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Sonographic assessment of mean pulmonary artery pressure and diaphragmatic excursion in chronic respiratory failure patient after using home non-invasive positive pressure ventilation

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Abstract

Background Non-invasive positive pressure ventilation (NIPPV) has emerged as a recognized and effective long-term therapeutic approach for individuals suffering from chronic respiratory insufficiency resulting from various diverse disorders. Nevertheless, providing home non-invasive positive pressure ventilation encounters several challenges, including compliance, training inadequacies, and limited resources. Therefore, the objective of this study was to observe the impact of home non-invasive positive pressure ventilation on patients with chronic respiratory failure, specifically focusing on its effects on the mean pulmonary artery pressure, diaphragmatic excursion, and associated complications.

Results The study included a total of 48 patients, consisting of 26 males (54.1%) and 22 females (45.9%). The baseline mean pulmonary artery pressure (PAP) was found to be 39.79 ± 7.51. Additionally, the baseline diaphragmatic excursion in quiet breathing was measured to be 1.80 ± 0.38 cm, while the baseline diaphragmatic excursion in deep breathing was recorded as 4.35 ± 0.99 cm. Following the implementation of home non-invasive ventilation, specifically bilevel-positive airway pressure or continuous positive airway pressure, significant improvements were observed in the aforementioned parameters. The most commonly reported complications among the patients included skin ulcers and aerophagia. Furthermore, mortality rate of 6.3% was observed.

Conclusions The use of home non-invasive positive pressure ventilation has been shown to be a successful and sustainable therapeutic approach for persons suffering from chronic respiratory failure. This therapy method has significantly improved PAP, diaphragm force, and overall mortality rates. The use of ultrasonography to evaluate pulmonary pressure or diaphragmatic excursion is considered a beneficial and straightforward approach for the follow-up of subjects with chronic respiratory failure.

Keywords Chronic respiratory failure, Mean pulmonary artery pressure, Diaphragm excursion

Background

Studies have shown conflicting findings regarding the effects of home-based non-invasive mechanical ventilation (NIV) on pulmonary function, exacerbations, quality of life, and survival in patients with hypercapnic chronic respiratory failure (HCRF) [1, 2]. Subsequent have shown

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improved outcomes in cachexia, pulmonary function, ventilatory parameters, and overall quality of life [3, 4]. Particularly concerning is the increase in people suffering from severe pulmonary hypertension (PH) brought on by alveolar hypoventilation [5, 6]. Hypoxia may lead to an increase in pulmonary arterial pressure (PAP); however, studies have shown significant interindividual variability in acute pulmonary vasoconstriction [7]. Though lung vasoconstriction is influenced by acidosis and carbon dioxide (CO2), all individuals with hypoventilation and hypercapnia may be experienced with PH [8, 9]. During an autopsy investigation, it was shown that persons classified as obese had a notable prevalence of pulmonary arterial and venous hypertension [10].

Furthermore, the precise impact of non-invasive ventilation (NIV) on the augmentation of alveolar ventilation remains uncertain, particularly during respiration. Hill et al. [11] (1993) were the first to propose a multifactorial mechanism for that phenomenon, which involves three distinct pathways. These pathways include the resensitization of respiratory centers to reduce hypercapnia, the enhancement of lung mechanics through increased lung compliance, and the improvement of respiratory muscle strength achieved through relaxation during the night period with the aid of respiratory support, thereby relieving the burden on the pulmonary musculature [11].

The measurement of transdiaphragmatic pressure through the esophagus is widely regarded as the most reliable method for evaluating diaphragm function. However, this process is invasive and not universally available in all healthcare facilities. Pulmonary function tests like the forced vital capacity (FVC) and maximum inspiratory pressure (MIP) examinations are easy and non-invasive but fail to be highly sensitive [12]. Diaphragmatic ultrasonography has significantly transformed the assessment of diaphragm function and morphology in real time due to its non-invasive nature, reproducibility, and ability to be performed at the bedside without any contraindications [13]. Consequently, it is possible to monitor patients without exposure to radiation [12, 13].

Methods

Aim of this study

In the present study, several individuals with HCRF were evaluated using transthoracic echocardiography and diaphragmatic ultrasonography to determine the effects of home NIV on PAP and respiratory muscle performance.

A prospective cohort research was conducted between June 2020 and May 2022 at the Chest Department of Zagazig University Hospitals. The Chest Department Administrative Council and Zagazig University Institutional Review Board (ZU-IRB no. 6193/2–6-2020) approved the study and its procedures. Informed written consent was collected from all participants.There were 48 cases of chronic respiratory failure (CRF), newly commencing home NPPV as a long-term treatment according to the Swiss Society of Pulmonology 2020 Recommendations for Long-Term Mechanical Ventilation [14].

Inclusion criteria

Individuals with CRF who were prescribed home non-invasive positive pressure ventilation (NPPV) as a long-term treatment according to guidelines [14] as follows:

Obstructive lung diseases Patients with stable severe chronic obstructive pulmonary disease (COPD) who are chronically hypercapnic(PaCO2 > 52.5 mm Hg). Also if hypercapnia (PaCO2 > 52.5 mmHg) continues for two to four weeks following an acute exacerbation of hypercapnic respiratory failure (AEHRF) [15].

Obesity hypoventilation syndrome (OHS) Within 3 months after an OHS-related AEHRF, a native sleep analysis must be performed. NIV was recommended when obstructive sleep apnea (OSA) was mild, moderate, or absent. A trial of continuous positive airway pressure (CPAP) is required when OSA is severe. When the arterial blood gas (ABG) confirmed the clinical suspicion of OHS in stable patients, a sleep investigation was conducted. If there was severe OSA, CPAP was the first-line treatment.

Sleep-related breathing disorders CPAP or bilevel inspiratory positive airway pressure (BIPAP) may be utilized based on the presence of obstructive sleep apnea, tolerance, and/or the need for elevated pressures to manage respiratory episodes.

Restrictive respiratory diseases other than obesity hypoventilation and neuromuscular disease (NMD) Long term NIV is initiated if there is daytime hypercapnia (PaCO2 ≥ 45 mmHg) or nocturnal hypoventilation, as determined by the American Academy of Sleep Medicine (AASM) guidelines, is defined as PCO2 ≥ 55 mmHg for more than 10 min, or an elevation in PCO2 ≥ 10 mmHg from the value when awake, and supine to a value over 50 mmHg for more than 10 min [16].

Exclusion criteria

Patients < 18 years old, with bulbar weakness,cognitive impairment,and/or uncooperative patients, uncontrolled comorbidities, and patients who failed to complete the study.

Study protocol

Reviewing and recording the following data of all studied patients:

Medical history, chest X-ray (posteroanterior or anteroposterior view); blood gases analysis; laboratory testing which includes complete blood count (CBCs), liver and kidney function assessments, and thyroid function tests; previous electrocardiography and echocardiography reports; and home NPPV (recording: type of the device, mode, setting, whether associated with oxygen supply, and usage hours per day).

All patients were evaluated for the following:

- Follow-up of sonographic measurements at baseline, 1, 3, 6, and 12 months, including follow-up by mean PAP (mean pulmonary artery pressure) by echocardiography probe [17, 18]
- Ultrasound assessment of DE (diaphragmatic excursion) at baseline, 1, 3, 6, and 12 months of the study in quiet and deep breathing.
- Using a Sonoscape SSI 4000 ultrasound system (China) (using 2 probes of same device echo probe for mean PAP and curvilinear probe for DE), participants were evaluated with a semi-recumbent position to assess the right diaphragm.

Statistical analysis

IBM SPSS v20.0 (IBM Corp., Armonk, New York, USA) was used to analyze the data that was entered into the computer. Utilizing numbers and percentages to represent qualitative data. Quantitative data were characterized by range (minimum and maximum), mean, standard deviation, median, and interquartile range (IQR). The significance of outcomes was determined using the 5% significance threshold. Utilized tests were chi-square analyses. Monte Carlo adjustment for chi-square was used to compare groups of categorical variables when more than 20% of the cells have an anticipated count < 5. Comparing two time periods required the use of paired *t*-tests.

Results

The current study included a total of 48 adults: a total of 26 males (54.1%) and 22 females (45.9%) with a mean ± SD of age 52.94 ± 13.39 years. Mean ± SD of body mass index (BMI) 31.37 ± 7.48 kg/m^2 were classified into five groups: 31.2% overlap syndrome (COPD and OSAS), 29.2% had OHS, 12.5% had COPD, 18.8% had restrictive thoracic diseases (RTD), (kyphoscoliosis-post-TB-thoracic cage), and 8.3% had neuromuscular diseases (NMDs), (myasthenia gravis-post-polio-chronic inflammatory-demyelinating polyradiculoneuropathy (CIDP).

During the study, eight patients left due to intolerance to NIV therapy, and three died as follows: in the third and sixth months of the study, two and three patients left due to intolerance to NIV therapy, respectively, while one patient died. In the 12th month, three new patients left due to intolerance, and two patients died. Accordingly, the total number of patients who left the study was 11.

Table 1 shows the baseline subjects' features and anthropometric measurements, mean PAP, DE in quiet and deep breathing, type of device used for NIV, and oxygen need. CPAP was used in 20% of overlap syndrome patients, 35.7% of OHS patients, and 11.1% of RTD patients, while BIPAP used in 80% of overlap syndrome patients, 64.3% of OHS patients, 100% of COPD patients, 88.9% of RTD patients, and 100% of NMD patients. Oxygen was needed in 66.7% of COPD patients, 33.3% of RTD, and 50% of NMD.

Table 2 depicts the values of mean PAP, showing a statistically significant decrease in mean PAP at different periods in overlap syndrome, OHS, COPD (at 3, 6, and 12 months), and RTD with p-value < 0.05.

By the end of the study, after 1 year of NIV therapy, the lowest mean PAP achieved in NMD patients mean ± SD 22.50 ± 3.54 mmHg while the highest in COPD patients mean ± SD 32.60 ± 8.71 mmHg.

Tables 3 and 4 illustrate the diaphragmatic excursion measurements in quiet and deep breathing, clarifying that DE in quiet breathing was significantly increasing in overlap syndrome, OHS, COPD, RTD, and NMDs at 3 and 12 months at different periods, and DE in deep breathing was significantly increasing in overlap syndrome at 3, 6, and 12 months and OHS, COPD, RTD, and NMDs at 6 and 12 months with p-value < 0.05.

Following 1 year of NIV therapy, overlap syndrome patients demonstrated the highest DE during both quiet breathing (mean ± SD 2.74 ± 0.32 cm) and deep breathing (mean ± SD 5.75 ± 0.55 cm). Conversely, patients with NMD achieved the lowest DE during both quiet breathing (mean ± SD 1.90 ± 0.14 cm) and deep breathing (mean ± SD 3.95 ± 0.354 cm) after undergoing NIV therapy.

Table 5 shows that most common complications notified were skin ulcers and aerophagia, with the following frequencies: 86.7% in overlap syndrome patients, 85.7% in OHS, 100.0% in COPD, 77.8% in RTD, and 100.0% in NMD. The mortality presented in 6.3% of patients (three

Table 1 Baseline subjects' features and anthropometric measurements, mean PAP, DE in quiet and deep breathing, type of device used for NIV, and oxygen need

Variables	Min.–max	Mean ± SD		Median (IQR)	
Age (years)	22.0–75.0	52.94 ± 13.39		55.0 (46.0–64.0)	
Male	Number	= 28		58.3%	
Female	Number	= 20		41.7%	
BMI (kg/m^2)	16.0–46.0	31.37 ± 7.48		32.0 (25.85–36.0)	
Pao^2 (mmHg)	38.0–60.0	53.41 ± 4.03		54.0 (52.0–56.0)	
Mean PAP (mmHg)	27.0–55.0	39.79 ± 7.51		40.0 (35.0–44.0)	
DE in quiet breathing (cm)	0.90–2.50	1.80 ± 0.38		1.80 (1.50–2.10)	
DE in deep breathing (cm)	2.50–6.30	4.35 ± 0.99		4.20 (3.60–5.20)	
Chronic respiratory failure on home NPPV	**Overlap syndrome**	**OHS**	**COPD**	**RTD**	**NMD**
Number (percent)	15 (31.2%)	14 (29.2%)	6 (12.5%)	9 (18.8%)	4 (8.3%)
Type of device					
CPAP n(%)	3 (20%)	5 (35.7%)	0 (0%)	1 (11.1%)	0 (0%)
BIPAP n(%)	12 (80%)	9 (64.3%)	6 (100%)	8 (88.9%)	4 (100%)
Oxygen need					
Yes n(%)	0 (0%)	0 (0%)	4 (66.7%)	3 (33.3%)	2 (50%)
No n(%)	15 (100%)	14 (100%)	2 (33.3%)	6 (66.7%)	2 (50%)

Table 2 Baseline and follow-up mean PAP analysis among studied patients with different etiologies of chronic respiratory failure

Mean PAP (mmHg)	Baseline	1st month	3rd month	6th month	12th month
Overlap syndrome	*n* = 15	*n* = 15	*n* = 14	*n* = 12	*n* = 11
Min.–max	30.0–55.0	25.0–55.0	25.0–52.0	20.0–52.0	20.0–35.0
Mean ± SD	39.93 ± 8.04	37.40 ± 9.44	34.36 ± 8.35	30.42 ± 8.60	24.64 ± 4.50
^{t}p		< 0.001*	< 0.001*	< 0.001*	< 0.001*
OHS	*n* = 14	*n* = 14	*n* = 14	*n* = 12	*n* = 11
Min.–max	28.0–55.0	26.0–54.0	26.0–50.0	24.0–50.0	22.0–50.0
Mean ± SD	39.43 ± 8.24	37.36 ± 8.71	35.14 ± 8.31	32.08 ± 9.41	28.18 ± 8.05
^{t}p		0.001*	< 0.001*	< 0.001*	< 0.001*
COPD	*n* = 6	*n* = 6	*n* = 5	*n* = 5	*n* = 5
Min.–max	35.0–45.0	33.0–45.0	30.0–45.0	28.0–45.0	24.0–46.0
Mean ± SD	39.50 ± 3.94	38.83 ± 4.45	35.40 ± 6.19	34.60 ± 6.58	32.60 ± 8.71
^{t}p		0.175	0.021*	0.020*	0.037*
RTD	*n* = 9	*n* = 9	*n* = 9	*n* = 9	*n* = 8
Min.–max	35.0–55.0	32.0–55.0	30.0–53.0	26.0–53.0	22.0–50.0
Mean ± SD	43.67 ± 6.38	41.33 ± 6.91	38.33 ± 7.04	34.78 ± 8.04	29.88 ± 8.82
^{t}p		0.002*	< 0.001*	< 0.001*	< 0.001*
NMDs	*n* = 4	*n* = 4	*n* = 4	*n* = 4	*n* = 2
Min.–max	27.0–40.0	27.0–38.0	25.0–40.0	22.0–40.0	20.0–25.0
Mean ± SD	32.25 ± 5.56	31.0 ± 5.23	30.0 ± 6.78	28.0 ± 8.12	22.50 ± 3.54
^{t}p		0.194	0.078	0.065	0.105

SD standard deviation, *t* paired *t*-test, *p p*-value for comparing between baseline and each other periods

* Statistically significant at $p \leq 0.05$

patients distributed as follows: one overlap syndrome patient, one OHS patient, and one NMD patient). Time spent on home NIV was best reported in OHS patients compared to other groups.

Discussion

The study aimed to evaluate the efficacy of home NIV in patients with CRF, specifically comparing CPAP and BIPAP treatments. The primary outcomes of interest

Table 3 Baseline and follow-up diaphragmatic excursion in quiet breathing among studied patients with different etiologies of chronic respiratory failure

Quiet breathing DE (cm)	Baseline	1st month	3rd month	6th month	12th month
Overlap syndrome	*n* = 15	*n* = 15	*n* = 14	*n* = 12	*n* = 11
Min.–max	1.25–2.40	1.20–2.50	1.40–2.80	1.50–3.0	2.40–3.30
Mean ± SD	1.81 ± 0.37	1.90 ± 0.42	2.11 ± 0.42	2.39 ± 0.40	2.74 ± 0.32
^{t}p		0.048*	0.001*	< 0.001*	< 0.001*
OHS	*n* = 14	*n* = 14	*n* = 14	*n* = 12)	*n* = 11
Min.–max	1.40–2.50	1.40–2.50	1.50–2.60	1.40–2.80	1.50–3.30
Mean ± SD	1.96 ± 0.35	2.04 ± 0.36	2.14 ± 0.37	2.22 ± 0.43	2.62 ± 0.50
^{t}p		0.003*	< 0.001*	< 0.001*	< 0.001*
COPD	*n* = 6	*n* = 6	*n* = 5	*n* = 5	*n* = 5
Min.–max	1.50–2.10	1.70–2.20	1.70–2.30	2.0–2.50	2.50–3.0
Mean ± SD	1.78 ± 0.23	1.93 ± 0.20	2.04 ± 0.23	2.34 ± 0.21	2.72 ± 0.19
^{t}p		0.007*	0.004*	0.004*	< 0.001*
RTD	*n* = 9)	*n* = 9	*n* = 9	*n* = 9	*n* = 8
Min.–max	1.50–2.20	1.50–2.40	1.60–2.40	1.60–2.50	1.70–2.70
Mean ± SD	1.87 ± 0.26	1.99 ± 0.31	2.08 ± 0.25	2.24 ± 0.28	2.39 ± 0.30
^{t}p		0.010*	0.001*	< 0.001*	< 0.001*
NMDs	*n* = 4	*n* = 4	*n* = 4	*n* = 4)	*n* = 2
Min.–max	0.90–1.30	0.80–1.40	1.30–1.60	1.10–2.0	1.80–2.0
Mean ± SD	1.13 ± 0.17	1.18 ± 0.26	1.45 ± 0.13	1.50 ± 0.39	1.90 ± 0.14
^{t}p		0.391	0.001*	0.072	0.049*

*Statistically significant at $p \leq 0.05$

Table 4 Baseline and follow-up diaphragmatic excursion in deep breathing among studied patients with different etiologies of chronic respiratory failure

Deep breathing DE (cm)	Baseline	1st month	3rd month	6th month	12th month
Overlap syndrome	*n* = 15	*n* = 15	*n* = 14	*n* = 12	*n* = 11
Min.–max	2.90–6.30	3.0–6.30	3.0–6.50	3.10–6.50	4.80–6.50
Mean ± SD	4.65 ± 1.04	4.70 ± 1.09	5.03 ± 1.04	5.39 ± 0.95	5.75 ± 0.55
^{t}p		0.318	< 0.001*	< 0.001*	0.001*
OHS	*n* = 14	*n* = 14	*n* = 14	*n* = 12	*n* = 11
Min.–max	3.40–5.80	3.50–5.80	3.50–6.0	3.50–6.10	3.80–6.30
Mean ± SD	4.43 ± 0.74	4.53 ± 0.74	4.69 ± 0.77	4.85 ± 0.82	5.26 ± 0.72
^{t}p		< 0.001*	< 0.001*	< 0.001*	< 0.001*
COPD	*n* = 6	*n* = 6	*n* = 5	*n* = 5	*n* = 5
Min.–max	3.40–4.80	3.60–5.0	3.60–5.30	4.0–5.50	4.0–6.10
Mean ± SD	3.90 ± 0.52	4.07 ± 0.55	4.38 ± 0.62	4.66 ± 0.61	5.16 ± 0.82
^{t}p		0.004*	0.012*	0.006*	0.003*
RTD	*n* = 9	*n* = 9	*n* = 9	*n* = 9	*n* = 8
Min.–max	2.50–6.0	2.50–6.0	2.60–6.10	2.60–6.20	2.70–6.50
Mean ± SD	4.68 ± 1.12	4.86 ± 1.14	4.96 ± 1.06	5.13 ± 1.09	5.30 ± 1.14
^{t}p		0.014*	0.012*	0.003*	0.002*
NMDs	*n* = 4	*n* = 4	*n* = 4	*n* = 4	*n* = 2
Min.–max	2.50–3.30	2.30–3.50	2.80–3.80	2.50–4.20	3.70–4.20
Mean ± SD	2.85 ± 0.34	2.90 ± 0.50	3.25 ± 0.44	3.23 ± 0.71	3.95 ± 0.35
^{t}p		0.604	0.006*	0.166	0.037*

*Statistically significant at $p \leq 0.05$

Table 5 Complications, mortality, and time spent on home NPPV among the studied patients for 12-month duration ($n=48$)

Complications	Chronic respiratory failure on home NPPV											
	Total	Overlap syndrome (COPD and OSAS) ($n=15$)		OHS ($n=14$)		COPD ($n=6$)		RTD ($n=9$)		NMDs ($n=4$)		
		No	%	No	%	No	%	No	%	No	%	
No	6 (12.5%)	2	13.3	2	14.3	0	0.0	2	22.2	0	0.0	
Yes	42 (87.5%)	13	86.7	12	85.7	6	100.0	7	77.8	4	100.0	
Aerophagia	14 (29.2%)	5	33.3	4	28.6	1	16.7	2	22.2	2	50.0	
Rhinitis	11 (22.9%)	3	20.0	3	21.4	1	16.7	2	22.2	2	50.0	
Conjuctivitis	6 (12.5%)	0	0.0	2	14.3	2	33.3	1	11.1	1	25.0	
Skin ulcer	19 (39.6%)	5	33.3	6	42.9	2	33.3	4	44.4	2	50.0	
Air leak	10 (20.8%)	3	20.0	3	21.4	2	33.3	2	22.2	0	0.0	
Claustrophobia	4 (8.3%)	1	6.7	1	7.1	1	16.7	1	11.1	0	0.0	
Mortality												
No	45 (93.7%)	14	93.3	13	92.9	6	100.0	9	100.0	3	75.0	
Yes	3 (6.3%)	1	6.7	1	7.1	0	0	0	0	1	25	$\chi^2=3.195$ $^{MC}p=0.598$
Time spent on NIV(Mean±SD)		5.93 ± 1.33 h		6.14 ± 1.46 h		5.67 ± 1.51 h		6.11 ± 1.27 h		5.75 ± 2.06 h		

were the impact of these interventions on PAP, diaphragmatic excursion, and the occurrence of complications. Concerning PAP, our findings indicate no discernible differences among the patients under investigation. However, when examining the mean PAP over various time intervals, significant enhancements were observed in different patient groups, except for those with COPD in the first month. Notably, individuals with NMD exhibited the lowest mean PAP, while those with RTD displayed the highest mean PAP.

This finding aligns with the research conducted by Held et al. [19], which explored the correlation between hypoventilation-induced pulmonary hypertension and exercise capacity, as well as the hemodynamic and functional changes observed during non-invasive ventilation. The patients in the study exhibited a PAP of 49 ± 13 mmHg. However, following a 3-month period of non-invasive ventilation, a significant reduction in PAP was observed, with a mean value of 31 ± 9 mmHg (− 18 mmHg; $p < 0.001$).

Kauppert et al. [20] evaluated clinically stable OHS subjects (NPPV lasting 3 months) by performing right-heart catheterization, echocardiography, and serum biomarker evaluations. They concluded that daytime sleepiness is associated with the severity of PH and is inversely related to daily NPPV usage [20].

Schönhofer et al. [21] studied two patient groups (individuals with restrictive lung disease and COPD) at baseline and after 1 year of NIV, showing a significant decrease in mean PAP after 1 year of NIV in subjects with restrictive lung disease. This decrease may be attributed to the advantage conferred by NIV in subjects with CRF due to restrictive lung disease. However, improvement in pulmonary hypertension was unreported in individuals with COPD following a year of NIV.

There was a difference between the studied patients in the DE in quiet breathing. The lowest DE was in NMD patients from baseline and in all studied periods compared to other patient groups. With follow-up DE in quiet breathing analysis after NIV therapy among studied patients, we observed a significant increase at different periods except in NMDs (at 1 and 6 months), with the highest DE in quiet breathing presented in OHS patients at (the 1 and 3 months) and in overlap syndrome patients at (6 and 12 months).

Nevertheless, the examination of diaphragmatic excursion during deep breathing revealed variations among the patients under investigation, except at the 12 months. The analysis of diaphragmatic excursion during deep breathing after NIV demonstrated that the patients experienced improvements in different time intervals. The patients with RTD exhibited the highest diaphragmatic excursion during deep breathing at 1- and 3-month intervals, while those with overlap syndrome showed the highest excursion at 6- and 12-month intervals. Conversely, patients with NMD consistently displayed the lowest diaphragmatic excursion during deep breathing across all studied periods.

Limited research has been conducted on NIV's impact on diaphragm excursion during quiet and deep breathing. In their study, Hernandez-Voth et al. [22] evaluated 30 patients with extremely severe COPD and assessed them at baseline and after 12 months for treatment adherence, quality of life, pulmonary function assessments, and diaphragmatic ultrasonography. They assessed diaphragm thickness (without excursion) at FRC (functional residual capacity), TLC (total lung capacity), and thickness fraction diaphragmatic ultrasonography verified an elevation in the thickening fraction of 14% ($p = 0.002$).

The mechanism by which NIV enhances the function of respiratory muscles remains uncertain. The studies conducted by Nickol et al. in 2005 and 2008 [23, 24] found no evidence of enhanced respiratory muscle strength based on both pulmonary function assessments and esophageal measurements of transdiaphragmatic pressure. The authors attributed the improvement in alveolar ventilation to the compensatory mechanism for elevated intrinsic end-expiratory pressure resulting from gas trapping, as well as the heightened responsiveness of the bulbar respiratory center to the hypercapnic response. Furthermore, only two out of ten patients had regular NIV utilization that exceeded 4 h per day when they assessed individuals 3 months after initiating home NIV.

In this study, DE was assessed at 1, 3, 6, and 12 months following home NIV utilization, with participants completing an average of more than 5.98 ± 1.39 h per day. The longer follow-up and more NIV treatment each day probably contributed to our reported pulmonary muscle strengthening.

The inclusion of diaphragmatic ultrasonography has been implemented as a method for assessing the diaphragm. There is evidence to suggest that prolonged adherence to high-intensity NIV for a duration exceeding 12 months can enhance the ability of the diaphragmatic muscles to contract. This is supported by documented observations of increased diaphragmatic excursion, indicating that the muscles do not undergo atrophy due to this intervention.

The study revealed that a significant proportion of patients (87.5%) experienced complications when using either BIPAP or CPAP for NIV. The most frequently observed complications were skin ulcers (39.6%) and aerophagia (29.2%), whereas claustrophobia was the least commonly reported complication

(8.3%). Yüksel et al. [25] consistently demonstrated that the detrimental effects of home non-invasive mechanical ventilation (HNIV) primarily manifest locally, specifically affecting the head and face due to the mechanical pressure exerted by the mask. Furthermore, these consequences are frequently unreported as potential life-threatening risks. Moreover, the extent of these regional complications undermines the adherence of patients to non-invasive ventilation (NIV) treatment. It is imperative to carefully consider and prudently manage the potential negative outcomes, as the effectiveness of HNIV primarily hinges on the level of compliance with NIV.

By prioritizing factors such as mask fit and tightness, pressure titration, patient comfort, and effective communication with the patient, it is possible to minimize the occurrence of non-life-threatening consequences. A range of strategies were employed to address issues related to mask discomfort and nasal ulcers. These included adjusting the mask tightness, readjusting the mask positioning, utilizing soft forehead spacers, and exploring alternative sizes or types of masks. The application of topical corticosteroids was employed to mitigate the facial erythema.

Regarding mortality, 6.3% of the studied patients died (three patients), two were incompliant with NIV therapy (one overlap patient and one COPD patient), and the last one was a compliant NMD patient concluding that increased mortality in noncompliant groups. Budweiser et al. [3] showed that survival rates in OHS on NPPV were about 97–70% within 1–5 years, respectively, with higher survival rates than patients who had untreated with NPPV. Duro et al. [26] found that death among COPD patients utilizing home NIV was 24.8% throughout the 3-year assessment period, and the total follow-up time following the initiation of NIV was 5.3 ± 4.3 years. Even though there was no control group in that retrospective investigation, the findings revealed a higher survival rate than in other reports. For individuals with ≥ 1 hospitalization per year in the year preceding to home NIV institution, admissions and the days spent in hospitals for pulmonary diseases significantly reduced following NIV institution. Sharma and Wolfe [27] concluded that NIV was a better therapeutic modality than tracheostomy for people with NMD because it increased survival, reduced symptoms, and was more agreeable. The number of NMD cases was anticipated to rise as the utilization of NIV extended childhood survival into adulthood.

Conclusions

Home NPPV was an effective long-term treatment for patients with chronic respiratory failure, as it improved PAP and diaphragmatic force and decreasing mortality. Using ultrasound to assess pulmonary pressure or diaphragmatic excursion is considered a helpful, easy method for follow-up of subjects with CRF.

Abbreviations

NIV	Non-invasive ventilation
HCRF	Hypercapnic chronic respiratory failure
PH	Pulmonary hypertension
PAP	Pulmonary artery pressure
FVC	Forced vital capacity
MIP	Maximum inspiratory pressure
NPPV	Non-invasive positive pressure ventilation
COPD	Chronic obstructive pulmonary disease
AEHRF	Acute exacerbation of hypercapnic respiratory failure
OHS	Obesity hypoventilation syndrome
OSA	Obstructive sleep apnea
CPAP	Continuous positive airway pressure
ABG	Arterial blood gas
BIPAP	Bilevel-positive airway pressure
DE	Diaphragmatic excursion
RTD	Restrictive thoracic disease
NMDs	Neuromuscular diseases
Mpap	Mean pulmonary artery pressure
FRC	Functional residual capacity
TLC	Total lung capacity
HNIV	Home non-invasive ventilation

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Authors' contributions

MEA, design of the work, data acquisition, analysis, and interpretation of data. WMS, conception, data acquisition, manuscript review, and supervision. DMG, design of the work, data acquisition, analysis, and interpretation of data. MEE, sample analysis, data analysis, methodology, and manuscript preparation. All authors have read and approved the manuscript.

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Availability of data and materials

All the data of the current study are available from the corresponding author upon reasonable request.

Declarations

Ethics approval and consent to participate

Institutional Review Board of the Faculty of Human Medicine, Zagazig University (Approval number: ZU-IRB no. 6193/2–6-2020).

Consent for publication

Not applicable.

Competing interests

The authors declare that they have no competing interests.

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