that day after a period of observation and rest. Her usual home management consisted of nocturnal Bilevel Positive Airway Pressure (BiPAP) (pressures of 18/6) with a physiotherapy chest clearance routine of nebulised 0.9% saline, mechanical insufflation-exsufflation (MIE) 3 times per day (settings +30/-35 ratio 1:1 with 5 breaths per cycle) followed by oropharyngeal suction. The patient had no supplementary oxygen requirement when well, had nasogastric feeds and was under investigation for Gastro-Oesophageal Reflux Disease (GORD). She had a history of recurrent Paediatric Intensive Care Unit (PICU) admissions for acute respiratory deterioration.

On admission to the ward, she was noted to have an increased work of breathing, increased oxygen requirement and pyrexia. The patient was initially managed with her usual chest clearance routine with the addition of manual chest clearance techniques (expiratory chest wall vibrations and percussion) but after 6 days with no improvement and difficulty in clearing thick secretions she was started on HFCWO using a Hill-Rom 'The Vest', Model 105 to try and increase secretion yield.

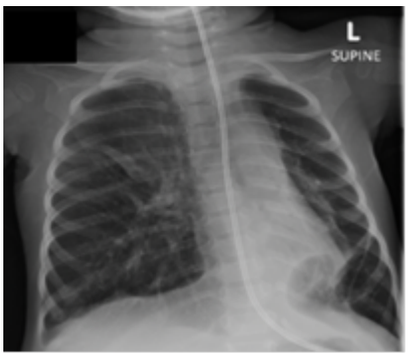
Treatment and Clinical Course

Admission (Day 1): The patient's normal chest clearance routine was used, with the addition of manual expiratory chest wall vibrations during MIE due to increased viscosity of secretions. Patient requiring 0.5-2L/min O2 to maintain saturations.
Day 2: Patient requiring continuous BiPAP (pressures of 18/6) with 0.5-2L/min 02, patient's normal chest clearance routine continued.
Day 3: Chest x-ray (CXR) demonstrated a complete left lung collapse (see CXR). Her normal physiotherapy routine was continued with the addition of percussion in right side lying, she was started on budesonide with a plan to keep her on BiPAP for 24hrs with settings increased to 20/8.



Day 4: Left sided collapse remained on CXR. Inspiratory breaths using pressures of +30 via MIE added to existing physiotherapy in right side lying at end of treatment. Requiring 2L/min O_2 throughout the day via BiPAP and having regular desaturations during physiotherapy treatment especially when starting to tire.
Day 5: CXR; almost complete left lung collapse, no clinical change since starting steroid treatment on day 3. Remained on BiPAP 20/8 with 2L/min O2, tolerating short periods off BiPAP on 2L/min O2 via nasal cannula.
Day 6: Patient started on high flow nasal cannula (HFNC) via Optiflow (Fisher & Paykel Healthcare Ltd, Unit 16, Cordwallis Park, Clivemont Road, Maidenhead, Berkshire SL6 7BU), 20L/min, FiO_2 0.35 as not tolerating BiPAP when awake (remained on nocturnal Bipap with previously increased settings), carbocisteine started. HFCWO commenced 3 times per day alongside physiotherapy routine of MIE and expiratory vibrations. The HFCWO was set at 14Hz and a pressure of 1 with a treatment time of 10 minutes and used to assist mobilisation of secretions prior to MIE and expiratory vibrations. (Note it is recommended that HFCWO is used in conjunction with positive pressure ventilation in this age group. However, in this case the patient was unable to tolerate her BiPAP when awake therefore the decision was made to treat on optiflow and a low pressure of 1 used on The Vest).
Day 7: Treatment continued as per day 6.
Day 8: FiO_2 via Optiflow weaned from 0.35 to 0.26. Secretion yield noted to be increased and patient requiring fewer cycles of MIE and suction to clear.
Day 9 & 10: Patient continued with 3 times daily physiotherapy treatment of HFCWO followed by MIE and expiratory vibrations.

Day 11: CXR showed partial improvement of left middle zone collapse, with left basal collapse thought to be chronic (see CXR fig 2.).
Day 12: FiO_2 via Optiflow weaned to 0.21.
Day 13: Flow weaned from 20L/min to 10L/min and nocturnal BiPAP reduced back to previous home settings 18/6. HFCWO discontinued and the patient's usual home physiotherapy routine was recommenced.



Discussion

New therapeutic options and an increased life expectancy for patients with SMA1 mean that we are seeing an increase in the hospital treatment of respiratory complications in these patients (Ali et al., 2019, Chen et al., 2020 Keating et al., 2011). Optimising secretion clearance is critical in this patient group, and it has been suggested that airway clearance should not be limited to conventional physiotherapy and use of MI-E alone (Keating et al., 2011).

HFCWO is not used in isolation, and in this case, it was used alongside MI-E, chest wall vibrations, suction and carbocisteine, we must therefore be cautious in attributing the clinical improvements to HFCWO alone. However, examination of the patient's progress reveals that there were few clinical improvements seen until the use of HFCWO and carbocisteine was commenced. Prior to starting HFCWO the patient had been having frequent desaturations during physiotherapy sessions, these were particularly evident as the patient started to tire following repeated cycles of MIE and suction. Following treatment with HFCWO secretion yield was noted to be greater with the patient needing fewer cycles of MIE and suction to clear their chest and a reduction in desaturations during treatment. After commencing HFCWO, FiO_2 could also be safely weaned, and improvements on x-ray became evident.

Although the initial cost of a HFCWO device is high (£8,394), a recent study has demonstrated that the use of HFCWO in adult neuromuscular disease patients with respiratory complications can decrease health care costs and hospital admissions in the long term (Lechtzin et al., 2016). The use of HFCWO in managing neuromuscular conditions such as SMA is also praised in the community as it provides a more standardised treatment than manual percussion for secretion management (NICE, 2018).

As previously highlighted, using HFCWO in isolation would have put the patient at risk of airway collapse due to having increased chest wall compliance. Additional positive pressure was therefore utilised to reduce this risk. The patient was too distressed and unable to synchronise with her own BiPAP using HFCWO and therefore completed her treatment on high flow nasal cannula oxygen and a low pressure of 1.

The use of HFCWO via the Hill Rom Vest is not licenced for children under the age of 2 (Hill-Rom, 2020). However, we have demonstrated its safe and effective use in a child of 14 months with the support of her respiratory consultant.

Conclusion

Safe and effective use of the High Frequency Chest Wall Oscillation device in a child under 2 years of age, has been demonstrated in this case when used in conjunction with conventional physiotherapy techniques to treat lung collapse. In this example, the addition of HFCWO and carbocysteine aided weaning of oxygen from high flow back to room air and a reduction of BiPAP settings from 20/8 to her baseline 18/6.

It is important that use of the HFCWO device in children under the age of 2 is a multi-disciplinary decision and used with the support of the child's treating consultant. When choosing to use this device, consideration must be given to the risk of loss of lung volume due to increased compliance of the chest wall in younger children, or patients with conditions such as SMA which can result in even more compliant chest walls. This is why it is recommended that whenever possible young children are placed on positive pressure ventilation when using HFCWO devices. These risks need to be weighed up against the potential benefits of improved secretion clearance especially when secretions are tenacious and requiring intensive physiotherapy sessions to clear, especially in patient groups such as SMA where patients can fatigue quickly.

Further assessment and research into the use of HFCWO in children less than 2 with SMA1 is therefore recommended to aid the future management of this condition.

Funding
This research received no specific grant from any funding agency in the public, commercial or not-for-profit sectors

Patient consent:
Written consent for patient's details and images for publication was obtained from her parent.

Acknowledgements
Dr Martin Samuels, Consultant Paediatric respiratory service, Staffordshire Children's Hospital at Royal Stoke

References

Ali I, Gilchrist F, Carroll W, Alexander J, Clayton S, Kulshrestha R, Willis T, Samuels M. Healthcare utilisation in children with SMA type 1 treated with nusinursen: a single centre retrospective review. BMJ Paediatrics Open 2019

Chen KA, Widger J, Teng A, Fitzgerald D, D'Silva A, Farrar M. (2020). Real-world respiratory and bulbar comorbidities of SMA type 1 children treated with nusinursen: 2-year single centre Australian experience. Paediatric Respiratory Reviews 2020 September

Hill-rom. (2020). *The vest System Model 105.* Available: https://www.hillrom.com/en/products/the-vest-system-105/. Last accessed 29th Jul 2020.

Keating, M., Collins, N., Bush, A. & Chatwin, M. (2011). High-Frequency Chest-Wall Oscillation in a Noninvasive-Ventilation-Dependent Patient With Type 1 Spinal Muscular Atrophy. *Respiratory Care* . 56 (11), p1840-1843.

Lechtzin, N., Wolfe, LF. and Frick, KD. (2016). The Impact of High-Frequency Chest Wall Oscillation on Healthcare Use in Patients with Neuromuscular Diseases. *AnnalsATS*. 13 (6), p904-909.

Lunn MR, Wang CH.(2008) Spinal muscular atrophy. Lancet 2008; 371:2120-2133.

Mantellini, E., Perrero, L., Petrozzino, S. , Gatta, A. & Bona, S.. (2012). Clinical Implications High Frequency Chest Wall Oscillation. *Working Paper of Public Health*. 10

NICE (2018). The Vest for delivering high-frequency chest wall oscillation in people with complex neurological needs.

Robinson V, Edel L, Grime C, Chan E and G (2018). A novel scoring system for the respiratory health of children with spinal muscular atrophy type 1. Archives of Disease in Childhood 2018;103:A25-A26.