

## Postoperative use of nasal intermittent positive pressure in a patient with spinal muscular atrophy type II

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**Abstract :** We report the successful use of nasal intermittent positive pressure ventilation (NIPPV) in the perioperative period of a 51 yr-old woman with a type II spinal muscular atrophy (SMA II). The patient was treated chronically with nocturnal NIPPV at home and scheduled for endoscopic retrograde cholangiopancreatography (ERCP) under general anesthesia. Some criteria of difficult intubation were present (forced mouth opening of 1.5 cm, short neck and thyromental distance of 5 cm). Nasal endotracheal fiberoptic intubation during spontaneous breathing under sedation with propofol was performed. The ERCP procedure was conducted without complications. At the end of the procedure, IPPV was maintained until recovery of respiratory function. After extubation, NIPPV was continued in the recovery room. The patient was discharged from the post-anesthesia care unit 4 hours after the procedure.

Management of patients with SMA remains a challenge and clinicians must be aware that the use of NIPPV may be a useful and life-saving tool in the perioperative period for these patients.

### INTRODUCTION

Spinal muscular atrophies (SMA) represent a group of neuromuscular disorders leading to progressive muscular impairment. The incidence is estimated to be 1 in 10 000 (1). Clinical classification may be divided in three subtypes but their evolution is unpredictable. Type II patients can initially sit alone but will never be able to walk and progressive degeneration of the medullar anterior horn leads to inability to sit as well. Restrictive lung disease may further complicate the clinical picture and death occurs in a large range of ages (from two years to adulthood) (1-2).

The genesis of SMA was hypothesized/ thought to be through Apoptosis. But more complex mechanisms are probably involved. Specific deletions on the survival motor neuron gene (5q-region) were incriminated (2).

There are few reports concerning anesthesia for patients with SMA, especially type II. While improving perioperative management of a rare disease is important, it is a real challenge to care for

such patients especially with advanced lung disease (3-5). Moreover, association with poor mouth opening and short neck has been reported causing potential difficult airway management, particularly endotracheal intubation (3).

Modern management of ventilatory failure caused by neuromuscular disease includes nasal intermittent positive pressure ventilation (NIPPV). Such non-invasive mode of ventilation may considerably simplify the postoperative course after general anesthesia (3).

### CASE REPORT

A 51 yr-old woman was referred to our institution from another hospital with an edematous pancreatitis from biliar lithiasic origin associated with hepatic cytolysis and cholestasis, and was to receive an endoscopic retrograde cholangiopancreatography (ERCP) under general anesthesia (GA). A first attempt to perform an awake ERCP failed due to poor mouth opening.

SMA II was diagnosed in this patient in early childhood. Respiratory function was described as quite good during several years. The sitting position was possible during childhood. Major striate muscular function impairment had confined our patient to bed rest for the last several years. NIPPV was introduced after an episode of pneumonia associated with respiratory distress and carbonarcosis and

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was in use for 19 months. Polysomnography described major abnormalities linked to severe fragmentation of the sleep and continuous hypoxemia. The apnea-hypopnea index was 99 (defined as the number of apneas and hypopneas per hour of sleep). Mean SpO<sub>2</sub> was 85%, 77% during NREM-sleep and 67% in paradoxical sleep. Initiation of NIPPV was well tolerated and efficacious. After three days, mean awake SpO<sub>2</sub> was 92%, 91% during NREM-sleep and 91% in paradoxical sleep. Apnea-hypopnea index was 1 (Parameter of ventilator : frequency 20 per min, inspiratory pressure : 18 cm H<sub>2</sub>O, expiratory pressure : 5 cm H<sub>2</sub>O).

Forced expiratory volume in 1 s (FEV<sub>1</sub>) was 0.52 L (26% predicted), and forced vital capacity (FVC) was 0.69 L (25%).

Her medical history included esophagitis, frequent urinary infections and left hypoacusia. Appendectomy was performed under general anesthesia at the age of 27 yr without problems. A subcutaneous intravenous port was placed under local anesthesia at the age of 47 years to allow frequent intravenous antibiotherapy linked to urinary tract infections.

Physical examination revealed an obese tetraparetic female (height : 145 cm, weight : 75 kg, Body Mass Index : 35.7). She was unable to sit and presented atrophic and deformed extremities. A marked dorsolumbar scoliosis was associated with a fixed cervical kyphosis. Poor mouth opening (1.5 cm), short neck and thyromental distance (5 cm) without cervical mobility (no more than 5° flexion or extension) were predictive of a difficult intubation.

Laboratory investigations showed a moderate iron deficiency anemia (Hemoglobin : 11.3 g/dL, serum iron 27 mcg/dL and ferritin 21 ng/mL), normal renal function (urea 17 mg/dL and creatinine (0.12 mg/dL), hepatic cytolysis (ASAT 118 IU/L, ALAT 219 IU/L) and cholestasis (alkaline phosphatase (1129 IU/L) and gamma-GT (1354 U/L). Amylase and lipase concentrations were initially increased (634 and 2714 IU/L resp.) before normalisation. C-reactive protein was decreased (4.8 to 1.9 mg/dL). Urine sediment showed 1163 leucocytes/ $\mu$ L and more than 100.000 *Escherichia coli*/mL (amoxicilline-clavulanate-sensible).

Her treatment consisted in omeprazole 20 mg twice a day, inhalation of formoterol fumarate 9  $\mu$ g in the morning, amoxicilline-clavulanate 1 g four times per day, subcutaneous enoxaparine 40 mg in the evening and physiotherapy two to three times per day. NIPPV parameters were stable during several months (frequency 17 per min, inspiratory

pressure : 15 cm H<sub>2</sub>O, expiratory pressure : 3 cm H<sub>2</sub>O).

Our radiologists considered chest X-ray as uninterpretable. Abdominal CT-scan and ultrasonography showed a choledocian dilatation but no lithiasis. MRI helped to locate a 4 mm intracholedocian lithiasis.

The risks and the modalities of the procedure were explained, understood and accepted by the patient. An intensive care unit bed was booked for postoperative care.

Routine monitoring included SpO<sub>2</sub>, 3-leads electrocardiography, non invasive blood pressure, end-tidal carbon dioxide -ETCO<sub>2</sub>- and capnography. Venous access was realized via the subcutaneous intravenous port with NaCl 0.9% infusion. Preoxygenation was administered for five minutes with facial mask. Sedation was induced during spontaneous breathing with a continuous infusion of propofol and ketamine 25 mg preceded by lidocaine 40 mg. Fibroscopy was easily performed and a deeper state of anesthesia was then obtained after a bolus of propofol. No muscular relaxant or opioid was administered. An nasotracheal tube (Mallinckrodt 7.0 with low pressure cuff) was inserted over the fiberoptic laryngoscope. Mechanical ventilation was started (Fabius Dräger : volume-controlled-mode, volume-tidal of 350 mL, eighteen cycles per minute, expiratory pressure 3 cmH<sub>2</sub>O) with an oxygen/air mixture. Peak (19 cmH<sub>2</sub>O) and mean (7 cmH<sub>2</sub>O) pressure obtained were accepted. No significant hemodynamic alterations were observed (i.e. no more than 20% from baseline). At the end of the procedure, while the patient was allowed to resume spontaneous respiration, IPPV was initiated on the patient's own machine after connecting it to the nasotracheal tube. To verify the adequacy of breathing, the inspired fraction of oxygen was reduced to 21%. SpO<sub>2</sub> remained stable i.e. > 95% and ETCO<sub>2</sub> < 38 mmHg (5.05 kPa). With a fully collaborating patient and after breathing 100% oxygen for five minutes, extubation was realized. NIPPV was then immediately reinstalled. Excellent breathing was confirmed with stable SpO<sub>2</sub> (> 96%) and ETCO<sub>2</sub> (< 39 mmHg, 5.19 kPa). The patient was transferred to the post-anesthesia care unit (PACU), where NIPPV was continued. The inspired fraction of oxygen was gradually decreased to 21% one hour after the end of the procedure. Excellent ventilatory parameters were registered. After one hour, hyoscine butylbromide 20 mg was intravenously administered for spasmodic-like abdominal pain. It was followed, 30 min later, by alizapride 50 mg and

tramadol 50 mg. Pain relief was then satisfactory without side-effects.

Four hours after the end of the procedure, the patient was discharged from the PACU. Nocturnal NIPPV was continued uneventfully. The patient stated that she was fully satisfied and had recovered from general anesthesia without any complication.

## DISCUSSION

We reported a case of general anesthesia in a SMA II patient with predicted difficult intubation. We used a nasal fiberoptic intubation technique during spontaneous breathing. Propofol was chosen for its short-lasting effect, independent elimination of the ventilation rate and lack of effects on respiratory muscular function. No opioid was used to avoid long-lasting effects on ventilatory function. No muscle relaxant was given to avoid unpredictable response (5). Sedation with propofol infusion may be controversial in a patient with documented esophagitis. Nevertheless, we considered that after several years of symptom-free treatment with omeprazole the technique was safe enough to be used. Sedation may permit to improve tolerance of intubation under fibroscopy.

One of the most important issues of our report is how NIPPV considerably simplified perioperative management. This outcome was facilitated by the patient's tolerance of this mode of ventilation for several months.

General anesthesia is rarely reported in SMA II patients and remains a challenge when no other acceptable solution is advisable. Reports of such cases may allow to improve perioperative management. At the best of our knowledge, only two cases of well-established general anesthesia in SMA II patients have been reported (3-4). NIPPV was used in an 11-yr-old boy after spinal surgery (3). It was established during five months before surgery. The trachea was intubated through a laryngeal mask airway using a fiberoptic laryngoscope. Vecuronium was given to facilitate intubation and the patient was electively ventilated for 24 hours. NIPPV was reinstalled after removal of the tracheal tube. A cough-assisted device was used to treat sputum retention prevented tracheostomy. A second report described general anesthesia in a SMA II patient, a 23-yr old woman for cesarean section (4). Good intubating conditions were described and no need of postoperative mechanical ventilation described. Our patient presented comparable preoperative val-

ues of FEV1 and FVC. Nevertheless the management of our patient was potentially more complicated due to long lasting and severe evolution of the pathology, obesity and the predicted difficult intubation.

SMA II patients usually survive into early adulthood in contrast to other types. In type I, patients remain unable to sit and death usually occurs at two years after the diagnosis. In type III, onset occurs after 18 months. Patients are able to walk. Death occurs later in adulthood. In type II, truncal and extremity weakness is typical. Cardiorespiratory failure is the main cause of death (1-2).

NIPPV is becoming standard therapy to treat hypoventilation in neuromuscular diseases. It reduces the incidence and severity of respiratory complications and improves quality of life (6). In the light of this report, further studies should confirm the potentially life-saving use of NIPPV in the perioperative period. It may be particularly illustrated in major surgery or with advanced disease. Ideally it should be optimised preoperatively.

In conclusion we report the case of general anesthesia for ERCP in a patient with SMA II. It was successfully managed with early postoperative extubation and use of NIPPV. Clinicians must be aware of this type of management facilitating such perioperative challenge.

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