

# Management of gastrointestinal and nutritional problems in children on home invasive mechanical ventilation

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## Abstract

In recent decades, the patient survival is increased due to the advances in intensive care units and development of modern mechanic ventilators. Unfortunately, it is not always possible to wean these children from mechanical ventilation. Recently, after placement a tracheostomy tube, they can support at home with non-invasive or invasive mechanical ventilation. Most of the children who need ventilation support at home have neurological impairment. The nutritional issues and gastrointestinal complications are well defined in critically ill patients, but there are very limited studies on the children with tracheostomy. Considering that majority of the patients has neuromuscular disorders, the nutritional and gastrointestinal problems of the children with tracheostomy are discussed, in the light of the knowledge on critically ill patients.

## Management of gastrointestinal and nutritional problems in children on home invasive mechanical ventilation

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## Abstract

In recent decades, the patient survival is increased due to the advances in intensive care units and development of modern mechanic ventilators. Unfortunately, it is not always possible to wean these children from mechanical ventilation. Recently, after placement a tracheostomy tube, they can support at home with non-invasive or invasive mechanical ventilation. Most the children who need ventilation support at home have neurological impairment. The nutritional issues and gastrointestinal complications are well defined in critically ill patients, but there are very limited studies on the children with tracheostomy. Considering that majority of the patients has neuromuscular disorders, the nutritional and gastrointestinal problems of the children with tracheostomy are discussed, in the light of the knowledge on critically ill patients.

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## **Management of gastrointestinal and nutritional problems in children on home invasive mechanical ventilation**

Advances in neonatal/pediatric intensive care units and mechanical ventilation (MV) over the last three decades have increased the patient survival in worldwide. Despite increased survival, the patients who couldn't be weaned from MV in intensive care units due to chronic respiratory failure caused by different etiologies, have been emerged the need of MV support at home. With the development of more modern and advanced home ventilators, home invasive mechanical ventilation (HIMV) has been used widely in children with chronic respiratory failure all over the world<sup>1-3</sup>.

The children who have progressive respiratory failure or who could not weaned from MV in intensive care units need long term mechanical ventilation. When a non-invasive MV support is not an option, invasive MV support via tracheostomy can be continued at home, after clinical stability have been established at hospital<sup>1,4</sup>. To be a clinically stable child the followings are required; a stable airway, stable oxygen requirement (< 40%), stable ventilator settings that haven't been changed over a period of time, the need for infrequent laboratory testings, a stable home care plan and an adequate nutrition to maintain growth<sup>1,4</sup>. A safe and stable enteral feeding is as important as a stable airway. While preparing the child to discharge, not just preparation for MV, but also a detailed nutritional assessment and adjustment should be done. The caregiver (the parents, the homecare providers) training should be performed not only for MV, but also for nutritional issues before discharge. Besides giving information for the acute life-threatening events related to MV and underlying disease, the importance of nutrition and negative effects of inadequate nutrition on growth, development and respiratory functions should be explained<sup>1,5</sup>.

In childhood HIMV is generally required for neuromuscular, central nervous system and cardiopulmonary disorders. The indications for HIMV support in children are given in Table 1. The majority of these children have neuromuscular disorders, therefore it is not possible to directly associate the gastrointestinal and nutritional problems with mechanical ventilation<sup>1-3</sup>.

### **Nutritional assessment and nutrition management of the children with tracheostomy**

The global problem of the world, malnutrition leads to loss of lean body mass, growth and developmental delay, muscle weakness (e.g., decreased respiratory muscles' and diaphragm strength), immune dysfunction (e.g., altered humoral and cellular immunity and increased bacterial adhesions in the lower respiratory tract, increased infections), delayed wound healing and prolong hospital stay<sup>6-8</sup>. To prevent and improve malnutrition, the children with tracheostomy and HIMV should be evaluated for the nutritional status at hospital and after discharge regularly. The frequency of the visits is determined by the patients' malnutrition degree, weight gain rate, presence of feeding intolerance or gastrointestinal symptoms. If the patients are stable, visits should be performed every 1 to 3 months for infants and every 3-6 months for older children<sup>1-3,6</sup>.

The nutritional assessment of children consists of a detailed dietary history, physical examination findings, anthropometric measurements and basic laboratory tests. During hospitalization and at every visit, patients' detailed dietary intake (including water) and nutritional requirements should be determined. The deficiencies of vitamin and trace elements may reveal some pathological physical findings such as angular cheilitis, keratitis, dermatitis, hair loss. Decubitus skin ulcers and poor peripheral circulation findings are the important physical examination findings related to undernutrition particularly in neurologically impaired patients. The anthropometric measurements should be recorded during hospitalization and should be routinely monitored in every visit. The length or height, body weight, head circumference measurements, weight for length and body mass index values should be evaluated due to age and gender of the patients and z-scores should be calculated. The growth charts and z-scores are very useful to monitor the growth of the children. The segmental measurements such as tibial and knee-heel length are useful in estimating height of the patients who cannot be measured due to severe contracture. In addition mid-upper arm circumference and triceps skin fold measurements are valuable in detecting acute malnutrition and also determining the effect of nutritional

support on follow-up. Malnutrition definition generally based on the antropometric measurements. The severity of malnutrition is defined according to z-scores. Weight for age, weight for height, body mass index are mostly used indices to define malnutrition. European Society for Pediatric Gastroenterology, Hepatology, and Nutrition (ESPGHAN) and Academy of Nutrition and Dietetics/American Society of Parenteral and Enteral Nutrition (ASPEN) recommendations for definition moderate-severe acute malnutrition are given in Table 2. Also Academy of Nutrition and Dietetics/ASPEN defined inadequate nutrient intake as a risk factor for severe and moderate malnutrition, when nutrient intake is [?]25% and 26%-50% of estimated energy/protein requirement, respectively<sup>7,9-11</sup>.

Routine laboratory evaluation including hemoglobin levels, renal function tests, albumin, liver enzymes, fasting blood glucose should be performed. ESPGHAN recommended routine screening of vitamins (vitamin D, vitamin B12, folate, vitamin A, vitamin E), macrominerals (calcium, phosphate, magnesium), trace elements (zinc, iron), ferritin and parathormon in children with chronic conditions under high risk of malnutrition. The measurements should be performed in the absence of systemic inflammation because blood levels of vitamins and trace elements may be affected from systemic inflammation. In addition, a simultaneous measurement of acute phase reactans and albumin level is required<sup>6,9,12</sup>.

The main purpose of nutritional support is to optimize growth and development. The nutritional requirement of every children changes due to own clinical condition and underlying disease. The patients who have undernutrition need higher energy requirements to catch-up growth.

Energy requirement of the children with neurological impairment (NI) differs due to the heterogeneity of clinical features. It depends on severity of the NI, muscle tone, physical activity level, mobility and presence of malnutrition. ESPGHAN recommended the use of dietary reference intake (DRI) standarts to estimate the energy need. But sometimes DRI may overestimate the energy needs of the children with NI. For instance total energy expenditure (TEE) / resting energy expenditure (REE) ratio is lower in quadriplegic cerebral palsy compared to children with normal activity (TEE/REE ratio is 1.1 and 1.5-1.6 respectively). Additionally the overestimation of energy need may result in obesity or being overweight. On the other hand the children with athetosis need more energy. At least in the beginning 'The Schofield equation' may be used to calculate basal metabolism rate (Table 3)<sup>13</sup>. On follow-up according to the weight gain velocity, activity and stress factors further dietary interventions can be made. In children with NI, recommended DRI for protein is same for healthy children. In case of decubitus ulcers or severely undernourished patients the protein intake may be increased to 2-2.4 g/kg/day<sup>6,9</sup>. The DRI values for energy and protein for healthy children are given in Table 4<sup>14</sup>.

In the patients with Duchenne muscular dystrophy (DMD), the Schofield equation considered as an accurate predictive equation and they should receive 100% of DRI for energy. The energy requirement should be specialized based on ambulation, physical activity and current weight of the patient. For instance, some reports suggested the estimated energy need is 80% of DRI in ambulatory DMD boys, and 70% of DRI in non-ambulatory boys with DMD. In the presence of weight loss, energy intake can be increased to 130%-150% of DRI in DMD boys. Similarly protein intake should meet the DRI for age<sup>15,16</sup>.

The accurate estimation of REE of the infants with spinal muscular atrophy (SMA) type 1 should be based on nutritional and ventilation status (spontaneous, non-invasive or invasive MV) of the patient. The tracheostomy and invasive MV leads to 50% of decrease in REE, but spontaneously breathing infants need more energy. The estimated amount of energy is recommended to be 9-11 kcal/cm/day by 'The International Standarts of Care for SMA'<sup>17</sup>.

The patients with cystic fibrosis may have increased REE and increased nutrient loss due to pulmonary, pancreatic and hepatic involvement To maintain growth, development and pulmonary functions, daily energy and protein requirements should be 120-150% of DRI and 150%-200% of DRI<sup>16</sup>. The recommended daily energy intake for the infants with bronchopulmonary dysplasia (BPD) is 120-150 kcal/kg<sup>18</sup>. Luo J<sup>19</sup> et al. reported a significant increase in weight and length z-scores of the infants with BPD and a decreased energy requirement within 4 weeks after tracheostomy placement.

The water intake is adjusted based on the healthy children's daily requirements and can be calculated by Holliday Segar formula (Table 5)<sup>20</sup>. Daily water need is equal to daily caloric expenditure and approximately 100 ml/kg water for every 100 kcal/kg is needed in normal physiologic conditions. The water losses due to pathological conditions should be given separately<sup>15,20</sup>.

Enteral tube feeding is required when the patients can't feed by oral route or oral intake is not sufficient for nutrition, hydration, growth and even medication intake. It may be performed through a nasogastric, nasojejunal, percutaneous endoscopic gastrostomy (PEG), or jejunal tube [PEG with a jejunal extension (PEG-J), percutaneous endoscopic jejunostomy (PEJ) or surgical jejunostomy]. When the patient need non-oral nutrition support more than 3-6 weeks, PEG is indicated<sup>21</sup>. There is no need to investigate the children for gastroesophageal reflux in the absence of related symptoms prior to PEG placement<sup>6,21</sup>. The long term MV requires a safe enteral feeding route. Goneidy A. et al<sup>22</sup>. suggested to insert a gastrostomy tube at time of tracheostomy formation in children receiving long term ventilation and have been already feeding with NG tube.

Percutaneous endoscopic gastrostomy placement should be performed under general anaesthesia by a trained staff. Aseptic cleaning (simple washing is sufficient) and sterile dry dressing of the wound site is recommended for one week. While site is healing, a clear discharge may be seen. The parents or caregivers should be trained for feeding tube and stoma care. They should be informed for possible complications such as tube removal, clogging. In daily care, infectious signs should be looked for the gastrostomy site. The parents or caregivers should be informed for the importance of nutrition. They should be trained for feeding techniques before discharge<sup>21,23</sup>.

Three hours after PEG placement, feeding can be started. Isoosmolar formulas are the best choice to start feeding. The first choice should be bolus feeding because it is more physiological. If the patient hasn't tolerated the bolus feeding via NG tube previously, the alternative will be continuous feeding. The targeted energy intake is determined by assessing the nutritional status of the children. The feeding volume should be increased gradually both in bolus and continuous feeding because excessive feeding may cause abdominal distension, which may affect respiratory function. A stepwise increase of feeding is useful to monitor feeding intolerance or the other gastrointestinal complications<sup>21</sup>.

Enteral feeding formulas are chosen according to the patients age, underlying disease and tolerance. Breast milk, infant formulas, isocaloric and hypercaloric formulas may be used for enteral feeding. Formula may be changed depend on calorie requirements or volume restriction. Also feeding type or content of formula can be changed due to feeding intolerance in follow-up<sup>21</sup>. Adjusting a safe feeding regimen and optimal formula before discharge is essential for the patient with tracheostomy,

### **Gastrointestinal problems in children on home invasive mechanical ventilation**

The gastrointestinal (GI) problems in critically ill patients who were given MV support in intensive care units have been demonstrated both in animal and human studies<sup>24-26</sup>. During MV, splanchnic hypoperfusion seems to play an important role in the pathogenesis of GI complications due to cardiocirculatory dysfunction. In particular, it was reported that high positive end-expiratory pressure (PEEP) levels (PEEP >15 cmH<sub>2</sub>O) cause an increased right ventricular afterload which lead to an increase in right atrial pressure, decreased systemic venous return and cardiac output and splanchnic perfusion respectively<sup>24-27</sup>. Also acute moderate increase in arterial carbondioxide level has been observed to increase hepatic and splanchnic blood flow in a biphasic manner; initially reducing of blood flow due to sympathetic stimulation, then increasing blood flow due to direct vasodilator effect of carbondioxide<sup>28</sup>. Despite there are conflicting results on the effect of PEEP on hepatic blood flow as decreased or no change. The decrease in portal flow may be compensated with increased arterial blood flow in order to maintain hepatic blood flow<sup>29,30</sup>. The gastro-esophageal reflux disease, dysphagia, delayed gastric emptying and constipation are the most common gastrointestinal problems of the children with tracheostomy on MV<sup>31-33</sup>.

### **Gastro-esophageal reflux disease**

Gastro-esophageal reflux (GER) is the passage of gastric contents into the esophagus, with or without regurgitation and vomiting. North American Society for Pediatric Gastroenterology, Hepatology, and Nutrition (NASPGHAN) and ESPGHAN defined pediatric gastro-esophageal reflux disease (GERD) as GER leads to troublesome symptoms which affect daily functioning and/or complications, such as hematemesis, dysphagia, erosive esophagitis, poor weight gain, feeding refusal, dystonic neck posturing, apnea, recurrent aspiration pneumonia, coughing or choking at the end of feeding in infants and children. The children who had specific underlying conditions such as neurological disease, pulmonary disease (e.g., cystic fibrosis), congenital gastrointestinal abnormalities (e.g., diaphragmatic hernia, esophageal atresia, intestinal atresia.), obesity or prematurity are more vulnerable for GERD<sup>34</sup>. Lower esophageal sphincter (LES), angle of His (angle between the esophagus and the gastric fundus), the crural diaphragm and phrenoesophageal ligament are the components of the anti-reflux barrier. The LES relaxes during swallowing and allows the pass of esophageal contents to the stomach. The postprandial gastric distension triggers transient LES relaxations (TLESRs) and the gastric contents move upward into the esophagus. Despite TLESRs are considered to be the major cause of GER episodes, delayed gastric emptying, decreased LES pressure, increase in the His angle, increased intraabdominal pressure may lead GER. Coughing, straining, increased respiratory effort, medications, obesity, lying on right side position may cause GER due to anti-reflux barrier impairment<sup>35-38</sup>.

In the study of mechanical ventilated critically ill patients, the basal LES pressure was found to be low and a minimal increase in abdominal pressure due to such as suctioning, straining, coughing may cause frequent reflux episodes. Also medications like anti-cholinergic drugs, calcium canal blockers, barbiturates may relax LES<sup>39</sup>. Enteral nutrition related gastric distension or delayed gastric emptying may increase the frequency of TLESRs and causes increased GER episodes in these patients<sup>31</sup>. Newton M et al<sup>40</sup>. reported that GERD was diagnosed in 20% of the children with tracheostomy and of these children 12.4% had tracheostomy related complications, with odds ratio of 1.5 for developing tracheostomy related complications. Ertuğrul A et al<sup>41</sup>. demonstrated GERD was the major co-morbidity (%28) in children dependent HIMV, most of them had an underlying a neuromuscular disease. In the study of Blanchi ET et al.<sup>42</sup>, the prevalence of reflux in the adults with tracheostomy was reported as 45.2% and there was no typical symptoms most of 24-hour pHmetry and manometry measurement.

In the presence of the GERD related symptoms in infants and children with MV, red flag symptom and signs should be investigated (Table 6). In the presence of these findings the appropriate tests and radiologic evaluation should be performed to rule out other pathological conditions (Table 7)<sup>34-38</sup>.

The treatment of GERD in otherwise healthy infants and children were detailed in ESPGHAN and NASPGHAN recommendations<sup>34</sup>. In suspicion of GERD and if there is no red flag symptom and/or signs, the first step in treatment should be feeding modification. The head elevation and positioning is not recommended in sleeping infants due to the risk of sudden infant death syndrome<sup>34</sup>, but there is no clear data for the infants with tracheostomy.

The nutritional interventions should be made according to patients underlying illness, concomitant symptoms or disorders, age and nutritional status. Food thickeners may be given to orally feeding patients. Reducing feeding volumes, increasing feeding frequency and avoiding overfeeding may relief the symptoms. If the infant fails to these modifications, an extensively hydrolyzed protein-based formula and in breastfed infants elimination of cow's milk from maternal diet should be considered for 2- to 4-weeks. If there is no response to extensively hydrolyzed protein based formula, an amino-acid based formula may be given. In children with NI who fed with tube whey based formula may be given, it also help to gastric emptying. If there is no improvement, acid suppression treatment should be considered for 4-8 weeks. The use of alginates may slightly improve the GERD symptoms and signs but ESPGHAN and NASPGHAN do not recommend use antacids/alginates for chronic treatment of infants and children with GERD. Also proton pump inhibitors (PPIs) may be considered in the treatment. If the symptoms improve in 4-8 weeks period, PPIs can be ceased<sup>6,34-38</sup>.

If there is no response to the optimal medical treatment in 4-8 weeks (refractory GERD) or weaning attempts of PPIs are failed within 6-12 months period, possible underlying conditions which may lead to GERD

symptoms and efficacy of treatment (e.g., drug usage, dosage, adherence, interactions) should be evaluated.

In suspect of congenital gastrointestinal abnormalities such as hiatal hernia, pyloric stenosis, malrotation, duodenal or antral web, duodenal or esophageal stenosis, or esophageal stricture, achalasia, extrinsic compression of esophagus barium/water soluble radiopaque swallowing follow-through studies should be performed. Abdominal ultrasound also may be helpful to detect the congenital abnormalities. Follow-through studies and abdominal ultrasound is not recommended to diagnose GERD<sup>34-38</sup>.

In the suspicion of GERD due to persistent troublesome symptoms, the association of the symptoms and gastroesophageal reflux events (both acid and non-acid) should be identified by pH-multichannel intraluminal impedance (MII). If it is not available only pH-metry can demonstrate the relation of symptoms only with acid GER events.

Esophagogastroduodenoscopy with biopsies should be performed to clarify the etiology of esophagitis (ie, eosinophilic esophagitis) or identify a mucosal disease as a complication of the GERD such as erosive esophagitis and Barrett esophagitis<sup>34-38</sup>.

In the presence of reflux-related erosive esophagitis in infants and children, the first-line treatment is proton pump inhibitors (PPIs). If PPIs are not available histamine-2 receptor antagonists (H2RAs) may be used. Baclofen may be considered before surgery in refractory GERD. Kawai et al<sup>43</sup>. reported a decrease in vomiting, number of acid refluxes with the gamma-aminobutyric acid type B receptor agonist baclofen. The use of domperidon, metoclopramide or any other prokinetics (e.g.,erythromycin) in infants and children with GERD is not recommended<sup>34</sup>.

Since the incidence of GERD in neurologically impaired children is high (up to 70%) for these clinically fragile patients, ESPGHAN recommended using whey-based formulas, thickening the enteral formulas, and PPIs treatment before diagnostic tests.. Despite GERD symptoms cannot be expressed by severely neurologically impaired patients, objective diagnostic tests such as esophageal pH-metry or pH/multichannel intraluminal impedance monitoring, and/or upper GI endoscopy will be beneficial to determine the further need of PPIs treatment<sup>6,34</sup>.

In the treatment of infants and children with GERD who are refractory to optimal medical treatment, transpyloric or jejunal feedings may be an alternative treatment to fundoplication, but there is no strong evidence. In neurologically impaired children both procedures didn't prevent aspiration pneumonia. This condition may be independent of the reflux events and related to their swallowing dysfunction<sup>34,44</sup>. The complication rates of transpyloric or jejunal feeding reported to be considerably high about 85%; including clogging, dislodgement, intussusception, and perforation<sup>45,46</sup>.

Anti-reflux surgery is recommended as follows; life threatening complication under optimal medical treatment, refractory GERD symptoms, underlying a chronic condition (e.g.,neurologically impaired, cystic fibrosis), need for chronic pharmacotherapy to control the signs and/or symptoms of GERD. Fundoplication leads to decrease the baseline pressure of LES, the number of TLESRs and increase the length of intra-abdominal esophagus. Despite different anti-reflux surgical procedures, Nissen fundoplication is accepted as gold standart technique, with shorter hospital stay and low incidence of perioperative complications and also morbidity. Unfortunately, no reduction of extraesophageal symptoms such as pneumonia and mechanical ventilation requirement was demonstrated in neurological impaired children after fundiplication<sup>44,47,48</sup>. Also, more than 75% of the patients couldn't cease acid suppression medications for at least one year after fundoplication<sup>49</sup>. After fundoplication gas-bloat, early satiety or pain after feeding, dysphagia, retching, worsening aspiration risk from esophageal stasis, unwrapping and redo fundoplication (2.2%-12.2%) are the other reported side effects<sup>50,51</sup>.

Total esophagogastric disconnection may be considered as a rescue procedure for neurologically impaired children with a failed fundoplication Since these complications may inversly effect the patients with tracheostomy, the parents should be carefully informed for the risks and benefits of the surgical interventions<sup>34</sup>.

## Dysphagia

The children with tracheostomy may be fed by oral route and/or by tube, depending on underlying conditions and swallowing abilities. Mainly, the severity and characteristics of neurological impairment are determinative of feeding route. For instance, the lack or diminished gag reflex, the disorders with bulbar involvement require tube feeding. Factors that may affect feeding of the children with tracheostomy are given in Table 8<sup>33,52</sup>.

Swallowing is a complex process which is controlled by brain stem, cortex and enteric nervous system that innervates esophagus smooth muscles. There are three phases of swallowing. The oral phase begins with opening of the mouth and continues with sucking, biting, chewing. The tongue movements and addition of the saliva allow the food bolus to pass into oropharynx. In pharyngeal phase, the velum elevates to close the nasopharynx. The hyoid and larynx elevate with anterior moving, epiglottis descends and vocal cords are closed. The laryngeal elevation protects the airway and anterior movement of the larynx helps open the upper esophageal sphincter. The contraction of the pharyngeal muscles moves the bolus from the pharynx to the esophagus. In esophageal phase, bolus passes into the stomach by esophagus peristalsis, gravity and LES relaxation<sup>53</sup>. The effect of tracheostomy tube on increasing the risk of aspiration and penetration is defined but there is no strong evidence. Tracheostomy tube fixes the larynx and prevents laryngeal elevation. Also it desensitizes larynx and leads an ineffective cough mechanism, because a positive subglottic pressure cannot be maintained during swallowing. The cuff of the tracheostomy tube may impinge on the esophagus during deglutition<sup>8,53</sup>. Streppel M et al<sup>54</sup>. reported 70% of the children with tracheostomy had swallowing disorders. The aspiration rate in the study group was 43% and half of them had silent aspiration. The underlying diseases may also contribute to the development of swallowing disorders<sup>54</sup>.

The aspiration, penetration and other swallowing problems can be detected by various diagnostic approaches. Penetration is passage of material into the laryngeal lumen, but remaining above the vocal cords, not passing to the glottis. Aspiration is the passage of material below the vocal cords. The Modified Blue Dye Test is a screening test for aspiration. After the patient swallow a methylene blue mixed drink or food, suctioning a colored secretion is considered as aspiration. Flexible Endoscopic Evaluation of Swallowing (FEES) and Videofluoroscopy (VFS) are the gold standart methods to evaluate swallowing dysfunction<sup>33,53,54</sup>. Pharyngeal phase of swallowing, penetration and aspiration can be detected by FEES. A speech and language therapist (SLT) and a pediatric Ear-Nose-Throat (ENT) specialist can perform FEES. A VFS demonstrate oral, pharyngeal and esophageal phases of swallowing, penetration, aspiration, obstruction, fistulas and motility problems can be observed. VFS is usually performed by a SLT and a radiologist. In the suspicion of an abnormality in the esophageal phase of swallowing, manometry may be considered. Swallowing the different consistencies of foods such as liquids, thickened liquids, purees and solids can be determined<sup>33,53,54</sup>.

Aspiration is a major problem that complicates the clinical status and respiratory condition of these children. Aspiration may lead hypoxemia, cough, respiratory distress or it may be silent. Untreated aspiration may result in bronchiectasis and decreased lung function<sup>33</sup>. On contrary, in recent years many centers change their feeding strategy to giving heavy-thickened liquids even if there is aspiration and/or penetration on VFSS. Particularly in patients with tracheostomy it is considered to be safer due to the chance of removing aspirates through the tracheostomy tube. Furthermore, improvement of the swallowing function depending on oral stimulation over time is possible<sup>55,56</sup>.

In the first 2-4 months of life, swallowing and sucking are reflexive, subsequently swallowing becomes more voluntary and it progressively develops in the first years of life. The development of proper oral feeding skills is negatively affected in premature infants who had long term intubation in intensive care units. Oral-motor and sensory feeding difficulties such as weak or incoordinate suck and swallow, oral aversion, hypersensitivity to oral stimuli (may be due to repetitive aspirations), refusal to taste food may occur<sup>32,53,54</sup>.

The early experience of oral feeding in tube fed children will be beneficial as follows; facilitates the oral motor skills such as chewing and swallowing, improves sensory skills such as taste and texture of the foods, and reduces oral aversion. A team work should be required to decide and carry out to transit oral feeding in tube-feeding children with tracheostomy. The team should be consist of an occupational therapist, speech language pathologist, physical therapist, respiratory therapist, psychologist, dietitian and clinicians

as pediatric pulmonologist, pediatric ear-nose-throat specialist and pediatric gastroenterologist<sup>32,53,54</sup>.

Following the instrumental tests, the patients who have a medical stability, swallow their oral secretions, need infrequent tracheal suctioning are seem to be ready to oral feeding. Oral feeding should be attempted under the actual mode of ventilation with following oxygen saturation. The aspiration and oral aversion risk may be reduced by tasting small amounts of food in extreme caution. Coughing, choking, increased oxygen demand or worsening pulmonary functions, presence or suctioning of food or liquid around the stoma or in the tracheostomy tube, sudden changes in the amount, constituency or color of the secretions are significant findings of unsuccessful swallowing<sup>32,55</sup>.

### **Gastrointestinal hypomotility**

The critically ill positive pressure ventilated patients had decreased gastric and duodenal contractility. This may be due to Cajal cell (GI motor activity control unit) dysfunction, pre-existing disease, endotoxin secretion, corticotropin releasing factor secretion during stress and drugs<sup>27,31</sup>.

### **Delayed gastric emptying**

Delayed gastric emptying is a common problem in intensive care units in the patients with MV. In the absence of mechanical obstruction, vomiting, bloating feeding intolerance and demonstrating the delayed emptying with 'gastric emptying scintigraphy', '<sup>13</sup>C-octanoate acid gastric emptying breath test' or 'acetaminophen absorption test' is sufficient to diagnose.

Supine position, infections, obesity, frequent suctioning, hiatal hernia, increased intracranial pressure and gastrostomy tube placement may lead delayed gastric emptying. Also electrolyte disturbances and hyperglycemia may worsen gastric emptying<sup>27,31,57-59</sup>. In tube fed children with neuromuscular disorders delayed gastric emptying has been reported. The children with DMA, the delay reported to be related to gastric smooth muscle involvement<sup>60,61</sup>. Also delayed gastric emptying reported in patients with SMA type 1<sup>62</sup>. In enterally fed children diagnosed mitochondrial disease gastric delay impairment was reported<sup>63</sup>.

Gastric decompression should be performed via nasogastric tube or gastrostomy tube. If it doesn't resolve a jejunal tube placement or total parenteral nutrition can be considered to achieve feeding<sup>31</sup>. Small volume, frequent, low-fat and low fiber content feeding may be given in stable patients<sup>58</sup>. The gastric emptying is significantly faster with whey based formula compared to casein based formula<sup>64</sup>. The use of prokinetic drugs such as erythromycin, domperidone, cisapride and metoclopramide is controversial because of their serious side effects and conflicting efficacy<sup>58</sup>.

### **Constipation**

Constipation is a common gastrointestinal complication in children with neuromuscular disorders. The cause of the constipation may be immobility, inadequate fluid taking, abdominal muscle weakness, smooth muscle involvements in neuromuscular disease, low fiber diet, gastrointestinal dysmotility (may be related to underlying disease). A careful history taking, together with abdominal and rectal examination will be sufficient to diagnose. Abdominal examination may reveal fecaloid masses. It is important to evaluate the perianal site for fissures, skin tags, anal tonus and sensation. Rectal digital examination should be performed to identify the placement and size of rectum, and also the consistency and amount of stool. In the suspicious of the surgical abdomen, abdominal radiography and ultrasound may be performed. The colonic transit time assessment may be used in treatment resistance patients. A slow colonic transit time was reported in patients with demonstrated in the children with brain lesions, DMD and tube feeding children<sup>6,15,17,65</sup>.

Simons JP et al<sup>66</sup>. hypothesized that the Valsalva maneuver increases in subglottic pressure, but tracheostomy tube may lead loss of this pressure and may predispose constipation. But they found no relation with tracheostomy and constipation in their study<sup>66</sup>.

Treatment of constipation should be individualized to the patient. If there is stool in the rectum, previously a fecal impaction should be performed. Enemas and polyethylene glycol (per oral in dosage of 1.5 g/kg/day) are given for 3 consecutive days and continue till a clear and liquid defecation. The fecal impaction treatment

may be prolonged to 6 days. In maintenance treatment, polyethylene glycol 0.8 g/kg/day or lactulose 1-2 ml/kg/day is required. The risk of aspiration pneumonia due to aspiration of mineral oil and polyethylene glycol should be kept in mind. Tube feeding patients should be reviewed for water intake and formula content. Fiber content formulas may be preferred in tube feeding patients<sup>65</sup>.

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