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CASE REPORT

Clinical Effectiveness of Mechanical Insufflation Exsufflation (MI:E) and Chest Physiotherapy Protocol for Airway Clearance in Children with Neuromuscular Diseases: A Case Study Report

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ABSTRACT

Children with Neuro Muscular Diseases (NMD) are hospitalised with respiratory distress; ineffective airway clearance mechanism, poor functional cough effort demands the need of Mechanical Insufflation Exsufflation (MI:E) therapy along with chest physiotherapy to clear secretions over deeper lung zones. This case study describes the effectiveness of MI:E therapy with chest physiotherapy on a 5 years old child with SMA type 2. Therapy frequency were 4th hourly in PICU and 6th hourly in ward based on severity of respiratory distress, throughout the hospitalisation for 10 days. MI:E therapy can be used as an effective airway clearance modality along with chest physiotherapy to reduce the respiratory distress and improve better clinical outcomes to decrease the duration of shifting out transitions from intensive care unit to ward thereby prepare plan for early hospital discharge.

Keywords: Neuromuscular diseases; Spinal muscular atrophy; Mechanical Insufflation Exsufflation therapy; Chest Physiotherapy; Respiratory distress in children

1 INTRODUCTION

Children with neuromuscular diseases are group of relatively common diagnosis that includes SMA, DMD, ALS, congenital myopathy with a prevalence of about 1 in 3000 globally[1] and 26% of the respiratory impairment in India[2].

Spinal Muscular Atrophy (SMA) is an autosomal recessive neurodegenerative disorder characterized by progressive muscle wasting due to motor neuron degeneration, secondary to mutations in the survival motor neuron 1 (SMN1) gene[3].Muscle weakness and poor muscle coordination can affect any part of the upper airway and cause breathing problems such as reduced lung function, hypoventilation, and weak cough, difficulty clearing mucus from the airways, upper airway obstruction and sleep apnoea[4].

Respiratory impairment is predominant with variable severity of chest wall distortion, impaired airway clearance and cough that worsens later in childhood[5]. Common respiratory consequences such as hypoventilation, upper airway obstruction, aspiration lung disease, secretion retention, lower airway infection finally to respiratory failure in children with neuromuscular disease[6].

Secretion retention is the major burden of lung disease due to a weak cough typically noted in children with neuromuscular disease. Airway clearance can eliminate secretions to improve survival and prevent unexpected hospital admissions[7].

Commonly practiced chest physiotherapy techniques such as Percussions, vibrations, positioning, adapted pos-

tural drainage, breathing exercises and manually assisted cough were effective airway clearance techniques. Traditional chest physiotherapy techniques are clinically tiring procedure for children and also precipitate episodic oxygen desaturation in children with SMA that demands lesser therapy duration [8].

Children with neuromuscular disease have a weak cough that limits their ability to get retained secretions; airway clearance therapies such as a cough assist device can help to maintain bronchial hygiene [1].

Mechanical Insufflation-Exsufflation device is a non-invasive procedure and effective airway clearance modality to simulate physiological cough. Positive air pressure (insufflation) is delivered to obtain a large volume of air within the lungs; quickly reversing the flow of air by shifting to negative air pressure (exsufflation) [9]. High expiratory flow "Asymmetric pressure values" helps mobilize retained secretions from deeper lung zones [10].

Visual Analogue Scale (VAS) determines user rated comfort during MI:E therapy with cut-off points were 0–3 not/slightly, 4–6 moderately and 7–10 very uncomfortable. (B.hov et al 2024) "Asymmetric pressure" values i.e. exsufflation pressure (Pe) greater than insufflation pressure (Pi) has better airway clearance efficiency in children with NMD but the effect of airway clearance using MI:E therapy with chest physiotherapy throughout hospitalisation course during respiratory distress were not reported [11]. This study intends to find out the clinical effectiveness of MI:E therapy as an effective airway clearance on respiratory distress during hospital stay in children with NMD.

2 CASE REPORT

This 5 years old child diagnosed to have SMA – type 2 by gene testing with birth history of triplet pregnancy delivered at 31 weeks of gestation by LSCS admitted at NICU for 16 days with initial diagnosis of Respiratory Distress Syndrome (RDS), child was on disease modifying therapy SPINRAZA at 4 years of age.

In Emergency ICU

Admitted with high grade fever with RDS with SPO_2 lesser than 80 % for further evaluation the child was shifted to Paediatric ICU.

In PICU

Admitted for aspiration risk evaluation, during 1st the child was on BiPAP (18/7) support with SPO_2 80 – 85 % radiograph findings were few air space opacities in both upper zone infective, no cardiomegaly, pulmonary vasculature is normal, B/L lower lobe consolidation. On the 2nd day of PICU the child developed apnoea with bradycardia needing bag and mask ventilation, serial arterial blood gas monitoring suggested respiratory failure; sleep study was abnormal with multiple hypopus and obstructive aponic episodes. Prolonged requirement of respiratory support and life-threatening events on the 3rd day of PICU stay tracheostomy and PEG tube intervention was done, post this procedure the child was on mechanical ventilation; hemodynamically stabilised. On the 5th day referred for intensive chest physiotherapy and Mechanical Insufflation Exsufflation therapy; assistive scoring tool was evaluated to decide every 4th hourly frequency of MI:E therapy.

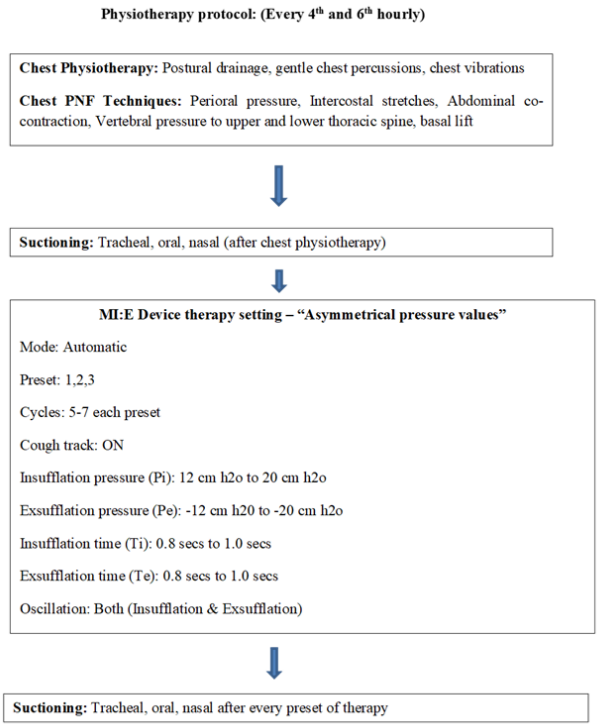

Fig. 1: Physiotherapy Protocol

The above mentioned therapy protocol was given every 4 hours once for another three days; Nebulisation was given before and after therapy. Visual analogue score was obtained with the parent support after every therapy session to evaluate the treatment comfortableness. Physiological parameters such as heart rate, respiratory rate, saturation was documented before and after the therapy. On the 8th day of PICU stay the child showed clinical improvements with reduction in the respiratory distress status, physiological parameters were within normal limits, chest radiograph findings were clear CP angles, domes of diaphragm are in normal position, tracheostomy tube is in place, as compared to the previous radiograph the lower lobe collapse has resolved with an opacity in the region of the left upper lobe, possible ectasis. Child was weaned off from the mechanical ventilation and shifted to ward with BiPAP support (15/7) continuously, chest physiotherapy with MI:E therapy was continued 6th hourly based on assistive scoring tool.

In WARD

During 9th,10th and 11th day of the ward stay parents were educated and trained for PEG tube feeding, BiPAP device handling, suctioning (oral, tracheal, nasal) basic chest physiotherapy techniques such as postural drainage, gentle chest percussions and MI:E therapy under supervision; visual analogue scores were obtained after every therapy. During this course child was clinically stable with respiratory distress resolved completely, chest radiograph findings were-CP angles are clear, B/L lower lobe collapse completely resolved, physiological parameters are within normal ranges. The child was discharged from the hospital to continue BiPAP support with chest physiotherapy, MI:E therapy and advised for Outpatient department follow-up after a week.

Table 1: Visual Analogue Scale (VAS) scores

Pre (PICU)	Post 1 (Ward)	Post 2 (Discharge)
3	1	1

Table 2: Physiological parameters

Vitals	Pre (PICU)	Post 1 (Ward)	Post 2 (Discharge)
Heart rate	102 beats/min	96 beats/min	86 beats/min
SPO_2	86%	99%	99%
Respiratory rate	36 breaths/min	20 breaths/min	18 breaths/min
Blood pressure	115/81 mmhg	114/88 mmhg	103/78 mmhg

3 DISCUSSION

Spinal muscular atrophy is the most common sequalae in children with neuromuscular diseases that worsens respiratory functioning remarkably. Quality of life is at most important factor of consideration by intervening early primary and secondary complications that may prolong the disease state. Poor functional and productive cough is compromised in children with neuromuscular diseases which results in retention of secretions over the deeper lung zones. Children with SMA type 2 are hospitalised with the respiratory distress and risk of aspiration pneumonia, this frames a major life-threatening reason for medical evaluation and management. Peak cough flow is the clinical outcome to decide the severity risk of aspiration pneumonia chances; children with NMD averages critical value lesser than 180 L/min. Medical management should strongly consider PEG tube insertion, effective airway clearance strategies to prevent further respiratory infections in children with NMD. Cough augmentation is crucial for poor and non-functional cough; children with NMD has difficulties in coughing effectively as their respiratory muscle strength are very weak and retention of secretions over the deeper lung zones this demands the need of MI:E therapy. MI:E therapy is the effective airway clearance modality that mobilises secretions from peripheral airways to the central airways to prevent aspiration and lung infections. Miguel et al 2012 concluded that inclusion of MI-E therapy reduces reintubation rates with consequent reduction in postextubation ICU length of stay also efficient in improving the efficacy of non-invasive ventilation. Similarly in this case report intensive chest physiotherapy combined with M:IE therapy had significant changes in maintaining bronchial hygiene thereby reducing the respiratory distress gradually from PICU admission day 5 to day 8 to wean off from ventilation support. MI:E therapy along with BiPAP support was clinically effective for this child to maintain hemodynamic stability and physiological parameters within normal limits to facilitate early discharge from the hospital that was anticipated to be prolonged (Table 2). Involvement of parents during the MI:E therapy was supportive to reduce the apprehension of the child and also to gain better clinical outcome. Visual analogue scores were ranging 1-2 "very slightly uncomfortable" when MI:E therapy was given by parents during hospital stay (Table 1) ensured that MI:E therapy is very comfortable to applied as a part of home care settings.

Strengths and limitations

Strength

Single case study followed up the effect of MI:E therapy and chest physiotherapy protocol throughout the hospital stay during respiratory distress along with multidisciplinary team that includes Paediatric emergency medicine team, PICU Intensivist team, Paediatric Pulmonologist, Paediatric neurologist, Paediatric ENT team, Paediatric surgery team Paediatric nurses and Paediatric physiotherapist.

Limitations

Assistive scoring tool was used to determine the frequency of MI:E therapy, scores where suggesting every 4th hourly and 6th hourly MI:E therapy during PICU and ward stay; after the 4th visit at night therapy was not given as the child's sleep pattern was disturbed and parents felt the child was not cooperative for the treatment.

4 CONCLUSION

In Children with Neuro Muscular diseases, chest physiotherapy with MI:E therapy have an excellent clinical effectiveness to reduce respiratory distress; also to decrease the duration of shifting out transitions from intensive care unit to ward thereby prepare plan for early hospital discharge.

Future scope and directions

Only one SMA type 2 was reported in this case report, future studies should be done on other diagnosis in children with Neuro Muscular diseases to find out better perspective of clinical outcomes on this chest physiotherapy MI:E therapy protocol.

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REFERENCES

1. American journal of respiratory and critical care medicine;vol. 195. 2017. Available from: https://www.atsjournals.org/loi/ajrccm?expanded=d2010&expanded=d2010.y2017&expanded=d2010.y2017.v195.
2. Mahesan A, Kamila G, Gulati S. Rare paediatric disorders in Indian healthcare settings with focus on neuromuscular disorders: Diagnostic and management challenges. *Journal of Biosciences*. 2024;49(1):15. Available from: https://dx.doi.org/10.1007/s12038-023-00403-w.
3. Prior TW. Neuromuscular Diseases. In: Molecular Pathology in Clinical Practice. Springer, New York, NY. 2007;p. 87–96. Available from: https://doi.org/10.1007/978-0-387-33227-7_7.
4. Voulgaris A, Antoniadou M, Agrafiotis M, Steiropoulos P. Respiratory Involvement in Patients with Neuromuscular Diseases: A Narrative Review. *Pulmonary Medicine*. 2019;2019:2734054. Available from: https://doi.org/10.1155/2019/2734054.
5. LoMauro A, Aliverti A, Mastella C, Arnoldi MT, Banfi P, Baranello G. Spontaneous Breathing Pattern as Respiratory Functional Outcome in Children with Spinal Muscular Atrophy (SMA). *PLOS ONE*. 2016;11(11):e0165818. Available from: https://dx.doi.org/10.1371/journal.pone.0165818.
6. Hull J, Aniapravan R, Chan E, et al. British Thoracic Society guideline for respiratory management of children with neuromuscular weakness. *Thorax*. 2012;67(Suppl 1):1–40. Available from: https://doi.org/10.1136/thoraxjnl-2012-201964.
7. Kinimi I, Sahoo A, Shinde SS, et al. Clinical Profile and Outcome of Children Using Mechanical Insufflation Exsufflation Device (Cough Assist) at a Tertiary Care Hospital in Bangalore, India. *Journal of Pediatric Pulmonology*. 2024;3(1):11–15. Available from: https://journals.lww.com/jpp/fulltext/2024/01000/clinical_profile_and_outcome_of_children_using.4.aspx.
8. Siriwat R, Deerojanawong J, Sritippayawan S, Hantragool S, Cheanprapai P. Mechanical Insufflation-Exsufflation Versus Conventional Chest Physiotherapy in Children With Cerebral Palsy. *Respiratory Care*. 2018;63(2):187–193. Available from: https://doi.org/10.4187/respcare.05663.
9. Chatwin M, Simonds AK. Long-Term Mechanical Insufflation-Exsufflation Cough Assistance in Neuromuscular Disease: Patterns of Use and Lessons for Application. *Respiratory Care*. 2020;65(2):135–143. Available from: https://dx.doi.org/10.4187/respcare.06882.
10. Lacombe M, Castrillo LDA, Boré A, Chapeau D, Horvat E, et al. Comparison of three cough-augmentation techniques in neuromuscular patients: mechanical insufflation combined with manually assisted cough, insufflation-exsufflation alone and insufflation-exsufflation combined with manually assisted cough. *Respiration*. 2014;88(3):215–222. Available from: https://doi.org/10.1159/000364911.
11. Chatwin M, Wakeman RH. Mechanical Insufflation-Exsufflation: Considerations for Improving Clinical Practice. *Journal of Clinical Medicine*. 2023;12(7):2626. Available from: https://dx.doi.org/10.3390/jcm12072626.