Editorial

Swallowing dysfunction in patients with esophageal atresia-tracheoesophageal fistula: infancy to adulthood

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Introduction

Esophageal dysfunction is a common problem in children with repaired esophageal atresia-tracheoesophageal fistula (EA-TEF) and considered as a long-term sequel of the cases. Impaired esophageal motility in EA survivors is multifactorial and is attributed to primary abnormality of esophageal innervation and vagal nerve damage during esophageal repair (1). Dysphagia, regurgitation, aspiration and chronic respiratory tract infections are considered as clinical findings of esophageal dysmotility (2). In addition to esophageal motility problems, 28% of patients with EA-TEF had oropharyngeal impairment causing swallowing dysfunction (SD) (2). Due to lack of objective criteria there is a wide variation in prevalence of SD in children with EA-TEF. Gibreel *et al*. reported that 82% of cases show some degree of SD in adulthood and most of the cases had mild symptoms that not affect the food choices of patients (3). Another study evaluating the dysphagia in adult EA survivors report dysphagia with a prevalence of 57% by using a symptom score test (4). Dysphagia is the most common symptom in patients with EA of all ages and incidence is varying depending on the definition. It has been shown that incidence is lower in infants than children and adults (5). Also, diagnostic methods that used to evaluate the dysphagia are responsible from the varying data. In this review, definition, diagnostic methods and clinical course of SD in patients with EA-TEF will be summarized from infancy to adulthood.

Definition of SD and dysphagia

Dysphagia is defined as swallowing disorder caused by sensory-motor dysfunctions or structural pathology of oral, pharyngeal and/or esophageal phases of bolus transport to the stomach (5). Gibreel *et al*. suggest that dysphagia mainly focus on difficulty of swallowing solid food and the term SD includes difficulty swallowing to all food consistencies (3). In the letter definition, difficulty of thin or thick liquids may also assessed as SD.

Incidence

Feeding difficulties are common in children with EA and they are more likely to eat slowly, refuse meals, cough and choke during eating and vomit with meals. Chetcuti *et al*. noted that these difficulties lessen with age and only 10% of them report these symptoms after age 15 (6). The incidence of dysphagia varies 15–52% in children (5) and 57% of adults with repaired EA complaint about dysphagia (4). A systematic meta-analysis by Conner *et al*. found that overall incidence of dysphagia is 50.3% (7).

Clinical findings of SD

Children present with SD show wide spectrum of clinical findings including gagging, vomiting and food refusal. Different clinical findings can be seen in different phases of deglutition (*Table 1*). By the age, most of the patients

Table 1 Different clinical findings according to the effected phases of deglutition

Phases	Disorders
Feeding behaviour	Feeding refusal, long mealtimes, late introduction to solid foods
Oral preperatory	Oral hypersensitivity, rumination
Oral	Oral hypersensitivity, oral motor impairment
Pharyngeal	Dyphagia, chocking, gagging, coughing, aspiration
Esophageal	Dysphagia, regurgitaion, dysmotility, bolus impaction, reduced peristalsis

adapted their eating habits by eating slowly, longer chewing on solid foods, drinking after having swallowed solid foods and avoiding dry and hard solids. Therefore, feeding difficulties show different clinical courses and causes variety of symptoms in different age periods.

Mechanisms of SD

Esophageal dysmotility is the main cause of SD in children with EA. The primary motility disorder relates either with the development of esophageal muscle or to the innervation of the esophagus (8). Neuronal abnormalities including both intrinsic and extrinsic neuronal defects are seen. Also, interstitial cells of Cajal cell counts was reduced in esophageal examinations in EA (9). Esophageal dysmotility is also secondary to surgery and gastroesophageal reflux. Extensive mobilization of esophageal pouches may cause myoneuronal damage and worsen esophageal motility (10). Esophageal motility studies show that postoperative motor functions were disturbed when compared with preoperative studies. Thus, the mechanism of SD is multifactorial and dysphagia is an ongoing problem in patients with EA with special consequences in different periods of life.

SD related respiratory problems

Respiratory problems are reported in 47% of children with EA (11). Structural anomalies of esophagus and trachea may also lead to aspiration and chronic respiratory complications (12). Tracheomalacia, gastroesophageal reflux and vocal cord paralysis are common structural problems in which tracheomalacia tends to be decrease with age and is not related with lower tract respiratory infections and chronic respiratory airway disease in adulthood (13). In addition to esophageal motility problems, oropharyngeal dysphagia (OPD) was observed in 28% of the EA cases and aspiration was detected in 55% of the patients with OPD (2). In patients with OPD, penetration and aspiration were also detected with higher PAS scores in 15% of all cases. It has been demonstrated that delayed pharyngeal swallowing was an indicator of aspiration and dysphagia is considered as a risk factor for respiratory problems (14).

Although, there is no direct correlation between the type of atresia and pulmonary complications, patients causing airway problems with long gap atresia or tensioned anastomosis often have respiratory problems and dysphagia. In addition to structural anomalies, tensioned anastomosis or extensive mobilization of both pouches alter hyolaryngeal mobilization and cause aspiration (15). Aspiration during swallowing is associated with esophageal dysmotility and/or gastroesophageal reflux and may lead to reduced pulmonary functions and bronchiectasis (2). Early detection and management of aspiration and thus protection of airways is mandatory to prevent pulmonary complications.

Diagnosis of SD

The evaluation of SD can be performed by diagnostic tests and/or by symptom scoring questionnaires. Contrast swallowing esophagograms, esophageal manometry and videofluoroscopic evaluation of swallowing function are common diagnostic methods. Esophageal motor functions can be assessed by both radiological esophagograms and esophageal manometry. The conventional diagnostic tools do not correlate with the clinical symptoms, however, high-resolution manometry (HRM) in combined with impedance meter is highly relevant to detect symptom related motor dysfunctions. Lemonie *et al*. described three patterns of disturbed motor function in children using esophageal pressure topography (EPT) in HRM according to Chicago classification; aperistalsis (38%), pressurization (15%) and various types of distal contractions (47%) (16). In this study, esophageal peristalsis was affected in all children including asymptomatic patients. These observations suggest that motor dysfunction may not be related with the symptoms. Secondly, van Wijk *et al*. found normal peristalsis but absent distal esophageal propagation in 6 of 20 children (17). However, lower esophageal sphincter relaxation was complete in 84% of patients.

Videofluoroscopy (VFS) is a dynamic method investigating the oral, pharyngeal and esophageal phases of

Table 2 VFS scale

VFS findings	Score
Oral phase (0–3)	
Lip closure	
Tongue elevation	
Tongue retraction	
Oral retention	
Pharyngeal phase (0–3)	
Delay in deglutitive reflex	
Touch of root of tongue to pharynx	
Velopharyngeal closure	
Hyopharyngeal elevation	
Closure of airway	
Vallecular retention	
Pharyngeal retention	
Retention in pyriform sinus	
Aspiration	
Silent aspiration	
Esophageal phase (0–3)	
Opening of UES	
UES retention	
Esophageal back-flow	
Motility problem	
Esophageal retention	
LES dysfunction	

Each parameter was marked with 0–3 points. Normal, within functional limits =0, mild =1, moderate =2, severe =3. Retentions points: no retention =0, minimal retention =1, moderate retention =2 and severe retention =3. UES, upper esophageal sphincter; LES, Lower esophageal sphincter.

deglutitive function in children. All phases of deglutition can be evaluated with different consistencies of food in this procedure. To compare the results, a scoring system can be used (*Table 2*) (2). During the exam, it is possible to evaluate the penetration and aspiration by a scale (*Table 3*) (18). Fiberoptic endoscopy evaluation of swallowing (FEES) test uses a transnasal flexible fiberoptic laryngoscopy to visualize the pharynx and larynx while swallowing. It can be used to evaluate aspiration in children and adults. Not in the esophageal parameters, but in laryngeal penetration and aspiration, it has highest specificity and positive predictive value when compared to VFS.

Finally, several symptom-scoring systems have been developed to evaluate the SD in children and adults. Dysphagia scoring (DS) system introduced by Dakkak *et al*. includes 9 item questions regarding with the difficulty in swallowing in different food consistencies (*Table 4*) (19). The total sum of DS was obtained by multiplying dysphagia frequency (presence of dysphagia =1, sometimes =1/2 and absent =0) with the row number. The patients with DS score 0 were considered as no dysphagia, between 1–44 were mild dysphagia, and more than 44 were severe dysphagia. The pediatric eating assessment tool-10 (pEAT-10) is a validated, self-administered, symptom-specific outcome tool that is commonly used in documenting initial symptom severity, monitoring treatment efficacy and to predict aspiration and aspiration risk in patients with dysphagia (20). It has been found that patients with EAT-10 scores higher than 10 had 2.2 times more risk of aspiration and scores higher than 3 is predictive for aspiration (*Table 5*) (21). Soyer *et al*. found that sensitivity and specificity of pEAT-10 to predict aspiration were 88% and 77% in children with EA (22). Gibreel *et al*. developed swallowing dysfunction questionnaire (SDQ) including swallowing total score and overall swallowing difficulty score for adult EA survivors (3). Functional oral intake score can be also used to define the ability of oral intake in patients (*Table 6*). Gatzinsky *et al*. also define SD in adults by use of DS (4). There results suggest that symptom-scoring systems are more common in the evaluation of SD in adults, whereas VFS and HRM were preferred in childhood.

Management of SD in EA

Management of SD includes reducing reflux and aspiration and to improve deglutitive skills to have full oral feeding. Thickened oral feeding reduce reflux into the oropharynx and prevent retching. Patients with OFD have more airway aspirations with liquids and safer swallowing can be seen with semi-solid foods. None of the studies have shown a difference between bolus versus continuous feeding to reduce the risk of aspiration. Other strategies such as changing feeding schedule or formula and transpyloric feeding can be used in patients with severe OFD (23). Conforti *et al*. recently defined the methods, which can be applied to children with insufficient airway closure with aspiration are nonnutritive stimulations (sham feeding and/or oral stimulations) (24). However, only oral sensory

Table 3 Penetration and aspiration scale

Number	Scale	Definition
1	No penetration and aspiration	No contrast material in the airway
2		Contrast material pass to airway, above the vocal cords, no contrast remnants
3	Penetration	Contrast material pass to airway, above the vocal cords, visible contrast remnants
4		Contrast material pass to airway, at the level of vocal cords, no contrast remnants
5		Contrast material pass to airway, at the level of vocal cords, visible contrast remnants
6		Contrast material pass to airway, below the vocal cords, no contrast remnants
7	Aspiration	Contrast material pass to airway, below the vocal cords, in addition to response to aspiration visible contrast remnants
8		Contrast material pass to airway, below the vocal cords, no response to aspiration

Table 4 Dysphagia score system

Food	Often (1 points)	Occasionally (1/2 points)	Never (0 points)	Total
Water				1×
Milk/soup				2×
Yogurt/fruit pure				3×
Jelly/jam				4×
Mashed potatos or scrambled eggs				5×
Boiled vegatables or fish				6×
Bread				7×
Fresh fruits				8×
Meat				9×

Table 5 Pediatric EAT-10 (0: no problem–4: severe problem)

pEAT-10	0	1	2	3	4
My swallowing problem has caused me to lose weight					
My swallowing problem interferes with my ability to go out for meals					
Swallowing liquids takes extra effort					
Swallowing solids takes extra effort					
Swallowing pills takes extra effort					
Swallowing is painful					
The pleasure of my eating is affected by my swallowing					
When I swallow food stricks to my throat.					
I cough when I eat					
Swallowing is stressful					

EAT-10, eating assessment tool-10.

Table 6 Parameters of FOIS

Number	Definition
1	No oral intake
2	Tube dependence with minimal/inconsistent oral intake
3	Tube supplements with consistent oral intake
4	Total intake of a single consistency
5	Total oral intake of multiple consistencies requiring special preparation
6	Total oral intake with no special preparation, but must avoid specific foods and liquid items
7	Total oral intake with no restrictions

FOIS, functional oral intake scale.

stimulation techniques are not sufficient to improve pharyngeal phase problems, which cause aspiration and related respiratory symptoms. Arslan *et al*. developed a swallowing rehabilitation protocol (SRP) in patients with EA (25). SRP includes neuromuscular electrical stimulation for 30 minutes to activate anterior neck muscles, thermal tactile application for 15 min to trigger swallowing reflex and hyolaryngeal mobilization for 15 min to support hyolaryngeal elevation. After 20 sessions of SRP, patients with EA-TEF showed improvement in penetration and aspiration scores, liquid and pudding aspiration, oral phase dysfunction, delay in swallowing reflex, and residue in vallecula and pyriform sinuses after pudding swallow. Therefore, SRP has been recommended in children with OFD in the early period.

Infancy to adulthood

SD is a common problem in children with EA and persists in some of the patients in adult ages. Limited studies have been performed about SD in adult EA survivors. As the patients become adolescents and young adults they overcome SD by adopting their feeding habits such as longer chewing, swallowing with liquids and longer meal times. Therefore, it is no possible to suggest that SD resolves with age. Despite better results, Gibreel *et al*. and Gatzinsky *et al*. report poor quality of life score in these patients when they are become adults. In adult studies, SD is usually evaluated by QOL questionnaires and dysphagia score systems. The incidence of SD, evaluated by symptom scores is higher in adults when compared to children. However, there is no objective data about the motility problems that correlating with the symptoms in adults. In conclusion, SD is a life-long consequence of EA/TEF that requires detailed diagnostic evaluation with limited therapeutic options.

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