effusion without parenchymal or mediastinal abnormalities. Two percutaneous Abrams needle biopsies of the pleura (with 3 weeks of interval) were negative for malignancy and infection, only demonstrating nonspecific fibrous thickening of the pleura. The thoracoscopy showed hyperaemia and thickening of the costal pleura, and small whitish nodular lesions, that were biopsied (Fig. 1). An apple-green birefringence was noted on Congo-red staining under polarized light, compatible with pleural amyloidosis, mostly with a perivascular pattern (Fig. 2).

Eight months after the thoracoscopy, the pleural effusion recurred. This time, it was bilateral with a moderate volume and a transudate. Despite initiating treatment for WD few months after the beginning of symptoms, the inflammatory process evolved to systemic amyloidosis, which was the major factor contributing to the progressive deterioration of our patient, with cardiac and renal failure. The patient died due to multiple organ failure, after 3 years of follow-up.

Although a definitive diagnosis of WD could not be made, the endoscopic characteristic findings and the response to the antibiotic therapy suggest it as the precursor of the AA amyloidosis.

Pulmonary amyloidosis may present as tracheobronchial infiltration, parenchymal nodules, persistent pleural effusions, and pulmonary hypertension.[3,4] Pleural involvement is very rarely reported, and it is usually associated with AL amyloidosis, which accounts for up to 80% of pulmonary amyloidosis.[5,6] Typical aspects of thoracoscopy consist of hyperaemia of the pleural surface, inflammation with nodular lesions or brown nodules of the parietal pleura.[5,7] Persistent pleural effusions occur in 1–2% of patients with systemic amyloidosis and are usually associated with poor prognosis and often refractory to treatment. Pleurodesis has been effective in some cases.[7,8]

This case illustrates the difficulty in diagnosing pleural amyloidosis, after two failed needle pleural biopsies. This is probably due to the fact that amyloid deposition is not uniform over the pleural surface. The pleural effusion in our patient may derive from increased fluid production induced by inflammation, interruption of lymphatic drainage caused by amyloid infiltration of the pleura, decreased resorption of fluid from the pleural space due to vascular deposition of amyloid and in the bilateral pleural effusion from congestive heart failure secondary to amyloid infiltration in the heart.[5,7]

Although in this case the previous diagnosis of amyloidosis supported and incited to actively search for amyloidosis, it should be always kept in mind, as pleural effusion is a rare manifestation of pulmonary involvement but could be the presenting manifestation of amyloidosis.

Conflicts of interest

The authors have no conflicts of interest to declare.

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C. Dias*, I. Tavares, A. Magalhães, N. Melo

Centro Hospitalar de São João, Porto, Portugal

* Corresponding author.
E-mail address: catarinadias1@gmail.com
(C. Dias).

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Use of the Intermittent Abdominal Pressure Ventilation to guarantee speech in a tracheostomized Amyotrophic Lateral Sclerosis patient

Dear Editor,

Amyotrophic Lateral Sclerosis (ALS) is a fatal, progressive, neurodegenerative disease. When Respiratory failure is too severe to be corrected with Non Invasive Ventilation and/or when bronchial secretions cannot be managed with noninvasive techniques, tracheostomy and invasive mechanical ventilation (IV) are an option.[1]

Tracheostomy ventilation significantly prolongs survival in ALS patients without effect on the disease progression. For this reason, patients with IV experience a worsening of disability and an increment of the dependency with a severe impairment of their quality of life.[2] One of the most important aspect of the multidisciplinary approach in ALS is to guarantee as long as possible the maintenance of the residual functions. In this context, the first and most widely used strategy to allow tracheostomized patients without severe bulbar involvement to speak is the simple cuff deflation, but, in a percentage of these patients, this technique fails

Table 1 Ventilation and cough machine settings (PS: pressure support; IP: inspiration pressure; EP: expiration pressure; RR: respiratory rate; Ti: inpiratory time; Te: expiratory time; Tp: pause time; Tr Insp: inspiratory trigger; IV: invasive ventilation; TOV: tracheal open ventilation; IAPV: intermittent abdominal pressure ventilation; APCV: assisted pressure-control ventilation; ST: spontaneous timed).

	IV	TOV	IAPV	Cough machine
Mode	APCV	ST	APCV	Auto
PS (cmH_2O)	13	16	40	
IP				40
Peep (cmH_2O)	4	4	0	
EP				40
RR	14	14	18	
Ti (s)	1.4	1.4	2.2 (Te)	3
Te (s)				2
Tp (s)				1
Tr Insp	High	High	Auto	
Rise time (ms)	400	400	600	

with the consequent impossibility of verbal communication and a severe reactive mood depression.[3]

Intermittent Abdominal Pressure Ventilation consists of an elastic inflatable bladder incorporated within a corset surrounding the abdomen. With bladder inflation by a ventilator, the abdominal content and diaphragm move upward, assisting expiration. With bladder deflation, inspiration occurs passively. There have been only scattered reports on the use of IAPV[4–7] and two publications concerning its use in large populations of patients, in a regimen of noninvasive[8] and invasive[9] ventilator support.

A 49 years old man with a diagnosis of definite ALS, according to El Escorial Criteria, was admitted for the first time to our Centre, having recently had tracheostomy and gastrostomy done because of worsening in respiratory function and swallowing, The patient complained of difficulty using home IV device and difficult management of the secretions. NIV was introduced in Spring 2014 after onset of wheezing during hospitalization in an intensive care ward of an other Centre. The patient reported intermittent use of non invasive ventilation due to poor tolerance until Autumn 2014. At that time there was a significant worsening of respiratory involvement due to a right lung pneumonia associated to weight loss and deterioration of swallowing and dysphagia.

During the hospitalization, we optimized both IV and secretion clearance by cough machine through the tracheostomy tube in association with tracheal aspirations. He could not sustain the spontaneous breathing, so, to permit speech, we introduced diurnal tracheal open ventilation (TOV) with cuff deflation and the concomitant usage of a speaking valve, but with poor patient tolerance due to discomfort with the unnatural experience of air coming in the upper airways and poor synchrony with the ventilation. The patient could not even manage secretions and saliva. Thus, to increase the feasibility and pleasure of speaking during mechanical ventilation and improve breathing comfort we introduced diurnal IAPV (*Pneobelt*™) associated with a tracheostomy speaking valve for spontaneous breathing. During tracheal ventilation, our patient was fitted while supine,

Figure 1 Pneobelt™ corset.

with the corset's horizontal upper border approximately two finger breadths below the costophrenic junction. Once positioned, the patient was placed in a wheelchair, his cannula wad deflated and a speaking valve was placed. *Pneobelt*™ was connected to the same portable ventilator used for tracheal ventilation but set with parameters ''*ad hoc*''for IAPV.

Table 1 shows IV, cough machine and IAPV settings (Fig. 1) shows Pneobelt™ with the corset and inner bladder.

Table 2 Pulmonary gas exchange (ABG: arterial blood gas analysis; IV: invasive ventilation; IAPV: Intermittent Abdominal Pressure Ventilation).

ABG first evaluation	IV	IAPV + speaking valve
PH	7.57	7.43
PaO_2	84.2	71
$PaCO_2$	36.3	40
HCO_3^-	27	27

Table 3 Respiratory assessment during IAPV (intermittent abdominal pressure ventilation)(§: 5 point scale; 1 poor tolerance–5 optimal tolerance).

	I evaluation	II evaluation	III evaluation	IV evaluation	V evaluation
Tidal volume (ml)	490	600	550	620	580
Transcutaneous CO_2 (mmHg)	30	25	32	28	31
Dyspnoea (Borg Scale 0–10)	1	0	0	0	0
IAPV tolerance after 1 h treatment§	5	5	5	5	5
IAPV usage (h)	4	6	6	8	8

The use of IAPV permitted optimal speech, an efficient diurnal ventilatory pattern, good pulmonary gas exchange (Table 2), without dyspnoea and with a significant improvement of the salivary secretions' management and a decrease in the tracheal aspiration need.

At time of discharge, our patient maintained IAPV for 3–4 h/day, while he was in a wheelchair, with great compliance to treatment. He felt his breathing was ''normal, like before tracheostomy''. To guarantee a safe discharge we trained the caregivers in the proper respiratory management and periodic supervision calls were made.

Table 3 shows the clinical follow-up of the patient performed every 3 months. Moreover, over time we checked the reappearance of a spontaneous respiratory activity without any need for ventilatory support. Nowaday our patient can maintain the spontaneous breathing for almost 2 h, without dyspnoea or tachipnoea. No major technical problems or adverse effects due to IAPV use were reported.

To the best of our knowledge, this case describes for the first time the use of IAPV in a tracheostomized ALS patient, a noninvasive ventilation mode that functions primarily by exerting force on the abdomen and indirectly causing motion of the diaphragm. As reported in the Literature we confirm that IAPV is simple to use, portable and without particular side effects related to its daily use.

A respiratory rehabilitative treatment for ALS tracheostomized patients should consist in optimizing IV and secretion management with cough machine and in verifying spontaneous respiratory trials, TOV and speaking valve use both during ventilation and spontaneous breathing.[3] In 1988, Miller et al., in a large, high quadriplegic patient series with tracheostomies, reported that the advantages if IAPV use included plugging of the tracheostomy tube. With IAPV speech was easier and louder.[9] Our patient had a good speaking competence, but serious difficulties to manage secretions and saliva during TOV. So, to optimize speech, we introduced the diurnal use of IAPV.

In our centre tracheostomy is proposed only for patients with severe bulbar involvement, because the indication for tracheotomy should be dependent on glottic function rather than on inspiratory or expiratory muscle failure,[1] but this patient arrived in our department having already had a tracheostomy. In our practice, ALS patients are not decannulated, to avoid the risk of near re-intubation due to disease progression and, or new episodes of acute respiratory distress syndrome; other authors,[10] instead, propose the use of continuous volume-cycled NIV and cough machine with oximetry feedback in ambient air to permit safe extubation of unweanable ALS non bulbar patients. Our combined, ventilatory approach with tracheal invasive ventilation and IAPV has guaranteed optimal speech and a safe respiratory management with an optimal patient acceptance of treatment. Furthermore, there has been spontaneous breathing activity, which demonstrates an improvement in the patient's respiratory condition. IAPV facilitates diaphragmatic motion and may be particularly useful in patients with bilateral diaphragmatic weakness or paralysis and permits plugging of the tracheostomy tube with cuff deflation for several hours during the day, with prevention of tracheal damage.

Our case suggests that Pneumobelt can be a safe and effective method of daytime ventilation, and, by improving speech and secretion management, it permits a better quality of life even in a tracheostomized ALS patient.

Ethical disclosures

Protection of human and animal subjects. The authors declare that the procedures followed were in accordance with the regulations of the relevant clinical research ethics committee and with those of the Code of Ethics of the World Medical Association (Declaration of Helsinki).

Confidentiality of data. The authors declare that they have followed the protocols of their work centre on the publication of patient data.

Right to privacy and informed consent. The authors have obtained the written informed consent of the patients or subjects mentioned in the article. The corresponding author is in possession of this document.

Conflicts of interest

The authors declare no financial support for the conduct of the study.

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E. De Mattia[a,*], M. Iatomasi[a], B. Garabelli[a], C. Lunetta[a], V.A. Sansone[a,b], F. Rao[a]

[a] *NEuroMuscular Omnicentre (NEMO), Fondazione Serena Onlus, Milano, Italy*
[b] *Department of Biomedical Sciences for Health, Università degli Studi di Milano, Milan, Italy*

* Corresponding author.
E-mail address: elisa.dematti a@centrocliniconemo.it
(E. De Mattia).

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Multifocal micronodular pneumocyte hyperplasia associated with tuberous sclerosis complex: A case report without lymphangioleiomyomatosis association

Dear Editor,

Multifocal Micronodular Pneumocyte Hyperplasia (MMPH) corresponds to the proliferation of type II pneumocytes, which is almost exclusive to tuberous sclerosis complex (TSC) and/or lymphangioleiomyomatosis (LAM).[1] The authors report on an uncommon case of MMPH in a patient with TSC, without LAM.

We present a case of a 48-year-old female, with previous history of seizures and TSC diagnosed 18 years earlier, based on the presence of skin lesions (facial angiofibromas) and cortical tubers on brain MR. Investigation of her family revealed that 6 relatives have the same condition. She is a non-smoker and has had no relevant occupational exposure. The patient presented asymptomatic, namely without respiratory complaints. The pulmonary function tests were normal: FEV_1 2.91 L (104% of predicted); FVC 3.6 L (110% of predicted); DLCO 81%. Abdominal CT were performed to assess renal angiomyolipomas; the lower parts of the lungs showed pulmonary nodules that were followed up with a chest CT. The chest CT revealed multiple ground-glass nodules scattered through both lungs, the largest one was 8 mm (Fig. 1). There were no cystic lesions suggestive of LAM. After 2 years of follow-up the nodules remained stable in size and number, compatible with a MMPH diagnosis.

Tuberous sclerosis complex is an autosomal dominant disease, which commonly affects various organs, typically the brain, skin, kidneys, heart and lungs. LAM is the most common pulmonary manifestation of TSC, with a reported prevalence 1–3% of the TSC patients, however it could be much higher (up to 35%) in women with TSC.[2]

MMPH, despite being considered rare, is the second most common cause of lung involvement by TSC. This disorder consists of multicentric, well-demarcated nodular proliferation of type II pneumocytes along alveolar septa, and has been reported in men and women with TSC (with or without LAM manifestation) and in women with sporadic LAM, with only 3 cases described without this association.[3] Clinically, MMPH might be associated with cough, dyspnea and moderate or asymptomatic hypoxemia.[2] Unlike LAM, treatment is generally not needed in MMPH, because it seems to be indolent and not progressive, moreover it has not been proven to have malignant potential.[4] On HRCT MMPH is characterized by multiple solid or ground-glass nodules, ranging from 2 to 10 mm, scattered through the lungs in a random distribution. The main radiological differential diagnosis includes atypical adenomatous hyperplasia, early Langerhans histiocytosis, hematogenous metastases and miliary tuberculosis. The differentiation between MMPH and these entities may be very difficult, however the history of TSC, the multiplicity and random distribution of the nodules, as well the stability of number and size of these lesions in the follow-up examinations, helps to provide the diagnosis MMPH.

To the best of our knowledge there are no established guidelines for management of MMPH in TSC. Taking into account the benign nature of this entity, we think that is reasonable to assume the diagnosis MMPH in this case, leaving histopathologic sampling as an option if there are suspicious lesions, nodule progression in dimension or number; or if the patient decides to follow this approach.[4] However, a long-term follow-up is advised to document the stability the nodules to corroborate this diagnosis.