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The effectiveness of non-invasive ventilation in neuromuscular patients in KAMC

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ABSTRACT

Introduction: The impact of non-invasive ventilation on neuromuscular disease (NMD) has become evident over the past two decades due to frequent use of this technique. It allows some patients with non-progressive pathology to almost live a normal life. The purpose of this study is to indicate the impact of NIV and its results in NMD patients.

Objectives: The primary objectives for the study were to find the incidence and outcome of NMD patient on NIV, to determine whether NIV can avoid intubation and Mechanical ventilation and to compare vital signs and arterial blood gas parameters before and after NIV.

Methodology: This retrospective study was conducted during a period from January 2010 to October 2017 in King Abdul-Aziz Medical city (KAMC). The data was collected from Medical Record Department. The collected data got statistically analyzed and results obtained by using SPSS software. A total number of 21 patients admitted in KAMC in adult ICUs, wards, and Emergency room for both males and females with neuromuscular diseases with an age above 18 years old were included. Parameters related to patients and NIV were collected and recorded in an excel sheet and there by SPSS software.

Results: A total of 21 patients enrolled in this study. There were 14 males and 7 females. The minimum age 19 years old and the maximum 81 years old with a mean age of 45. Arterial Blood Gas done Pre and Post NIV and compared using paired't' test. The pre and post pH on NIV was significant with a P value (p: 0.029). The pre and post PaCO2 on NIV with a P value (P: 0.034) and oxygenation in pre and post NIV with a P value (P: 0.008) was also significant.

Conclusion: In this study, since most of the results indicates poor prognosis, it is difficult to say that NIV improve the quality of life. This can be due to the limitations and bias in the study like the heterogeneity of the diseases and bias can be due to the hospital and its policies.

Keywords: neuromuscular disease, NMD patient, Mechanical ventilation, non-progressive pathology, non-invasive ventilation

INTRODUCTION

Neuromuscular disease (NMD) is an interesting disorder that needs more investigation. Unlike other diseases, NMD does not affect a certain age. However, the patient's age plays a huge role in the effectiveness and benefit of the treatment. When the therapy was used with young patients the outcome was better. There are many methods that used in order to treat NMD. One of these method is noninvasive ventilation. The impact of noninvasive ventilation (NIV) in clinical practices on neuromuscular disease (NMD) has become prevalent over the past two decades & is being vastly used all over.

REVIEW OF LITERATURE

The effect of noninvasive ventilation (NIV) in neuromuscular disease (NMD) has become evident over the past two decades & is being widely used all over. It helps to extend the survivability in certain diseases like muscular dystrophy, rapidly deteriorating disease like amyotrophic lateral sclerosis etc. without further reduction in mortality rate [1]. Many youngsters and children with NMD survive to adulthood by using noninvasive ventilation [2]. Since NMD accounts for about 1% of the world population; it is considered one of the rarest diseases in the world [3]. Neuromuscular diseases impact the nerves that control our voluntary muscles [4]. It affects the muscles either directly or indirectly through the

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central nervous system which can cause spasm and some degree of paralysis depending on the location and the origin of the problem. It can affect respiratory muscles which can contribute to respiratory failure [4, 5]. On the other hand, non-invasive ventilation is a technique which adapts positive pressure to help respiration and to reduce endotracheal intubation. It mainly includes two modes which is continuous positive airway pressure (CPAP) and noninvasive pressure support ventilation (NIPSV) [6, 7]. NIPSV mode depends on a person's inspiration and expiration. In addition, NIPSV has an expiratory pressure called EPAP or PEEP and inspiratory pressure called IPAP, resulting in a bi-level pressure modality (BIPAP) which has shown a reduced intubation rate [8]. In NMD there is a mismatch between the muscular system and the neuronal system that lead to loss of muscle control which leads to muscle fatigue & there by difficulty in breathing [9]. This weakness in respiratory muscles make shallow breathing when lying or at the time of sleep & can also lead to carbon dioxide retention [10]. Nocturnal hypoventilation is yet another problem associated with NMD which is characterized by nocturnal shallow breathing, hypercapnia & hypoxemia which can lead to tiredness during daytime [11, 12]. NIV has proven its effectiveness in improving the breathing quality in NMD where the symptoms start from dyspnea and it could worsen until it causes death due to respiratory failure. It decreases the incidence of shallow breathing, decreasing the carbon dioxide and increasing the oxygen in the blood. [13, 14, 16]. Even though there are studies which shows its effectiveness, there are many other studies which shows that NIV cannot be used for patients who have acute neuromuscular disorders like Guillain–Barre syndrome (GBS). In these kind of diseases the upper airway gets affected more and NIV does not treat this area, but it could make it worse by increasing the probability of pulmonary aspiration [15]. Respiratory insufficiency occurs slowly and follows a predictable rate of worsening in patients with chronic, progressive neuromuscular diseases such as muscular dystrophies and amyotrophic lateral sclerosis (ALS) [17]. There are types of neuromuscular disorders with an acute or sub-acute onset, and some other types with sudden exacerbations, thus presenting with acute respiratory failure [17, 18]. This respiratory failure in these patients may not be initially recognized because they do not have frank abnormalities on auscultation or severe cyanosis. But certain signs such as paradoxical abdominal movement, use of accessory respiratory muscles, or becoming breathless while talking should raise a warning [18]. An investigation for NMD such as vital capacity, mouth pressures, arterial blood gases, chest x-ray and sometimes overnight respiratory monitoring is important [19]. Studies have shown that long-term noninvasive ventilation improves symptoms, gas exchange, quality of life and survival [20]. Neuromuscular diseases are those diseases that cause prolonged ventilator dependency [21].

Since NMD though not very common but can lead to respiratory failure, morbidity, mortality & can affect the quality of life as well, the purpose of this study is to find out the number of patients who got admitted with neuromuscular disorder who received noninvasive ventilation in King Abdul-Aziz medical City (KAMC). This study also emphasis the effectiveness of NIV & the outcome of patients who had undergone treatment with NIV.

MATERIALS AND METHODS

We conducted our research based on the patients' files in King Abdul-Aziz Medical city, Medical Record Department. Then, we collected and analyzed the results using statistical methods. We conducted a retrospective study by using convenient sampling technique during a period from January 2010 to October 2017. We studied 21 patients of KAMC in adult ICUs, wards, and Emergency room for both males and females with neuromuscular diseases. Inclusion criteria included all adult Patients 18 years old and above. Exclusion criteria included all patients less than 18 years old, Patients over 81 years of age, and neuromuscular disease patients who breathe in room air. The data collection sheet had variables such as age, gender, vital signs, NMD Type, Duration of NIV, NIV Parameters, NIV Outcomes, ABG test results,. The data entered in Microsoft Excel sheets and subsequently uploaded into SPSS software. The statistical analysis of the variables carried out using SPSS software. Bar charts used to compare the outcome variable.

RESULT

After meeting the inclusion and exclusion criteria, a total number of 21 patients got enrolled in this study. Out of the selected subjects, the males were 14 (n= 14), which constitutes 66.70% and the females were 7 (n= 7), which constitutes 33.30% as shown in **Figure 1**. The minimum age of the subjects was 19 years old and the maximum age was 81 years old with a mean age of 45years old (n=45) (**Table 1**). The predominant neuromuscular condition in this study subjects was Guillian Barrie syndrome (GBS) which was (28.5%) followed by Myasthenia Gravis (MG) (14.3%), Duchene muscular dystrophy (9.5%). Other conditions such as congenital myotrophy, neuropathy, radiculopathy and other types of

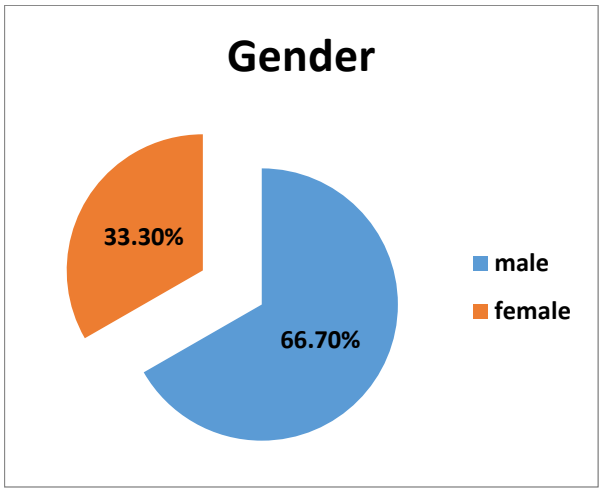

Figure 1: *Gender distribution of study subjects*

Table 1: *Descriptive Statistics on Age*

	N	Minimum	Maximum	Mean	Std. Deviation
Age	21	19	81	45.05	20.742
Valid N (list wise)	21				

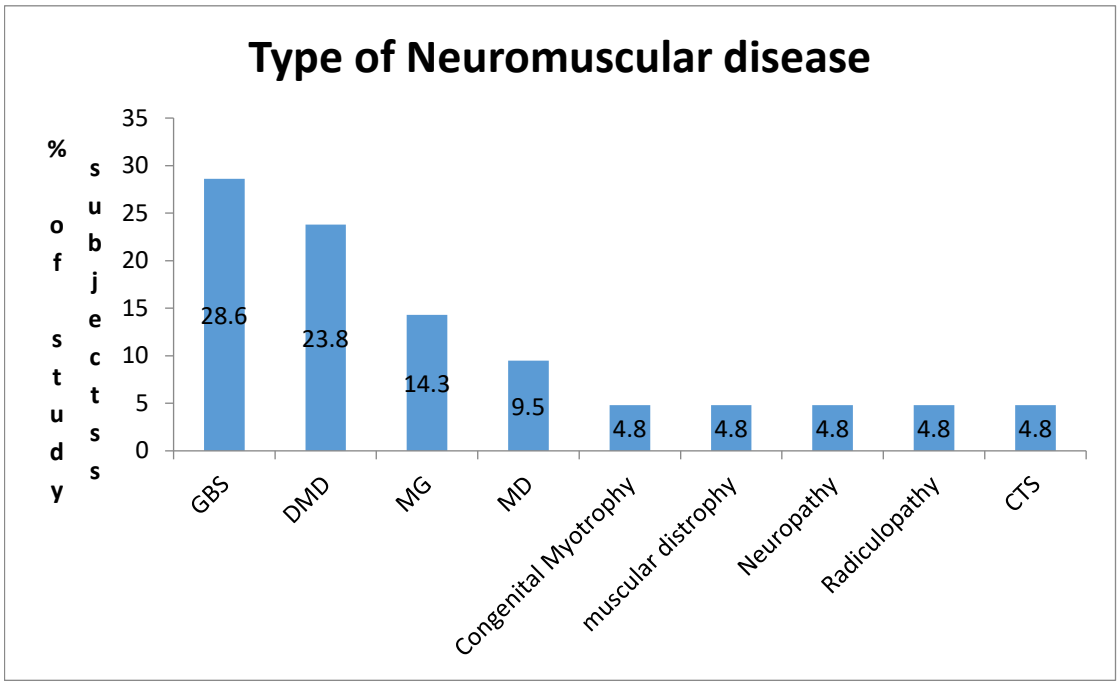

Figure 2: *Bar diagram showing the distribution of neuro muscular disease in the study subjects*

muscular dystrophies were (4.8%) as mentioned in **Figure 2**. All the selected subjects got admitted as inpatients in different Intensive Care Units (ICUs) of King Abdulla Medical City (KAMC) Riyadh.

The subjects in this study were connected with Non Invasive Ventilator (NIV) with oro nasal mask as interface. They were connected with Bi-level Positive Airway Pressure (BIPAP) which has a parameter Inspiratory Positive Airway Pressure (IPAP) and Expiratory Positive Airway Pressure (EPAP). In this study, the minimum duration of NIV was 2hours and maximum duration was 10 hours 40 minutes with minimum IPAP settings 10cm of H2O and maximum settings of 18cm H2O. The minimum Expiratory Positive Airway Pressure (EPAP) was set at 4cmof H2O and maximum of 8cm of H2O. The minimum duration of NIV with oxygen was 20 minutes were as the maximum was 40 minutes (**Table 2**).

Table 2: *Mean and Standard Deviation for various parameters*

	N	Minimum	Maximum	Mean	Std. Deviation
Duration of NIV by hours	21	2	624	123.67	197.985
NIV Parameters inspiratory positive airway pressure	21	10	18	13.24	2.406
NIV Parameters expiratory positive airway pressure	21	4	8	5.95	1.322
NIV Parameters oxygen	21	.20	40.00	2.2576	8.64867

Table 3: *Comparison of pre and post ABG parameters after NIV use*

		Mean	N	Std. Deviation	T value	P value
Pair 1	pre-NIV Ph	7.3177	21	.07833	-2.356	0.029*
	post-NIV Ph	7.3700	21	.05854		
Pair 2	pre-NIV carbon dioxide	65.0190	21	23.89597	2.269	0.034*
	post-NIV carbon dioxide	54.9619	21	13.45126		
Pair 3	pre-NIV oxygen	63.0857	21	23.15045	-2.933	0.008*
	post-NIV oxygen	90.5571	21	33.90349		
Pair 4	pre-NIV HCO3-	30.7429	21	8.14976	0.649	0.524
	post-NIV HCO3-	29.6762	21	7.29533		

Table 4: *NIV Outcomes*

		Frequency	Percent
Valid	Room air	3	14.3
	shifted to NC	5	23.8
	Intubated	12	57.1
	Tracheotomy	1	4.8
	Total	21	100.0

An Arterial Blood Gas (ABG) analysis was done Pre and Post NIV and was compared using paired 't' test. The analyzed and compared ABG parameters were pH, arterial pressure of carbon dioxide (PaCo2), arterial bicarbonate ($HCO3^-$) and pre and post values of partial pressure of arterial Oxygen (PaO_2). The mean pH pre NIV (7.31) and post NIV (7.37) with a P value (p-0.029) showed statistical significance. The mean PaCO2 pre NIV (65.02mmHg) and post NIV (54mmHg) with a P value (P - 0.034) and the mean oxygenation pre NIV (63.9mmHg) and post NIV (90.55mmHg) with a P value (P -0.008) was also statistically significant as shown in the **Table 3**. There was a slight difference in arterial bicarbonate in pre and post NIV.

The outcome of NIV in this study was not that effective. Out of the total 21 subjects, majority of the subjects (n=12) got intubated and mechanically ventilated (57.1%), some patients (n=5) was connected to nasal cannula (23.8%), while others (n=3) made into room air (14.3%) and tracheostomised (n=1) (**Table 4**).

DISCUSSION

The study was conducted to see the effectiveness of Non-invasive ventilation on the survival and improvement of patients with neuromuscular disease. In this study, we collected data of 21 neuromuscular disease patients all of whom used NIV. The age varies between 18 and 81 with the mean of 56 year-old. We analyzed, retrospectively their data and the outcomes were 57.1% got intubated, 4.8% deteriorated and got tracheostomy, 23.8% got better and shifted to nasal cannula, and 14.3% were on room air. From this result we can deduct that most patients got their condition worsen and the NIV treatment did not make a major effect. But that can be due to the patient own health. Since sample of patients we have contains a lot of elderly patients and some of whom had co-existing illness. They can develop respiratory failure because of the weak muscles. Most importantly though, this does not mean the NIV therapy will directly be good for young people. However, another study about the outcome of non-invasive positive pressure ventilation in pediatric neuromuscular conducted by The Hospital for Sick Children, Toronto, Canada. [22] Showed that there is notable improvement in children health. The time spent during hospitalizations was significantly lower. [22] Children spent 85% fewer days in hospital mean pre-NIV 48.0 days, the mean post-NIV 7.0 days. [22] Unfortunately, this does not support our previous clinical impression and reported findings in adult.

CONCLUSION

In conclusion, it is hard to say that NIV improve the quality of life in our study. Since most of the results indicates otherwise. But other studies show vast improvement. This can be due to the limitations and biases in the study. The limitations are the heterogeneity of the diseases and the progressive ones. The bias can be due to the hospital and its policies. In the end we think we need more studies to show if there is an association between the progressiveness of neuromuscular disease and improvement of the patient with respiratory problems at the time of using NIV.

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APPENDICES

Appendix I

King Saud Abdulaziz University for health sciences

College of Applied Medical Sciences

Research unit

CAMS 411 & 412 Research methodology 1 & 11

Research title: The Effectiveness of Noninvasive Ventilation in Neuromuscular Disease Patients in King Abdul-Aziz Medical City.

Data collection sheet

Data collection format

Serial Number	Age	Gender	Vital Signs pre and post				NMD Type	Duration of NIV	NIV Parameters	ABG Parameters pre and post				NIV Outcomes
			T (°C)	BP	HR	RR				Ph.	$PaCO_2$	PaO_2	HCO_3^-	
1														
2.														
3.														
4.														
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