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Neuromuscular diseases in the pediatric intensive care unit: 11 years of experience from a tertiary children's hospital

[1]Gamze SARIKAYA UZAN, [1]Pınar EDEM, [2]Tolga BESCI, [1]Cem PAKETÇI, [2]Gültaç EVREN, [1]Semra HIZ KURUL, [2]Murat DUMAN, [1]Uluç YIŞ

[1]Division of Child Neurology, Department of Pediatrics, [2]Division of Pediatric Critical Care Medicine, Department of Pediatrics, Dokuz Eylül University Faculty of Medicine, İzmir, Turkey

Abstract

Background & Objective: We present 11-year data of patients with neuromuscular disease (NMD) that were treated in the pediatric intensive care unit of a tertiary children's hospital. *Methods:* The data of all cases followed-up in the pediatric intensive care unit (PICU) were retrospectively analyzed. Patients were evaluated in terms of age, gender, diagnosis, PRISM, neuroanatomical localization, hospitalization time-age, cause of admission to pediatric intensive care unit, clinical course, complications and clinical discharge status. *Results:* A NMD was detected in 43 of the 1,411 patients admitted within the study period and accounted for approximately 3% of pediatric intensive care unit admissions. NMD consisted of genetic (n= 35, 74.8%), acquired (n=6, 13.8%) and metabolic (n=2, 4.6%) causes. The diagnoses included spinal muscular atrophy type 1 (n=12, 27.9%), Duchenne and Becker muscular dystrophy (n=7, 16.2%), congenital myopathy (n=6, 13.9%), congenital muscular dystrophy (n=5, 11.6%), Guillain-Barre syndrome and its variants (n=2, 4.6%), spinal muscular atrophy associated with respiratory distress type 1 (n=2, 2.3%), critical illness neuropathy (n=2, 4.6%), acute flaccid myelitis (n=2, 4.6%), congenital myasthenic syndrome (n=1, 2.3%), peripheral neuropathy associated with disorder of riboflavin transporter (n=1, 2.3%), juvenile amyotrophic lateral sclerosis (n=1, 2.3%), stress-induced childhood-onset neurodegeneration with ataxia and variable seizures (n=1, 2.3%), and metabolic myopathy (n=1, 2.3%). Respiratory complications (n=31, 72%) were the most common reasons of admission to the pediatric intensive care unit. Seven (16.2%) patients have the NMD diagnosis confirmed during their first admission in PICU. These included both genetic cause of NMDs (n=4) and acquired NMDs (n=3). Mortality rate was 6.9% (n=3). Forty-three patients diagnosed with NMD had 75 times PICU admissions. The disease with the highest rate of re-admission to the PICU was SMA type 1, and the most common reason for re-admission was respiratory reasons.
Conclusion: Accurate diagnosis of NMD and knowledge of causes of admission to PICU is crucial for increasing awareness, sensitivity and effectiveness when treating these diseases.

Keywords: Neuromuscular disease, pediatric intensive care unit, acute flaccid myelitis, spinal muscular atrophy

INTRODUCTION

Improvement in both pediatric intensive care units (PICUs) and treatment options of neuromuscular diseases (NMDs) has led to an increase in the number of patients with NMDs seen in PICUs. Common reasons for admission to PICU are rapid progression of the disease, respiratory failure, complications due to a primary diagnosis, or postoperative care. Yates *et al*.[1] have reported that most children with progressive NMD that were admitted to the PICU recover and are discharged without the need for prolonged invasive ventilation. However, further PICU admissions are likely due to frequent needs of non-invasive home mechanical ventilation (NIHMV) support. Other studies evaluating acute NMDs in PICU have emphasized the importance of timely diagnosis of acute and acute-onset chronic

Address correspondence to: Gamze Sarıkaya Uzan, Dokuz Eylul University Faculty of Medicine, Department of Pediatric Neurology, İzmir, Turkey. e-mail: gamzeuzan36@gmail.com

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neuromuscular diseases. Early recognition of these diseases would minimize the complications that might develop due to diagnostic delays and ensure effective management.[2,3] Our goal in this study was to contribute information regarding the management of NMDs by presenting 11 years of data from PICU of a tertiary children's hospital.

METHODS

The electronic medical records of all cases treated at Dokuz Eylül University Faculty of Medicine Department of Pediatric Intensive Care Unit between March 2009 and December 2019 were retrospectively reviewed and patients with NMDs were included in the study. Our PICU is a 6-bed intensive care unit in a tertiary-care children's hospital, accepting children between 1 month-18 years of age with medical, surgical, oncological, and trauma indications. Extracorporeal therapies (continous renal replacement therapy, therapeutic plasma exchange) along with invasive ventilators are available when indicated. Patients diagnosed with NMDs only by genetic analysis, muscle biopsy or EMG were included in the study. The subjects were evaluated in terms of age, gender, diagnosis of the primary NMD, neuroanatomical localization, genetic etiology, cause of admission to PICU, clinical course, mortality score, the length of hospital stay, number of hospitalizations, treatment for the primary disease, ventilatory support, tracheostomy and percutaneous endoscopic gastrostomy (PEG) status, presence of complications during hospitalization, clinical discharge status, cause of death (if applicable), and re-admission to PICU in the after discharge. In the course of the research, the first hospitalization of the patients who were admitted to the PICU multiple times was taken into account. We retrospectively calculated the Pediatric Index of Mortality (PIM) of each patient to assess the clinical status of the patients. While calculating PIM, we used Pediatric Risk of Mortality Score 4 (PRISM4) for inpatients as of 2016 and the Pediatric Risk of Mortality Score 2 (PRISM2) scale for inpatients before 2016. While evaluating the patients, 2 main grouping methods were used. Firstly; they were divided into 4 main groups according to the neuroanatomical localization: diseases that involve the anterior horn of the spinal cord, peripheral nerve and nerve root, neuromuscular junction and muscle. Secondly; according to their hospitalization dates, they were divided into two subgroups, consisting of patients followed up between 2009 and 2014 (subgroup 1) and followed up between 2015 and 2019 (subgroup 2). There have been changes in treatment opportunities and proactive care in PICU over the 11-year period. In this period, there have been some changes in the diagnosis and treatment approaches of neurological diseases (such as the use of nusinersen in SMA and ataluren in DMD, increased the use of plasmapheresis in autoimmune neurological diseases, the increase in genetic diagnosis possibilities). In addition, the use of respiratory physiotherapy, high flow nasal cannula O2 therapy (HFNC) and the number of PICU physicians per patient increased in our hospital during this period. For this reason, we examined our patients by grouping them according to time periods. The data obtained in the study were entered into the database created in the SPSS (Statistical Package For Social Sciences) 18.0 program and statistical analyzes were performed with the same program. Mean and st. deviation, categorical variables were presented as numbers and percentages. Conformity of continuous variables to normal distribution was investigated by considering graphical research, normality tests and sample size. Categorical independent variables were presented in cross-tables as frequencies and percentages, their distributions were compared by ''Chi-Square'' test methods and ''Fisher's Exact Test''. In all statistical comparison tests, the margin of error for type 1 was determined as α: 0.05 and double-tailed test If $p<0.05$, the difference between the groups was considered statistically significant. All procedures performed in accordance with the Dokuz Eylül University ethics committee. Consent received from Dokuz Eylül University ethics committee (2020/16-12).

RESULTS

A total of 1411 patients were admitted to PICU during the study period. Sixty-one children had NMDs and suspected NMDs. Nineteen of those patients were diagnosed with rhabdomyolysis. One patient had rhabdomyolysis secondary to an underlying NMD and was included in the study. The other 18 patients were excluded because the cause of rhabdomyolysis was not a neuromuscular disease. Therefore, of 1411 subjects, 3% (n=43) were diagnosed with NMDs and included in the study. The median age was 2 years (1 month-17 years) and 53.4% (n=23) of the patients were male. While 19 (44.1%) patients had muscle involvement, 17 (39.5%) had involvement of anterior horn of the spinal cord, 6 (13.9%) had involvement of peripheral nerve

and nerve root, and 1 (2.3%) had involvement of the neuromuscular junction. The most frequent diagnoses are spinal muscular atrophy (SMA) type 1 (n=12, 27.9%), Duchenne (DMD) and Becker (BMD) muscular dystrophy (n=7, 16.2%) (Table 1). In addition, the patients also were classified as etiological. It consisted of genetic (n= 35, 74.8%), acquired (n=6, 13.8%) and metabolic (n=2, 4.6%) causes. The etiological features of our patients are given in Table 2.

Respiratory failure (n=31, 72%) was the most common cause of admission to the PICU. Respiratory support was given to 41 patients. Four patients received O2 support via face mask, 37 patients invasive mechanical ventilation (IMV). Three of 37 patients died, 1 of them switched to spontaneous respiration extubated. We used the term 'spontaneous' to describe spontaneous breathing in room air. Thirty three patients were tracheostomized. Tracheostomy was performed in 26 patients in our PICU. Median tracheostomization time was 16 days (1-57 days). The remaining 7 patients were already tracheostomized in another hospital. Fourteen (42.4%) patients were discharged as succesful decannulation. The respiratory support status of our patients is given in Figure 1. Eight (18.6%) cases received treatments specific to their NMD diagnosis (Table 1). The median hospitalization period was 26 days (1-100). The median PRISM of our patients was 5.86% (0.3-39.3). The discharge status of our patients is given in Table 1. The median value of the PRISM mortality index of our patients was 1.6 (0.3-39.3). The mortality rate was 6.9% (n=3). Three patients died in the PICU. The patients were diagnosed with SMA type 1, SMARD1, and myotubular myopathies. First patient was a 9 years old male patient diagnosed with SMA type 1. The patient was admitted to the PICU due to respiratory failure. Second patient, that were followed-up due to respiratory distress associated with pneumonia were unable to be weaned from mechanical ventilation support and were diagnosed with SMARD1. The other patient, a female patient who was previously diagnosed with myotubular myopathy while being examined as a hypotonic infant at the age of 6 months, was admitted to the PICU because of respiratory failure associated with pneumonia. They died in PICU due to respiratory and multisystem failure.

The clinical and demographic characteristics of the patients by their hospitalization dates is shown in Table 3. The most common diseases in the first group (2009-2014) were congenital myopathy (n=5) and SMA type 1 (n=4). In the second group (2015-2019) SMA type 1 (n=8) and DMD/BMD (n=4). We compared PRISM scores of the two groups. We found that the PRISM score was lower in the second group (Table 3). There was no significant difference between the PRISM scores, length of hospitalization and age of hospitalization of the two groups (p=0.190; 0.104; 0.545).

Seven (16.2%) patients were diagnosed with NMD during their follow-up in the PICU. These diseases were SMA type 1, SMARD1, Ullrich congenital myopathy, critical illness neuropathy, glycogen storage disease type 5 (GSD5) and Guillain Barre syndrome (GBS) (n=2). A history of upper respiratory tract infection acquired 7-10 days ago, unconsciousness, sore throat, difficulty in swallowing, dysarthria, symmetrical paralysis starting from the lower extremities and absent DTR were the common findings of GBS cases. In the first case, Bickerstaff's brainstem encephalitis (BBE) was considered due to the additional findings such as asymmetrical mydriasis and total ophthalmoparesis, although without pleocytosis in cerebrospinal fluid. She was admitted to the PICU because of her increasing requirement for respiratory support. There were no abnormal laboratory findings other than positive adenovirus polymerase chain reaction test. The second case was admitted to the PICU due to respiratory arrest while being observed at another hospital for the diagnosis of encephalitis. Acute motor axonal neuropathy was considered owing to progressive muscle weakness, respiratory muscle involvement, dysarthria, dysphagia, electroneuromyography findings, and autonomic dysfunction. The patient diagnosed with BBE received intravenous immunoglobulin (IVIg) and steroid. The other patient received only IVIg treatment. At the time of discharge, they had muscle strength of 5/5 in the upper extremities and 3/5 in proximal lower and 4/5 in distal lower extremities according to the Medical Research Council Manual Muscle Testing scale.[4] A patient diagnosed with GSD5 male patient was admitted to the PICU for the treatment of acute renal failure arising from recurrent rhabdomyolysis attacks after exercise. He recovered with hemodialysis. He was diagnosed with GSD5 based on muscle biopsy and genetic analysis *(PYGM gene, p.M1V rs267606993)*.

Sixteen (37.2%) cases were re-admitted to the PICU in the first year after discharge (Table 1) [SMA type 1; (n=11), SMARD1; (n=1), RYR1 myopathy; (n=1), SEPN1 myopathy; (n=3)].

Table 1: Clinical and demographic characteristic of the patients

Primary disease		Age (mean/ years)	Gender and patient number	Reason for admit	Spesific treatment	Respiratory support	Status of discharge	Recurrent hospitalization on first year	PICU days (mean)
Disorders involving the anterior horn of the spinal cord (n= 17)	**SMA type 1 n=12**	9,1	5F, 7M	Respiratory failure (n=12)	n=1 Nusinersen	NIMV(n=2) IMV(n=10)	NA and IMV(n=9) NA and NIMV(n=1) NA and Spontaneous resp. (n=1) Ex(n=1)	n=11	36.9
	SMARD1 n=2	5	1F, 1M	Respiratory failure (n=2)		IMV(n=2)	NA and IMV(n=1) Ex(n=1)	n=1	7.5
	AFM n=2	9,5	2F	Bulbar inv. Rapidly progression(n=2)	IVIg+Steroid+ Plasmapheresis (n=2)	IMV(n=2)	NA and NIMV(n=2)	n=0	14.5
	Juvenile ALS n=1	15	1M	Respiratory failure (n=1)		IMV(n=1)	IMV, NA (n=1)		
Disorders involving the peripheral nerve and nerve root (n=6)	**Riboflavin associated neuropathy n=1**	7	1F	Respiratory failure (n=1)	Riboflavin(n=1)	IMV(n=2)	Spontaneous,ambulatory(n=1)	n=0	22.8
							IMV, NA (n=1)		
	CONDSIAS n=1	6	1F	Postoperative(n=1)			NIMV, amb.(n=2)		
	Cr.illness neuropathy n=2	15.5	2M	Respiratory failure (n=2)			IMV, NA (n=1)		
	BBE n=1	13	1F	Bulbar involvement(n=1)	IVIg		Spontaneous,ambulatory(n=1)		
	AMAN n=1	11	1M	Respiratory failure (n=1)	IVIg		NA, NIMV(n=1)		
Muscular Disorders	**CMD n=5**		3F,2M	Respiratory failure (n=4) Postop(n=1)		IMV(n=5)			43.6
	DMD/BMD n=7	11,7	7M	Respiratory failure (n=1) Postoperative(n=5) Heart failure(n=1)		IMV(n=5) NIMV(n=1) Spontaneous (n=1)	NA and spontaneous (n=1)	n=0	7.2
							Spontaneous,ambulatory(n=1) (n=4)		
							NA and NIMV(n=1)		
							NA and IMV(n=1)		
	TTN n=1	1F	1F	Respiratory failure (n=1)		IMV(n=6)	NIMV, NA (n=3)	n=4	32.6
	Myotubular n=1	0.75	1F	Postoperative(n=1)1			Ex (n=1)		
	RYR1 n=1	0.75	1F	Respiratory failure(n=1)			IMV, NA (n=1)		
	SEPN1 n=3	4.2	2F, 1M	Respiratory failure(n=3)			IMV,NA (n=1)		
	Glycogene storage disease type 5 n=1	13	1M	Acute renal failure	Hemodialysis	Spontaneous (n=1)	Spontaneous,ambulatory(n=1)	n=0	3
Neuromuscular Junction Disorders	**Congenital myasthenia**	1	1F	Respiratory failure (n=1)	Pyridostigmine(n=1)	IMV(n=1)	Spontaneous,ambulatory(n=1) (n=1)	n=0	11

SMA: Spinal muscular atrophy SMARD: Spinal muscular atrophy associated with respiratory distress type 1 CMD: Congenital muscular dystrophy AFM: acute flaccid myelitis GSD: Glycogene storage disease ALS: Amyotrophic Lateral sclerosis CONDSIAS: Stress-induced childhood-onset neurodegeneration with variable ataxia and seizures BBE: Bickerstaff's brainstem encephalitis AMAN: Acute motor axonal neuropathy DMD/BMD: Duchenne and Becker muscular dystrophy NIMV: Noninvasive mechanical ventilator IMV: Invasive mechanical ventilatory NA:Nonambulatory IVIg: Intravenous immunoglobulin

Table 2: Classification of our patients according to etiology

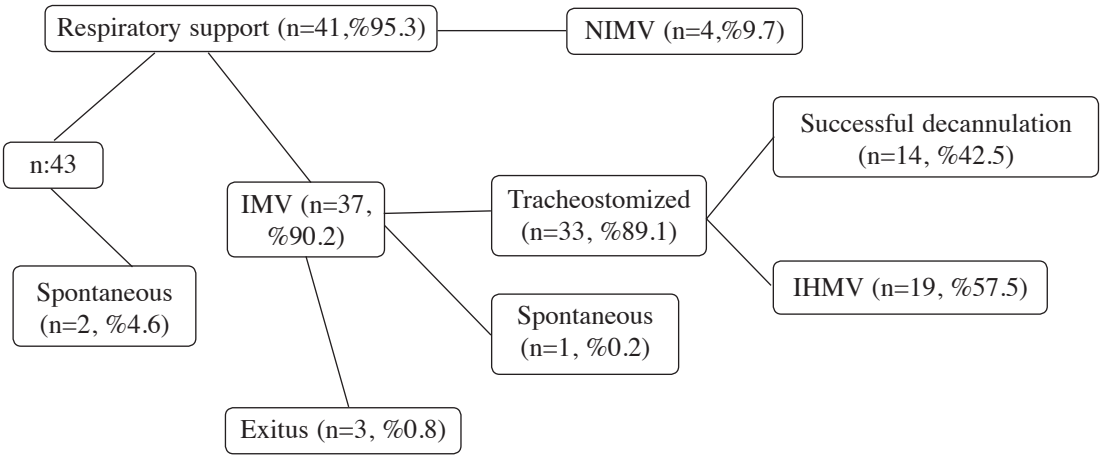
Etiology of neuro-muscular disorders	Disease		Gene
A. Genetic etiology (n=35, 74.8%)	Anterior horn cell diseases (n=15, 34.8%)	5q SMA: SMA type 1 (n=12, 27.9%)	*SMN1* (n=12)
		Non 5q SMA: SMARD (n=2, 4.6%)	*IGHMBP2* (n=2)
		Juvenile ALS (n=1, 2.3%)	*FUS* (n=1)
	Muscle diseases including myopathies or muscular dystrophies (n=18, 41.7%)	DMD and BMD (n=7, 16.2%)	*Dystrophin* (n=7)
		Congenital myopathy (n=6, 13.9%)	*RYR1* (n=1) *SEPN1* (n=3) *TTN* (n=1) *MTM1* (n=1)
		Congenital muscular dystrophy (n=5, 11.6%)	— (clinical and muscle biopsy findings)
	Neuromusclar junction disorder (n=1, 2.3%)	Congenital myasthenic syndrome (n=1, 2.3%)	*CHRNE* (n=1)
	Disorders involving the peripheral nerve and nerve root (n=1)	Stress-induced childhood-onset neurodegenertion with ataxia and variable seizures (n=1, 2.3%)	*ADPRHL2* (n=1)
B. Metabolic etiology (n=2, 4.6%)	Metabolic myopathy (n=1, 2.3%)	Glycogen storage disease type 5	*PYGM* (n=1)
	Peripheral neuropathy associated with disorder of riboflavin transporter (n=1, 2.3%)		*SLC52A2* (n=1)
C. Acquired etiology (n=6, 13.8%)	GBS and its variants (n=2, 4.6%)		—
	Acute flaccid myelitis (n=2, 4.6%)		—
	Critical illness neuropathy (n=2, 4.6%)		—

SMA: Spinal muscular atrophy SMARD: Spinal muscular atrophy associated with respiratory distress type 1 CMD: Congenital muscular dystrophy AFM: acute flaccid myelitis ALS: Amyotrophic Lateral sclerosis DMD/BMD: Duchenne and Becker muscular dystrophy

During the 11-year period, 43 patients diagnosed with NMD had 75 times PICU admissions. 21.3% (n=16) of this hospitalization rate occurred within the first year after discharge. More than half of the recurrent hospitalizations consisted of patients with SMA type 1 (n=30, 40%) and congenital myopathy (n=13, 17.3%).

DISCUSSION

Pediatric NMDs accounted for 3% of the patients admitted to the PICU over approximately eleven years in our center. In a study by Harrar *et al*. they found the prevalence of acute or acute-onset chronic NMDs in their PICU was 0.1%, whereas in another study from Egypt this rate



NIMV: Noninvasive mechanical ventilator IMV: Invasive mechanical ventilator
NIHMV: Noninvasive home type mechanical ventilator IHMV: Invasive home type mechanical ventilator

Figure 1. Respiratory Support Status of Our Patients

was around 5%.[2,3] Harrar *et al*. attributed their low rate to inclusion of acute NMDs only. The other study, on the other hand included chronic NMDs in their evaluation, and their results were similar to ours.[3] Although SMA Type 1, DMD, BMD, and congenital myopathy were the most common diagnoses in our study, GBS was the most common diagnosis in other studies.[2,3] The fact that we encountered chronic NMDs more frequently may be because our clinic is a tertiary intensive care unit (ICU) and is a center where both undiagnosed and ICU patients get referred to.

In our cohort, we encountered disorders affecting all components of the neuromuscular system. The most common reasons for admission to the PICU were post-operative care in diseases involving muscle cells, cardiopulmonary arrest in diseases involving anterior horn cells, cardiopulmonary arrest and respiratory failure in diseases involving peripheral nerves and nerve roots, and respiratory failure in diseases with neuromuscular junction involvement. Therefore, we conclude that cases with NMD were most often admitted to or followed up in the PICU for

Table 3: Findings of the patients by years

Patient Characteristics	2009-2014 (group 1)	2015-2019 (group 2)
Age (mean)	4.7 (0.2–16)	7 (0.1–17)
Gender	11F/7M	9F/16M
Location of NMD involvement (n)	– Anterior horn: 7 – Muscle: 8 – Muscle nerve junction: 0 – Dorsal root and peripheral nerve: 3	– Anterior horn: 10 – Muscle: 11 – Muscle nerve junction: 1 – Dorsal root and peripheral nerve : 3
NMD course (n)	Acute: 0 Chronic: 1 Progressive: 15 In the PICU: 2	Acute: 6 Chronic: 0 Progressive: 19 In the PICU: 0
Location of diagnosis (n)	PICU: 4 Out of PICU: 14	PICU: 3 Out of PICU: 22
MV (n)	IMV: 15 NIMV: 2	IMV: 23 NIMV: 2
Tracheostomy (n)	13	19
PRISM2 (%) (median)	7.82 (0.43–39.3)	4.49 (0.2–24.94)

F: Female M: Male NMD: Neuromuscular disorders PICU: Pediatric intensive care unit MV: Mechanical ventilatory
NIMV: Noninvasive MV IMV: Invasive mechanical ventilatory PRISM: Pediatric risc score index of mortality

respiratory disorders. As far as we know, one of the most important concerns of PICU clinicians regarding NMDs is the difficulties in the transition of patients to spontaneous breathing. In a study conducted in the United Kingdom, an underlying NMD was identified in 27% of all PICU patients requiring respiratory support longer than 28 days after an acute illness.[5] In our study, respiratory failure was the cause for admission to the PICU in 36.3% of the cases. Moreover, invasive or non-invasive mechanical ventilation support was required in all patients (n=41, 95%) except for two. One of these two patients was diagnosed with GSD5 after hospitalization for rhabdomyolysis and the other was a BMD patient having myocarditis. In one study, approximately 50% of the cases were admitted to PICU for respiratory reasons and received invasive or non-invasive respiratory support.[2] We believe that higher rates of ventilation support requirement in our study is related to the inclusion of patients with chronic progressive NMDs. In another study published in 2004, it was reported that nearly half of the cases with neuromuscular disease needed home respiratory support at any given stage of the follow-up period.[1] In this study, 86.8% of the patients were treated with a tracheostomy and were discharged with an IHMV, while in another study that evaluated acute NMDs in PICU, this rate was 20%.[2] We think that including chronic NMDs has resulted in higher tracheostomy rates in our study. This result indicates that the need for long-term respiratory support is high, despite how well they are managed under the PICU conditions. In addition, although the PRISM median value of our patients is not very high, we think that highly values such as 39.3%, 24.9% contribute to this result. The reason why most of our patients were palliative care patients was their advanced age and advanced level their disease. The reason for this result was the absence of pediatric pulmonologist and polysomnography in our hospital. This limitation is an important problem of both our unit and our country**.** Thus, the care of pediatric NMD is managed in a multidisciplinary by the departments of pediatric neurology, pediatrics, physical therapy and rehabilitation, orthopedics in our hospital. General pediatrician is responsible for the respiratory care of tracheostomized patients after they were discharged from PICU. In addition to the treatment methods developed for NMDs, we believe that these areas need to developed for increase the quality of life of patients.

In our cohort, a complication was reported in 4.5% (n=2) of patients during hospitalization (pneumothorax and venous thrombosis), but no complications associated with given treatments were observed. We believe that increasing awareness of NMDs over the past years, as well as developing new treatment methods for chronic NMDs, and improving PICU conditions will further reduce complication and mortality rates by increasing survival rates.

Of the 43 subjects, 6 (13.9%) were not previously diagnosed with NMDs, but were diagnosed for the first time in the PICU. These diseases were SMA type 1, SMARD1, Ullrich congenital muscular dystrophy, critical illness neuropathy, acute motor axonal neuropathy type of GBS and GSD5. It should not be forgotten that both acute and chronic NMDs can be diagnosed in PICU. Admission to the PICU may be necessary during the course of some NMDs such as types of metabolic diseases with recurrent attacks of rhabdomyolysis.[6] Moreover, it should be noted that cardiac arrest due to autonomic dysfunction may develop in GBS, which may be observed to a lesser extent with early diagnosis.

Since treatment of NMDs is often limited to supportive care, general care and minimization of complications are vitally important. In this study, 20.4% (n=9) of the NMDs had treatment options that targeted the underlying pathophysiological process. When initiated immediately, these treatments reduce the duration of hospitalization and the need for intensive care management. An example of this rapid initiation is IVIg or plasmapheresis, which speeds up the recovery in patients with GBS, or the use of drugs known to improve respiratory difficulties, such as nusinersen, in treating SMA.[7-9] Moreover, clinical findings such as hearing loss, sensorial ataxia, and progressive sensorimotor peripheral neuropathy should raise suspicion for disorders associated with defect of riboflavin metabolism, in which rapid initiation of treatment without waiting for the genetic test results can be life-saving.[10]

At the time of discharge from the PICU, 9 patients were breathing spontaneously and 6 were ambulatory. Although patients with NMDs may be staying immobilized and receiving respiratory support longer than other groups of diseases in the PICU, early recognition and effective patient care management can produce positive clinical results. Therefore, we want to highlight the importance of early diagnosis of NMDs, avoiding unnecessary examination and ensuring timely initiation of supportive therapy, even in some NMDs without a specific treatment option.[11]

In conclusion, despite the fact that NMDs

are often suspected and diagnosed by pediatric neurologists and pediatricians, they can also be diagnosed for the first time by pediatric intensive care specialists, especially if the conditions are acute. For this reason, neurological evaluation is very critical in PICU. Regardless of their relative rarity, early recognition of these diseases will have a positive impact on survival by ensuring the implementation of effective management strategies and minimizing complications. Even though most treatments are supportive; nusinersen, IVIg, plasmapheresis, pyridostigmine and riboflavin can be life-saving. However, in the near future, as new treatments for NMDs will extend the patients' life spans, PICU specialists will face more of these diseases.

DISCLOSURE

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