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Original Research Article

Respiratory morbidity in children who had undergone primary repair of esophageal atresia with tracheo esophageal fistula in a tertiary care centre in India

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ABSTRACT

Background: Objective of the study was to assess the prevalence of respiratory and related GI morbidity in children who had undergone repair of oesophageal atresia/tracheoesophageal fistula (TEF) in the neonatal period.
Methods: Patients operated during the period 2008–2015 were evaluated in 2017 by clinical evaluation, radiological imaging (chest X-ray, barium swallow study and high-resolution computed tomographic scan) and laryngoscopy examination.
Results: Of the 50 neonates who underwent primary repair of TEF during the 8 year period, 39 were discharged successfully. Twenty-six of them at a Mean age of 64 months were included in the study. At the time of assessment, 65% of subjects registered body mass index (BMI) less than 10th centile. Abnormal respiratory system findings in 63% and digital clubbing in 27% were noted. Unilateral asymptomatic vocal cord was demonstrated in 7.7%. Radiological evaluation showed abnormal oesophageal motility/reflux in 54% and bronchiectasis in 19%. In addition, 62% had needed hospitalization for recurrent pneumonia in infancy. Environmental smoke exposure was reported in 38% and 73% had no regular follow up with a pediatrician.
Conclusions: This comprehensive evaluation of respiratory morbidity in children who had been operated for TEF in the neonatal period, demonstrated a high prevalence of reported respiratory morbidity in the first year of life. Poor nutritional outcomes and abnormal respiratory system examination were observed in a significant proportion. One-fifth of these patients had already developed bronchiectasis by a mean age of 75.4 months. Asymptomatic unilateral vocal cord palsy was another important finding.

Keywords: Oesophageal atresia/tracheo esophageal fistula, Respiratory morbidity and sequelae

INTRODUCTION

Among the congenital anomalies of foregut, esophageal atresia with or without tracheo esophageal fistula (OA/TEF) is one of the common defects with an incidence of 1 in 2500 per births.[1] With advancements in the fields of surgical techniques and peri operative intensive care, the survival rate after TEF repair has reached over 90%.[2] With improved survival, it is important to focus on prevention of long term morbidity and complications.

It has been established that respiratory morbidity of survivors of this surgery is high. Medical literature from developed countries shows that patients require multiple admissions for respiratory and gastrointestinal symptoms in infancy and beyond.[3,4] Sequelae of this respiratory involvement have been shown to persist to adulthood with

20% demonstrated restrictive lung disease and 41% having airway disease on pulmonary function tests. However there is a paucity of data on the lung morbidity of these patients who live in developing countries.

In India, where post-surgery follow up is suboptimal, operated OA/TEF patients tend to present to the clinician after irreversible lung damage has occurred. In this study, we aimed to assess the burden of respiratory and related GI morbidity in those who were operated for TEF in the neonatal period.

METHODS

This study was carried out in Christian Medical College, Vellore, India after obtaining approval from institutional ethics committee. Patients who were discharged home after neonatal surgery for OA/TEF during the period January 2008 to December 2015were identified and parents were contacted. Cross sectional observational study on this cohort was conducted between December 2016 and June 2017 after obtaining written consent.

All recruited subjects underwent clinical evaluation by the investigator. History pertaining to respiratory system and GI system in infancy was obtained and verified with hospital records and patient's own medical records wherever available. Assessment of nutritional status and physical examination was done. Radiological studies included chest X-ray and barium swallow studies for all patients. High resolution computed tomography (HRCT) scan of the chest was done for children who reported ongoing respiratory symptoms or had abnormal findings on plain radiography. All studies were reported by a single radiologist. Detailed ear, nose and throat (ENT) examination and flexible laryngoscopy was done by a single pediatric ENT surgeon.

Data analysis was done using STATA/IC 15.0. The data were expressed as number along with percentage. Association between bronchiectasis and selected risk factors were analyzed using Fischer exact test and the estimate of effects were presented as relative risk (95% CI).

RESULTS

This cohort of 26 patients, whose mean age was 64 months (range 13-103 months), included 12 boys. Analysis of the peri-operative data obtained from hospital records showed that 27% of these babies were born preterm and median birth weight was 2470 grams. Forty two percent were born elsewhere and then referred to our centre for management. All 26 subjects had type C-TEF according to Gross classification i.e. blind upper esophagus with lower end connected to the trachea and three fourth of them underwent corrective surgery within day two of life. Other congenital anomalies were observed in this group as shown in Table 1. In many, post-operative period was complicated by blood stream infection (23%), prolonged ventilation (27%) or anastamotic leak (27%). None required surgical re-exploration or tracheostomy.

Table 1: Associated anomalies in the subjects detected during neonatal period (n=26).

	Number	Percentage (%)
Cardiac	19	73
PDA	15*	
Others	14	
Vertebral	6	23
Single umbilical artery	2	7
Imperforate Anus	4	15
Renal anomaly	4	15
Cloacal anomaly	3	11
VACTERL	6	23

*- Preterm PDA: 5

Morbidity in the first 2 years of life

Analysis of the historical information pertaining to the period of infancy showed that there was significant respiratory and gastro intestinal morbidity in the first year of life. Wheeze, prolonged cough of more than 2 weeks or stridor was reported by majority of them and 62% required hospitalization for respiratory illness before the first birthday. In the second and subsequent years notable reduction in frequency of respiratory symptoms was observed.

Gastro–esophageal symptoms in the first year varied from simple regurgitation to food bolus impaction. Half the patients had significant GER in infancy documented by barium swallow and had received medical treatment. One third of children had undergone fundoplication by the second birthday.

The other commonly noted upper GI problem was dysphagia secondary to esophageal stricture. While 50% had reported dysphagia, 31% required at least one esophageal dilatation procedure for food bolus impaction during the period of infancy. Interestingly, while occurrence of serious respiratory morbidity needing hospitalization and GI symptoms like vomiting decreased by 8% and 6% respectively after the first year, dysphagia symptoms and incidence of food bolus impaction worsened by 8% after the first birthday. This is likely due to the introduction of solid food at this stage.

Assessment of current morbidity

At the time of assessment (age range 13–103 months), 62% of subjects reported to have ongoing significant respiratory symptoms like prolonged cough, wheeze or recurrent pneumonia. However only 27% were under regular follow up with a pediatrician. There was high rate of exposure to household cigarette smoke or wood fire smoke (38%).

Nutritional assessment revealed that height and weight for age was less than 10th centile.[5] Body mass index (BMI) was less than 10th centile in 65% of the patients when plotted on CDC chart. Significant physical findings as shown in Table 2 included clubbing and abnormal auscultatory findings.

Plain radiography revealed abnormal findings in 69%, which included bronchiectasis and collapsed lung segments. Barium swallow study did not demonstrate aspiration into the airway tract in any patient, but was abnormal in terms of esophageal motility, reflux or presence of stricture.

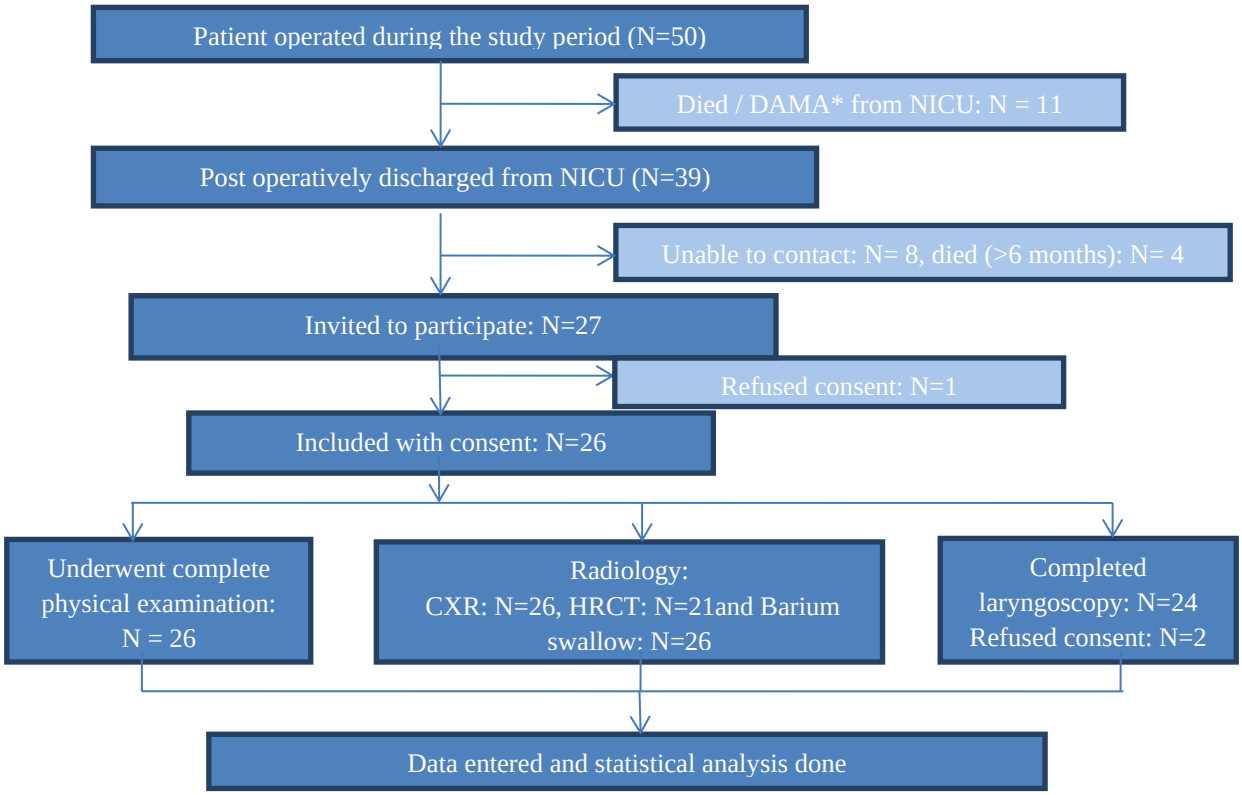
Out of 21 patients on whom high resolution computed tomography (HRCT) of chest was done for clinical indications, five children had bronchiectasis giving an overall prevalence of 19% in the cohort.

Though this cross sectional study was not designed to assess the risk factors for development of sequelae like bronchiectasis, we analyzed the data for hypothesis generation and to look for possible associations. Data from this small cohort of patients suggest that current gastro esophageal reflux demonstrated on barium swallow study may be a risk factor for development of bronchiectasis (RR=1.75, CI=0.42-7.35).

Table 2: Important findings on physical examination and radiological imaging (n=26).

Clinical examination	N	Percentage
Increased work of breathing	10	38%
Adventitious sounds on auscultation	16	61%
Clubbing	7	27%
Chest X Ray	N=26	
Volume loss/collapse	8	30%
Bronchiectasis	3	11%
Normal	8	30%
Barium Swallow	N=26	
Abnormal swallowing	9	34%
Reflux	11	42%
Oesophageal stricture	10	38%
Esophageal diverticulum	3	11%
HRCT – chest finding	N =21	
Tracheal diverticulum	15	57%
Bronchiectasis	5	24% **
Collapse	2	7%
Oesophageal diverticulum	2	7%

**(19 % of whole cohort)



*DAMA - Discharge against medical advice.

Figure 1. The algorithm of the study.

Table 3: Comparison of the results of the study with selected published data.

Results of study	Our study (n=26)	Literature review	No. of subjects	Author, country year of publication (ref)
Base line data				
Prematurity (%)	27	32	97	Seo, Korea, 2010[6]
Birth weight (gm)	2336	2552	371	Deurloo, Netherland, 2002[7]
Cardiac anomalies (%)	73.1	65.6	463	De Jong, Netherland, 2008[8]
First year morbidity				
Recurrent respiratory symptoms in early childhood (%)	96	68	334	Chetcuti, Australia, 1993[9]
Hospital admission for respiratory illness in the first year of life (%)	62	48	307	Schneider, French registry, 2014[10]
History of GERD (%)	39	34	169	Rintala et al, Finland, 2017[11]
Current morbidity				
GERD (%)	42 (current)	48	69	Little et al, Indiana, 2003[12]
Bronchiectasis (%)	19 (n=26)	17	41	Banjar, Saudi Arabia, 2005[13]
Vocal cord palsy (%)	13 (n=24)	7.7	65	Robertson, Columbus, Ohio, 1976[14]

DISCUSSION

This study done in a tertiary care centre in south India provides results of comprehensive respiratory evaluation in survivors of neonatal OA/TEF surgery.

In developing countries like India, burden of respiratory morbidity in operated OA/TEF patients, especially in infancy is underrecognized and our study has estimated the prevalence to be much higher than that reported from other countries. Lack of adherence to a structured follow up by a respiratory specialist or pediatrician in spite of ongoing symptoms is alarming.

Diagnosis of GERD and surgical intervention is less in our group compared to 59% fundoplication rate reported by other authors. This difference is partly due to under diagnosis as a result of poor adherence to follow up plan. Stricture formation is a known complication of TEF surgery developing in up to 64%, while only 34.6% of patients in this cohort had symptomatic stricture. This difference may be due to selection bias as there were a number of patients who could not be followed up.

Development of bronchiectasis is an irreversible damage to lung which negatively impacts the overall health of the child. Though we limited HRCT evaluation to those who had respiratory symptoms or abnormal chest X-ray, it is likely to be the correct estimate of the prevalence in the whole group studied. Prevalence of 19% is similar to reports by other researchers.

An interesting observation is that in 38%, parents considered that their children have no reportable respiratory problems, while in fact only 3 (11%) children were completely normal by examination and diagnostic evaluation. This highlights the need for a structured follow up plan (irrespective of perceived need) to be discussed with the parent at the time of discharge from neonatal or surgical ward. In this cohort, physical examination and plain radiography at primary care level would have identified children at risk who needed specialist evaluation.

In our cohort, tracheal diverticulum at the site of surgery was reported in 58% of the CT scans. Tracheal diverticulum in patients with bronchiectasis has been reported in anecdotal cases in adults. Presence of a diverticulum may interfere with normal drainage of secretions predisposing to bronchiectasis. Significance of this observation needs confirmation by further studies with bronchoscopy in children operated for TEF.

Though barium swallow study is not a gold standard test for GE reflux, we used this modality because of its ability to identify incordinate swallowing, reflux and stricture in these patients. These may contribute to silent micro aspiration which worsens the lung damage.

Asymptomatic unilateral vocal cord palsy was another important finding, but it is not clear if this has contributed to respiratory morbidity.

Limitations

Of those who underwent neonatal TEF surgery and discharged from hospital, we could recruit only 74%. Data collection about early life morbidity could be done only in a retrospective manner from history and medical records. Bronchoscopy evaluation and infant pulmonary function tests were not done as part of the study.

What is already known: Western studies have shown high prevalence of ongoing respiratory morbidity in survivors of neonatal TEF surgery.

What this study adds: This study is first of its kind from India providing data on the high burden of respiratory morbidity and sequelae among children operated for TEF in neonatal period. It high lights the need for structured

follow up to identify risk factors and early indicators of lung damage.

CONCLUSION

In conclusion, this comprehensive evaluation on survivors of neonatal TEF surgery from a tertiary care centre in south India shows that there is high prevalence of respiratory and upper GI morbidity, especially in the year following surgery. One fifth of these patients had already developed bronchiectasis when evaluated at a mean age of 75.4 months. Ongoing micro aspiration due to oesophageal dysmotility, gastro esophageal reflux or asymptomatic vocal cord palsy has to be systematically evaluated in these patients. One fifth of these patients had already developed bronchiectasis when evaluated at a mean age of 75.4 months. Lack of regular follow up care by a pediatrician was a matter of concern in the majority.

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Conflict of interest: None declared
Ethical approval: The study was approved by the Institutional Ethics Committee

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