Long-term pulmonary consequences of esophageal atresia and tracheoesophageal fistula

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Abstract

This study represents the experience of a tertiary care center in Saudi Arabia including long-term effect on the lungs.

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

A total of 41 patients. twenty-six (63%) males and 15 (37%) females. EA/ TEF were 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 post-op complications including Pneumothorax, recurrent TEF, leakage at operation site and Empyema. More than 2/3 of the patients required prolonged ventilation. 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 PFT showed abnormal values of moderate obstructive and restrictive lung disease.

Patients with TEF and long gap EA, prematurity and congenital anomalies should be referred early to a tertiary care center to prevent long term complications

Introduction

The incidence of esophageal atresia (EA) and tracheoesophageal fistula (TEF was reported to be 1 in 4000-5000 live birth [1], with both sexes affected equally. 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, gastroesophageal 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]. Prognostic classification of Waterston and Montreal types have been used to determine survival which involve birth weight, presence or absence of pneumonia and minor or major congenital anomalies and the need for ventilation before surgical intervention [5,6]. Division of TEF and primary anastomosis of esophageal end is beneficial in healthy full term infants with no significant pulmonary disease and stable cardiovascular status. Delayed primary repair for short period of time (2-4 weeks) is preserved for premature patients or patients with minor congenital anomalies. Staged procedure is preserved for unstable patients with prematurity, severe respiratory distress and major congenital anomalies with long gap EA. In the later group, patients may need to have GT insertion, esophageal suctioning followed by retropleural or transpleural division of TEF,

correction of life threatening anomalies and later esophageal anastomosis, which is done through a second thoracotomy (6). In this report we present the different factors that contributed to morbidity and mortality of a tertiary care center in Saudi Arabia.

Materials and 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 Nov. 1993- Oct. 2004 at King Faisal Specialist Hospital and research center (KFSH&RC) in the Riyadh region which is considered the main tertiary care center for referral of complicated cases in Saudi Arabia. Demographic, TEF types, surgery type, associated anomalies that involve any part of the body either minor as limbs anomalies or major as cardiac, renal, gastrointestinal and central nervous system anomalies that required invasive procedures or surgical interventions, morbidities that required emergency visits or hospitalization, and pulmonary complications were collected.

Gastroesophageal reflux was diagnosed by barium swallow studies and or milk scan. None of the patients underwent esophagoscopy to diagnose esophagitis.

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.

Definitions

Esophageal stricture

It requires > 3 dilatations to improve swallowing.

Group (1)

Patients who were diagnosed and had their esophageal anastomosis at KFSH&RC.

Group (2)

Patients who had their final anastomosis at a local hospital and then referred to KFSH&RC for treatment of chest complications.

Group (3)

Patients who had their initial surgery such as esophageal spit 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.

Results

A total of 41 patients, 5 (12%) in group (1), 10 (25%) in group (2), and 26 (63%) in group (3). 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 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. Major and minor congenital anomalies were associated in 28 (68%) of the patients. Cardiac anomalies were found in 11 (27%). Large ventricular septal defect (VSD) in 2 patients, 2 patients had a combination of VSD and atrial septal defect (ASD), secundum

type, Dextrocardia in one patient, Tetralogy of Fallot in 3 patients, PDA in 2 patients, and one patient with Coarctation of the aorta.

Gastrointestinal (GIT) in 8 (20). Two patients with duodenal atresia, one patient with jejunal atresia, 2 patients with imperforate anus, one patient with duplicate ileal cyst and congenital villous atrophy, and 2 patients with colonic stricture that required colostomy tube insertion. Respiratory system anomalies in 12 (30%). Two patients with congenital pneumatocele, 4 patients with right lung hypoplasia, 5 patients with severe tracheo-bronchomalacia hat required aortopexy, and one patient with right diaphragmatic hernia. Renal in 7 (17%), One patient with lobulation of right kidney in combination with renal calcification and renal stones, 4 patients with severe vesicoueretric reflux grade 3 and 4 with Hydronephrosis and one of them required nephrectomy, one patient with hypospadia and one patient with urethral stricture. Skeletal in 12 (30%. Ten patients with rib anomalies with fusion of at least 2 ribs, 2 patients with spina bifida in addition to sacral anomalies, vertebral anomalies and scoliosis. Chromosomal in 7 (17%). Five patents with VATER association (vertebral, anal, tracheal, esophageal, renal and radial association), one patient with VACTREL association (vertebral, anal, cardiac, tracheal, renal, esophageal and limb association), one patient with Down's syndrome. Central nervous system anomalies in 11 (27%) of patients. One patient with hypoplastic left cerebellum, 4 patients with hydrocephalus that required ventriculo-peritoneal shunt, 2 patients with cerebral hypoplasia and seizure disorders. 4 patients with microcephaly. Twenty-one (51%) had a primary repair, 14 (34%) had delayed primary and 6 (15%) had staged repair. More than 1/3 of the patients had post-op 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 gastroesophageal reflux (GER) devel oped in > 90% of the patients. 60% of the patients required Nissen fundal plication for GER (Table 1). Esophageal stricture that required > 3 dilatations developed 16 (46%) of the patients, and all of them from group 2 and 3, but none form group (1) (P= < 0.02). 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. Two of them after gastric tube replacement of esophagus, 1 after colonic replacement, and 4 developed after primary repair. Two of the 4 patients with primary repair were premature, another one with multiple ribs, vertebral anomalies in addition to VSD and the 4th one with recurrent fistula, esophageal diverticulum and VSD. 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). Patients in group 2 and 3 developed more morbidity, required longer period of time for final anastomosis compared to patients in group (1) ($P < 0.05$) (Table 3).

Discussion

Many studies have shown that mortality increases with major congenital anomalies [2,4-8]. Rejjal et.al [8] (from the same study center) described 89 patients with TEF diagnosed at the neonatal period with 44 (49%) had associated anomalies. Thirteen of 89 (15%) patients died in the neonatal period from cardiac and chromosomal anomalies with a total survival of 85% [8]. Long-term pulmonary complications have been described before [3, 9-11]. Couriel et al [9] described Bronchitis for more than 8 years in 5/20 patients (25%), and denoted that lung disease improves with time. Chetcutti et.al [10] described asthma development in 40/ 155 (26%) patients after TEF repair, with restrictive lung changes in 18 (12%) of the population. Delius et al [3] showed that 31/ 68 patients (46%) developed recurrent pneumonia that required 1-10 admission to hospital for treatment. Robertson et al. [11] 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. 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.

GER and esophageal stricture have been described in many studies [7,12,13] as a complication post TEF repair in approximately 40-70% of patients. It is estimated to be secondary to tension at the anastomosis site, dysmotility of the lower esophagus, and altered angle of Hiss due to distal esophageal mobilization during surgery. Fundoplication may be required in about half of the patients if medical treatment failed [13]. 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 (1-30). Esophageal stricture developed mainly in those patients who referred to KFSH&RC for complication, but did not develop in those who had their primary repair in a tertiary care center.

Bronchiectasis developed in 7 (17%) of the 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 complications. Prematurity and complicated clinical course have contributed significantly in the cause of bronchiectasis in our population. Another important cause is the replacement of the esophagus with gastric tube or colon, which need to be addressed again as the last option in staged surgery unless spontaneous elongation of esophagus is failed. Such patients need to be followed for undetermined period of time to ensure that such complication is avoided and to be treated early.

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].

Our report has shown that a significant morbidity developed in more than 2/3 of the patients who had TEF repair and needed a prolonged follow up due to chest symptoms. Mortality of patients who survived the neonatal period is less if managed carefully in a specialized center. Our survival was 97.5% as only one patient died after TEF repair with VACTREL association.

In summary: Patients with EA/ TEF with congenital anomalies and morbid factors such as long gap esophagus, and prematurity should be referred early to a tertiary care center to prevent long term complications.

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/ Hyperreactive 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
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
Failure to thrive	32	78
Pneumothorax (post-operative)	10	24
Recurrent TEF	10	24

Leakage at Operation Site	16	39
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 3: COMPARISONS OF OUT COME IN BETWEEN 3 HOSPITAL SETTINGS (TOTAL 41 PATIENTS)

Variable Total patients	Group (1) 5	Group (2) 10	Group (3) 26	P Values
Diagnosis to initial Surgery (months)				
Median	0.066 (2 days)	4.3	5.2	0.027
Range	0.00 – 0.27	0.07 – 60	0.03 – 128	
Mean	0.106 ± 0.1	14.5 ± 20	15.3 ± 28	
Premature	4 (80%)	4 (40%)	6 (23%)	0.04
Full term	1 (20)	6 (60)	20 (77)	0.04
Infection (blood)	2 (40)	1 (10)	14 (54)	0.05

Atelectasis	5 (100)	7 (70)	25 (96)	0.04
Asthma/Hyper reactive airway disease	4 (80)	10 (100)	26 (100)	0.025
Dysmotility	3 (60)	10 (100)	24 (92)	0.04
GER surgery	1 (20)	4 (40)	19 (73)	0.034
Esophageal stricture	0.0	2 (20)	15 (56%)	0.02

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

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