



## CASE REPORT

# PEP mask therapy for the rehabilitation of a pre-term infant with respiratory distress syndrome: a case report

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### ABSTRACT

**BACKGROUND:** Preterm infants can develop many complications related to organs underdevelopment. Respiratory distress syndrome (RDS) is considered the most important cause of morbidity and mortality in these patients. Traditional therapies for severe RDS, such as mechanical ventilation, come with a potential risk for pneumothorax and bronchopulmonary dysplasia while evidence on chest physiotherapy in preterm infants are controversial in terms of feasibility, tolerability and safety. The use of the positive expiratory pressure (PEP) mask is known in the pediatric field especially in cystic fibrosis for the removal of secretions and lung re-expansion. However, no literature exists on the application and effectiveness of this treatment modality for the respiratory rehabilitation of preterm infants. In this study, we aimed to assess the efficacy of a respiratory rehabilitation protocol based on PEP mask in a preterm infant with respiratory distress syndrome.

**CASE REPORT:** A Caucasian girl born at 26 + 5 weeks of gestational age with respiratory distress syndrome was treated with mechanical ventilation, oxygen therapy and PEP-mask.

**CLINICAL REHABILITATION IMPACT:** Three weeks of PEP mask led to a significant clinical and radiological improvement of the lung's function with progressive reduction of the oxygen supplement and mechanical ventilation until complete weaning off. Given the absence of literature on this subject, further studies should be conducted to confirm these preliminary observations.

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**KEY WORDS:** Infant, premature; Thorax; Physical therapy modalities; Positive-pressure respiration; Masks.

Modern neonatal care techniques in neonatal intensive care units (NICU) have enhanced the chances of survival of a growing number of preterm newborns with multiorgan pathologies due to neonatal underdevelopment.

Preterm newborns represent approximately 11% of the births worldwide and are classified according to their gestational age as mild preterm (32-36 weeks), very preterm (28-31 weeks) and extremely preterm (<28 weeks).<sup>1</sup>

Respiratory distress syndrome (RDS) is the single most important cause of morbidity and mortality in this popula-

tion.<sup>2</sup> Reduced production of surfactant in immature lungs alters the respiratory dynamics causing poor alveolar compliance and increased breathing's work that leads to rapid hypoxemia, hypercapnia and death if corrective measures are not applied.<sup>2</sup>

The standard of care for infants with respiratory failure may range from oxygen administration and continuous positive airway pressure (CPAP) to invasive procedures like mechanical ventilation and surfactant administration that increase the risk of pneumothorax and bronchopulmo-

nary dysplasia (BPD).<sup>2</sup> Besides this, chest physiotherapy including gravity-assisted drainage postures, chest percussion and vibration are sometimes added to promote physical recovery.<sup>3</sup> Chest wall percussion and vibration create changes in the intrapleural pressure that assist in dislodging secretions. Drainage postures involve placing the patient in specific positions (including semi-recumbent) which enables gravity to drain excess secretions from bronchopulmonary segments.<sup>4</sup>

While for compliant adults many chest physiotherapy techniques are applicable, for the pediatric population the choice much depends on the patient's age, clinical status, and factors that may influence the treatment adherence.<sup>4</sup> Moreover, the type of respiratory physiotherapy must be carefully selected to minimize the risk of adverse events or aggravation of comorbidities. Given the paucity of research in the pediatric population, the principles of airway rehabilitation mainly follow those applied for adults with chronic obstructive pulmonary disease (COPD) and children with cystic fibrosis (CF).<sup>4</sup> In premature infants, however, literature evidence on chest physiotherapy techniques appears controversial in terms of tolerability, safety, and clinical outcomes.<sup>3</sup>

Positive expiratory pressure (PEP) and in particular PEP mask therapy is an alternative modality widely used in children with CF that may represent a safe alternative for preterm newborns. PEP mask therapy uses a one-way valve that allows unrestricted inspiration and a resistance to expiration.<sup>4</sup> In the presence of small airways obstruction, the positive pressure increases the functional residual capacity and prevents premature airway collapse during expiration.<sup>4</sup> Moreover, the positive pressure creates a pressure gradient behind secretions that forces them toward larger airways.<sup>4</sup>

Previous studies have highlighted a good clinical efficacy and feasibility of this treatment. Long-term use of PEP mask was associated with improved gas exchange in children with multiple severe disabilities<sup>5</sup> and reduced exacerbation rate in children with CF, HIV and cerebral palsy (CP).<sup>6-8</sup>

Another positive aspect is that PEP therapy does not require the patient's cooperation<sup>4, 5</sup> and can be easily performed home by the caregiver.

Despite the lack of specific contraindications, there are no studies in the literature on the application of PEP mask in preterm infants.

Therefore, here we report the case of a premature infant affected by RDS who underwent respiratory rehabilitation with the use of PEP mask during her hospitalization in the neonatal intensive care unit (NICU).

## Case report

A Caucasian female child (M.M.) was born at 26 + 5 weeks of gestational age from a G3P3 woman. The pregnancy was uneventful until the 19<sup>th</sup> week of gestation when a preterm premature rupture of the membranes (PPROM) occurred. The woman underwent three amnioinfusions and adequate steroid prophylaxis. The delivery was uncomplicated although the baby presented in breech position. Birth weight was 915 g, head circumference 24.7 cm and length 35.5 cm. Right after delivery, the child developed generalized cyanosis and cardiorespiratory depression that required intubation and supplemental oxygen. Apgar Score was 3 after 1 min, 6 after 5 min and 7 after 10 min.

The infant spent several months in the Neonatal Intensive Care Unit (NICU), during which she developed respiratory distress syndrome (RDS), pulmonary hypertension, left pneumothorax and multiple cases of pneumonia.

During hospitalization in the NICU, the newborn underwent a total of 112 days of oxygen therapy: 9 days of high-frequency mechanical ventilation, 26 days of conventional mechanical ventilation, 26 days of inhaled nitric oxide therapy and 23 days of nasal continuous positive pressure with high flow nasal cannulas (HFNC). Then, she was transferred to the neonatal subintensive care unit where she still needed respiratory assistance and oxygen supplement for another 28 days.

To improve her post-ICU physical condition, the newborn underwent a respiratory rehabilitation protocol with PEP mask as usual care.

Before starting the physiotherapy, the baby was first examined by a multidisciplinary team that included a NICU pediatrician, a physiatrist and a physiotherapist. A specific respiratory training protocol with PEP mask was chosen according to the criteria described in the literature,<sup>5, 6</sup> our clinical experience and the characteristics of the newborn. The child's parents were informed about the procedure and signed informed consent.

The physical examination included an evaluation of the posture, respiratory pattern, skin tone, chest shape and mobility, signs of respiratory distress, heart rate, respiratory rate, saturation, temperature, muscle tone and abdomen conformation. The spontaneous behavior of the newborn including the response to environmental stimuli was assessed with the Assessment of Preterm Infant Behaviour Scale (APIB).<sup>9</sup> Finally, the Silverman Andersen Scale was applied for the evaluation of the respiratory distress.<sup>10</sup>

### Respiratory rehabilitation protocol

Starting from 37 + 4 weeks of gestational age, M.M. underwent PEP Mask (PEP/Rmt, Astra Tech, Mölndal, Sweden) treatment twice a day, 5 days a week for 3 weeks, for a total of 30 treatments. Each session included 6-8 repetitions of 30 seconds each, with 1 minute of rest in-between.

The PEP mask was applied by a physiotherapist specialized in developmental neurorehabilitation and preterm rehabilitation. To counteract the thrust on the face generated by the operator, the nape of the child was placed against the therapist's body and the child was wrapped as shown in Figure 1.

The therapy was applied between meals (1 hour before or two hours after the meal) while the baby was awake. A pressure gauge measuring the pressure generated by the newborn's exhalation against resistance verified the effectiveness of the procedure with a reference value set between 10 and 20 mmHg. Heart rate, respiratory rate and oxygen saturation were monitored throughout the procedure and the treatment was temporarily interrupted in case of significant variation of the vital signs or crying.

To assess the effects of the PEP mask on the infant's pulmonary function a clinical and radiological evaluation was performed at different points in time. The volume of oxygen needed (L/min and % FiO<sub>2</sub>) was assessed at baseline (T0) and after one (T1), two (T2) and 3 weeks (T3) of treatment. A chest X-ray was also performed at baseline and after 3 weeks of treatment (T3). Finally, the Silverman Andersen scale was applied at baseline and at discharge for the evaluation of the level of respiratory distress.

Before starting the rehabilitation protocol, the newborn was under 7 L/min (32% FiO<sub>2</sub>) of oxygen supplement with HFNC. The Silvermann Andersen Score was 2, with lower and upper chest retraction, and the ABIP Score was 3 which was indicative of a well-modulated and organized behavioral regulation.



Figure 1.—Application of the PEP-mask in a preterm infant.

TABLE I.—Newborn oxygen requirement during the PEP-mask rehabilitation protocol.

	FiO <sub>2</sub>	L/min
T0 - baseline	32%	7 L/min
T1 (day 7 <sup>th</sup> )	28%	7 L/min
T2 (day 14 <sup>th</sup> )	28%	6 L/min
T3 (day 21 <sup>st</sup> )	24%	6 L/min
T4 (day 28 <sup>th</sup> )	24%	5 L/min, at 3h intervals
Day 29 <sup>th</sup>	Complete weaning from oxygen therapy	

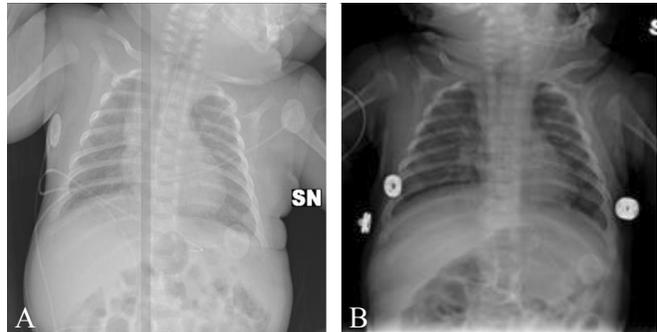


Figure 2.—Chest X-ray showing the lung interstitial involvement before (A) and after 3 weeks (B) of treatment with PEP-mask.

Throughout the treatment protocol, a progressive recovery of the lungs function was observed and complete weaning from respiratory support was possible after 29 days of sub-intensive care (Table I).

A progressive improvement of the clinical picture was also demonstrated on radiological assessment. After 3 weeks of treatment with PEP-mask a net improvement of the bilateral interstitial lung involvement was noticed (Figure 2). Moreover, the Silverman Anderson score at discharge was 0.

### Discussion

Respiratory distress syndrome represents a significant problem for preterm infants with immature lungs. Respiratory rehabilitation in this field mainly focuses on chest physiotherapy to reduce bronchial obstruction, atelectasis and improve lung ventilation.<sup>3</sup> Although many techniques exist, a single preferred method has not yet been identified. Moreover, strong doubts remain about the safety and tolerability of this practice especially for very low birth weight infants (VLBW), due to the risk of brain damage.<sup>3</sup> For this reason, lung physiotherapy with PEP-mask represents an innovation in the rehabilitation of preterm infants with respiratory distress syndrome. Several studies have discussed the effect of this technique on the pulmonary outcomes of

patients with cystic fibrosis, HIV, CP, and obstructive lung diseases.<sup>6-8</sup> To the best of our knowledge, this is the first study on the use of PEP mask in preterm infants.

In this case report, we assessed the effect of three weeks of PEP-mask on the lung's function of a preterm infant affected by respiratory distress syndrome.

Our results showed that regular application of positive expiratory pressure led to a significant improvement of the newborn's respiratory function with a progressive reduction of the ventilatory support and oxygen need. These results are in line with the study of Lagerkvist *et al.* which showed a significant improvement of transcutaneous oxygen tension (tcPO<sub>2</sub>) immediately after PEP treatment in children with multiple severe disabilities. The author also reports a good reproducibility of the treatment among several patients.<sup>5</sup>

In our case, PEP therapy was also efficacious in promoting airway clearance and atelectasis reversal as observed by the chest X-ray performed before and after three weeks of treatment and by the Silverman scores at baseline and at discharge.

This observation is in line with the study of Plebani *et al.* which showed a significant improvement in lung function tests (FEV<sub>1</sub>, FVC) and a lower rate of lower respiratory tract infections in a group of HIV children treated with PEP mask compared to a subgroup of similar patients who did not undergo PEP therapy.<sup>7</sup> A reduction in the number of respiratory infections was also observed in a retrospective study on CP patients by Di Pede *et al.*<sup>8</sup>

In terms of safety and tolerability, the child did not experience any side effect throughout the procedure and the device was well tolerated. This observation is in line with the results of Lagerkvist,<sup>5</sup> Plebani<sup>7</sup> and Di pede<sup>8</sup> which showed that compared to traditional chest physiotherapy, PEP therapy demonstrated a greater safety and tolerability in children with multiple severe disabilities, CF and CP respectively.

## Conclusions

PEP-mask therapy in the rehabilitation of preterm infants suffering from respiratory distress syndrome represents a new therapeutic opportunity. In our case, PEP therapy determined a progressive improvement of the respiratory function with good results in terms of feasibility, safety

and tolerability. Given the paucity of literature on this subject, further studies should be conducted to confirm these preliminary observations. Moreover, optimization of the treatment protocol according to the developmental stage, pulmonary symptoms and lung function of the newborn should be attempted. This study could open new scenarios in the management of premature newborn as well as other fragile uncooperative patients for which chest manipulation or drainage posture may result impossible or overly risky. Improved management of RDS in premature children may then lead to reduction of days spent in the hospital, better life expectancy and cost reduction for the health system.

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### Conflicts of interest

The authors certify that there is no conflict of interest with any financial organization regarding the material discussed in the manuscript.

### Authors' contributions

All authors read and approved the final version of the manuscript.

### History

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