Pneumothorax in chronically ventilated neuromuscular and chest wall restricted patients: A case series

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INTRODUCTION: Pneumothorax occurs in chronically ventilated neuromuscular and chest wall-restricted patients. Non-invasive home mechanical ventilation (HMV) and airway clearance techniques are standard care, but are perceived to put patients at increased risk of pneumothorax. Malnutrition as reflected by low body mass index was identified as an association with pneumothorax in this case series.

METHODS: Retrospective chart review of six cases of pneumothorax in patients with neuromuscular and chest wall diseases receiving chronic HMV.

RESULTS: Six cases were reviewed. Median duration of HMV at time of the pneumothorax was 3 years (range 0.25 to 9 years). Five patients used airway clearance techniques. The mean forced vital capacity (FVC) was 1.0L (27% predicted). Patients had a low mean body mass index (BMI) of 16 ± 3 kg/m2. Five of 6 patients required chest drain and 4 of 6 had recurrent pneumothoraxes. One patient received chemical pleurodesis.

CONCLUSION: Pneumothorax occurs in neuromuscular patients receiving HMV and using airway clearance techniques. Management can include reducing ventilator pressure settings, holding airway clearance techniques and use of a chest drain. Chemical or surgical pleurodesis were felt to be high risk. Recurrence of pneumothorax was common. Low BMI in this case series raises the possible association between malnutrition and risk of pneumothorax in patients receiving HMV.

Key Words: *Pneumothorax; Neuromuscular disease; Chest wall restriction; Home mechanical ventilation; Lung recruitment; Airway clearance*

Pneumothorax in chronically ventilated neuromuscular and chest wall restricted patients has been documented previously (1-3), although the risk has been felt to be very low. Current guidelines for home ventilation include recommendations for ventilation and airway clearance techniques, but do not address how to prevent or manage pneumothorax in this patient population (4). A descriptive case- series is presented here.

METHODS

Consecutive cases of chronically ventilated patients with neuromuscular disease or chest wall restriction presenting with pneumothorax in southern Alberta were included. The population and referral base is approximately 2 million people. A retrospective chart review was performed to assess the characteristics of patients presenting with pneumothorax, and the diagnosis, treatment and outcomes.

RESULTS

Patient characteristics

The mean age of patients was 47 ± 18 years. The diagnoses of patients were: Amyotrophic Lateral Sclerosis, Limb Girdle Dystrophy, CAP myopathy, Collagen 6 disease, Myotonic dystrophy type 1 and kyphoscoliosis with ankylosing spondylitis. Mean FVC at time of most recent pulmonary function prior to the event was 1.00 ± 0.94 L (27 percent of predicted). The mean patient weight was 43 ± 13 kg with BMI of 16 ± 3 kg/m^2.

The median duration of non-invasive ventilation prior to first presentation with pneumothorax was 3 years (range 0.25 to 9 years). All patients received non-invasive ventilation using bilevel positive airway pressure devices with nasal or full-face mask. Settings at the time of first presentation with pneumothorax were: mean IPAP 20 ± 3 cm H_2O and EPAP 6 ± 2 cm H_2O, with back up rate of 18 ± 3 breaths per minute. The median daily usage of non-invasive ventilation was 9 hours (range 8-22 hours). Of the six patients two had had prior surgical fixation of the spine with Harrington rods.

All patients were prescribed and had been given instruction in airway clearance techniques including lung volume recruitment (LVR) or maximum insufflation-exsufflation (MIE) in accordance with Canadian Home Mechanical Ventilation guidelines (4). One of six patients reported that she was not actually using LVR prior to presentation with pneumothorax; however the remaining five patients did use airway clearance strategies on a regular basis (Table 1).

Pneumothorax characteristics

Three patients had small pneumothorax (<2 cm) on initial presentation and three had large pneumothorax (>2 cm) using the BTS guidelines definition for spontaneous pneumothorax (5).

CASE REPORTS

Case 1

A 61-year-old male with ALS presented with a right pneumothorax (Figure 1A). The patient was treated with a chest drain and upon removal of the tube, a small loculated basilar right pneumothorax persisted, as documented on CT (Figure 1B). The patient was ventilator-dependent at night, and continued using NIV throughout his treatment for pneumothorax. Airway clearance maneuvers (LVR) were held until radiographic stability was achieved on CT of the chest.

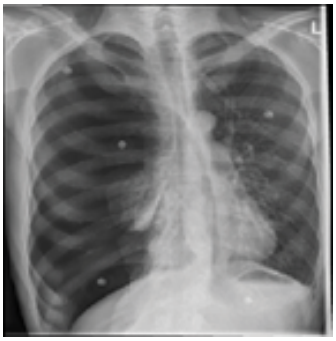

Figure 1A) *Chest x-ray of ALS patient at presentation with large right pneumothorax*

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TABLE 1
Description of the cases

Patient	Age (yr)	Gender	Diagnosis	FVC (L)	Wt (kg)	BMI	IPAP	EPAP	Back up rate	Daily hours	Ventilation (yrs)	Airway Clearance	Compliant with LVR or MIE	Size of initial pneumothorax (cm)	Diagnostic modality
1	61	M	Amyotrophic Lateral Sclerosis	2.9	65	19	22	10	16	9	0.25	LVR	Yes	12	CXR AP
2	24	F	Limb Girdle Dystrophy	0.54	50	19	20	4	20	8	4	LVR QID	Yes (qid)	<0.5	CXR PA
3	53	F	CAP myopathy	0.6	42	16	24	5	16	22	2	LVR	No	<1	CT
4	26	F	Collagen 6 disease	0.41	27	13	20	4	20	8	6.5	LVR BID	Yes (bid)	1.3	CT
5	55	F	Myotonic Dystrophy 1	0.68	36	15	16	6	14	15	0.5	MIE + LVR	Yes	2.5	CXR PA, CT
6	64	F	Kyphoscoliosis and Ankylosing spondylitis	0.83	36	14	20	6	20	9.5	9	LVR	Yes	large (with adhesions)	CXR PA

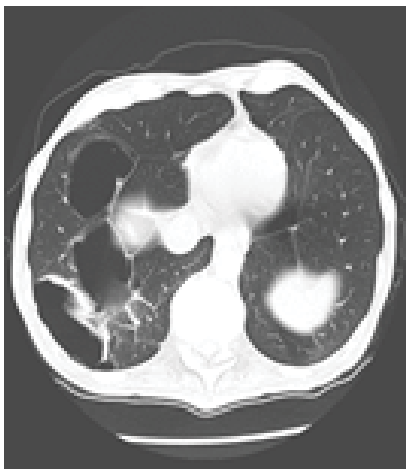

Figure 1B) *Follow up CT of ALS patient with residual blebs at right lung base*

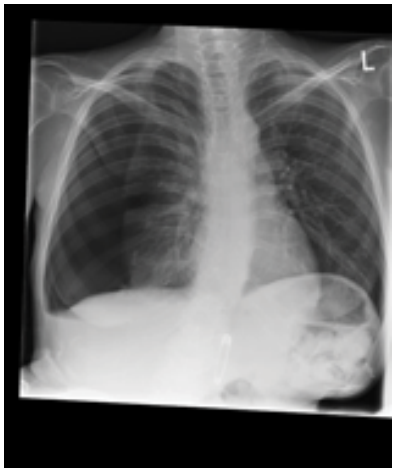

Figure 2B) Large *right pneumothorax in a patient with limb girdle muscular dystrophy, presenting 2 months after initial left pneumothorax*

Case 2

A 24-year-old female with limb girdle dystrophy presented with a sentinel event, a tiny left apical pneumothorax (Figure 2A) while using nocturnal NIV (BPAP 18/10) and LVR. This was accompanied by symptoms of increased dyspnea and orthopnea that had started abruptly in the days prior to assessment. No chest tube was placed at that time, but LVR was suspended. Two months later, the patient represented with a large right pneumothorax (Figure 2B). A chest tube was required. After hospital discharge one month later the patient had a large left pneumothorax. Surgical pleurodesis was considered but declined by the patient. Another pneumothorax on the right occurred two months later and pigtail chest tube was inserted. A doxycycline pleurodesis was performed on the right at that time. This procedure was well-tolerated although precautions were taken by admitting the patient to intensive care and intubating her prior to pleurodesis (Figure 2C). The patient was successfully extubated and no subsequent pneumothorax occurred.

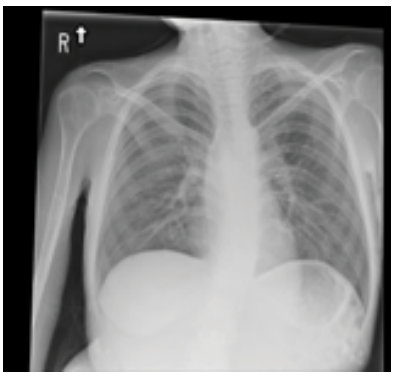

Figure 2A) *Radiograph demonstrating the sentinel event in patient with limb girdle muscular dystrophy, a left apical pneumothorax measuring <1 cm*

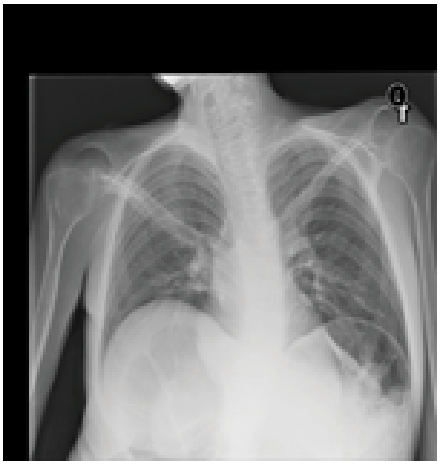

Figure 2C) *One-week post doxycycline pleurodesis of the right pleura in patient with limb girdle muscular dystrophy*

Case 3

A 53-year-old female with CAP myopathy and coexisting anorexia and anxiety disorder developed progressive respiratory failure with high level NIV dependence (22 hours per day). The patient had been prescribed but was not using lung volume recruitment. After a change in respiratory status, which was subsequently felt to be related to an aspiration event, the patient underwent a CT scan (Figure 3). The patient was found to have a <1 cm apical right pneumothorax and pneumomediastinum. Management was conservative; NIV was continued, with no chest tube drainage. There was no recurrence of pneumothorax in this case.

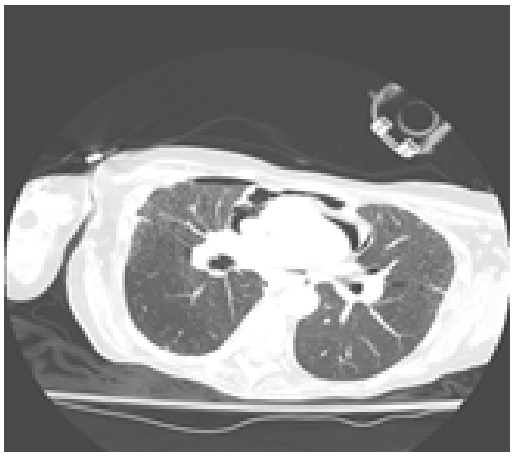

Figure 3) *CT scan at presentation with pneumomediastinum and small apical right pneumothorax in patient with CAP myopathy*

Case 4

A 24-year-old female with collagen 6 disease who had had a history of scoliosis and Harrington rod placement with multiple revisions. The patient was given a single trial of mouthpiece ventilation for persistent hypercapnia despite nocturnal ventilation and daytime dyspnea. Settings on a Legendaire ventilator were VT 600 mL, PEEP 0 cm H_2O, waveform sine, rate of 5 breaths per minute and I time of 25%. Pressure alarms were set at minimum 2 cm H_2O and maximum 38 cm H_2O. The patient experienced right-sided chest pain later in the evening and night after the mouthpiece ventilation trial. Upon presentation to the emergency department she had a small 1.3 cm right sided pneumothorax (Figure 4). An 8 French chest tube was initially used and then replaced with an 18 French chest tube due to persistent air leak. NIV settings were maintained. After eventual re-expansion of the lung and clamping of the chest tube it was withdrawn, however pneumothorax recurred within 6 hours, necessitating a third tube. The total duration of chest tube therapy was 62 days. The patient went on to use bi-level non-invasive positive airway pressure ventilation increasingly through the day and night time and did not re-trial mouthpiece ventilation subsequently.

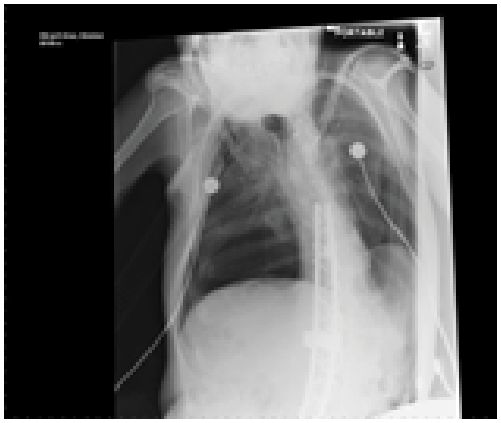

Figure 4) *Chest x-ray at presentation with right pneumothorax several hours after initial trial of mouthpiece ventilation in a patient with collagen-6 myopathy*

Case 5

A 55-year-old female with myotonic dystrophy type 1 had a history of recurrent aspiration and interstitial lung changes related to this. The patient had declined enteral nutrition with gastrostomy and so continued to maintain oral intake despite known aspiration risks. The patient was using nocturnal NIV 12 hours and periodically during the daytime as well as lung volume recruitment and MI-E for airway clearance. The patient presented with a large pneumothorax (Figure 5A) which was treated with a 10 French chest tube, with resolution and removal of chest tube in 16 days. During this time MI-E was held. The patient did experience a recurrence 7 months later after resuming her usual NIV settings, LVR and mouthpiece ventilation (Figure 5B). At this time both chemical and surgical pleurodesis were considered, but declined due to the patient's overall condition.

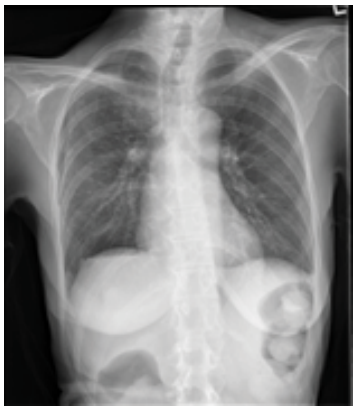

Figure 5A) *Chest x-ray at presentation of patient with myotonic dystrophy and right pneumothorax*

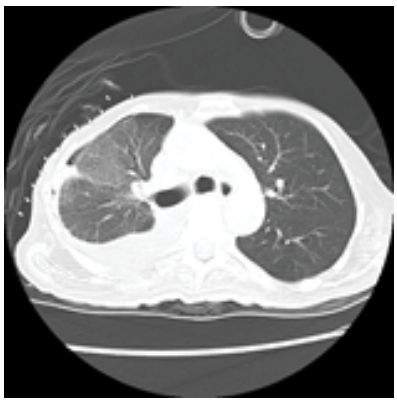

Figure 5B) *CT scan at time of recurrent right pneumothorax in a patient with myotonic dystrophy, demonstrating chronic right upper lobe inter- and intra-lobular septal thickening and right pleural effusion (autopsy investigation confirmed chronic aspiration pneumonitis)*

Case 6

A 64-year-old female had ankylosing spondylitis and kyphoscoliosis with Harrington rod placement. She had no known interstitial lung disease. This patient was using nocturnal NIV with daytime LVR airway clearance technique. The patient presented with a large right pneumothorax with adhesions (Figures 6A and 6B) and underwent a CT-guided chest tube placement of a 12 French catheter. This chest tube remained in for 27 days until pneumothorax was felt to be resolved. LVR was held during this time. The patient had a recurrence in four months' time after resuming usual care, including LVR. Pleurodesis was considered on both occasions but due to the high risk of respiratory failure, intubation and invasive ventilation that could follow an ARDS-like reaction to chemical pleurodesis, this was declined. The second pneumothorax resolved and the patient has remained stable for 2 years subsequently (Figure 6C).

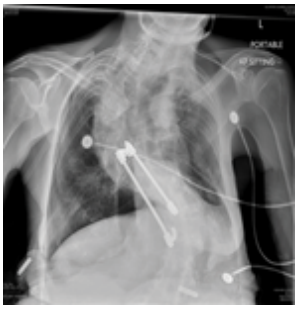

Figure 6A) *Chest x-ray at presentation with right pneumothorax in a patient with ankylosing spondylitis and kyphoscoliosis chest wall disorder. The diagnostic possibilities of the chest x-ray changes include possible large hiatal hernia. CT scan was performed to confirm and assess extent of pneumothorax*

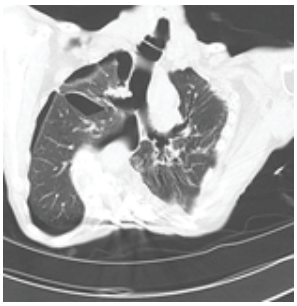

Figure 6B) *CT scan demonstrating large right pneumothorax in a patient with ankylosing spondylitis and kyphoscoliosis chest wall disorder, showing focal tethering of the superior segment of the right lower lobe*

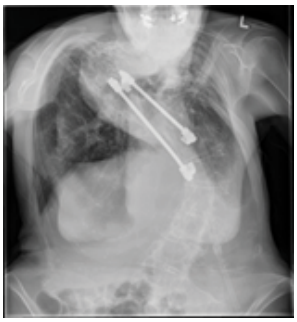

Figure 6C) *Chest x-ray documenting resolution of pneumothorax in a patient with ankylosing spondylitis and kyphoscoliosis chest wall disorder*

DISCUSSION

Pneumothorax occurs in chronically ventilated neuromuscular patients and those with chest wall restriction. This is the largest case-series to be reported to date.

Diagnostic imaging of pneumothorax in neuromuscular patients can be complicated by the chest wall abnormalities and scoliosis. CT was commonly used in addition to chest x-ray for diagnosis and chest tube management in this patient population. Of note, coexisting interstitial lung disease did not appear to be a predisposing factor to pneumothorax in these patients.

The majority of pneumothoraces occurred on the right side. Bilateral sequential pneumothorax was seen in some patients. Recurrence was common, occurring in 4 of 6 patients.

Pneumothorax during positive pressure ventilation is predominantly caused by barotrauma when lung compliance is low (6,7) for example in ARDS. It has been observed that high peak airway pressure may be associated with pneumothorax during invasive ventilation (8,9), and high PEEP has been variably associated with pneumothorax (10,11). A high tidal volume has been reported to cause pneumothorax (11) and animal studies support over-distension of the alveolar units as causative in pneumothorax (12,13). During chronic non-invasive ventilation, over-distension of regional alveolar units may be possible although steps are taken to minimize peak pressures and control volume.

Lung volume recruitment with a resuscitation bag or utilizing a volume-cycled ventilator to breath stack is typically performed to help a patient achieve an improved or normalized maximal insufflation capacity and peak cough flow (14-16). Airway clearance techniques have been reported to decrease rates of invasive tracheostomy and bronchoscopy or deep suctioning for secretion management (17) and use of these techniques may slow the decline in FVC and PCF over time (18). Our own clinical investigation of the lung volume recruitment maneuver using resuscitation bag and one-way valve has demonstrated that patients do not exceed 50 cm H_2O pressure (as measured during the recruitment breaths with open glottis and end-inspiration). Maximal insufflation-inexsufflation devices can provide better improvements in peak cough flow than other techniques (15). With MI-E the inspiratory and expiratory pressures are controlled. However, as with positive airway pressure ventilation, it is possible that during either LVR, breath-stacking using a volume-cycled ventilator or MI-E technique regional variability in the lung could cause areas of over-distension leading to pneumothorax.

It is interesting that pneumothorax was observed in these stable neuromuscular patients in whom lung compliance is assumed to be normal. Low BMI was seen in this patient group. Malnutrition has been associated with spontaneous pneumothorax in patients with anorexia nervosa (19). Decreased total lung protein may play a role in the pathophysiology of pneumothorax in a state of malnutrition as described in an animal model (20) and therefore may play a significant role in the development of pneumothorax in neuromuscular patients.

CONCLUSION

The management of pneumothorax in neuromuscular patients dependent on chronic non-invasive ventilation requires special consideration. Removing positive airway pressure ventilator support is not an option, and this is perhaps why early chest tube management and prolonged chest tube duration was seen in this group. Lung volume recruitment maneuvers or MI-E were suspended until pneumothorax was resolved, but with careful monitoring to ensure de-recruitment and retained airway secretions did not further compromise the patient. These maneuvers were typically restarted in 4-6 weeks following the pneumothorax event.

Talc pleurodesis has associated with an ARDS-type reaction, dyspnea and increased oxygen requirements in patients, although studies have been mixed as to the incidence of these side-effects (21,22). In patients with chronic respiratory insufficiency and NIV dependence, it is important to consider the risk of precipitating acute respiratory failure with pleurodesis. One patient in this series did undergo successful chemical pleurodesis with doxycycline in the ICU, without adverse sequelae. This patient did not experience recurrence on this side.

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