Gastroesophageal reflux following repair of esophageal atresia and tracheoesophageal fistula

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ABSTRACT

Objectives: This study represents the experience of a tertiary care center in the Kingdom of Saudi Arabia (KSA) on the long-term effect on the lungs of esophageal atresia (EA) and tracheoesophageal fistula repair (TEF), and to emphasize the magnitude of gastroesophageal reflux (GER) post-EA or post-TEF repair.

Methods: A retrospective review of all patients referred to the pulmonary clinic with EA/TEF or re-operative evaluations from the period 1993-2004 at King Faisal Specialist Hospital and Research Centre, Riyadh, KSA.

Results: Forty-one patients with confirmed EA/TEF (26 males and 15 females) were included in the study. Congenital anomalies were associated in 28 (68%). Gastroesophageal reflux developed in 39 (95%) of patients, 24 (59%) needed Nissen fundoplication. Esophageal stricture that required more than 3 dilations developed in 16 (46%) patients, esophageal dysmotility in 37 (90%) and hiatal hernia in 11 (27%). Pulmonary complications developed in >70% of the patients including persistent atelectasis, chronic aspiration pneumonia, asthma and chronic lung disease that required oxygen for more than one month. Tracheomalacia in 12 (29%) and bronchiectasis in 7 (17%). Eighty-eight percent of patients who were able to do pulmonary function test showed abnormal values of moderate obstructive and restrictive lung disease.

Conclusion: Pulmonary complications cause significant and prolonged morbidities post EA/TEF repair. Gastroesophageal reflux is a common complication after EA/TEF repair and causes significant morbidity that needs a prolonged follow up. Patients with GER may need Nissen fundal plication to improve respiratory problems.

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The incidence of esophageal atresia (EA) and tracheoesophageal fistula (TEF) was reported to be one in 4000-5000 live births,[1] with both gender affected equally. Type C or EA and distal TEF have been described to be the most common type which affect 87% of the patient population.[2] Recurrent aspiration pneumonia is the most common complication described according to different mechanisms due to spill over of secretions through TEF or esophageal pouch, esophageal dysmotility, gastroesophageal reflux (GER) and absence of ciliated epithelium in the trachea which impairs clearance of secretion.[3] Gastroesophageal reflux was reported to be the most common cause of respiratory complications in these patients.[3] Other factors that may contribute to the severity of the respiratory complication are tracheomalacia[4] and recurrent TEF.[4,5] In this report, we evaluate the contribution of GER to the magnitude of respiratory morbidity in a tertiary care center in the Kingdom of Saudi Arabia (KSA).

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Methods. A retrospective review of charts for all EA/TEF patients that were referred to the pulmonary clinic for evaluation of recurrent chest infection and preoperative evaluation during the period from November 1993 to October 2004 at King Faisal Specialist Hospital and Research Center (KFSH&RC), Riyadh, KSA, which is considered the main tertiary care center for referral of complicated cases in KSA. Patients who were referred directly to the surgeons only but not evaluated by pulmonary service were not included. The demographic, TEF types, surgery types, type of anomalies, morbidities and pulmonary complications were collected.

Esophageal stricture is considered "stricture" when it requires >3 dilatations to improve swallowing. Gastroesophageal reflux diagnosis was based on barium swallow study or nuclear medicine scan. Tracheomalacia diagnosis was based on bronchoscopy study. Group 1 includes those patients who were diagnosed and had their esophageal anastomosis at KFSH&RC. Group 2 are those patients who had their initial surgery such as esophagostomy or gastrostomy (GT) with or without esophageal anastomosis at a local hospital and then referred to KFSH&RC for redo anastomosis and treatment of surgical complications.

The statistical Package for Social Sciences program was used for data analysis. Chi square was used to compare categorical variables, and Kruskal Walis was used to analyze continuous variables.

Results. A total of 41 patients with confirmed EA/TEF (26 males and 15 females) were included in the study. The mean age of patients at referral was 15 ± 29 months and at follow-up was 5 ± 3.8 years. Five (12%) in Group 1 and 36 (88%) in Group 2. Forty patients (98%) are alive and one (2%) died. Fourteen (34%) were premature and 27 (66%) were full term. Tracheoesophageal fistula was diagnosed at birth in 34 (83%) of the patients. Patients were referred to KFSH&RC at 15 ± 29 months. The period of follow up was 5 ± 3.8 years. Diagnosis of TEF was based on nasogastric tube coiling (NGT) in 38 (93%) and by dilated blind esophagus in chest x-ray in 40 (98%) patients. Esophageal atresia and distal TEF were found in 37 (90%) patients, isolated EA in 2 (5%) and H-type fistula in 2 (5%) patients. Congenital anomalies were associated in 28 (68%) patients. Cardiac anomalies were found in 11 (27%), gastrointestinal (GIT) in 8 (20%), respiratory system anomalies in 12 (30%), renal in 7 (17%), skeletal in 12 (30%), and chromosomal in 7 (17%). Twenty-one (51%) had a primary repair, 14 (34%) had delayed primary for 2-4 weeks, and 6 (15%) had staged repair. Thirty (73%) presented with pneumonia and required prolonged ventilation. Pulmonary complications developed in >70% of the patients including persistent atelectasis, chronic aspiration pneumonia, asthma or hyper reactive airway disease, and chronic lung disease that required oxygen for more than one month (**Table 1**). Leakage at operation site developed in 16 (39%), the leakage in 11/16 patients was mentioned in the referral hospital report, but its degree was unknown whether it was minor or major leakage and only 2/16 needed redo anastomosis. Recurrent fistula developed in 10 (24%), 2 were defined as contained sinuses, one resolved spontaneously, 3 had esophageal diverticulum, 2 had recurrent fistula in the local hospital before referral, and one needed redo anastomosis. Esophageal dysmotility was found in 37 (96%), Hiatal hernia in 11 (27%) and GER developed in 39 (95%) of the patients. Twenty-four (60%) of the patients required Nissen fundoplication for GER (**Table 1**). Esophageal narrowing at anastomosis site developed in 25 (61%) of the patients and 21 (51%) of them required 1-12 dilatations. Esophageal stricture that required >3 dilatations developed in 17 (41%) patients (**Figure 1**). Gastroesophageal reflux was significantly related to development of atelectasis, dysmotility, and aspiration pneumonia ($p<0.05$), but not related to surgery type or development of chronic lung disease ($p>0.09$) (**Table 2**). Gastrostomy tube feeding was required in 26 (63%) and jejunostomy tube feeding in 9 (22%) of the patients. Esophageal stricture developed mainly in those patients who were referred to

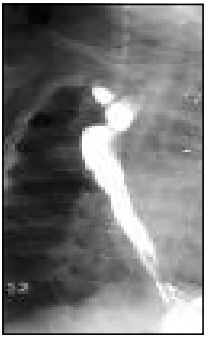

Figure 1 - Upper gastrointestinal series showing esophageal stricture in the upper third of the esophagus with approximately 5 mm of stenotic area and a lumen of 3 mm. There is also an additional pre-stenotic dilatation of the esophagus. There is mild gastroesophageal reflux up to the lower third of the esophagus.

Table 1 - Tracheoesophageal fistula repair pre- and postoperative complications (N=41).

Complication type	n	(%)
Pneumonia at diagnosis	30	(73)
Ventilation required pre and post surgery	28	(68)
Persistent atelectasis	37	(90)
Chronic aspiration pneumonia (radiology)	40	(98)
Asthma/hyper-reactive airway	40	(98)
Chronic lung disease/O_2 requirement	36	(88)
Tracheomalacia (bronchoscopy)	12	(29)
Bronchiectasis by CT chest	7	(17)
Esophageal narrowing (anastomosis site)	25	(61)
Esophageal dysmotility (radiology)	37	(90)
Gastroesophageal reflux (by Barium swallow)	39	(95)
Gastroesophageal reflux surgery (Nissen fundal plication)	24	(59)
Hiatal hernia	11	(27)
Esophageal dilatation required (1-12 times)	21	(51)
Gastrostomy feeding	26	(63)
Jejunostomy feeding	9	(22)
Failure to thrive	32	(78)
Pneumothorax (postoperative)	10	(24)
Recurrent fistula	10	(24)
Leakage at operation site	16	(39)
Infection (blood)/empyema/mediastinitis	17	(42)

Table 2 - Correlation of gastroesophageal reflux (N=41).

Variable	Gastroesophageal reflux n	(%)	*P* value
Atelectasis	36	(92)	0.04
Dysmotility	3	(60)	0.04
Aspiration pneumonia	39	(100)	0.0001
Esophageal stricture	16	(46)	0.023
Esophageal narrowing (radiology)	25	(64)	0.07
Asthma/hyper-reactive airway disease	38	(97)	0.8
Surgery type			0.36
Primary	19	(49)	
Delayed repair	14	(36)	
Staged repair	6	(15)	
Chronic lung disease/O_2	35	(90)	0.09
Bronchiectasis	7	(18)	0.5
Pneumothorax	9	(23)	0.38

KFSH&RC for complication, but did not develop in those who had their primary repair in a tertiary care center (KFSHRC) ($p \leq 0.022$). Tracheomalacia developed in 12 (29%) patients, 5 of them required aortopexy and one required tracheal stent. Bronchiectasis developed in 7 (17%) patients, 2 of them had gastric tube replacement, one had colonic interposition for long gap EA, and 4 had primary anastomosis. The pulmonary function test (PFT) was carried out in 16 (40%) patients who were able to comprehend the test maneuver. Eighty-eight percent of patients who performed PFT showed abnormal values: obstructive PFT changes in 3 (7%), restrictive in 8 (20%), combined obstructive and restrictive changes in 3 (7%) and normal in 2 (4%).

Discussion. Respiratory complications affect a significant number of patients after repair of EA with TEF. The incidence of postoperative respiratory complications is the third most frequent postoperative complication, with an incidence of 30%.[3,5] Although these symptoms typically diminish in frequency and severity, as the patients grow older, they are of concern as long-term abnormalities of respiratory function have been documented.[3,5] Most reports have implicated GER as the primary cause of respiratory symptoms in these patients,[3] which is understandable as GER and other functional abnormalities of the esophagus are remarkably common in these patients.[3,5]

Gastroesophageal reflux and esophageal stricture have been described in many studies[5-7] as a complication post TEF repair in approximately 40-70% of patients. It is also unclear whether lower esophageal sphincter incompetence is part of a generalized esophageal neuro-motor disturbance or relates more to technical issues such as obliteration of the normal esophageal cardiac angle or reduction in the length of the intra-abdominal esophagus by traction on the lower esophageal segment.[7] In either event, reflux may cause significant problems and can be responsible for life-threatening events in the first months following repair. In some instance, significant reflux may lead to chronic aspiration or reactive airway disease.[8-11] Some patients with GER may respond to medical management, but a significantly higher percentage require operative intervention than patients with GER who did not have EA.[7] Owing to the dysfunctional motility of the distal esophageal segment, a partial-wrap fundoplication is preferable to a complete wrap to avoid functional obstruction of the esophagus.[12] Failure rates for the partial wrap are lower than for the complete wrap fundoplication.[12] The morbidity of complete wrap fundoplication in patients following repair of EA has been increased compared

with patients in the same institution having the same fundoplication unrelated to EA.[12]

Our study has shown a high number of patients: 39 (95%) who developed GER and 24 (59%) required Nissen Fundoplication (**Table 1**). Gastroesophageal reflux was significantly related to development of atelectasis, dysmotility, and aspiration pneumonia ($p<0.05$), but not related to surgery type or development of chronic lung disease ($p>0.09$). Our findings are similar to previously reported studies.[1-3] Distorted anatomy and dysmotility of the esophagus in these patients may make these children poor candidates for Nissen fundoplication. Because most patients who underwent an antireflux procedure had a gastrostomy tube in place, it is difficult to determine if a gastrostomy tube has a role in managing recurrent respiratory symptoms after TEF repair. Despite the presence of a gastrostomy tube; however, a significant number of our patient population who were initially diagnosed with GER had persistent symptoms after an antireflux operation.

The increased respiratory effort due to tracheomalacia can produce increased intra-abdominal pressure with resulting reflux episodes.[13,14] The incidence varies from 10-20%.[12-14] Operation is not always necessary if the symptoms are mild, and patients with tracheomalacia typically show improvement as they become older and the trachea increases in diameter. In the most severe cases an operation is required, and aortopexy has become the standard procedure for this problem. Aortopexy was uniformly successful in increasing the size of the tracheal lumen in our patients. The similarity between the symptoms of tracheomalacia and GER makes bronchoscopy vital for distinguishing between the 2 different diagnoses.[12-14] Tracheomalacia formed a significant morbidity in 12 (29%) of the patients in our study and required surgical intervention in 50% of them. It has also been described to cause apnea, cyanosis and prolonged respiratory infection.[4]

A persistent of recurrent fistula is also a common cause of respiratory symptoms and can be a particularly difficult diagnosis to make. Particular care has to be taken during barium swallow if this diagnosis is to be made, and maneuvers such as laying the patient prone may be necessary.[3,15] In our study, 10 (24%) of the patients developed this complication, and only 3 of them required surgical repair. Bronchiectasis developed in 7 (17%) of the patients, which are described for the first time in the literature post TEF repair. Half of the patients had gastric or colonic replacement for long gap EA. This may warrant the necessity of using the esophageal ends for final anastomosis and delay the use of gastric or colonic replacement. Such patients will require long life follow up and antibiotic prophylaxis. A high percentage of patients who were ventilated needed ventilation in the pre- and postoperative period [28 (68%)]. The incidence may have been over estimated as the policy of the surgeon in our center to ventilated patients in the postoperative period to reduce tension at the anastomosis site for few 3-4 days.

In summary, although GER is the most common cause of respiratory problems in patients who have undergone TEF repair, other factors are often responsible for these symptoms and should not be overlooked. Persistent or recurrent respiratory symptoms are common and can be the result of misdiagnosis of the original problem, concurrent contributing factors, or failure of the original management to treat the underlying cause of the respiratory symptoms adequately. Careful follow-up of these patients is necessary to detect respiratory problems. Any respiratory symptoms after TEF repair should initiate a complete workup. In some cases, repeated surgical efforts may be necessary to resolve the underlying problem causing respiratory symptoms. Patients who develop GER as a complication of EA/TEF may need to have Nissen fundoplication to improve their respiratory conditions and reduce morbidity.

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