

Respiratory Muscle Aids During an Episode of Aspiration in a Patient With Duchenne Muscular Dystrophy

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We report the case of a Duchenne muscular dystrophy patient with good bulbar function but severely decreased forced vital capacity (9%) and spontaneous peak cough flow (PCF) (2.35 L/s). The patient needed continuous noninvasive ventilation (NIV) consisting of a volumetric ventilator with a nighttime nasal mask and a daytime mouthpiece. He also required application of manually assisted coughing techniques by insufflation with a resuscitation bag and chest thrust (manually assisted PCF after maximum insufflation capacity of 4.33 L/s). An episode of serious food aspiration was resolved by his main caregiver through NIV and manually assisted coughing. Bronchoscopy under sedation using NIV with a lip seal connection to his volumetric ventilator later revealed that no material remained. This case exemplifies the potential role of skilled respiratory management in some neuromuscular diseases.

Key words: *Neuromuscular diseases. Noninvasive ventilation. Assisted cough. Fiberoptic bronchoscopy. Aspiration.*

Ayudas a los músculos respiratorios durante un episodio de aspiración en un enfermo con distrofia muscular de Duchenne

Presentamos el caso de un enfermo afectado de distrofia muscular de Duchenne con buena función bulbar, pero con disminución importante de la capacidad vital forzada (9%) y del pico de flujo de tos espontáneo (2,35 l/s) que le obligaba a mantener ventilación no invasiva (VNI) continua (ventilador volumétrico, mascarilla nasal nocturna y pieza bucal en vigilia) y a utilizar ayudas manuales para la tos mediante insuflaciones con ambú y compresión torácica espiratoria (pico de flujo de tos asistida tras máxima capacidad de insuflación de 4,33 l/s). El paciente presentó una aspiración importante de material digestivo que se resolvió gracias a la asociación de las ayudas manuales para la tos proporcionadas por su cuidadora principal y a la VNI. En un segundo momento, la VNI con una boquilla sellada como conexión a un ventilador volumétrico permitió realizar una broncoscopia bajo sedación, en la que se comprobó que no había material retenido. Este caso es un ejemplo de la utilidad potencial del manejo cualificado de los problemas respiratorios en algunas enfermedades neuromusculares.

Palabras clave: *Enfermedades neuromusculares. Ventilación no invasiva. Tos asistida. Fibrobroncoscopia. Aspiración.*

Introduction

Respiratory problems are the leading cause of death in some chronic neuromuscular diseases.^{1,2} Fatal respiratory failure is usually the result of progressive deterioration of the respiratory function (parallel to the loss of muscular strength) compounded by the devastating effect of a lung infection that at first may seem harmless.² On the other hand, an acute episode can result in death directly related to choking which overly weakened respiratory muscles cannot resolve through coughing.³

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Respiratory muscle aids have proven useful in cases where respiratory muscles can no longer function adequately.⁴ We report the case of a man with good swallowing function but serious deterioration of the chest muscles: he was unable to produce an effective spontaneous cough and was practically unable to breathe without mechanical ventilation. Manually assisted coughing and noninvasive ventilation (NIV) prevented a potentially serious problem related to food aspiration. NIV and sedation later facilitated fiberoptic bronchoscopy with minimum risk and no discomfort.

Clinical Description

A 29-year old man with Duchenne muscular dystrophy required continuous NIV with a volumetric ventilator (PV 501, Breas Medical, Mölndal, Sweden) because of respiratory failure (less than 4 hours per day without NIV). He used a

daytime mouthpiece and a nighttime nasal mask (Mirage, Resmed, San Diego, California, USA). Moreover, he had severely diminished cough capacity (requiring manual assistance for effective coughing).⁵ The last examination prior to the episode (2 months earlier) revealed lung function values consistent with his clinical situation: forced vital capacity (FVC), 0.51 L (9%); forced expiratory volume in one second (FEV₁), 0.48 L (11%); FEV₁/FVC, 0.95; peak cough flow (PCF), 2.35 L/s; maximum insufflation capacity (MIC), 1.63 L; and assisted PCF after MIC (PCF_{MIC}), 4.33 L/s.

The patient was reading while lying in his normal position in bed (Figure 1) when 30 minutes after supper he experienced epigastric pain followed by nausea and vomiting with evident food aspiration. The patient remained conscious. His mother immediately began applying manually assisted cough techniques. She used a resuscitation bag to reach MIC by delivering the air volume the patient could not inhale spontaneously. This insufflation technique was coordinated with chest thrusts, which substituted for the respiratory muscles in effecting the expulsion phase of a cough. By alternating these techniques with periods of NIV, the patient was able to expel first liquid material, followed by some remaining food (chewed lentils), and finally, saliva only around 25 minutes after starting assisted cough maneuvers.

The patient remained calm after the maneuvers; no dyspnea, nausea, or feeling of foreign bodies in the airways was noted. Oxygen saturation was 98% with a fraction of inspired oxygen (FiO₂) of 0.21 with peaks of maximum inspiratory pressure of 15 cm H₂O in the ventilatory cycles. Tired and drowsy, the patient asked for his nasal mask and fell asleep. During the night he woke up with chills and fever (peak temperature, 39°C) but did not go to the hospital until the next morning.

Upon arrival at our hospital the patient was alert and oriented. He was connected to the ventilator (assist-control mode) with a lipseal mouthpiece, at 16 cycles per minute (untriggered). Heart rate was 110 beats per minute; blood pressure was 110/70 mm Hg; and temperature, 37°C. Heart and lung sounds were rhythmic, and vesicular sounds were normal (with NIV). No anomalies were noted. Blood gases (connected to ventilator with FiO₂ of 0.21) analysis showed a pH of 7.40, PaO₂ of 93 mm Hg, and PaCO₂ of 42 mm Hg. The white blood cell count was 16 700 (92% neutrophils). The chest x-ray showed no alveolar flooding or atelectasis.

Later that morning fiberoptic bronchoscopy, under sedation (fentanyl) and using NIV with a lipseal connection to the volumetric ventilator (Figure 2), confirmed that no foreign bodies remained and airways were clear. Oxygen saturation remained above 96% (with FiO₂ set at 0.21) throughout the procedure.

The patient was treated with amoxicillin-clavulanic acid. No complications were evident, and he was discharged 24 hours after admittance.

Discussion

This case demonstrates how the coordination of 2 noninvasive respiratory aids (manually assisted coughing and NIV) was crucial for resolving a life-threatening situation in a patient with Duchenne muscular dystrophy

Currently, 2 approaches are recommended to manage conditions in which the chest muscles cannot function properly in patients with some neuromuscular diseases: *a*) the conventional approach,⁶ which uses an invasive



Figure 1. Daytime mechanical ventilation with simple mouthpiece.



Figure 2. Fiberoptic bronchoscopy under sedation via the nasal route using a volumetric ventilator with lipseal connection.

access to the airways to remove secretions and to obtain an appropriate alveolar ventilation in cases where unassisted coughing or breathing is not possible, and *b*) the noninvasive approach, which uses noninvasive techniques⁷ to achieve the same objective provided that the patient's bulbar function is good.⁸ Our patient, who had a PCF of only 2.35 L/s, would have been unable to expel the aspirated material without manual cough assistance. And without simultaneous NIV support, he could not have performed the necessary exertion that assisted cough maneuvers require. Good bulbar function as well as active, coordinated participation of patient and caregiver is necessary to achieve a MIC and PCF_{MIC} strong enough to eliminate secretions. In cases

of severe choking, the CoughAssist (J.H. Emerson Co., Cambridge, Massachusetts, USA) should be more effective than manually assisted coughing in resolving such a life-threatening situation because it has greater extractor capacity and does not require active cooperation.⁸ However, few patients outside of the USA have access to such a device for cough assistance. Therefore, manual techniques are the only available options in most cases.

Fiberoptic bronchoscopy was used to assess our patient's airways⁹ as well as to remove any fragments that might have remained. The procedure was carried out with a lipseal mouthpiece connection that allowed us to provide effective sedation.

There is evidence to support the use of NIV during procedures requiring sedation in neuromuscular disease patients with severe respiratory muscle impairment.^{10,11} Although neurologists recommend carrying out percutaneous endoscopic gastrostomy before the FVC falls below 50% of the expected value because of associated respiratory risks,¹² recent studies show that percutaneous endoscopic gastrostomy with NIV is a safe technique even when the FVC is clearly less than 50%.^{10,11} Given that FVC provides information regarding the "unassisted" functional situation, its possible predictive value should only be taken into account when adequate patient management is impossible. In an appropriate environment, however, FVC loses its prognostic role,¹³ yielding to other measures of function (MIC and PCF_{MIC}) whose quantitative values are directly related to the clinical effectiveness of manually assisted coughing. In our patient, for example, the FVC of 0.51 L (9%) increased by 300% to result in an active MIC of 1.63 L. In turn, an ineffective PCF (2.35 L/s) became a PCF_{MIC} of 4.33 L/s, allowing for an effective cough.

Fiberoptic bronchoscopy can also be safely carried out in hypoxemic patients with the help of pressure support or continuous positive airway pressure and different masks (perforated helmet, perforated oronasal mask, nasal mask).^{14,15} The interest of the case we present lies in the patient's disease (Duchenne muscular dystrophy treated with continuous ventilatory support), in the type of mask (lipseal), and in the use of a volumetric ventilator. The lipseal device facilitates the technical procedures of fiberoptic bronchoscopy by leaving the nasal passageway—the most common access route—free for use. At the same time, the seal prevents possible ventilator air leaks. Alveolar ventilation in our patient remained within the appropriate range during fiberoptic bronchoscopy (without additional oxygen therapy) with the chosen mask and ventilator combination.

In conclusion, the case of our neuromuscular disease patient demonstrated that good bulbar function, cooperation along with good caregiver-patient coordination, and a combination of manually assisted coughing and NIV enabled him to expel aspired material from his airways. Later, volumetric NIV with a lipseal interface maintained appropriate alveolar ventilation during fiberoptic bronchoscopy under sedation. This case shows the potential benefit of skilled respiratory management in patients with neuromuscular diseases.

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