Clinical Characteristics of Pediatric Patients with Esophageal Strictures

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Background: Esophageal stricture is commonly encountered in adults. Yet, it is not uncommon among children.

Objective: To evaluate the clinical presentations and causes of esophageal strictures in children.

Design: A Retrospective Cross-Sectional Study.

Setting: Pediatric Department, Salmaniya Medical Complex, Bahrain.

Method: A review of medical records of patients diagnosed with esophageal strictures between 1995 and 2019 was performed. The patients were diagnosed based on clinical, radiological and endoscopic findings. Data including patients' characteristics, clinical presentations, stricture etiologies and proton pump inhibitors use were documented.

Result: Forty-six pediatric patients had esophageal strictures. Twenty-five (54.3%) were males. Thirty-six (78.3%) patients were Bahraini. Thirty-two (69.5%) patients were infants. Thirty-two (69.5%) presented with dysphagia and 27 (58.7%) with vomiting. Anastomotic post-esophageal atresia/tracheoesophageal fistula (EA/TEF) repair strictures was the main cause and found in 35 (76.1%) patients. Twenty-two (47.8%) patients had associated diseases; 10 (21.7%) had congenital heart disease. Twenty-six (56.5%) had esophageal stricture of the upper esophagus. Twenty (43.5%) patients received proton pump inhibitors. The Median follow-up period was five years.

Conclusion: Esophageal stricture is not a rare disease in children. It is more common in males. Dysphagia and vomiting are the most frequent clinical presentations. The most common cause of esophageal strictures in children is anastomotic post EA/TEF repair. Congenital heart diseases are the most commonly associated anomalies.

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Esophageal stricture is commonly encountered by gastroenterologists in adults[1]. However, it is not an uncommon condition among children[2]. It is a serious disease in the pediatric age group and requires meticulous management[3,4].

Esophageal stricture has different causes[5]. In adults, the etiology of esophageal strictures could be either malignant or benign[1]. In children, it could be congenital esophageal strictures, anastomotic strictures post esophageal atresia/ tracheoesophageal fistula (EA/TEF) repair, caustic ingestion, gastroesophageal reflux disease (GERD) or peptic stricture and eosinophilic esophagitis[1-3,5-9]. The clinical presentation includes dysphagia, recurrent vomiting, or food impaction[2].

The diagnosis of esophageal stricture is confirmed by barium swallow or endoscopy[10,11]. Esophageal strictures and their underlying etiologies can be identified by performing upper gastrointestinal endoscopy[1].

To the best of our knowledge, there were only three studies

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of esophageal strictures in children from the Gulf region, two from Saudi Arabia and one from Iran[2,5,9]. There has not been any study about esophageal strictures in children from Bahrain.

The aim of the study is to evaluate the clinical presentations and causes of esophageal strictures in pediatric patients.

METHOD

A review of electronic and paper-based medical records of all patients diagnosed with esophageal strictures between 1 August 1995 and 31 August 2019 was performed. Patients above 18 years of age at presentation were excluded from the study. Patients were diagnosed to have esophageal stricture based on clinical, radiological and endoscopic findings.

Personal characteristics including gender, nationality, gestational age, delivery mode, birth weight, age at presentation, diagnosis, and age were documented. Associated congenital anomalies such as vertebral, anal, cardiac, renal, limbs (VACTERL) were documented. The number and types of surgical interventions were recorded.

Results of radiological imaging, specifically gastrografin studies and endoscopic findings were reviewed. The use of proton pump inhibitors and patients' follow-up period were documented.

Data were entered into an Excel sheet then transferred to the Statistical Package for Social Sciences (SPSS) version 21 program for analysis. Frequencies and percentages were calculated for categorical variables. The patients' ages were classified into five age groups. Continuous variables were checked for normal distribution using Kolmogorov-Smirnov. Group data were presented as mean and standard deviation (SD) for normally distributed variables or median and range for non-normally distributed variables.

RESULT

Forty-six pediatric patients had esophageal strictures. Twenty-five (54.3%) were males and 21 (45.7%) were females. The presenting clinical symptoms were dysphagia, vomiting, cyanosis, food impaction and hematemesis. Thirty-six (78.3%) patients were Bahraini and ten (21.7%) were non-Bahraini (two Yemeni, two Pakistani, one Saudi, one Turkish, one Indian, one Syrian, one Singaporean, and one not specified), see table 1.

Post EA/TEF repair anastomotic stricture was the main cause for esophageal strictures found in 35 (76.1%) patients followed by GERD, seven (15.2%) patients, see figure 1. One patient with EA/TEF had a history of mild polyhydramnios. One patient was diagnosed by antenatal ultrasound. Twenty-two (47.8%) patients had associated diseases; the majority of them, 10/22 (45.5%), had associated congenital heart disease.

All patients underwent a gastrografin study and esophagoscopy. The site of stricture was documented in 35 (76%) patients; 26 (56.5%) had upper stricture, eight (17.4%) patients had a middle stricture and one (2.2%) had a distal stricture.

Table 1: Personal Characteristics of Patients with Esophageal Strictures

Personal Characteristics			Patients number & %
Gender	Male		25 (54.3%)
	Female		21 (45.7%)
Total			**46**
Nationality	Bahraini		36 (78.3%)
	Non-Bahraini		10 (21.7%)
Total			**46**
Gestational age	Term		29 (63%)
	Preterm		10 (21.7%)
			39⁂
Type of delivery	NVD*		25 (54.3%)
	LSCS†		14 (30.4%)
Total			**39⁂**
Birth weight (kilogram), mean (SD‡)			2.65 kg
Presentation age category (year)		0-1	32 (69.6%)
		1-2	3 (6.5%)
		2-3	0(0.0%)
		3-4	1 (2.2%)
		4-5	1 (2.2%)
Total			**37⁂**
Age at time of the study (years), mean			8.3 years (6.92)
Age at time of study category (year)		0-4	16 (34.8%)
		5-9	15 (32.6%)
		10-14	10 (21.7%)
		15-18	1 (2.2%)
		>18	4 (8.7%)
Total			**46**
Clinical presentations	Dysphagia		32 (69.6%)
	Vomiting		27 (58.7%)
	Cyanosis		8 (17.4%)
	Food impaction		2 (4.3%)
	Hematemesis		1 (2.2%)
Surgical interventions			**36 (78.3%)✧**
	EA/TEF** repair		35 (76.1%)
	Pyloric stenosis repair		2 (4.3%)
	Tetralogy of Fallot repair		2 (4.3%)
	Gastrostomy/gastrojujensotomy		2 (4.3%)
	Duodenal atresia repair		1 (2.2%)
	Duodenostomy		1 (2.2%)
	Aortoplexy		1(2.2%)
	Adenoidectomy		1 (2.2%)
	Fundoplication		1 (2.2%)
	Diaphragmatic hernia repair		1 (2.2%)
	Imperforated anus repair		1 (2.2%)
Total			**48**

*normal vaginal delivery, †lower segment cesarean section, ‡Standard deviation, §interquartile range, **esophageal atresia/tracheoesophageal fistula,⁂Missing Data, òSome patient had more than one procedure.

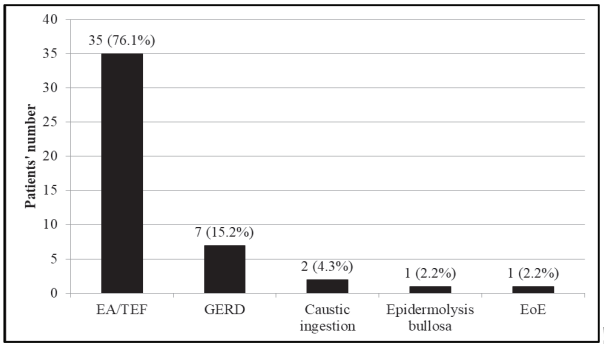
Table 2: Associated Diseases with Esophageal Strictures

Associated diseases	Patients N & %
Cardiac system (total)	**13 (28.3%)**
Ventricular septal defect	4 (8.7%)
Atrial septal defect	3 (6.5%)
Tetralogy of Fallout	2 (4.3%)
Patent foramen ovale	2 (4.3%)
Patent ductus arteriosus	1 (2.2%)
Tricuspid valve disease	1 (2.2%)
Gastrointestinal system (total)	**10 (21.7%)**
Pyloric stenosis	2 (4.3%)
Duodenal atresia	2 (4.3%)
Diaphragmatic hernia	1 (2.2%)
Helicobacter pylori gastritis	2 (4.3%)
Hiatal hernia	2 (4.3%)
Anal stenosis	1 (2.2%)
Airway and lung	**5 (10.9%)**
Tonsillitis	1 (2.2%)
Adenoid hypertrophy	1 (2.2%)
Tracheomalacia	1 (2.2%)
Bronchial asthma	1 (2.2%)
Right hypoplastic lung	1 (2.2%)
Syndromes	**5 (10.9%)**
Trisomy 21	2 (4.3%)
VACTERL†association	2 (4.3%)
Dysmorphysim	1 (2.2%)
Genitourinary system	**5 (10.9%)**
Horseshoe kidney	1 (2.2%)
Left kidney dysplasia	1 (2.2%)
Hydroureter	1 (2.2%)
Undescended testicle	1 (2.2%)
Mental retardation	1 (2.2%)
Skeletal system	**3 (6.5%)**
Skeletal dysplasia	1 (2.2%)
Spina bifida occulta	1 (2.2%)
Flat feet	1 (2.2%)
Genetic/hematology	**1 (2.2%)**
Sickle cell disease	1 (2.2%)
Missing data	**3 (6.5%)**

*some patients had more than one associated diseases. †vertebral defects, anal atresia, cardiac defects, trachea-esophageal fistula, renal anomalies and limb abnormalities.

Two patients died; one was female, at the age of 48 days postnatally due to prematurity (birth weight 1.2 kg), dysmorphism, right hypoplastic lung and EA/TEF. The second patient was a preterm female born by lower uterine segment Cesarean section (LSCS) with TEF type C; the cause of death was not clear. Twenty (43.5%) patients received proton pump inhibitors. The median follow-up period was five years and the interquartile range was 6.55 years.

Figure 1: Causes of Esophageal Strictures



DISCUSSION

This study showed that more males had esophageal strictures compared to females, a ratio of 1.2:1. This is similar to other pediatric and adult studies[2-4,8,9,12-21]. The majority of the patients, in our study, had EA/TEF which is known to occur more in males[2]. Shawyer et al showed that more males had EA/TEF than females[22]. Davari et al study showed that 60% were males and 40% were females[16]. However, Weintraub et al study on children with benign acid-peptic esophageal strictures found more females than males[7]. Dehghani et al found esophageal strictures more in females than males[5].

This study reveals that term infants had more strictures compared to preterm infants. Yang et al study found that term babies were more than preterm[15]. Narasimman et al on patients with EA/TEF showed that term newborns were more compared to preterm newborns[13]. Davari et al study also showed that terms were more than preterm[16]. Generally, premature babies have an increased complications rate compared to term babies[15]. This might be attributed to the fact that preterm infants with EA/TEF are usually associated with other congenital anomalies that might lead to stillbirth or abortion before the development of any esophageal strictures[25]. Moreover, the mortality risk is higher in premature babies with EA/TEF compared to term babies[13].

In our study, the majority of patients with esophageal stricture presented during infancy, within the first year of life. This early presentation could be explained by the presence of high percentage of patients with EA/TEF which most frequently presents soon after birth[2,3,7,10,19,21]. EA/TEF can be suspected antenatally if polyhydramnios was present[26]. Weintraub et al reported a median age of 1.25 years (range from two weeks to 17.5 years)[7]. Reinders et al showed that the median age at presentation was 17.7 months (range 21 days to 12 years and three months) with 51% were younger than 18 months[19]. Lakhdar-Idrissi et al found that the age ranged between 10 months and 17 years[4]. The median age at presentation was two years (range, 1-16 years) in the study from Saudi Arabia[2]. Dehghani et al study from Iran reported a mean age at presentation of 3.95 ± 0.4 years (range, 15 days -14 years)[5].

Our study revealed that EA/TEF was the main cause of esophageal stricture in children followed by GERD. EA/TEF is one of the most frequent congenital anomalies seen by pediatric surgeons[26,27]. The incidence of EA/TEF abnormalities is approximately 1 in 3,500 neonates/year[11,27]. Esophageal

stricture is considered the most common complication post-EA/TEF repair despite the advances in operative practices[11,28]. Al Sarkhy et al study from Saudi Arabia reported post-TEF repair anastomotic strictures as the most common cause of stricture followed by GERD[2]. Reinders et al found that EA/TEF is the main cause followed by caustic ingestion[19]. Similarly, Alshammari et al study showed that EA/TEF accounts for 49%[21]. Weintraub et al, Allmendinger et al, and Yeming et al showed that EA/TEF was the main cause[7,23,24]. However, Cakmak et al reported an equal number of esophageal strictures caused by EA/TEF and corrosive strictures[8].

In our study, GERD was the second cause of esophageal stricture. Esophageal strictures are more likely to develop if the patient suffers from GERD[10,19]. GERD-related strictures, especially in patients with a prolonged disease, can be severe and challenging to manage[2]. Lakhdar-Idrissi showed that peptic stricture is the most common cause[4]. Peptic strictures are considered the end stage of chronic reflux esophagitis and account for 90% of benign esophageal strictures[29].

Corrosive was the third cause of strictures in our study. However, it is considered as the second cause in several other studies[7,19,21,23,24]. Dehghani et al reported that caustic ingestion was the most common[5]. One patient in our study had eosinophilic esophagitis (EoE). Esophageal stricture secondary to EoE is not an uncommon complication in children with untreated longstanding EoE[9].

Our study revealed that the main clinical presentation of esophageal strictures was dysphagia followed by vomiting. Other studies revealed similar findings[2,21]. Zouari et al showed that dysphagia was the most frequent presenting symptom[29]. AlHussaini et al also reported that dysphagia was the main symptom with esophageal stricture secondary to EoE[9]. However, Lakhdar-Idrissi et al reported vomiting as the main symptom of patients with esophageal stricture followed by dysphagia[4]. Similarly, Dehghani et al reported vomiting as the main symptom followed by dysphagia[5]. Usually, formal evaluation for an esophageal stricture is performed only for patients with symptomatic dysphagia[14]. Consequently, patients with mild symptoms or those with a high degree of tolerance to dysphagia might be overlooked and the overall incidence of strictures will be underestimated[14].

Our study showed that 47.8% of the patients had associated congenital anomalies. The most common associated anomaly was congenital cardiac disease. Similarly, Gupta et al study reported associated diseases in 37.5%, most of them were cardiac anomalies[26]. Yang et al reported that 80% of patients had associated anomalies, most of them were cardiac[15]. Narasimman et al reported 30% associated cardiac anomalies[10]. van der Zee et al reported 29.4% were associated cardiac anomalies[30]. Shah et al study on 100 children with EA/TEF found that 29% had associated anomalies mainly of VACTERL association[31]. VACTERL association was found in two patients (4.3%) in our study. The presence of associated congenital anomalies significantly affects the outcome of infant post-EA/TEF repair[13]. Patients without associated anomalies had a better survival rate compared to patients with associated anomalies[13,26].

Our study is limited due to missing data related to the patients' characteristics. Another limitation is that the number of patients in this study is relatively small. However, it is comparable to that reported from many neighboring countries and worldwide. Despite these limitations, this study is important being the first study to shed light on esophageal strictures in the pediatric age group and can be the foundation for any future studies.

CONCLUSIONS

Esophageal stricture is not a rare disease in children. It is more common in males. Dysphagia and vomiting are the most frequent clinical presentations. The most common cause of esophageal strictures in children is anastomotic post-EA/TEF repair. Congenital heart diseases are the most common associated congenital anomalies. Further studies are needed to investigate the safety and efficacy of esophageal stricture dilatations and the long-term impact of esophageal strictures on patients' quality of life.

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