Non-invasive ventilation (NIV) of acute respiratory failure in infants with spinal muscular atrophy (SMA) type I

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Disease

- Spinal Muscular Atrophy type I
- Werdnig-Hoffman Disease

- Defect of chromosome 5
- Recessive autosomic trasmission

- Healthy bearers: 1 out of 40
- Incidence of illness: 1 out of 25.000

Spinal Muscular Atrophy type I

Symptoms

- Precocious appearance, often in uterus or immediately after birth
- Generally clear signs between the ages of 6 months and 2 years
- Cardinal sign is weakness
- Inability to sit up without support
- Inability to cough with consequent accumulation of secretions in the lung
- Frequent and easy development of atelectasis and pneumonia
- Repetition of episodes of lung infection and respiratory insufficiency, leading to hospitalization and PICU admission

Spinal Muscular Atrophy type I

Characteristics

- Progressive respiratory failure connected with insufficient ventilation
- Inability to expel secretions due to inefficacy of cough
- Difficulty in swallowing

Complications connected with

1. Insufficient ventilation due to muscle impairment and paradoxic chest movements
   - Formation of disventilated and atelectatic area due inefficacy of ventilation and accumulation of secretions
   - Bronchopneumonia and pneumonia, due to infection in non ventilated areas
   - Aspiration of regurgitated gastric material which leads to airway obstruction, chemical pneumonia and ARDS
2. Inability to swallow
   - Easy tracheal aspiration of saliva, food and gastric content

Spinal Muscular Atrophy type I

Reluctance to intubate and to initiate intensive care treatment

- Loss of spontaneous breathing
- Difficulty in returning to spontaneous breathing

Outcome of invasive ventilatory treatment

- Tracheotomy and home care mechanical ventilation
- Child is allowed to die in case of subsequent respiratory failure after extubation

Study design

Aim of the study

- Evaluate possibility and efficacy of non-invasive ventilation in children affected by SMA I. Propose home assistance with non-invasive ventilatory support during the night

Patients

- 10 infants, 8 females and 2 males - age 6 to 12 months

- 1st admission: not home ventilated
- 2nd admission: not home ventilated or night-time ventilated

- Total admissions: 20

Non Invasive Ventilation using nasal mask

Criteria of admission

1. Not severe and recent appearance of respiratory failure

2. Not presence of additional signs of risk, such as
   - Bradicardia / asistolia
   - Severe cardiovascular instability and shock
   - Upper airway instability and/or obstruction
   - Lack of protective pharyngeal and laryngeal reflexes
   - Severe neurological compromission

Ventilatory mode

Non-invasive Ventilatory Mode

- Controlled Volume/Pressure Ventilation

- Synchronized + pressure support ventilation

  (back up: 2-3 breaths more or less than patient's current frequency)

Ventilatory mode

1. BiPAP 24 hours per day

2. Expulsion of secretions
   - Cough-Assist
   - RTX Respirator, Secretion Clearance mode

BiPAP setting

At admission

Back up	3-5 breaths more than patient's current frequency
EPAP	8-10 cm H_2O
IPAP	18-22 cm H_2O

Disconnection from ventilator

Back up	3-5 breaths less than patient's respiratory frequency in normal condition
EPAP	6 cm H_2O
IPAP	18 cm H_2O

Home care (nocturnal assistance per 6-8 hours)

Back up	14-16 breaths
EPAP	6 cm H_2O
IPAP	18 cm H_2O

BiPAP setting and weaning

Setting at admission was adapted in order to maintain

- $paCO_2 \leq 45$ mmHg
- Complete synchrony of patient with ventilator
- Limited or no oxygen support

Disconnection from ventilator

Progressive reduction of ventilatory support according to improvement in lung pathology until reaching

- Nocturnal support, and 3-5 hours day time support (noon to 4 pm)
- Nocturnal support for 6-8 hours

Expulsion of secretions

RTX Respirator

- Time: max 1 hour
- HFO 600 Hz –10 cmH_2O
- Cough –24 +12 cmH_2O I:E = 5:1

Cough- Assist

- Time: max 5 min
- +25÷30 cmH_2O –30÷35 cmH_2O

Expulsion of secretions

- Cough-assist had been used only in 2 infants until RTX Respirator became available. The machine appears unsuitable for use with children under two years as during the negative phase it favours alveolar collapse due to high closure volume of small airways and formation of inhomogeneous lung ventilation

- RTX Respirator, Secretion Clearance mode, has been continuously used as cough support with encouraging results. Secretion mobilization is obtained using HFO mode, and expulsion of secretion from trachea is achieved using artificial cough

- Secretion clearance was performed initially 6-8 times per day according to patient's need, then stabilized at 4 times per day and when necessary. When BiPAP was interrupted, effective secretion clearance was achieved before connection and immediately after disconnection from ventilator

Problems related to BiPAP and nasal mask

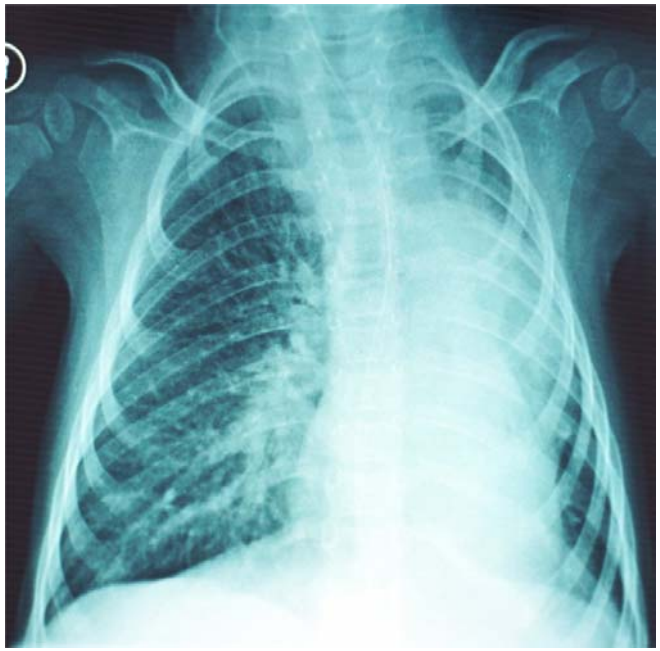
- Difficulty in synchronization between infant and ventilator
- Difficulty in obtaining suitable size of facial mask
- Easy dislocation of mask and difficulty to avoid leakage

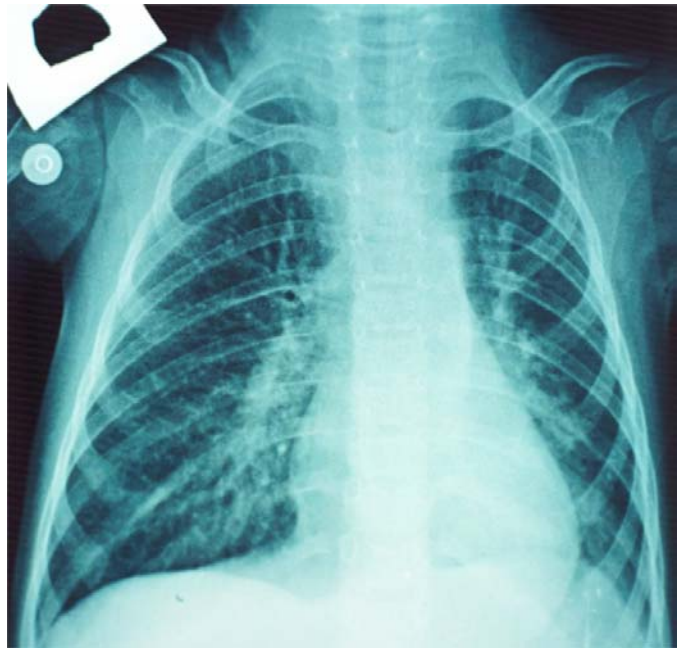
- Accumulation of secretion in dependent lung areas
- Unstable PEEP level maintenance according to leakage from mouth

Complications

- Severe necrosis connected with facial mask
- Gastric distension
- Hypoventilation
- Difficulty in feeding

Clinical cases





- Chest X-Ray – Before and after treatment – Case A

Clinical cases

- Chest X-Ray – Before and after treatment – Case B

Results

- All infants survived and at present are under home care assistance with nocturnal BiPAP, except one who maintains a good respiratory autonomy 3 months after discharge. No child needs home care oxygen support
- Re-admission was more frequent until infants could avail of home care BiPAP and a secretion removal machine
- PICU stay was between 5 and 55 days, with a mean of 20 days
- Improvement of chest x-ray was achieved on average after 4 days, depending on the severity of case and whether pathology was recent or consolidated
- Withdrawal of day-time BiPAP was possible between 4^{th} and 12^{th} day of PICU stay

Results

- Gastric distension was observed frequently and conditioned feeding of the infant, and therefore total parenteral support was often necessary
- 2 infants developed aspiration syndrome which was treated by increasing ventilatory support and RTX Respirator frequency (8-10 times /day)
- Supplementary oxygen was suspended generally immediately after initiation of ventilatory support. Only two infants needed FiO_2 0.3 until 48 hours after admission
- Moderate oxygen desaturation (SpO_2 between 86-90%) was observed sporadically in relation to need for secretion removal and during the suctioning manoeuvre

Conclusion and comments

- SMA I infants need some ventilatory support during night-time when respiratory insufficiency episodes become frequent and require hospital admission. Nocturnal ventilatory support becomes indispensable after first ICU admission, except in rare cases (1 case out of 10 in our study). Night-time non-invasive ventilation enables an improvement in patient's quality of life, reduces lung infections and re-admission to hospital

- In home care assistance a non-invasive ventilatory machine is indispensable, as well as equipment which favours the removal of secretions

- Physiotherapy gives good results in delaying admission to hospital, but is ineffective when respiratory insufficiency is more marked or in the presence of inflammation. In this situation physiotherapy represents a supplementary support and in any case a machine to remove secretions in the absence of cough is needed