Abstract:
Objective
Neuromuscular disorders (diseases of the motor unit), can cause respiratory problems such as impaired cough reflex, chest deformity, recurrent pneumonia and acute respiratory failure; these are the worst most common complications of these diseases and the leading cause of death in such patients (1, 2). Their management hence, very often, entails admission to the Pediatric Intensive Care Unit (PICU) (3,4) and during this phase, endotracheal intubation is almost always necessary, to maintain the patency of airways and to apply Positive Pressure Ventilation (PPV). However, endotracheal intubation is always temporary, and its success or failure depends on the timely decision of its termination to restore the normal respiration or to avoid the risk of recurring respiratory failure (5, 6). We designed this study to evaluate the role of neuromuscular disorders in causing extubation failure as compared to that of other risk factors.

Materials & Methods
In an analytical cross-sectional study, the risk factors of reintubation and duration of mechanical ventilation in two groups of 30 patients each, was compared, the first successful extubation and the second with extubation failure.

Results
Neuromuscular disorders (including Spinal Muscular Atrophy, Guillain-Barre' Syndrome, Congenital Myopathies and Muscular Dystrophies) were the main underlying diseases in extubation-failure group (P= 0.0002). Hypercapnia (PaCO2>50mmHg) was shown to be the most common cause of both the first intubation (P=0.001) and reintubation (P=0.004) in the group of patients who failed extubation. The mean duration of intubation and mechanical ventilation was longer in patients with neuromuscular disorders who had extubation failure (P= 0.01).

Conclusion
This study showed that, as underlying problems, neuromuscular disorders are the most common causes of prolonged intubation which defeat weaning from the ventilator and result in reintubation by inducing hypercapnia. Therefore the weaning process needs to be done gradually in these patients, and in conjunction with supportive measures, such as close observation for at least for 72 hours following extubation to monitor any possibility of recurrence of hypercapnic respiratory failure.

Key words: reintubation, neuromuscular disorders, respiratory failure
Introduction

Extubation failure is defined as a need for reintubation up to 72 hours after planned extubation (7), and may occur in 2-25% of all planned endotracheal extubations (8,9); endotracheal intubation is performed to maintain the airway patent and provide a route for positive pressure ventilation (PPV). Although a major therapeutic measure in PICUs and occasionally the sole way to save the patient’s life in a critical life-threatening situation, endotracheal intubation is always a temporary measure taken and should be discontinued apropos. The decision for timely of extubation, to restore patient’s normal respiration, is of crucial importance and constantly accompanied by some risk of recurrent respiratory failure, making reintubation necessary. The adverse effects of reintubation are increased risk of nosocomial pneumonia, prolonged ICU stay and finally, increased morbidity and mortality rates. Neuromuscular disorders are diseases of the motor unit, which consists of a single motor neuron, its axon, the neuromuscular junction and all muscle fibers innervated by it. These disorders include: Spinal Muscular Atrophy, Guillain-Barre’ Syndrome, Congenital Myopathies and Muscular Dystrophies (1,2). Respiratory problems such as recurrent pneumonia, chest deformity and acute respiratory failure are the worst complications and leading causes of death in these patients, making admission to PICU, mostly a supportive measure, a very common event in their medical management process (3, 4). We undertook this study to investigate the role of neuromuscular disorders in causing extubation failure as compared to that of other risk factors.

Materials & Methods

This is an analytical cross-sectional study involving 60 patients consecutively admitted to the PICU of Tabriz Children’s Hospital between January and November 2005; cases were selected using the convenience sampling method, divided into two groups of 30 patients each, one with successful extubation and the other with extubation failure. Inclusion criteria included age over 30 days and below 14 years (infants and children); PICU admission; endotracheal intubation (for positive pressure ventilation) and planned extubation for all patients; moreover, need for endotracheal intubation and mechanical ventilation, again, was considered for extubation-failure group. Data collected for all patients included age, sex, underlying disease which caused first intubation and necessitated mechanical ventilation, duration of mechanical ventilation before extubation and blood gas analysis results before first intubation and extubation. Variables such as age and sex were matched between these two groups, and their risk factors for reintubation compared. Causative factors of extubation failure and blood gas analysis results before reintubation were determined in patients of related group. Incidental extubation was considered as exclusion criteria. Finally all data collected was statistically analyzed using the T-test to compare the risk factors of reintubation in PICU patients.

Results

In the first or ‘successful extubation’ group there were 17 males and 13 females with a mean age of 18.3+1 months and in the extubation failure group there were 17 males and 13 females with a mean age of 17.7+9 months (P = Not Significant). Underlying disorders, in the successful extubation group included: Aspiration pneumonia in 10 patients (33.3 %); congenital heart diseases with or without pneumonia in 9 (30%); shock in 5 (16.7%); neuromuscular disorders in 3 (10%); apnea in 2 (6.7%) and drug poisoning in 1 patient (3.3%). In second or extubation failure group, underlying disorders included: Neuromuscular disorders in 15 patients (50%); congenital heart disorders with or without pneumonia in 5 (16.7%); neuromuscular disorders in 3 (10%); apnea in 2 (6.7%) and drug poisoning in 1 patient (3.3%). As is mentioned in table 1, neuromuscular disorders are the most important underlying causes of extubation failure (Table1).
"Hypercapnia" (increased PaCO2 to more than 50 mmHg) at the time of first intubation and reintubation is the dominant type of respiratory failure in the patients with failed extubation. Mean duration of endotracheal intubation and mechanical ventilation in the patients with successful extubation was 60 ± 73.8 hours, whereas the same period for extubation-failure group was 123.5 ± 115 hours, (P= 0.01).

**Discussion**

Worldwide medical literature documents neurological disorders, congenital heart diseases and inborn errors of metabolism to be the most frequent underlying disorders of ventilator dependency and extubation failure(7,8). The main mechanisms that result in extubation failure include imbalance between strength of respiratory muscles and labor of breathing, hypersecretion into airways, upper airway obstruction, insufficient cough reflex and deficient respiratory center drive. The final manifestation of these mechanisms may be either hypoxemia or hypercapnia (8,10,11).

Our study shows that neuromuscular disorders as underlying diseases, are the most important cause of defeat in the process of weaning from the ventilator and the main cause of reintubation, findings that are compatible with the studies of Edmunds et al. and Epstein et al (11,12). In a similar study Fraser et al. claimed that 27% of children requiring mechanical ventilation for more than a month had a neuromuscular disorder (13).

It was revealed that in any patient, the longer the duration of mechanical ventilation, the greater the likelihood of extubation failure and the need for reintubation; these results have also been reported by Edmunds et al (12). The present study shows that patients who undergo mechanical ventilation for the first time, because of hypercapnic respiratory failure, are more susceptible to dependency on mechanical ventilation, increasing hence the risk of extubation failure in such patients. Besides, they are more likely to have to undergo further reintubation and mechanical ventilation because of relapsed hypercapnia (table 1).

Ventilator weaning and extubation of these patients should be planned very cautiously; ventilatory support may be diminished gradually and tapered off by small decrements. After completion of weaning and extubation, it is also necessary to continue or even increase the respiratory care such as upper airway toilet, suction of secretions, chest physiotherapy, non-invasive oxygen therapy and so on (14). All these measures were applied.

**Table 1. Comparison of underlying disorders and blood gas imbalances in two groups of patients with successful and with failed extubation**

<table>
<thead>
<tr>
<th>Disorders &amp; Imbalances</th>
<th>Neur-</th>
<th>At the time of first intubation</th>
<th>At the time of reintubation</th>
<th>P Value</th>
</tr>
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<tr>
<td></td>
<td>omuscular Disorders</td>
<td>Hypercapnia PaCO2&gt;50 mmHg</td>
<td>Hypoxemia PaO2&lt;50 mmHg</td>
<td>Hypercapnia PaCO2&gt;50 mmHg</td>
</tr>
<tr>
<td>Successful</td>
<td>3</td>
<td>27</td>
<td>1</td>
<td>29</td>
</tr>
<tr>
<td>Failed</td>
<td>15</td>
<td>15</td>
<td>18</td>
<td>12</td>
</tr>
<tr>
<td>Total</td>
<td>18</td>
<td>42</td>
<td>19</td>
<td>41</td>
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<tr>
<td></td>
<td>P = 0.001</td>
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equally for all patients in this study to obviate confounding factors such as insufficient care or a hasty weaning process. In similar studies, it has been declared that complications such as nosocomial and ventilatory associated pneumonias, prolonged ICU stay and need for tracheostomy are more common in patients who had sustained reintubation. Also reported were poor final outcomes, increased morbidity and mortality rate (8,9,10,11,12,15).

**Conclusion**
The highest risk of extubation failure and need for reintubation are seen in the hypercapnic type of respiratory failure, which predominantly complicates neuromuscular disorders. It is therefore, better to wean gradually and prudently, along with accurately implementing supportive measures such as clearing of secretions and oxygen therapy.

**References**