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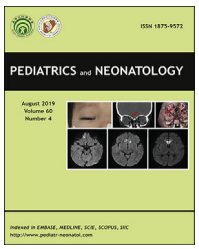

Original Article

Emergency room visits and admission rates of children with neuromuscular disorders: A 10-year experience in a medical center in Taiwan

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emergency room;
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Background: No previous studies have explored emergency medical care for children with chronic neuromuscular disorders (NMDs). We aimed to determine the major reasons for the emergency room (ER) readmission of pediatric patients with NMDs and suggest changes to the care plan to decrease readmissions.
Methods: Children with chronic NMDs (aged <18 years) who visited a medical center-based ER between January 2005 and January 2015 were included. The following data were extracted from the patient's ER records: presentations; demographic data, including sex and age; NMD diagnosis; triage classification; emergency examination; initial management and outcomes. The outcomes were death inside or outside the ER, admission to the ward or pediatric intensive care unit (PICU), uneventful discharge, and repeated ER visits.

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Results: In 10 years, 44 children with heterogeneous NMDs (boys/girls: 30/14, mean age: 9.9 years) visited the ER for a total of 204 times. Repeated ER visits and readmissions occurred in 56.8% and 55.6% of the patients, respectively. Most NMD children belonged to triage class 3 (35.3%), with underlying congenital hereditary muscular dystrophy (44.1%). The major symptoms were usually multiple and concurrent, and primarily respiratory (62.3%) or gastrointestinal (28.9%). The most common causes of hospitalization were pneumonia (48.5%) or acute gastritis (20.4%), and approximately half of the ER visits required further hospitalization, of which 28.2% involved PICU admission. Twenty of the 36 children admitted to the ER required readmission. The most commonly prescribed examinations were complete blood count (38%) and C-reactive protein (38%), and the most common therapy was intravenous fluid administration (34%). Although respiratory compromise caused most ER visits and admissions, pulmonary assessments, including chest films (28%), pulse oximetry (15%), and blood gas analysis (11%), were performed in a relatively small proportion.
Conclusion: The ER staff must recognize patients' unmet needs for respiratory and gastrointestinal care related to underlying NMDs.

1. Introduction

Children with chronic, degenerative, neuromuscular disorders (NMD) comprise a chronically debilitated population with extensive healthcare needs, high levels of health care utilization, and substantial costs of care. Since diminished muscle mass may increase the risk of acute illnesses, hospital readmissions are likely to be more common among these children.[1,2] Recurring emergency room (ER) visits are likely, and they should be actively anticipated. In particular, most children with heterogeneous underlying NMDs experience frequent acute exacerbations, usually presenting with acute neuromuscular respiratory failure requiring emergency assistance. These cases represent a diagnostic challenge in the acute care settings, especially in a busy ER, where clinical examinations are often limited by patients' inability to communicate and cooperate due to sedation, pharmacological paralysis, as well as interference from equipment necessary for respiratory support, including endotracheal tubes. As a result, few studies have examined the characteristics and subsequent flow of children with NMD following visits to the ER.

Although standards of care have been established for several subtypes of NMD,[3–6] a wide range of factors must be taken into account on the presentation of a patient with NMD to the ER because of the involvement of different systems in patients with heterogeneous NMD. Moreover, the evaluation of critically ill children with degenerative NMD may be confounded by concomitant conditions such as infections, metabolic disturbances, and overwhelming weakness/fatigue. Even though a reliable NMD prevalence rate is not available in Taiwan, a notable number of children with different NMD have sought help from the Kaohsiung Medical University Hospital because clinical trials and standard of care for NMD have been provided there for many years.[7–10] This study sought to identify the unmet needs of children with NMD in the ER settings by characterizing the presentation, examinations provided, management, subsequent flow to discharge or hospital admission, and outcomes.

2. Methods

We received approval to conduct this study from the institutional review board of Kaohsiung Medical University Hospital. The data were collected between January 2005 and January 2015. A list of pediatric emergency patients younger than 18 years with a definite diagnosis of degenerative NMD was compiled. Degenerative NMD was defined as presentation with any of the following conditions: spinal muscular atrophy, congenital hereditary muscular dystrophy, benign congenital myopathy, or hereditary progressive muscular dystrophy causing weakness. We used International Classification of Diseases, Ninth Revision (ICD-9) codes, including 335.10, 335.19, 359.0, and 359.1, from the institution's database. Furthermore, to protect the confidentiality of the study participants and to comply with the Personal Information Protection Act of Taiwan, all personal identifiers were removed.

Patient demographic data including sex, age and the number of emergency visits and admissions were collected retrospectively from electronic medical records for each ER visit. We further analyzed the following parameters by collaborating with the statistical analysis laboratory: number of ER visits, chief complaints/diagnosis/symptoms, received ER examinations/managements, number of admissions (including pediatric intensive care unit [PICU]), and mortality in ER. The data of pediatric emergency triage classification system documented in each ER visit of enrolled cases were also collected.[11] Finally, the correlations between patient's subtype of NMDs, major diagnosis/symptoms, ER assessments, and number of admissions were analyzed using the chi-square test/Fisher's exact test/Wilcoxon rank-sum test. A p value <0.05 was considered statistically significant. The statistical analysis was performed using the statistical analysis system (SAS) statistical package (version 9.3) (SAS Institute, Cary, North Carolina, USA).

Table 1 Patients' characteristics during their ER[a] visit and admissions.

NMDs[b]	n[c] (%)	Age (years) Mean ± SD[d]	Number of ER[a] visits (%)	Number of ER[a] admissions (%)
Spinal muscular atrophy	15 (34.1)	8.5 ± 5.8	60 (29.4)	31 (30.1)
Congenital, hereditary muscular dystrophy	12 (27.2)	10.3 ± 6.0	90 (44.1)	35 (34.0)
Hereditary, progressive muscular dystrophy	17 (38.6)	10.8 ± 5.1	54 (26.5)	26 (25.2)
Total	44	9.9 ± 5.6	204	103[e]

[a] ER, emergency room.
[b] NMDs, neuromuscular disorders.
[c] n: number of patients.
[d] SD, standard deviation.
[e] 11 admissions (10.7%) were unclassified NMDs among the 103 visits.

3. Results

Forty-four children with NMD received emergency medical care from the hospital between January 2005 and January 2015. Tables 1 and 2 present the NMD patients' characteristics (i.e., NMD subtypes, sex, and age) and the distribution of the types of NMD among ER visits or admissions to the hospital from the ER. Differences in sex and age distributions were not statistically significant between each NMD. Of the 44 enrolled children with NMD, 36 children required admissions (81.8%), and 20 of them had been readmitted (55.6%).

Although 204 emergency outpatient visits occurred, 112 triage classification results were collected, and the most common triage classification among the patients was triage 3 (35.3%). Of the 204 emergency visits, 103 (50.5%) patients were admitted to the hospital, of whom 74 (71.8%) were admitted to general wards and 29 (28.2%) were admitted to the PICU. Eleven admissions from the ER were unclassified due to limitations of the database used, but these admissions were believed to belong to those with three subgroups of NMDs. One hundred and one patients (49.5%) were discharged from the ER. One patient experienced acute respiratory failure that resulted in mortality within the 10-year period. The results revealed that the proportions of the emergency inpatients (50.5%) and outpatients (49.5%) were both approximately 50%; 19 out of 44 (43.2%) pediatric patients with NMD required only one ER visit, whereas 25 (56.8%) required more than one ER visit.

The major diagnoses/symptoms that led to ER visits, as well as causes of admission, are presented in Table 3. The major symptoms for which patients presented to the ER were usually multiple and concurrent, particularly involving the respiratory system (62.3%), including pneumonia, common cold, and respiratory failure; and gastrointestinal complications (28.9%), including acute gastritis, abdominal pain, and gastrointestinal hemorrhage. Similarly, respiratory and digestive complications accounted for most hospitalizations. For all the patients with NMD, only the number of cases with pneumonia ($p = 0.047$), respiratory failure ($p = 0.001$), and gastrointestinal hemorrhage ($p = 0.047$) were significantly different between the patients who visited the ER and those who were admitted to the hospital.

After analyzing variables between the cause of admissions and NMD subgroups, we found that acute gastritis was the statistically significant cause of admission among 12 out of the 35 hospitalized patients with congenital hereditary muscular dystrophy ($p = 0.0074$, chi-square test; software output data not shown). The results remained statistically significant after Fisher's exact test analysis for acute gastritis and congenital hereditary muscular dystrophy ($p = 0.0043$, Fisher's exact test; software output data not shown). However, neither pneumonia-associated ER visits nor admission rates were statistically significantly associated with ER admission among the NMD subgroups ($p = 0.7605$, analysis of variance and chi-square test; data not shown).

Table 4 presents the examinations and treatments arranged for children with NMD in ER. The most commonly used laboratory tests performed were complete blood count/differential count and examination of C-reactive

Table 2 Sex and age distribution in the ER[a] records.

	Sex			Age (years)		
	Male (%)	Female (%)	Fisher's test[b] *p*-value	Median	IQR[c]	Wilcoxon test[d] *p*-value
Number of patients visiting ER (44)	30 (68.2)	14 (31.8)	—	11.1	10.6	—
Number of patients admitted to the Ward/ICU[e](36)	25 (69.4)	11 (30.6)	0.695	10.7	10.6	0.494
Number of patients discharged from the ER (8)	5 (62.5)	3 (37.5)	Ref[f]	12.0	6.0	Ref[f]
Number of patients admitted to the ICU[e] (20)	11 (55.0)	9 (45.0)	0.999	11.3	13.5	0.666

[a] ER, emergency room.
[b] Fisher's test, Fisher's exact test compared to Ref.
[c] IQR, interquartile range.
[d] Wilcoxon rank-sum test compared to Ref.
[e] ICU, intensive care unit.
[f] Ref, reference.

Table 3 Symptoms/Diagnosis of ER[a] visits and hospitalization of pediatric patients with NMD[b].

	Number of ER visits (x)	%	Number of admission from ER (x)	%	Chi²[c] *p*-value
1. Pneumonia	75	36.8	50	48.5	0.047
2. Acute gastritis	47	23.0	21	20.4	0.597
3. Common cold[d]	41	20.1	14	13.6	0.161
4. Respiratory failure	11	5.4	17	16.5	0.001
5. Heart failure	9	4.4	3	2.9	0.522
6. Fever	8	3.9	0	0.0	0.055[e]
7. Abdominal pain	8	3.9	≦2[f]	—	0.281[e]
8. Gastrointestinal hemorrhage	4	2.0	7	6.8	0.047[e]
9. Seizure/epilepsy	4	2.0	≦2[f]	—	0.667[e]

[a] ER, emergency room.
[b] NMD, neuromuscular disorders.
[c] Chi², chi square test.
[d] For the study's purposes, common cold included acute pharyngitis, acute bronchitis, and acute tonsillitis (because secondary diagnoses occurred, the sum of the percentage exceeded 100%).
[e] Fisher's exact test.
[f] When the number is less than or equal to two, it must be concealed to ensure compliance with the requirements of the data provider.

Table 4 ER[a] evaluations and treatments received by pediatric patients with NMD[b].

	Number (n)	Percentage (%)[h]
1. Blood routine	78	38
2. CRP[c]	78	38
3. IV[d] hydration	69	34
4. GOT/GPT[e]	68	33
5. Electrolyte/sugar	67	33
6. BUN[f]/Creatinine	62	30
7. Chest film	57	28
8. Aerosol therapy	36	18
9. Oximetry	30	15
10. Blood culture	25	12
11. Plain abdomen film	24	12
12. CK[f]	22	11
13. Blood pH/gas analysis	23	11
14. CK-MB[g]	13	6

[a] ER, emergency room.
[b] NMD, neuromuscular disorders.
[c] CRP, C-reactive protein.
[d] IV, intravenous.
[e] GOT/GPT, glutamic oxaloacetic transaminase/glutamic pyruvic transaminase.
[f] BUN, blood urea nitrogen; CK, creatinine kinase.
[g] CK-MB, creatinine kinase-MB.
[h] Due to multiple evaluations conducted at each ER visit, the sum of the percentages exceeded 100%.

protein levels to assess the infectious condition, while the most commonly prescribed intervention was intravenous fluid supplement, followed by aerosol/oxygen therapies. However, even though pulmonary compromise resulted in most ER visits and admissions, pulmonary complication assessments, including chest films (28%), pulse oximetry (15%), and blood gas analysis (11%), were performed in a relatively small proportion of cases.

4. Discussion

Only two previous studies have evaluated the patterns of healthcare use among children and young adults with muscular dystrophy.[1,2] However, neither study discussed other NMD subgroups, including motor neuron disease, such as spinal muscular atrophy (SMA). To our knowledge, this is the first study seeking to identify unmet needs of children with NMD by characterizing the presentation, examinations, interventions, subsequent flow to discharge or admission, and outcomes in the ER setting. The symptoms of NMD may be misunderstood in the ER, because emergency care providers are not familiar with NMD; therefore, an ER visit may result in inappropriate care for these patients, such as unnecessary tracheostomy when an ER physician emphasizes aggressive care.[12] Indeed, a prior study showed that among patients with neuromuscular respiratory failure, those without known diagnosis before admission had poorer outcomes.[13] In our study, we analyzed emergency cases of pediatric NMD in an effort to assist pediatric emergency physicians in enhancing the quality of emergency care provided to this patient population. The chief concerns of the children with NMD presenting at each ER visit were usually multiple and concurrent, particularly involving respiratory (62.3%) and gastrointestinal issues (28.9%). More than half of the children with NMD who visited the ER required hospitalization or had been admitted repeatedly. Infections of the respiratory system, particularly pneumonia, accounted for the primary cause of hospitalization in 48.5% of the cases, followed by acute gastritis and its potential complications (27.2%). Correspondingly, a recent study of young adults with muscular dystrophies showed significantly greater ER use for acute conditions, including respiratory and digestive problems.[2] Notably, we further identified a discrepancy between ER visit and admission rates for severe respiratory or gastrointestinal decompensations in children with NMD, especially those with respiratory failure, pneumonia, or gastrointestinal hemorrhage. This finding suggests that ER physicians may play a pivotal role in identifying potential risks/exacerbating factors as well as in providing timely interventions to efficiently avert the progression of NMD-related complications in children with NMD presenting to the ER with an acute illness.

In the present study, majority of the patients were boys aged 9—10 years, although girls outnumbered boys among those who were admitted to the PICU. The larger number of boys possibly accounts for the proportional dominance of some specific NMDs with X-linked recessive inheritance; however, this was not observed in the SMA group. These results may be further explained by the course of common NMDs, specifically Duchenne muscular dystrophy, in which

more complications may occur after boys are 10 years old.[14,15] Alternatively, the most common subtype of NMD associated with ER visits and admissions was congenital hereditary muscular dystrophy, suggesting that this NMD subgroup not only comprises a more heterogeneous spectrum, but that its associated complications may require more frequent emergency care. It has been proposed that children with muscular dystrophy use ER services more frequently and have higher expenditures of acute care service than those with other special healthcare needs.[1] Furthermore, a recent study also indicated that adolescents with muscular dystrophy, including congenital muscular dystrophies, required more frequent ER visits and hospitalizations, especially as they became older.[2] Our data also suggest that more unmet ER care needs might exist in this subgroup of patients with NMD.

Respiratory complication is the primary cause of morbidity and mortality in children with NMD. In particular, children with NMD represent a distinct subgroup, and they are characterized by a slow decrease in muscle function, often precipitated by acute respiratory events.[16] As muscular atrophy and weakness ensue, these children may experience ineffective cough and clearing of airway secretions, aspiration of salivary and oral contents, atelectasis, frequent recurrent respiratory infections, pneumonias, airway obstruction, and gas exchange abnormalities.[17,18] Multiple factors contribute to the progression to respiratory failure, including poor clearance of lower airway secretions, dysphagia with aspiration, recurrent pneumonias, scoliosis, and poor nutritional status.[3–6] Viral respiratory tract infections, mostly known as common cold, can increase respiratory muscle weakness and secretions, which can hasten the time course of progression to acute respiratory failure among patients with NMD.[19] As a result, it is possible that some of the cases in our cohort were admitted to the hospital with a simple diagnosis of common cold due to a concern of deleterious progression raised by a clinical judgment for younger age groups. These patients are ideal candidates for non-invasive ventilation, which should be performed as early as possible, especially if signs that suggest respiratory failure become apparent.[16,17,20] Previous studies also showed that the use of non-invasive ventilation combined with assisted cough machine could significantly reduce hospital admission for respiratory complications in children with NMD.[8,18]

Our data further suggest that the laboratory investigations and interventions for pulmonary compromise should be performed early and be actively used in the ER settings among patients who present with acute respiratory illness. However, we were surprised to find that the laboratory tests and interventions for pulmonary complications in the ER had not been prescribed as frequently as we would have anticipated. Chest radiography, which can provide an instant clue to mucus-complicated atelectasis or pneumonia, was only ordered in 28% of the ER visits, and in less than 36.8% and 48.5% of the pneumonia-associated ER visits and admissions, respectively. Blood gas analysis and non-invasive pulse oximetry, which are recommended for acute respiratory events in patients with NMD,[4,8,20] were performed only in 11% and 15% of the ER visits, respectively. Emergency physicians should perform blood gas analysis or at least noninvasive oximetry to more thoroughly, conveniently, and efficiently understand the severity of respiratory distress in pediatric patients. Early or prophylactic use of empirical antibiotics in the ER should be considered in children with NMD to prevent progression of pneumonia[3,5,16,18]; this may also decrease the number of readmissions to the ER. However, antibiotic administration was not evaluated in this study. Further studies are warranted to identify whether antibiotics can be used early in the ER in children with NMD exhibiting pneumonia.

In this study, we demonstrated that 18% of the pediatric patients received aerosol therapy in the ER. Aerosol therapy involves delivering oxygen or bronchodilator/mucolytics to patients with NMD to enhance expectoration of secretion and relief of respiratory distress. However, emergency physicians may not fully perceive the pathophysiology of respiratory distress in patients with NMD, which is usually associated with hypercapnia complicated by hypoventilation or secretion-plugging, rather than by oxygen deficits. Using oxygen alone can worsen alveolar hypoventilation; hence, supplemental oxygen is not the first line of treatment in patients with NMD experiencing respiratory distress.[18,20] Therefore, supplemental oxygen should be provided only if it is concurrent with the support of non-invasive positive pressure ventilation, with or without assisted cough, which can be adjusted as needed to maintain pulse oximetry at $>$93%, except in an acute respiratory crisis.[8,18,20]

We found that gastrointestinal complications (specifically acute gastritis) are second to pneumonia as the major diagnosis during ER visit and hospitalization among children with NMD. It has been proposed that patients with muscle weakness are prone to digestive problems, for example, gastroesophageal reflux, delayed gastric emptying, gastrointestinal dysmotility, and constipation.[4,5,9] Interestingly, we demonstrated that there is a statistically significant correlation between acute gastritis-related admissions and patients with congenital hereditary muscular dystrophy. Indeed, feeding and swallowing difficulties have been identified as significant problems for some patients with congenital muscular dystrophy.[3] Further research is needed to examine whether this NMD subgroup is more susceptible to acute gastritis-related complications. Nevertheless, during acute illness, nutrition and hydration should be adequately maintained.[4,5,20] The frequency and volumes of tube feedings may be adapted to reduce the risk of gastroesophageal reflux and meet fluid and nutrient requirements. Symptoms or signs of gastritis or gastroesophageal reflux should trigger the prompt initiation of treatment with proton-pump inhibitors and/or H2-antagonists. Lastly, recent studies have also suggested that gastrostomy placement in patients with NMD may decrease gastroesophageal reflux and decrease episodes of aspiration pneumonia and hospitalizations.[16,17]

This study has some limitations. It was conducted in a single tertiary Taiwanese medical center, and it was uncontrolled and nonrandomized. Although the sample data were collected within a 10-year period, the study population was still limited. Nevertheless, we propose to study the pediatric group exclusively. Some data may have been missing or overlapping in the hospital database, and the prevalence of the disease might have been underestimated. In addition, NMD can be caused by autoimmune disease or inflammatory

disorders; however, this study only focused on genetic/hereditary disorders. Unfortunately in this study, we could not identify the subclassification of NMD for all the enrolled patients due to the retrospective nature of the ER record review, which could provide only limited information from the ICD-9 coding system. These limitations should be considered when interpreting the results.

In conclusion, children with NMD require a documented emergency plan which specifies a clear point of access for emergency care, even though investigation for and management of complicated NMD in the ER have not been performed as frequently as anticipated. It is critical to ensure that the ER staff recognizes the unmet needs of children with NMD during an emergent circumstance. A specifically designed alert card that advises ER physicians of the special needs of children with NMD in the scenario of acute illness would be worth advocating in the future. In addition, follow-up visits and intervention are also crucial to reduce the frequency of ER visits and admissions and to maintain a better quality of life for these debilitated children.

Conflict of interest statement

No conflicts of interest.

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