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Lung Function Abnormalities Following Repaired Esophageal Atresia and Tracheoesophageal Fistula

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Introduction: **Esophageal atresia and tracheo-esophageal fistula (EA/TEF) is common in the neonatal period and survival depends on the severity of the associated anomalies, prematurity and pre-morbid factors.**

Objective: This study represents the experience of a tertiary care center in Saudi Arabia of pulmonary function test abnormalities (PFT) after repair of (EA/TEF) including long-term effect on the lungs.

Methods: A retrospective review of all patients referred to pulmonary clinic with EA/TEF and or Pre-operative evaluations from the period 1993-2004.

Results: A total of 41 patients. Twenty-six (63%) males and 15 (37%) females. EA/TEF was diagnosed at birth in 34 (83%). EA and distal TEF were found in 37 (90%) of the patients. Congenital anomalies were associated in 28 (68%). More than 1/3 of the patients had postoperative complications including pneumothorax, recurrent TEF, leakage at operation site and empyema. More than two-third of the patients required prolonged ventilation. Pulmonary complications developed in > 70% of the patients including persistent atelectasis, chronic aspiration pneumonia, tracheomalacia in 12 (29%) and bronchiectasis in 7 (17%). Eighty-eight percent of patients who were able to do PFT showed abnormal values of moderate obstructive and restrictive lung disease.

Conclusion: (EA/TEF) form significant PFT abnormalities and cause significant morbidities that may last for a long period of time.

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The incidence of esophageal atresia and tracheoesophageal fistula (EA/TEF) was reported to be 1 in 4000-5000 live birth, with both sexes affected equally[1]. Type C or EA and distal TEF has 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, gastro-esophageal reflux (GER) and absence of ciliated epithelium in the trachea which impairs clearance of secretion[3]. Congenital anomalies have been described in 50% of patients with TEF, which determine survival and alter treatment approach[4]. Division of TEF and primary anastomosis of esophageal end is the ultimate goal in treating this abnormality. Respiratory complications are a common source of morbidity[3]. In fact, abnormalities in lung function several years after repair of EA/TEF have been documented[5,6]**.** Most commonly these symptoms have been attributed to aspiration resulting from GER. However, there are many other causes of respiratory complications including esophageal dysmotility, tracheomalacia, anastomotic stricture, and recurrent or double fistula[7]. In this report we present the long-term PFT abnormalities and the different factors that contributed to such abnormalities in a tertiary care center in Saudi Arabia.

METHODS

A retrospective review of charts for all EA/ TEF patients referred to the pulmonary clinic for evaluation of recurrent chest infection and pre-operative evaluation during the period from November 1993-October 2004 at King Faisal Specialist Hospital and Research Center (KFSH&RC). Patients' characteristics, TEF types, surgery types, and type of anomalies, morbidities and pulmonary complications were collected. PFT was done in all patients above 5 years of age and who are capable to do the test maneuver. Forced vital capacity (FVC), forced expiratory volume in 1 sec (FEV1), forced expiratory flow between 25% and 75% of vital capacity (FEF 25-75%), total lung capacity (TLC), residual volume (RV), and functional residual capacity (FRC) were measured in a pressure-compensated integrated flow plethysmographic (Sensormedics system 6200 Autobox DL) in a seated position, using standard technique[8]. All results were expressed as percent predicted for height, sex, and age and compared to normal standards[8,9].

Statistical analysis: SPSS 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. Twenty-six (63%) males and 15 (37%) females. Fourty patients (98%) are alive and 1 (2%) died. Fourteen (34%) were premature and 27(66%) were full term. TEF was diagnosed at birth in 34 (83%) of the patients. Patients were referred to KFSH&RC at 15 ± 29 months. Period of follow up was 5 ± 3.8 years. Diagnosis of TEF was based on nasogastric tube coiling (NGT) and by dilated blind esophagus in chest x-ray in 40 (98%) of the patients. EA and distal TEF were found in 37 (90%) of the patients, isolated EA in 2 (5%) and H-type fistula in 2 (5%) of the patients. Congenital anomalies

were associated in 28 (68%) of the 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%). More than one-third of the patients had postoperative complications including pneumothorax, recurrent TEF, major leakage at operation site and empyema (Table 1). Thirty (73%) presented with pneumonia and required prolonged ventilation. Esophageal dysmotility and gastro-esophageal reflux (GER) developed in > 90% of the patients. Twenty-four (60%) of the patients required Nissen fundal plication for GER (Table 1). Esophageal stricture that required > 3 dilatations developed in 16 (46%) patients. GER was significantly related to development of atelectasis, dysmotility, and aspiration pneumonia ($p < 0.05$), but not related to surgery type or its timing, or development of chronic lung disease ($p > 0.09$). 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). Tracheomalacia in 12 (29%) of the patients, 5 of them required aortopexy and one required tracheal stent. Bronchiectasis developed in 7 (17%) of the patients. Pulmonary function test (PFT) was done 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%) (Table 2).

Table 1: TEF Pre And Post-Operative Complications (Total 41 Patients)

Complication Type	**No.**	**%**
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 / O2 requirement	36	88
Tracheomalacia (Bronchoscopy)	12	29
Bronchiectasis by CT chest	7	17
Esophageal Narrowing (anastomosis site)	25	61
Esophageal Dysmotility (by Radiology)	37	90
Gastro-esophageal reflux (by Barium swallow)	39	95
Gastro-esophageal reflux Surgery (Nissen Fundal Plication)	24	59
Failure to thrive	32	78
Pneumothorax (post-operative)	10	24
Recurrent TEF	12	29
Leakage at Operation Site	20	49
Infection (blood) / Empyema/ Mediastinitis	17	42

Table 2: Pulmonary Function Test In Patients With EA/TEF (Total 16 Patients)

Variable	Mean	SD
FVC	67	16
FEV1	68	15
FEV1 / FVC	103	10
MMEF 25-75%	51	19
PEF	62	17
% Ventolin	39	25
FRC	82	15
RV	110	18
TLC	77	11
RV / TLC %	37	6
RV / TLC	134	25

Table 2 - Legend:
FVC- Forced vital capacity
FEV1- Forced expiratory volume in one second
MMEF- Maximum mid expiratory flow
PEF- Peak expiratory flow
% Ventolin- percentage of change in FEV1 values after administration of Ventolin
FRC- Functional residual capacity
RV-Residual volume
TLC- Total lung capacity
RV/ TLC- The ratio of RV/TLC in percentage and actual values

DISCUSSION

Long-term pulmonary complications have been described before (Table 3)[3,10-12]. Couriel et al described bronchitis for more than 8 years in 5/20 patients (25%), and denoted that lung disease improves with time[10]. Chetcutti et al described asthma development in 40/155 (26%) patients after TEF repair, with restrictive lung changes in 18 (12%) of the population[11]. Delius et al showed that 31/68 patients (46%) developed recurrent pneumonia that required 1-10 admission to hospital for treatment[3]. Robertson et al performed PFT in 25 patients with TEF repair and their siblings and found that: although PFT values were within normal limits, but were significantly different compared to their siblings[12]. The later study also showed that 6/25 patients had positive methacholine challenge test as a sign of obstructive airway disease and 9/25 had restrictive pattern (Table 3).

Table 3: Pulmonary Function Test In Patients With EA/TEF

Author	Couriel	Chetcutti	Delius	Robertson	Banjar
Year of publication	1982	1992	1992	1995	2004
Country name	Australia	Australia	USA	Canada	KSA
Population	20	155	68	25	41
Bronchitis > 2yr	5 (25)	40 (26)	4 (21)	18 (72)	36 (88)
Pneumonia (1-10 admission)	17 (85)		31 (46)	11 (44)	40 (98)
Asthma/Hyper reactive airway	5 (25)	40 (26)	31 (46)	6 (24)	40 (98)
Abnormal PFT				13 (52)	14/16 (88)
FVC	85 ± 11	- 0.9 (1.1) ♠		91 ± 14	67 (16)
FEV	83 ± 11	- 1.4 (1.1) ♠		89 ± 10	68 (15)
FEV / FVC	86 ± 7				103 (10)
MMEF (25-75%)	87 ± 27	- 1.2 (0.86) ♠		97 ± 29	51 (19)
FRC	103 ± 18				82 (25)
TLC	95 ± 11	- 0.5 (2) ♠		92 ± 13	77 (11)
RV	110 ± 21				110 (15)
RV / TLC		+ 1.95 (2) ♠		23 ± 5	37 (6)
Restrictive		18 (12) ♠		9 (36)	8 (50)
Obstructive				3 (12)	3 (19)
Mixed				1 (4)	3 (19)

Legend:
♠ - (SD below the mean)
PFT - (as mentioned in the text)

GER is the most prevalent cause of respiratory symptoms in this and several other reports[13,14]. GER in these patients has been attributed to a variety of causes, including dysfunction of the distal pouch and decreased lower esophageal sphincter pressure[3]. In many centers the appearance of respiratory symptoms in these patients prompts a diagnostic workup to rule out GER. However, because GER is common in this population and not all patients with GER have respiratory symptoms, the finding of GER in a patient with pulmonary symptoms does not definitively establish cause and effect. Delius reported 20 patients were initially found to have reflux[3]. In each case initial management was predicated on this finding. However, eight patients continued to have respiratory symptoms and four of these patients were ultimately found to have an entirely different cause of the symptoms such as tracheomalacia and recurrent fistula, underscoring the need for a thorough evaluation before recommending an antireflux procedure[3]. The incidence and relative significance of these multiple causes of respiratory symptoms are unclear.

Our study has shown a high number of patients 39 (95%) who developed GER and 24 (59%) of them required Nissen Fundoplication (Table 2). GER was significantly related to development of atelectasis, dysmotility, and aspiration pneumonia ($p < 0.05$), but not related to surgery type or its timing, or development of chronic lung disease ($p > 0.09$).

Our findings are similar to previously reported studies[7-16]**.** Esophageal stricture developed mainly in those patients who were referred for complication, but did not develop in those who had their primary repair in a tertiary care center.

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]. Cineradiography is also helpful in making this diagnosis. The workup of this problem should be circumspect, and the pediatric radiographer should take special care to rule out a persistent or recurrent fistula when performing the initial barium swallow. Patients should also undergo diagnostic bronchoscopy to rule out tracheomalacia even if the barium swallow shows GER[3]. Patients with a normal barium swallow and a normal bronchoscopic evaluation should undergo pH study to rule out GER not detected by barium swallow. If the result of this study is normal and respiratory symptoms persist, a recurrent fistula should be strongly suspected and cineesophagography should be repeated. If the cineesophagography is still normal, double endoscopy (simultaneous esophagoscopy and bronchoscopy) with instillation of methylene blue dye should be performed[3].

Bronchiectasis developed in 7 (17%) of our patients, which is described for the first time in the literature post TEF repair. It required long life follow up and antibiotic prophylaxis. This could be explained in view of recurrent infections, persistent atelectasis and recurrent aspirations due to GER that masked the early recognition of such complication.

Tracheomalacia formed a significant morbidity in 12 (29%) of the patients and required surgical intervention in 50% of them. It has been described to cause apnea, cyanosis and prolonged respiratory infection[14].

CONCLUSION

TEF repair is associated with significant morbidities that need to be recognized and managed early before significant and irreversible complications develop.

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