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Restoring Functional Status: A Long-Term Case Report of Severe Lung and Ventilatory Muscle Pump Dysfunction Involving Recurrent Bacterial Pneumonias

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ABSTRACT

Background and Purpose: Prolonged mechanical ventilation contributes to immobility and deconditioning making efforts to safely discontinue ventilator support desirable. This case report documents how implementing physical therapy treatment interventions, based on the *Guide to Physical Therapist Practice*, can help to restore a person's functional status even after multiple years of mechanical ventilation dependency. **Case Description:** A patient (female; aged 63 years) with severe restrictive and obstructive ventilatory impairment has survived 34 recurrent pneumonias involving 6 bacterial pathogens while being mechanically ventilated at home. A 3-year study was approved and informed consent obtained for a home exercise program of resistive extremity and inspiratory muscle training along with exercise reconditioning. Tolerable distances walked, maximal inspiratory and expiratory pressures, hours spent on versus off mechanical ventilation, activities performed within and around her home, and community excursions taken were charted. **Outcomes:** Daily time tolerated off the ventilator improved from less than one to 12 hours, distance walked in 6 minutes increased 33%, and maximal inspiratory and expiratory pressures improved 62% and 9.6% respectively. These improvements made out-of-home social excursions possible. **Discussion and Conclusions:** This patient's functional status improved following multiple physical therapy interventions dictated by the evaluation of initial physical therapy examination findings according to the *Guide to Physical Therapist Practice*. Long term mechanical ventilator dependency in the home environment did not exclude this patient from achieving clinically significant gains in functional status even when having severe restrictive and obstructive ventilator impairment.

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Key Words: restrictive and obstructive ventilatory impairment, pneumonia, inspiratory muscle training, ventilator weaning, functional status

INTRODUCTION AND PURPOSE

Individuals who are elderly, very young, or immunocompromised are predisposed to developing pneumonia,[1] the eighth leading cause of mortality in the United States.[2] This infectious process within the lungs is also a threat to individuals who are already diagnosed with a respiratory condition that is obstructive (eg, asthma) [3] or restrictive (eg, sarcoidosis)[4] in nature, particularly if they smoke[3-5] or are intubated and on mechanical ventilation.[6]

A variety of microbial pathogens may be involved in pneumonia including gram negative and gram positive bacteria, viruses, fungi, mycoplasmas, and parasites. Their preferred location within the lungs varies, and each microorganism has its own unique set of virulence factors that contribute to primary infection and secondary complications (eg, fibrosis). Of particular concern are bacteria classified as gram negative rods because of their production of endotoxin, exotoxins, and enzymes. For instance, *Pseudomonas aeruginosa* is particularly pathogenic because it produces an adherent and protective capsule, endotoxin, multiple enzymes, and exotoxin A which may be necrotizing to lung parenchyma. It is also inherently and adaptively resistant to a multitude of antibiotics.[7-9]

Patients with acute respiratory failure secondary to pathogenic causes that produce inadequate ventilatory function and/or gas exchange capabilities of their respiratory system, decreased respiratory drive, deficient cardiovascular function, and/or psychological duress may become dependent on mechanical ventilator support (ie, invasive or noninvasive methods) on a continuous (ie, 24 hours, daily) or discontinuous (ie, brief periods of ventilator muscle pump respite, daily) basis. This dependency may become permanent if the reason(s) for initiating mechanical ventilation (eg, increased work of breathing secondary to airway obstruction and pulmonary fibrosis) cannot be resolved.[10]

Sustaining the work of breathing necessary to support life, let alone functional activity, falls largely upon the status of the ventilatory muscles as a pump (VMP) and/or the lungs as a gas exchanging organ. Dysfunction or failure in either the VMP or the lungs are dependent upon multiple factors relating to the interplay between energy supplies (eg, nutritional substrates) and demands (eg, sepsis) as well as the loads to be overcome (eg, lung elasticity) and the neuromuscular competence (eg, VMP strength) in order to do so.[10-12] Functionally- based treatment interventions that collectively take these factors into account must consider the delicate balance between patient tolerance and respiratory insufficiency, especially when recurrent insults of pneumonia are superimposed.[10-12]

Strategies for discontinuing (ie, weaning) patients from prolonged (ie, > 24 hours) mechanical ventilator support and restoring functional status have been identified in the intensive care unit (ICU)[13,14] and long term acute care hospital (LTACH) settings.[15] To reverse the effects of immobility (eg, sedation-induced) and resultant deconditioning while on mechanical ventilator support, these treatment strategies often impose a prescribed challenge to the VMP[13] and/or provide extremity strengthening and progressive upright positioning with functional activity (eg, assisted ambulation) in mind.[13-15]

Recent clinical practice guidelines for recommending pulmonary rehabilitation (PR)[16,17] target a patient population that has primarily one variety of lung disease (eg, COPD) or another (eg, asthma). The degree to which a patient who has multiple pulmonary diseases of advanced severity may benefit from component parts of PR while requiring mechanical ventilator assistance is not known. The purpose of this case report is to describe the use of physical therapy interventions which, based on the *Guide to Physical Therapist Practice*, incorporated PR components to help improve the functional status of a patient with severe restrictive (ie, idiopathic scoliosis and pulmonary fibrosis), obstructive (ie, bronchiectasis), and restrictive/obstructive ventilatory impairment (ie, recurrent pneumonias) while on long-term mechanical ventilation at home.

CASE DESCRIPTION

Patient History

The patient of this case report is a 63-year-old female non-smoker (160.0 cm and 52.7 kg) who has severe idiopathic dextro-levo-scoliosis (Cobb angle of 90°-Figure 1) for which her spine was surgically fused at the age of 11. Since then as an adult, advanced multi-level disc degenerative changes and facet arthropathy, particularly at L4/L5 bilaterally, were identified by magnetic resonance imaging for the thoracic and lumbar spines.

By the age of 30 her breathing had become symptomatic and required nocturnal oxygen supplementation of 0.5 liters per minute via nasal cannula. At age 39, noninvasive ventilatory assistance was provided by a Pneumobelt system[18] (18 breaths/minute from 10 p.m. to 8 a.m. daily) secondary to chronic hypoventilation and cor pulmonale. Beginning with mechanical ventilator use (Puritan-Bennett 2800) on a nocturnal basis, the patient eventually became dependent on continuous mechanical ventilator use (settings of assist control at a respiratory rate of 12, a tidal volume of 450 cc, and an FiO_2 of 0.30) by age 46. This was the year when tracheostomy was performed to treat her respiratory distress secondary to chronic hypoventilation and corpulmonale with failure to thrive being evident (ie, a BMI of approximately 15 kg m^{-2}).

Figure 1. Representative PA and lateral plain-film radiographs.

Bronchiectasis, an obstructive ventilatory impairment (OVI) related to recurrent episodes of pneumonia causing pulmonary fibrosis, is superimposed on this patient's severe restrictive ventilatory impairment (RVI). Her most recent spirometry tests at age 56 reported an FEV_1 of 37% predicted and an FEV_1/FVC % of 90%. No pulmonary volumes were taken but her pulmonologist interpreted her flow-volume loop as suggesting very severe restrictive ventilatory impairment. Maximal inspiratory (PI max) and expiratory pressures (PE max) were markedly decreased at -19 cmH_2O and +15 cm H_2O, respectively. A 6 minute walk distance (6MWD) of 243.9 meters on room air was completed (no rest pauses taken) with SpO_2 dropping from 95% to 90% and heart rate increasing from 94 to 110 beats per minute.

A retrospective analysis of her medical record identified 4 documented diagnoses of respiratory failure and 34 confirmed or suspected episodes of pneumonia based on sputum cultures, signs and symptoms, and/or by chest radiographs. Six different gram-negative bacterial pathogens of increasing virulence caused 12 hospitalizations for a total of 70 days in the time span since having a tracheostomy performed in 1990. They are listed in Table 1 according to the year(s) in which they were isolated from her sputum to cause pneumonia and the number of hospital days, if any, that were required for effective medical treatment for each respiratory exacerbation.

Table 1. Documented Diagnoses of Respiratory Failure (n=4) or Bacterial Pneumonia (n=34) by Year

Year	Pathogen	Respiratory Exacerbations (n= 38)	Days Hospitalized (n=131)
1989	Respiratory Failure (no pathogens isolated)	Two	23
1990	Respiratory Failure (no pathogens isolated)	Two	38
1993	*Proteus mirabilis (Prm)*	One	0
1994	*Prm*	One	0
1998	*Pseudomonas aeruginosa (Pa)**	Three	0
	Pa	Two	0
2001	*Moraxella catarrhalis (Mc)*	One	11
2002	*Stenotrophomonas maltophilia (Sm*)-LLL**	Two	11
	Pa	Three	6
	Pa +Morganella morganii (Mm)	One	0
	Pasteurella multocida (Pam)	One	0
	*Pa**	One	0
2003	*Pa**	One	3
	Pa	Two	8
	Sm	One	0
2004	*Pa + Prm*	Two	7
	Pa	Three	6
	Pam	One	0
2005	*Pa*	Two	10
	Prm	One	0
2006	*Pa*	Two	2
	Pam	One	0
	Pa + Pam	One	3
2007	*Pa + Prm*	One	3

**Pa* refers to two phenotypes of this bacteria; **all bacterial pathogens produced a Right Lower Lobe (RLL) pneumonia unless designated as a Left Lower Lobe (LLL) pneumonia

With assistance provided by her husband, home treatments for mechanical ventilator support and bronchial hygiene purposes included use of the Vest and aerosolized nebulization of bronchodilators under the guidance of her pulmonologist and respiratory therapists.

Initial Physical Therapy Examination

Following hospitalization and treatment for her 21st episode of bacterial pneumonia, the patient was discharged home in August 2004, and a physical therapy treatment referral from her pulmonologist was received to provide inspiratory muscle training. An initial physical therapy examination was conducted in her bi-level home (one step entry and a staircase of 13 steps between floors). The patient reported spending her daily awake time, except for approximately one hour, in bed tethered to her mechanical ventilator. Although independent in feeding, bathing, toileting, and dressing activities, this patient relied upon assistive equipment in the bathroom (eg, tub transfer bench) and self-pacing (ie, 30-45 minutes at a time off the ventilator) with multiple rest pauses as necessary. Meal preparation, laundry, and other household chores were performed by her husband. Her medication list included digoxin, theophylline, zolpidem tartrate, mometasone furoate monohydrate, naproxen, and acetaminophen.

The patient was unable to perform a pelvic bridge and required minimal assistance to perform a sit-to-stand transfer from bed-to-chair and return. No orthostatic drop in blood pressure from 98/60 mmHg occurred with upright positioning with a pulse rate in the 70s along with a regular rhythm. Her breathing pattern at rest demonstrated an active expiratory phase and asymmetrical lateral costal expansion, decreased right compared to the left, without inspiratory paradox. Her respiratory rate was 12 breaths per minute. Breath sounds were generally tubular and diminished on the right as compared to vesicular on the left. Standing posture revealed a higher left shoulder and an uneven sacral base, left iliac crest higher than right. Her active range of motion for all 4 extremities equaled her active-assistive range. Hand held dynamometry force readings for muscle groups at the shoulders, elbows, hips, and knees were less than for women with advanced lung disease her age.[19] No dependent pedal edema was noted for either lower extremity. She demonstrated a positive Trendelenburg sign bilaterally in single limb support and ambulated 30 feet from her bed to the bathroom holding onto furniture or the wall without pause, pulse and respiratory rates remaining less than 100 beats and 20 breaths per minute, respectively. Pulse rhythm remained regular with pulse oximetry values on room air dropping from 98% to 92%.

Clinical Impression

Upon completion of her initial physical therapy examination, a list of physical therapy diagnoses (Table 2) involving all 4 of the preferred practice patterns (PPP) was determined according to the *Guide to Physical Therapy Practice*.[20] Documented evidence specific to her Cardiovascular and Pulmonary PPP identified deconditioning based on her prolonged immobility (Pattern B). The diagnosis of bronchiectasis, presence of excessive central air-

Table 2. Physical Therapy Diagnoses Determined from the Initial Physical Therapy Examination

Cardiovascular/Pulmonary Preferred Practice Pattern
Pattern B: Impaired Aerobic Capacity/Endurance Associated With Deconditioning
Pattern C: Impaired Ventilation, Respiration/Gas Exchange, and Aerobic Capacity/Endurance Associated With Airway Clearance Dysfunction
Pattern D: Impaired Aerobic Capacity/Endurance Associated With Cardiovascular Pump Dysfunction or Failure
Pattern E: Impaired Ventilation and Respiration/Gas Exchange Associated With Ventilatory Pump Dysfunction or Failure
Pattern F: Impaired Ventilation and Respiration/Gas Exchange Associated With Respiratory Failure
Musculoskeletal Preferred Practice Pattern
Pattern B: Impaired Posture
Pattern C: Impaired Muscle Performance
Neuromuscular Preferred Practice Pattern
Pattern A: Primary Prevention/Risk Reduction for Loss of Balance and Falling
Integumentary Preferred Practice Pattern
Pattern A: Primary Prevention/Risk Reduction for Integumentary Disorders
Pattern B: Impaired Integumentary Integrity Associated With Superficial Skin Involvement

way secretions, having a tracheostomy, and decreased PE max value placed attention to airway clearance dysfunction (Pattern C). Radiographic evidence of cardiomegaly and a 6MWD of 243.9 meters described impaired aerobic capacity/endurance related to the cardiovascular pump (Pattern D). Breathing pattern abnormalities along with decreased PI max and PE max values indicated ventilatory pump dysfunction (Pattern E). Arterial blood gas values at the time of intubation for mechanical ventilator support with subsequent tracheostomy and long term ventilator dependency linked her to impaired ventilation and respiration/gas exchange associated with respiratory failure (Pattern F).

Whether or not functional status might be restored in a person homebound on long-term mechanical ventilation with all 4 PPP impaired was a specific therapeutic outcome of interest. It was decided that IRB approval and informed consent would be obtained should this patient's documented case be eventually shared in the literature.

Phases of Physical Therapy Intervention

A chronological description of this patient's physical therapy treatment plan over 3 years is shared in 5 phases. Each phase reflects the clinical decisions made as the patient progressed or regressed in her functional status as she experienced recurrent bouts of pneumonia, any one of which could have caused her death.

Phase One of this patient's physical therapy treatment plan was directed at her level of deconditioning. Functional strengthening was initiated with the patient supine or side-lying in bed on an every other day, then daily basis as follows: resistive exercises using yellow then red elastic bands applied around the ankles for the hip flexors and abductors (3-5 repetitions up to a maximum of 10 for each leg); pelvic bridging for the hip extensors (3-5 repetitions up to a maximum of 15); sitting at the edge of her bed for the knee extensors (5-10 repetitions for each leg); and repetitive rising to stand and return to sitting from a bedside chair with the standby supervision of her spouse. Chair armrest use was employed until she could do so independently without the assistance from her upper extremities (beginning with 5 repetitions up to a maximum of 15). Progression was made to daily tasks within her home including meal preparation, laundry that necessitated the use of a flight of stairs to the laundry room (13 steps with one rail), light house-cleaning, along with increasing upright sitting time.

A walking route on the main floor of her home was mapped out for performance of 6-minute walk tests. This route was 29.9 meters long and served as a walk-way for ambulation exercise that employed intervals using a one-to-one walk: rest ratio (eg, walk 2 minutes then rest 2 minutes).

Phase Two of treatment intervention began the process of providing a strengthening stimulus to the VMP. The PI max and PE max pressures at the mouth were determined using a manometer as described by Black and Hyatt[21] as well as by Leith and Bradley[22] while the patient was seated. A Bugle dynamometer[23] was used to confirm measurement accuracy of the high pressure inspiratory force meter (model 4101 Boehringer Laboratories, Inc., PO Box 870, Norristown, PA) used for this case report. A threshold resistive inspiratory muscle training (IMT) device described by Sprague and associates[13] was set to provide a resistive load at the mouth for twice daily home use. The lowest resistance setting for the device of -7 cm H_2O was dialed in and progression was made by increasing the number of repetitions (from 3-5 up to a maximum of 25) on a very conservative basis as tolerated. Relying on the patient's self-report of no adverse symptoms, the next highest resistance setting would be dialed in thereafter as a strategy for applying progressive overload. Within 5 months, the patient was tolerating 25 repetitions at -11 cmH_2O resistance, twice daily. Whenever recovery from a pneumonia exacerbation occurred, PI max and PE max values were re-established with an average of 6 trials and/or the best of 6 trials recorded as noted in Table 3.

Also during this same phase, an attempt was made to target the diaphragm specifically for strength improvement instead of focusing alone on the group action of the inspiratory muscles that the threshold IMT device affords. A method for applying a resistive load to the diaphragm as illustrated by Kigin[24] and described by Block and colleagues[25] was added for one of the twice daily training sessions. The resistance amount began with a 2.3 kg weight placed over the patient's abdomen while supine and displaced upward with each full inspiratory excursion of a diaphragmatic breath to a tolerable limit of 10 repetitions (10 RM). The abdominal weights would be increased at intervals, 0.45 kg at a time, once the patient could comfortably achieve 20 repetitions without reporting any adverse symptoms.

Considering that any device that contacts the mouth has the potential to introduce pathogens to the upper respiratory tract, the threshold IMT device was cultured and found to be void of any harmful microbes. This confirmed the meticulous attention followed by the patient in effectively disinfecting the respiratory related equipment used. Nevertheless, the patient was vulnerable to pathogen exposure as evidenced, on 3 occasions, by having pneumonia caused by the microbe *Pasteurella multocida* that was suspected to come from the oropharyngeal secretions of her pet dog.[26] To eliminate one possible source of pathogen exposure, it was decided to target the diaphragm for strengthening only using free weights applied over the abdomen rather than including the use of the threshold IMT device at the mouth as well. This was the rationale behind Phase Three of the physical therapy treatment plan.

Late in the course of her inspiratory muscle training home program, Phase Four of the physical therapy treatment plan was implemented by adding supported upper extremity exercise for potential benefit in conditioning the VMP[27-29] while simultaneously providing an aerobic conditioning stimulus. A NuStep recumbent cycle ergometer was used for this purpose which permitted the necessary stabilization required for her scoliotic torso while unloading her lower extremities from weight-bearing. She was reporting to be experiencing non-radiating hip joint pain, bilaterally, that she attributed to her progressive ambulation. In the beginning, the ergometer was used with zero load for 3 sets of 1 minute intervals at 30 to 40 revolutions per minute (rpms) interspersed with 2 minutes rest (1:2 work-to-rest ratio) with the arms contributing ~25% toward each revolution as compared to the legs at ~ 75%, 5 times per week. Progression was made to 3 sets of 3 minute intervals at a 1:1 work to rest ratio at zero load, 30-40 rpms, arms/legs at ~ 50%/50% contribution per revolution, 5 times per week. However, the reciprocal action of the supported extremities contributed to painful symptoms (ie, bilateral lower lumbar pain radiating into each gluteal area; rated 3/10) secondary to her lumbar facet arthropathy and was therefore discontinued after completing 25 training sessions over a 7-week period. Subsequent symptom reduction was obtained with the application of 2-channel transcutaneous electrical nerve stimulation (ie, "continuous" @ 100 pps switched to "burst" mode at 1-10 pps and a pulse width of 250-300).

Once the IRB approved, 3-year study period was completed, the patient was encouraged to continue her performance of functional tasks in and around her home which necessitated at least twice daily use of her staircase and paced ambulation to her tolerance (Phase Five). Further IMT was not emphasized since she continued to comfortably tolerate up to 10 hours off the mechanical ventilator during waking hours on a daily basis. At the time of this writing, the patient reports that she has been able to maintain her functional status in and around her home.

Outcomes

Prior to the initiation of physical therapy treatment at the age of 60, the available data from the patient's medical chart review had her spending less than one hour on a daily basis while awake off mechanical ventilation, ambulating to tolerance a distance of 243.9 meters in 6 minutes while on room air, and displaying PI max and PE max pressures of -19 cm H_2O and + 15 cm H_2O, respectively in 2001 (Refer to Table 3).

Phase One of physical therapy intervention was geared toward reversing the state of deconditioning that was apparent and to improve her VMP dysfunction (Cardiovascular/Pulmonary Preferred Practice Patterns B and E in Table 2). Within the first 3 months of this year-long phase, the patient felt strong enough to host friends at her home for the first time in over one year. Other favorable changes that occurred during this phase included improvements in her PI max, 6MWD, and her daily time off mechanical ventilation by 15%, 20%, and 350%, respectively.

Phase Two of physical therapy intervention resulted in PI max, PE max, and time off mechanical ventilation improving by 61%, 39%, and 26%, respectively, although 6MWD declined by 9%. These changes occurred at a time period when two episodes of bacterial pneumonia took place, one of which required 6 days of hospitalization.

Phase Three of physical therapy intervention was complicated by 4 episodes of bacterial pneumonia necessitating two hospitalization periods totaling 5 days during this 8-month time period. Nevertheless, improvements of 85%, 58%, 22%, and 10% were realized in PI max, PE max, 6MWD, and time off mechanical ventilation, respectively.

Phase Four of physical therapy intervention did not permit a comparison of change in VMP performance and 6MWD (patient refusal) due to the development of acute low back pain during this 7-week period. On a positive note, daily time off mechanical ventilation remained unchanged from the previous phase.

Phase Five of physical therapy intervention spanned the final 19 weeks of this 3-year study period. The patient was not able to tolerate baseline testing at the start of this phase due to dyspnea related to her recovery from a recent episode of bacterial pneumonia that required 3 days of hospitalization.

Over the course of this 3-year study period, this patient with severe restrictive and obstructive lung disease demonstrated a consistent body weight and an ability to thrive despite enduring recurrent bouts of pneumonia while on mechanical ventilation. This represents a noteworthy gain, unrelated to fluid retention, from previously documented weight values which is significant since having a BMI < less than 21 kg m^{-2} is one predictor of increased mortality in patients with COPD.[30]

Table 3. Outcomes According to Phases of Physical Therapy Intervention over the Study Period

Date(s)	Physical Therapy Intervention Phases	MIP max (-cm H_2O)	MEP max	6MWD (+cm H_2O) (meters)	Daily Hours off Ventilator
7/25/97	None	-77	+142	237.8	14
1/18/01	None	-19	+15	243.9	<1
7/21/04 to 6/20/05	Phase 1: Functional Reconditioning Exercises in Bed and Bedside plus Threshold Inspiratory Muscle Training	-40 to -46	+90 to +76	274.4 to 328.7	2 to 9
6/21/05 to 10/13/05*	Phase 2: Threshold Inspiratory Muscle Training + Targeted Diaphragmatic Strengthening with Weights Over Abdomen	-46 to -74 (-72.0)	+76 to + 106 (+94.7)	328.7 to 298.8	9-10 to 12
2/01/06** to 10/07/06	Phase 3: Targeted Diaphragmatic Strengthening Only with Weights Over the Abdomen	-40 to -74 (-59.0)	+90 to +142 (+134.7)	298.8 to 365.9	9-10 to 10-11
1/17/07 to 3/10/07	Phase 4: Supported Upper Extremity Exercise Using Recumbent Cycle Ergometer	-64 (-57.3) to NT	+94 (+88.7) to NT	350.9 to NT	10-11 to 10-11
5/03/07 to 9/24/07	Phase 5: Homemaking Tasks as Work Hardening	NT to -62 (- 59.0)	NT to +114 (+92.0	NT to NT	10 to 10

Values in parentheses represent the mean of 6 trials; NT refers to "Not Tested." The time period between the * and ** denotes minimal PT intervention provided due to recovery from a bacterial pneumonia requiring hospitalization.

In summary, from the beginning of this 3-year study period to the end, Table 3 reveals the progress in functional status made by the patient. She improved her peak 6MWD to a total of 365.9 meters or an increase of 91.4 meters. Her exit PI max and PE max values of -62 cm H_2O and +114 cm H_2O remained 55% and 27% above her entry values at the time of the initial physician referral to begin physical therapy. Tolerable time off mechanical ventilation during waking hours improved from less than one hour to a peak of 12 hours with daily time consistently within 10 to 11 hours. Collectively, these gains translated into functional achievements of preparing meals, hosting dinner guests, doing laundry, gardening outdoors, attending festivals, and celebrating special events at favorite restaurants, to name a few.

DISCUSSION AND CONCLUSIONS

According to evidence-based guidelines, patients with moderate or severe COPD (ie, $FEV_1 < 50\%$ predicted) participating in PR programs that are community- or home-based, are likely to experience improvement in respiratory symptoms, quality of life, and 6MWD.[17] With an FEV_1 of 37% predicted, the patient in this case report would be a candidate to engage in PR intervention according to these guidelines, which often include exercise reconditioning and inspiratory muscle training components.

In a report by Menard-Rothe and colleagues[31] pertaining to functional ambulation for patients having end-stage emphysema, 28 females (mean age 63.4 years; mean FEV_1 of 22.7 % predicted) ambulated a mean distance of 231.1 meters in 6 minutes with an average time actually spent walking of 5.7 minutes. The patient in this Case Report achieved a personal best 6MWD of 365.9 meters, nonstop, and did so while on room air without displaying a SpO_2 desaturation response.

In the National Emphysema Treatment Trial (NETT) that involved 1,218 patients (mean age 67.1 years; 472 females) with advanced emphysema (mean FEV_1 of 26.9% predicted), the mean 6MWD was 348.2 meters with actual time spent walking versus resting unreported. Forty-two percent of these patients performed this test without supplemental oxygen and none were reported to have either co-existing restrictive lung disease or to require mechanical ventilation assistance as did our patient in this Case Report.[32]

A study by Salhi and associates[33] on 31 patients (mean age 57 years) having restrictive lung disease (11 with congenital kyphoscoliosis) reported a mean 6MWD of 118.9 meters with no mention of actual time spent walking versus resting. Patients with obstructive lung disease were excluded from this study. The patient in our Case Report has very severe restrictive lung disease secondary to kyphoscoliosis and pulmonary fibrotic changes due to recurrent pneumonias in addition to advanced obstructive lung disease. Despite her severely compromised pulmonary status that requires 10 to 12 hours of daily mechanical ventilatory assistance, she was able to improve her 6 MWD from 243.9 to 365.9 meters.

Ambulation distance in a 6-minute test is also known to be a predictor for mortality and morbidity, especially for COPD patients having tolerable distances less than 274 meters.[34,35]At the beginning of physical therapy intervention, this patient was below this critical level for ambulation and increased to greater distances over the next 3 years despite recurring insults of pneumonia. Her peak tolerable distance reached 365.9 meters, an increase of 91.4 meters. Clinically significant improvement in 6MWD is reported to be at least 54 meters.[36,37] This was an important accomplishment for our patient's functional status.

Finding the delicate balance point between VMP fatigue/failure and patient tolerance is of crucial importance when discontinuing (ie, weaning) mechanical ventilator support.[10] Diaphragmatic fatigue during the weaning process in patients with COPD is a key limiting factor.[38] Efforts to discontinue mechanical ventilation early and as safely as possible is advantageous for a plethora of reasons, one of which is to spare the diaphragm from undergoing disuse atrophy, particularly of the type IIa fast oxidative-glycolytic muscle fibers.[39] Whether applied at the mouth with a threshold loading device and/or by free weights over the abdomen to specifically target the diaphragm, inspiratory muscle resistance during Phases Two and Three of physical therapy intervention for this patient contributed to improved VMP competence that translated to increases in daily time tolerated off mechanical ventilation.

The improvement realized in the daily time tolerated off mechanical ventilation impacted favorably on this patient's life quality as well as her functional status. The extent to which this improvement was made possible is likely multifactorial in nature, including improved ventilator muscle pump competence,[12,13,28,29,40,41] desensitization to dyspnea,[15,28,35,37,38,40,41] and improved functional reconditioning.[15,28,29,35,37,38,40,41] As a consequence, this patient was now able to enjoy day trips by car to nearby cities, attend birthday and anniversary dinners at favorite restaurants, entertain holiday guests, and be present at special family celebrations along with her supportive husband.

Shortcomings of this case report include not using inventories for documenting changes in life-quality or dyspnea index and not determining PI max and PE max values at the beginning of each of the 5 phases of physical therapy intervention due to the patient's subjective report of breathing discomfort. In further reflection, it would have been valuable to have quantified the 10 RM weight-load targeted to the diaphragm with that of the 10 RM for the threshold IMT device.

Insight to be gained through future inquiry (eg, controlled, randomized clinical trial in patients having OVI and/or RVI) would be to determine the extent to which the unconventional use of concentric and eccentric strength training of the diaphragm using resistance of free weights applied over the abdomen may have on conditioning of the VMP and functional status. Targeting the diaphragm with a strengthening prescription using free weights applied over the abdomen may be an effective alternative to applying inspiratory resistance at the mouth, thereby eliminating the potential risk of transferring pathogens from a mouthpiece to the respiratory tract.

SUMMARY

The patient in this Case Report has survived 34 confirmed or suspected episodes of bacterial pneumonia from 6 different pathogens of increasing virulence superimposed upon medical diagnoses of bronchiectasis, pulmonary fibrosis, pulmonary hypertension, cor pulmonale, and severe idiopathic scoliosis. These conditions contributed to impairments spanning all 4 PPPs from the *Guide to Physical Therapist Practice* in the span of 17 years since her tracheostomy. She has managed to improve her functional status and enhance her opportunities for socialization events within and outside her home while on long term mechanical ventilation.

Early detection of lower respiratory tract infections, promptness in specific treatment intervention, a supportive spouse, meticulous attention to respiratory muscle rest therapy, bronchial hygiene practices (eg, the Vest, hand-held nebulization treatments, disinfection of airway interface equipment, frequent hand-washing), and exceptional patient compliance with prescribed physical therapy plans of care (eg, threshold inspiratory muscle training, targeted resistive exercise to the diaphragm, progressive upright activity challenge including ambulation over level surfaces and stairs, extremity strengthening, and aerobic exercise reconditioning) have collectively made this possible.

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