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C O M I N G T O G E T H E R

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Theme 9 – Young Investigator Session – Abstracts

[TH9-1] Prevalence of dumping syndrome after a surgery for oesophageal atresia type c without fundoplication

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Introduction: Dumping syndrome (DS) is a complication of antireflux surgery. We recently reported 2 cases of DS in 2 children with oesophageal atresia (OA) but without fundoplication, suggesting the oesophageal anastomosis and/or OA *per se* be responsible of accelerated gastric emptying.

Aim: To assess the prevalence of DS in infants operated for OA.

Method: We conducted a prospective multicentric study including infants with type C OA. Before the age of 3.5 months and when they weigh more than 4.150 kg, patients underwent an oral glucose tolerance test (OGTT). Glycemia and insulinemia were measured every 30 minutes for the first 2 hours and every hour until 4 hours after 1.75 g/kg oral glucose intake. The test was stopped if children presented with clinical symptoms and/or hypoglycemia. DS was defined as early hyperglycemia (more than 1.8 g/L until 30 minutes, > 1.7 g/L between 30 minutes and 2 hours and > 1.4 g/L between 2 and 3 hours) and/or late hypoglycemia (less than 0.6 g/L after 2 hours).

Conclusion: 30 patients were included to date in the study, in 3 patients OGTT was not possible because of technical problems. Among the 27 infants who completed OGTT, 11 (41%) presented DS: 5 with early hyperglycemia (2.03 [1.74-2.98]), 4 with late hypoglycemia (0.5 [0.39-0.56]) and 2 with early hyperglycemia and late hypoglycemia. Only 1 of the 6 patients with hypoglycemia at OGTT had clinical sign of hypoglycemia during the test. Parents reported before OGTT frequent abdominal pain in 5 children, bloating in 2 children, diarrhoea in 1 child, and agitation and sweat in 1 child. These preliminary results suggest that dumping syndrome in children operated at birth for OA type C is frequent and independent of antireflux surgery, underdiagnosed because asymptomatic or presenting with nonspecific clinical symptoms. We suggest a systematic screening of infants operated for type C OA.

[TH9-2] High prevalence of Barrett's esophagus, esophagitis and esophageal cancer after esophageal atresia repair: an update

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Introduction: Improved life expectancy, high incidence of gastroesophageal reflux, and young esophageal cancer cases, raise concerns about a possible increased risk of Barrett's esophagus (BE) and esophageal carcinoma in esophageal atresia (EA) patients. Similar genetic pathways may be involved in the development of EA, BE and esophageal carcinoma.

Aim: To assess the prevalence of BE and esophageal carcinoma in EA patients .

Methods: Prospective cohort study in a tertiary referral center (May 2012-April 2016) in adult EA patients. EA patients (age \geq 17 years) were invited for gastroscopy with random biopsies of the distal esophagus at the level of the gastro-esophageal junction (GEJ) and standard 4-quadrant biopsies in case of BE. All clinical, endoscopic and histological data were prospectively registered in a dedicated database.

Results: We included 93/118 patients (60.2% male; median age 22.2 (range 16.8-55.9) years). History of reflux was found in 58%, esophageal dilation in 66.7%, and 29% of the patients had undergone fundoplication surgery. Endoscopy revealed a normal esophagus in 69.9%, esophagitis in 4.3%, and columnar-lined esophagus in 25.8% of the patients. Histology revealed inflammatory changes within normal limits in 48.4%, esophagitis in 23.7%, gastric metaplasia in 17.2%, and intestinal metaplasia without dysplasia in 8.6% (median age 27.8 (range 17.9-45.3) years). One male patient (0.8%) developed esophageal squamous cell carcinoma twice (at ages 44 and 60 years). No clinical predictors for BE or metaplasia were found.

Conclusions: Prevalence of BE in young EA patients is 5.4 fold higher than in the general population. This signifies relevant clinical implications including transition of young adults from pediatric care units to adult gastroenterology departments for lifelong endoscopic surveillance. Genetic predisposition may play a role in BE development and carcinogenesis in EA patients. Future studies are needed to assess genetic profiles helping to identify those at risk and personalize screening and surveillance regimes.

[TH9-3] Prevalence of laryngo-tracheal abnormalities in esophageal atresia patients

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Introduction: Esophageal atresia (EA) patients frequently present respiratory disorders secondary to laryngo-tracheal abnormalities (LTA), whether congenital or acquired after esophageal surgery. To date, different authors suggest preoperative tracheobronchoscopy in patients with EA as a valuable tool to evaluate vocal cord motility, assessing laryngeal and tracheal anatomy, while preoperatively measuring esophageal GAP. Nonetheless, the overall incidence of LTA in EA infants is not well known.

Aim: to evaluate the prevalence of laryngo-tracheal problems in patients treated for esophageal atresia.

Methods: All patients who had an hospital discharge with a IDC-9-CM code 7503 (tracheoesophageal fistula and esophageal atresia), between January 2008 and June 2016 entered the present study. LTA were defined as presence of moderate to severe tracheomalacia, AND/OR subglottic stenosis AND/OR vocal cord paralysis (mono or bilateral), other major LTA (cleft, major complex anomalies). Patients were divided based on the presence/absence of LTA. Clinical files of all patients treated were evaluated.

Results: During the study period 163 patients were treated. LTA were observed in 52 patients (33%) out of 163. Table 1 summarized main outcomes.

Conclusions: Present data suggest a high prevalence of congenital LTA in patients affected by EA. Most of the abnormalities were congenital and a high proportion of patients (24%) with LTA required tracheostomy (12/52). Mortality significantly correlates with the presence of LTA.

		LTA + 52 pts	LTA - 111 pts	p
GA: weeks. median (IQR)		37.5 (35.25-39)	38 (36-39)	0.81
BW: ar. median (IQR)		2505 (2125-2990)	2625 (2313-	0.32
EA Type A-B/C-D		5/44	19/88	0.52
EA Type E		3	4	0.68
Referral		11	17	0.51
Long-gap EA		17	23	0.11
Associated anomalies		16	21	0.11
Congenital heart abnormalities		9	9	0.10
LTA	Moderate-severe tracheomalacia	33	-	-
	Subglottic stenosis	11	-	-
	Vocal cord paralysis	11	-	-
	Other major LTA	31	-	-
Tracheostomy		12	-	0.0001
Deaths		9	2	0.0005

[TH9-4] Intellectual, academic, and social-emotional outcomes in school-aged children with oesophageal atresia

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Introduction: As mortality associated with a diagnosis of oesophageal atresia (OA) at birth has significantly improved, there has been increasing interest in long-term morbidity and its associated challenges.

Aim: To investigate the intellectual, academic and social-emotional functioning of children diagnosed with OA at key points in childhood and to explore the medical and sociodemographic variables predicting outcome.

Method: Children treated for OA in Victoria between 2003 and 2010 were invited to attend a neurodevelopmental follow-up clinic at the Royal Children's Hospital. At ages 5 or 8 years, children were assessed with measures of intellectual functioning (WPPSI-III; WISC-IV), academic skills (WRAT4) and social-emotional outcome (parent questionnaire: BASC2). Scores were compared to normative data (mean 100±15 for intellectual and academic measures; 50±10 for social-emotional variables) using t-tests. Length of stay, VACTERL diagnosis, maternal education, type of OA and birth weight were entered as potential predictors of outcome in regression analyses.

Results: The groups' descriptive characteristics are outlined in Table 1. No significant findings were revealed for IQ in the 5-year age range. However, anxiety ($p < .001$), somatisation ($p < .001$) and social skills ($p = .023$) were rated as significant concerns by parents. In contrast, significantly poorer performances were found in the 8-year-old group for Full Scale IQ (FSIQ; $p = .043$), processing speed and working memory (both $p = .004$), and mathematics ability ($p = .001$). In addition, parents rated activities of daily living ($p = .017$) and functional communication ($p = .026$) as significantly reduced. Increased somatisation symptomatology was predicted by a positive diagnosis for VACTERL in the 5-year-old group ($p = .030$). In the 8-year-olds, lower maternal education predicted lower FSIQ ($p = .017$).

Table 1. Descriptive characteristics of samples

	5 year-old sample (n=51)	8 year-old sample (n=32)
Male (%)	30 (59%)	18 (56%)
Gestation (mean; SD)	37 (3.5) weeks	36.9 (3.1) weeks
Birth weight (mean; SD)	2742 (821) grams	2662 (774) grams
Type of OA Repair (%)	Short= 46 (90%); Long= 3 (5.9%); Delayed= 2 (4%)	Short= 25 (78%); Long= 4(12.5%); Delayed= 3(9.4%)
Length of Stay (median, IQR)	30 days (17-41)	31.5 days (12.5-92)

Age at assessment (mean; SD)	5.4 (0.3) years	8.5 (0.3) years
Number of oesophageal dilatations (median, IQR)	3 (0-6.25)	2 (1.5-6.5)
VACTERL Diagnosis (%)	5 (9.8%)	6 (18.8%)
Secondary Diagnosis	1 syndromic	1 mild autism; 1 syndromic
Maternal Education (completed High School)	64.7%	84.4%

Conclusion: The findings suggest that internalizing difficulties are more significant at younger ages with IQ, and mathematics and functional difficulties emerge with increasing age. Long-term follow up into at least middle childhood is warranted in infants treated for OA with intervention targeting different areas at different time points.

[TH9-5] Gastroesophageal reflux in young children with esophageal atresia: when is it time to stop the ppi?

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Introduction: Clinical course in esophageal atresia (EA) may be complicated by gastroesophageal reflux and formation of anastomotic strictures. Patients are therefore treated systematically with proton pump blockers (PPI) according to published guidelines.

Aim: The aim of this study is the evaluation of PPI treatment in a prospectively followed cohort of EA patients at a tertiary center with determination of predictive factors for long-term treatment.

Patients and methods: Children born with EA from September 2005 to December 2014 were included in the study. First pH-study at 1 year and esophagoscopy within 4 years after surgery were performed. Evaluation of the current PPI treatment was made at time of last follow-up.

Results: 86 patients (36 girls) were included, 10 type A, 73 type C, 3 type D (11 long gap with delayed anastomosis). 7 patients died and were not analyzed. All 79 patients but 2 (H2RA) were treated with PPI.

Median age at follow-up was 57 months (range 17-130).

At time of last follow-up acid suppressive treatment was stopped in 43 (54%). In this group, pH-study was performed in 38 patients and was normal in 27 (71%). Among the 11 with abnormal pH-study, 7 had an esophagoscopy (5 normal, 2 with recurrent anastomotic strictures).

35 patients (44%) had still ongoing treatment with PPI at time of follow up (9 with long gap). In this group, 16 patients presented with recurrent anastomotic strictures (6 with long gap). 7 pH-studies were performed in this group, 4 were normal. 28 patients underwent esophagoscopy: 4 patients were diagnosed with eosinophilic esophagitis, 4 presented with gastric metaplasia and 2 with intestinal metaplasia.

Conclusion: PPI treatment could be stopped in 53% of EA patients. Long gap, anastomotic stricture, gastric or intestinal metaplasia and eosinophilic oesophagitis seem to be predictive factors for long-term PPI treatment.

Poster walk abstracts

[P005] Long term Esophageal patency after Magnetic anastomosis (Magnamosis)

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Introduction: Use of sutureless compression anastomosis was described by Murphy more than 100 years ago.

Objective: To evaluate long term patency of sutureless Esophageal Magnetic compression anastomosis (Magnamosis)

Material & methods: Between June 2001 and July 2015, 14 patients (9 males and 5 females) underwent Magnamosis in a medical center in Argentina. 6 patient with Esophageal atresia (EA) type A and 8 with EA type C. We lost follow up of 2 patients. From the remaining 12 patients, five patients with EA type C were treated with discoid type magnets and one with cylindrical magnets all of which presented severe post-surgical stenosis, refractory to balloon dilatation, and seven (6 type A and 1 type C) were treated with catheter based cylindrical magnets to perform a primary anastomosis.

Results: Anastomotic revision with discoid or cylindrical magnets for EA type C was achieved in 6 days on average and the age at the moment of the treatment ranged between 2 months and 5.9 years. All of them are under full oral diet; five out of the six patients are still in need for one esophageal dilatation per year. Primary Magnamosis with catheter based cylindrical magnets was achieved in an average of 5.2 days. Average age at the moment of placement was 4 months. All patients are under full oral diet with no need for dilatation for the last 24 months.

Conclusion: Esophageal primary Magnamosis and Anastomosis revision was feasible with either catheter based cylindrical or discoid magnets. Long term patency is sustained with one esophageal dilatation per year in 5 of the 6 patients who were treated with discoid or cylindrical magnets for esophageal anastomosis revision. Long term patency was achieved in all patients with primary Magnamosis in which cylindrical magnets were applied. Esophageal patency was stable for the last 24 months.

[P026] Fifty years of OA/TOF care at one institution: More than just survival

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Introduction: The prevalence of oesophageal atresia with trachea-oesophageal fistula (OA/TOF) is estimated to be varying from 1 in 2500 to 1 in 4500 live births, with data from Australia suggesting the incidence to be 2 in 10,000 births. The first repair was performed in 1965 at Sydney Children's Hospital (SCH), with the oldest survivor being 51 years old. Surgical techniques and management have changed greatly over the years with survivals reaching 100% in absence of major associated anomalies or extreme prematurity. The 50 years' operative experience of children with OA/TOF at SCH, will be presented.

Aim: To review the historical data from 1965 to 2014 with respect to the type, associated anomalies, and the evolution of surgical techniques and complications of OA/TOF operated at SCH.

Methods: The study included 373 consecutive infants with OA from 1965 to 2014. The cohorts were divided into three eras based on surgical evolution, management techniques and the establishment of a dedicated TOF clinic. The first era from 1965 to 1984 had 151 infants of which 137 had proximal OA with distal TOF. Amongst 84 male and 67 female infants, forty-one were preterm and 121 weighed more than 2000gms. Nineteen of them had VACTERL association and 42 had other anomalies. There were 68 anastomotic leaks and a mortality rate of 34.4%. The second era consisted of 89 infants with OA/TOF of which 79 were proximal OA with distal TEF. Fifty-seven were preterm and 25 of them weighed less than 2000gms. There were 21 anastomotic leaks with a mortality rate of 16.9%. The third era consisted of 133 infants of which 118 were proximal OA with distal TEF. There were 37 premature infants and 20 were < 2000gms. Seven infants had VACTERL association. There were 3 anastomotic leaks and no mortality. The detailed surgical evolution, analysis of the techniques, complications and outcomes will be pictorially represented as a comparison of the three eras.

Conclusion: The three eras captured the evolution of OA/TOF care over the half century. A significant improvement in survival rates from 34.4% to nil mortality, with progressive changes in operative techniques and postoperative management strategies were noted. The emphasis moved from mere survival in the early years towards a distinct focus on long term outcomes, multidisciplinary care and quality of life in the new millennium.

[P028] Exhaled breath condensate & salivary pepsin as non-invasive markers of reflux in children with oesophageal atresia and tracheo-oesophageal fistula

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Introduction: Children with Oesophageal Atresia and Tracheo-oesophageal Fistula (OA-TOF) may suffer from reflux aspiration secondary to gastroesophageal reflux disease (GORD). There are currently no non-invasive tests for the diagnosis of reflux aspiration in children. We hypothesised that exhaled breath condensate (EBC) and salivary pepsin are potential non-invasive markers of reflux aspiration.

Aim: To measure pepsin in EBC and saliva and to correlate the presence of pepsin with: objective measures of GORD and pulmonary function and validated gastrointestinal and respiratory symptom questionnaire.

Method: EBC and saliva was collected from patients aged between 5 and 18 years attending the OA-TOF clinic. Samples collected were analyzed using two specific monoclonal antibodies against human pepsin A (Peptest). These results were correlated with:

- Parent/child completed gastrointestinal paediatric quality of life questionnaire (PedsQL)
- Parent completed Liverpool respiratory symptom questionnaire (LRSQ)
- Patient symptoms as reported to clinician
- Results of pH-impedance monitoring, esophageal biopsy where performed
- Pulmonary function testing (PFT) results

Results: EBC was collected from 12 OA-TOF patients, 9/12 also provided salivary samples. Pepsin levels in all EBC samples were below the level of detection (>16ng/mL). However, pepsin was detected in 4 (44%) of the salivary samples. The mean levels of pepsin were higher in patients who reported symptoms of regurgitation and/or vomiting [171.8 ng/mL (SD=143.6)] than those who did not (11.0 ng/mL (SD=24.6)) (p=0.04). Pepsin levels also significantly correlated with overall child gastrointestinal PedsQL scores (r²=0.65) (p=0.03). However the presence of salivary pepsin did not significantly correlate with objective measures of pulmonary function and GORD, LRSQ, respiratory symptoms or history of multiple strictures or fundoplication (Table 1).

Conclusion: Salivary pepsin was detected in nearly half (44%) of children with OA/TOF in this study. It's role as a potential non invasive marker of reflux aspiration needs to be validated with further studies.

Table 1: Correlation of Salivary Pepsin (n=9) with symptoms, GORD investigation and Lung Function Test Results

	Positive	Negative	p-value
Chronic cough*			

Yes: n (%)	3 (75)	1 (25)	0.20
No: n (%)	1 (20)	4 (80)	
Hoarse voice*			
Yes: n (%)	2 (50)	2 (50)	1.0
No: n (%)	2 (40)	3 (60)	
Wheeze*			
Yes: n (%)	4 (67)	2 (33)	0.16
No: n (%)	0 (0)	3 (100)	
Fundoplication ever*			
Yes: n (%)	2 (40)	3 (60)	1.0
No: n (%)	2 (50)	2 (50)	
Multiple strictures requiring dilatations*			
Yes: n (%)	3 (50)	3 (50)	1.0
No: n (%)	1 (33)	2 (67)	
Recurrent chest infections*			
Yes: n (%)	4 (57)	3 (43)	0.44
No: n (%)	0 (0)	2 (100)	
Respiratory admissions in the last 12 months*			
Yes: n (%)	1 (50)	1 (50)	1.0
No: n (%)	3 (43)	4 (57)	
Oesophageal Impedance-pH Monitoring (n=8)*			
Abnormal: n (%)	2 (50)	2 (50)	1.0
Normal: n (%)	2 (50)	2 (50)	
Histological findings (n=5)*			
Reflux Oesophagitis: n (%)	4 (100)	0 (1)	0.20
Normal: n (%)	0 (0)	1 (100)	

Spirometry (n=6)*			
Abnormal: n (%)	2 (67)	1 (33)	1.0
Normal: n (%)	1 (33)	2 (67)	
Fractional exhaled nitric oxide (FeNO) (n=7)*			
Abnormal: n (%)	1 (50)	1 (50)	1.0
Normal: n (%)	3 (60)	2 (40)	
Lung clearance index (LCI) (n=4) [§] : median (IQR)	6.85 (6.51-7.18)	6.43 (6.17-6.68)	0.67
LRSQ Overall Score [§] : median (IQR)	52.8 (44.5-52.8)	30.5 (21.3-78.0)	0.89

*Fisher's Exact Test; [§]Mann-Whitney U Test

[P030] Gastric emptying and myoelectrical activity in children with oesophageal atresia-tracheoesophageal fistula.

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Introduction: Abnormalities of gastric function in children with oesophageal atresia-tracheoesophageal fistula (OA-TOF) could potentially contribute to the gastrointestinal morbidity in this cohort.

Aims: To determine gastric myoelectrical activity and emptying in children with OA-TOF and to correlate these results with gastrointestinal symptoms.

Method: In this prospective study, gastric myoelectrical activity and gastric emptying were studied using surface electrogastrography (EGG) and ¹³C-octanoic acid breath test (OBT) respectively. EGG was considered abnormal if there was <75% of waves in 2-4cpm range during post-prandial period and/or post-prandial/fasting power ratio <1. Gastric emptying was defined as normal/abnormal based on comparison with matched control data. A validated PedsQL gastrointestinal symptoms questionnaire was completed by each parent and child >5 years. Correlations between EGG and OBT parameters, PedsQL scores, demographic factors, and results of GORD investigations was done.

Results: Details of the fifteen patients studied are shown in Table 1. Abnormal gastric myoelectrical activity in EGG and delayed gastric emptying in OBT was seen in 66.7% and 46.7% of patients respectively. Mean PedsQL scores as reported by parent and child were 74.88 and 71.55 respectively. There was no significant correlation between abnormal EGG and OBT results. There was also no significant correlation between EGG and OBT results with PedsQL scores. A significant correlation was found between a prior history of strictures and abnormal postprandial/fasting power ratio on EGG (p=0.04). EGG and OBT results did not significantly correlate with demographic factors, GORD investigations or prior fundoplication.

Conclusion: In this pilot study, abnormal gastric myoelectrical activity was seen in majority (66.7%) and delayed gastric emptying was seen in nearly half (46.7%) of children with repaired OA-TOF. There was no significant correlation between objective measures of gastric function as determined by EGG and OBT, nor did these results significantly correlate with subjectively reported gastrointestinal symptoms as determined by PedsQL.

Table 1. Demographic characteristics of patients		
Sex		
Male : Female	4 : 11	
Age (mean years)	6.42	(0.25 – 16)
Birth weight (mean g)	2693.64	(1900 – 4210)
Type of OA		
A	2	13.3%
C	12	80%
D	1	6.7%
Anti-reflux medications use	10	66.7%
Fundoplication	3	37.5%
Gastrostomy (closed)	6	40%
Strictures needing dilatation	9	60%
Endoscopy results		

Performed	14	93.3%
Normal	4	28.6%
Abnormal (reflux oesophagitis)	10	71.4%
Impedance results		
Performed	9	60%
Abnormal	6	66.7%

[P065] Alterations in hyolaryngeal elevation after esophageal anastomosis: a possible mechanism for airway aspiration

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Introduction: Tensioned esophageal anastomosis (EAN) may alter anatomical relation of airway and upper esophagus and may be attributed to airway problems in children with esophageal atresia-tracheoesophageal fistula (EA-TEF).

Aim: To evaluate anatomical alterations and hyolaryngeal elevation (HE) by videofluoroscopy (VFS) in patients with EA-TEF.

Methods: Patients operated for EA-TEF were evaluated for age, sex, type of atresia and time to EAN. VFS evaluations were performed by the same deglutitionist who was blind to the study. Penetration-aspiration scale (PAS \geq 7 is considered as aspiration), distance between upper esophageal sphincter and 2nd cervical vertebrae (UES-C2) and HE were evaluated. The results of EA-TEF patients were compared with healthy children.

Results: Eighteen patients with EA-TEF and 10 healthy controls were included. The median age was 16 months (12-36) in EA-TEF and 18 months (13-51) in controls. Male to female ratio was 5:4 and 4:1 respectively. 66.7% of cases were isolated-EA, 5.65% were EA-proximalTEF and 27.8% were EA-distalTEF. Half of the cases had primary EAN and others underwent delayed esophageal repair. Early oral feeding was also started in 9 patients (50%) whereas others had delayed oral feeding. VFS showed aspiration in 10 cases (55%) and PAS were higher than 7 in 5 cases (27.7%). The median distance between UES-C2 was 3.04 cm (min:2.17-max:3.94) in EA and 4.17 cm (min:3.45-max:6.24 cm) in controls. Median distance for HE was 0.37 cm (min:0.18-max:1.1) in EA and 1.15 (min:0.61-max:1.06 cm) in CG. When measurements of UES-C2 and HE compared with healthy controls, distance between UES-C2 is significantly lower than controls ($p<0.05$). HE is decreased in EA-TEF without any statistical significance. In conclusion, children with EA-TEF had shortened distance between airway and upper-esophagus. HE may be inefficient to protect airway during deglutition. Anatomical alterations after EAN suggest that airway problems may be related with shortened HE in children with EA-TEF.

Poster Abstracts

[P001] A seven-year experience in esophageal atresia with or without tracheoesophageal fistula

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Introduction: The management of newborns with esophageal atresia with or without tracheoesophageal fistula has evolved considerably over the years. Currently an overall survival of 80% to 95% has been reported from developed countries. In the developing countries like our country, several factors contribute to higher mortality rates. We describe our initial experience with management of 80 consecutive cases.

Aim: This study is retrospective analysis of esophageal atresia with or without tracheoesophageal fistula patients in our department during 7 years.

Method/Results: Retrospective analysis of neonates admitted with diagnosis of esophageal atresia with or without tracheoesophageal fistula in our neonatal and paediatric surgery department between April 2009 and April 2016. Medical records were reviewed for age at diagnosis, sex, birth weight, associated anomalies, aspiration pneumonia, treatment, post operative complication and outcome. Total 80 babies (52 male and 28female) were studied. The mean birth weight was 2300grams and mean gestational age was 37weeks. 20% were premature neonates. Age at diagnosis ranged from birth to 9 days. At the time of admission 40% had aspiration pneumonia. Associated anomalies were seen in 60% of the patients. Out of 80 patients 85% survived in that 40% had no associated anomaly. Of the 12 deaths 60% were patients with major anomalies and 40% in those were with more than 2 associated anomalies. Post operative complications were seen in 14% (mainly gastroesophageal reflex, stricture, anastomotic, recurrent TEF-,tracheomalacia), similar to those of developed counties but overall operative mortality 18% was high.

Conclusion: Overall mortality was high. Major congenital anomaly and sepsis was the most frequent cause of death. Factors contributing to mortality includes prematurity, low birth weight, delay in diagnosis and referral, aspiration pneumonia, Further effort must be taken to reduce mortality and morbidity.

[P002] Endoscopic removal of unusual foreign bodies esophagus in operated case of esophageal atresia: a case report

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Introduction: Gastrointestinal foreign bodies(FB) are common and potentially serious cause of morbidity and mortality in children. Impaction of FB in the esophagus occurs most commonly in the pediatric age group between 6 months to 6 years. Impacted meat or other food bolus is the most common esophageal FB in adults, but is relatively rare in children. Predisposing esophageal disease like esophageal strictures, particularly those related to esophageal stricture after surgery (as in our case), are a common predisposing factor to recurrent esophageal foreign body retention in infants and children.

Aim: To report cases unusual foreign body (Tamarind seed) causing obstruction of esophagus due to esophageal stricture secondary to esophageal atresia surgery. Foreign body was successfully removed endoscopically.

Case report: An 18 months male, presented with difficulty in swallowing solid food, vomiting and weight loss. There was history of esophageal atresia surgery in neonatal period, lost follow up after surgery.

On examination vitals were stable. Chest and abdomen examination was within normal limit. A barium swallow revealed narrowing at the junction of upper two-thirds and lower one-third of esophagus. The patient was taken up for upper gastrointestinal endoscopy under anaesthesia. On endoscopy, food particles were seen proximal to the obstruction. Tamarind seed was struck to esophagus, proximal to esophageal stricture. After removal of tamarind seed endoscopic dilatation of esophageal stricture was done. Post-procedure period was uneventful. The patient is doing well in follow up and taking solid and liquid foods.

Conclusion: Flexible esophagoscopy remains the safest method of esophageal foreign body removal in infants and children. High index of suspicion is required when history of foreign body ingestion is not clear for proper diagnosis and management.

[P003] A critical analysis to facilitate rational decisionmaking in the assessment, investigation and early management of oesophageal atresia (oa)

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Introduction: The rarity and anatomical variants in infants born with suspected OA has contributed to a lack of consistency in the way these infants are managed after birth. Paediatric surgeons sometimes struggle to develop a logical approach to their initial assessment and decision-making (clinical and operative) for these cases, leading to significant variation in their early management. This is likely to be a contributing cause of suboptimal outcomes, and often exposes these infants to unnecessary investigations and operations.

Methodology: A critical review of decision-making around the initial assessment and management of infants born with OA, including those with no distal fistula, was conducted.

Results: A clear understanding of the critical information that influences decisions around management enables a more consistent and rational approach to the assessment and investigation of these infants. OA without a distal fistula is a group that is associated with marked variation in early management, for whom the early clinical and operative decisions have significant implications for long-term outcomes.

Conclusions: A logical progression from: 1. Confirmation of atresia, 2. Determination of anatomical type (presence of fistula), 3 recognition of associated abnormalities that might influence management, 4 identification of factors that influence pre-operative investigations and timing of surgery, and 5. a rational process around operative decision-making, should be reflected in fewer procedures and improved outcomes. The key information required for each step is identified.

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Introduction: Children born with EA-TEF often have feeding difficulties which cause considerable worry and stress for parents. These feeding difficulties tend to diminish over time, though in some cases last for many years.

Aim: Our aim was to determine how parental perceptions of their children's mealtime difficulties changes over time, and to compare these perceptions with parental reports of their school-age children's ingestion ability at mealtimes.

Method: Parents of 25 children followed in our EA-TEF clinic from 2012-2016 completed the Montreal Children's Hospital Feeding Scale (MCH-FS) which reflects parental perception of feeding or mealtimes, at 6-10 months, 12-18 months, 2 years, 3 years and 5 years. At the same time, a "Self-Questionnaire of Digestive Symptoms" (SDQS), developed by the Quebec Network for Esophageal Atresia was administered, as part of an ongoing data base for all children in the province with EA-TEF. It includes several questions related to ingestion of food (e.g. difficulty swallowing, food getting stuck, needing to drink during meals, persistent cough, pain and being the last to finish meals) as well as digestive symptoms. The total MCH-FS scores at initial and school age were compared. As well, we compared MCH-FS score with the SDQS ingestion related items at school age.

Conclusion: Completed data is available for fourteen patients (71% type C, 1 Vacterl). All except one were eating table food by school age. The mean scores of the MCH-FS decreased over time. Seventy percent of parents report their child is the last to finish meals, and frequently need to drink during meals. Although parents of children find mealtimes initially difficult and worrisome, despite dysmotility issues associated with EA-TEF during meal times, parental worries lessen significantly by school age.

Prof Ali Raza Brohi

Peoples university of medical & health sciences nawabshah sind pakistan

Introduction: Esophageal atresia is rare congenital anomaly, which is traditionally repaired by open thoracotomy. Nowadays with the advent of minimal invasive neonatal thoracoscopy, high definition imaging, smaller size instruments & improvement in paediatric anesthesia it is possible to correct this problem thoracoscopically as there were lot of problems like musculoskeletal and others related to open thoracotomy.

Material & methods: This is a retrospective study done between May 2015 to June 2016 at peoples university of medical & health sciences Pakistan in which 10 patients included having type C esophageal atresia. Neonates having major cardiac problems & having other GIT association were excluded. The objective of study is to develop final technique for successful thoracoscopic esophageal repair after going through few steps of initial learning curve.

Results : Out of 10 patients first three patients were underwent video assisted technique through single incision of 2 cm to visualize the exact videoscopic anatomy & to do few steps if possible. In next 3 cases three-port technique done with ligation of azygos vein & fistula identification and it's clipping then conversion to open for completion of rest of steps .In last four cases steps were completed with both ends mobilization & intracorporeal single layer interrupted stitching of esophagus. Postoperative period was good in most of cases and 3 patients required ventilator support, few complications encountered in 3 patients like esophageal leak, stenosis & reflux in one patient each.

Conclusion: Thoracoscopic esophageal repair is challenging & advance technique in paediatric minimal invasive surgery, which requires skills & learning undersupervision of experienced surgeon. Learning curve for this procedure can be achieved step-by-step starting from observation to expert person, practice on models, video assisted method & then final pure thoracoscopic technique keeping in mind patients safety & life.

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[P007] Management of anastomotic strictures following repair of oesophageal atresia

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Introduction: Anastomotic stricture is one of the most common complications of oesophageal atresia (OA) repair, affecting 20-50% of patients. Anastomotic leak and gastro-oesophageal reflux have each been implicated as risk factors for stricture formation. Oesophageal stricture is an important cause of morbidity, including the need for often-repeated subsequent intervention. Oesophageal dilatation techniques include bougie, balloon catheter and, in highly select patients, stent placement.

Aim: To describe the rate, risk factors and outcomes for OA patients with oesophageal strictures.

Method: Single-centre retrospective review was performed of neonates undergoing operative repair of OA over a 17-year period (1999 – 2015). Data recorded included OA type, perinatal details, OA operative approach, oesophageal anastomosis outcome, stricture management and survival. Key endpoints were anastomotic leakage, fundoplication, oesophageal dilatation technique, episodes and complications. Stricture was defined as those patients with radiological and/or intra-operative findings of narrowing at the oesophageal anastomosis.

Conclusions: 289 patients were admitted for OA, of which 267/289 (92%) underwent operative repair and survived to discharge (Table 1). VACTERL association was present in 74/267 (27%), anastomotic leak occurred in 48/267 (18%), and fundoplication was performed in 35/267 (13%). Dilatations occurred in 154/267 (58%). The median number of dilatations was 4 (range 1 – 97). Techniques employed included bougie-alone (97/154, 63%), variable combinations of balloon dilatation and bougie (51/154, 33%), balloon-alone (6/154, 4%), and stent insertion (3/154, 2%). Complications following dilatation occurred in 11/154 (7%): 10 oesophageal perforations and 2 clinically significant aspirations. All complications were successfully managed conservatively. Long-gap OA (type A) was a risk factor for having 1 or more oesophageal dilatations (OR 4.0, CI 1.2-14.1, p 0.05).

Table 1

	OA type	Primary repair	Gestation (wks)*	Birth weight (g)*	Patients having dilatation (n)
A	15/267 (6%)	-	36 (34 – 38)	2383 (1780 – 2892)	12/15 (80%)
B	7/267 (3%)	-	37 (31 - 38)	2600 (1436 -3106)	7/7 (100%)

C	228/267 (85%)	215/228 (94%)	38 (28 – 42)	2780 (765 – 4450)	131/228 (58%)
D	6/267 (2%)	6/6 (100%)	38 (37 – 40)	3240 (2135 – 3605)	4/6 (67%)
E	11/267 (4%)	11/11 (100%)	37 (34 – 40)	2990 (2684 – 4512)	0/11 (0%)
Total	267	232 / 267 (87%)	38 (28 – 42)	2730 (765 – 4512)	154 / 267 (58%)

* data expressed as median (range)

Naomi Casimir

This paper is about the experiences and viewpoints of young people who live with a TOF/OA condition. I will draw on the experiences of myself, an 18-year-old girl, two teenage boys, 15 and 19 respectively, and a teenage girl, 13. This will allow me to show a wider range of opinions of the patient experience than just that of myself. I have chosen this age group because they are old enough to talk about subjects I feel should be of interest to doctors, nurses and other hospital staff. The first subject is the memories of hospitalisation and hospitals that we, as TOF/OA patients, have of our experiences from the ages of birth to 10 years. This will provide a base understanding of the way we saw and felt things as children, and will help to explain our reactions as adolescents. The second subject is the experience of transition for adolescent TOF/OA patients, an aspect of the hospital situation which I feel is neglected. By talking about this, I hope to increase awareness that the transition experience doesn't factor in the eventual adult issues of this condition. The third subject of this paper is the teenage experiences from the ages of 10 to 16 outside of the hospital. This will allow me to examine the effects that the TOF/OA condition and hospitalisation have on the private and public lives of a person. Overall, this paper aims to provide new information to doctors and others in the medical field about the ups and downs of actually living with TOF/OA.

[P009] Trends in the management of delayed repaired esophageal atresia infants

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Introduction: The management of long gap esophageal atresia (LGEA) is unique due to the requirement of delayed or staged esophageal repairs.

Aim: To review medical, surgical and nutritional management of LGEA patients at our institution.

Method: Retrospective review of LGEA patients from 2007-2015 who underwent delayed esophageal repair at The Hospital for Sick Children. Age, gap length and weight at time of surgical repair; total length of hospital stay; nutrition support modality at discharge; progression to oral intake; and frequency of esophageal stricture dilatations were reviewed.

Results: Delayed esophageal repair was required in 14 cases (64% male, 86% premature). 72% were pure LGEA, 9% had proximal fistulas and 18% distal fistulas. VACTERL association was confirmed in 36%. Time to surgery was 153.9 ± 43.4 days and weight at repair was 5.4 ± 1.4 kg. Gastric pull-ups occurred in 14%, primary anastomosis (\pm Collis gastrotomy) in 86%. At hospital discharge, 43% had gastrostomies (GT); 21% jejunostomies (JT); and 29% had both GT and JT insitu. 93% were discharged on antacid therapy and 79% on promotility medications. Total length of stay was 213 ± 62 days. At time of data collection, 57% of patients were exclusively fed orally; 14% were taking nothing by mouth. On average 3.4 ± 3.1 esophageal dilatations were required, excluding one patient who required 30+ dilatations.

Conclusion: The significant heterogeneity of these patients makes data pooling and comparison very challenging. From a quality of life perspective, parents want to know how their infant will integrate into society. Based on this review, the majority of LGEA patients at our institution are able to progress to oral feeds by school age but require ongoing support during their transition.

[P010] Outcomes of multi-gestational pregnancies affected by esophageal atresia.

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Introduction: The prognosis for multi-gestational vs. singleton pregnancies affected by esophageal atresia +/- tracheoesophageal fistula (EA/TEF) has not been reported.

Aim: To report the mortality and morbidity outcomes for singleton and multi-gestational EA/TEF patients.

Method: A single institution retrospective review of EA/TEF patients from 1999 to 2013 was performed including patient demographics, gestational age (GA), birth weight (BW), associated anomalies, EA/TEF type and gap length, complications and mortality with IRB approval (#1000032265).

Conclusion: Of 236 (1%) of EA/TEF patients, 22 were from multi-gestational pregnancies; all were twin births (Table 1). Compared to singletons, EA/TEF twins were significantly more premature ($p < 0.01$), had lower BW ($p < 0.01$), were more frequently affected with duodenal atresia ($p < 0.05$) and had higher mortality ($p < 0.05$). In multi-variate analysis, EA/TEF twins with low BW ($< 2000\text{g}$) or GA < 35 weeks ($p < 0.05$) were significantly more likely to require gastrostomy tube for feeds at discharge ($p < 0.05$). Compared to term births, the length of hospital stay for the premature infants was significantly longer ($p < 0.05$). EA/TEF infants of multi-gestational pregnancies have worse outcomes compared to singletons. Further review may provide greater insight into the underlying causes of the morbidities outlined here. Appropriate counselling to parents is needed to inform and assist families of potential morbid outcomes.

Table 1. Demographic and patient characteristics

	Singleton EA/TEF (n = 214)	Multi-gestational EA/TEF (n = 22)	p-value (Chi square tests)
Male/Female	116/98	11/11	>0.05
Gestational age in days (mean, range)	260.97, 182 - 294	235.82, 161 - 276	<0.01
Birth weight in grams (mean, range)	2695.64, 790 - 4390	1766.36, 540 - 2852	<0.01
VACTERL association (n, %)	122, 57.01%	12, 54.55%	>0.05
Duodenal atresia (n, %)	12, 5.61%	4, 18.18%	<0.05
Gastrostomy requirement (n, %)	71, 33.18%	9, 40.91%	<0.01

[P011] Feeding and swallowing disorders in esophageal atresia patients: a 3-year study

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Introduction: Children operated on for esophageal atresia (EA) often experienced feeding and swallowing disorders (FD/SD).

Aim: Aims of the present study were to describe FD/SD in patients treated for long-gap EA (LGEA) vs. non LGEA, evaluating the role of rehabilitative treatment to allow appropriate development of feeding abilities.

Methods: Patients with history of isolated EA and prospectively evaluated by our Speech-Language Pathologists (SLP) between January 2013 and January 2016, were included in the study.

General behavior at mealtime and specific swallowing phases were evaluated. Modified Asha NOMS scale was used to describe feeding levels. Fisher exact test and Mann-Whitney test were used as appropriate; $p < 0.05$ was considered significant.

Results: During the study period 47 patients were treated for EA and prospectively evaluated into our SLP clinics: 19 patients were affected by LGEA, 28 were not. Table summarized main results

Conclusion: SLP's evaluation of children operated on for EA demonstrates disorders in all phases of feeding/swallowing mechanism, with similar distribution between the two groups. LGEA patients had lower feeding level before SLP therapy. Nonetheless, final feeding levels were comparable between the two groups after an appropriate SLP rehabilitation program. Early (since pre-operative period) stimulation of suction and swallowing can contribute to promote and preserve these activities. Parental counselling should focus also on feeding/swallowing disorders, since they may have an impact on the quality of surgical treatment and patients' life.

Behaviour		Overall 47 pts	LGEA 19 pts	Non LGEA 28 pts	p
General	Poor oral intake, n. (%)	32	18	14	0.001
	Mealtime>45min, n. (%)	40	17	23	0.68
	Refusal food, n. (%)	39	16	23	1.0
	Need for distraction, n. (%)	24	7	17	0.14
Oral preparatory phase	Poor management secretion, n. (%)	18	6	12	0.54
	Food loss, n. (%)	21	8	13	1.0
	Oral hypersensitivity, n. (%)	43	18	25	1.0
Oral phase	Bolus stasis, n. (%)	25	8	17	0.24

	Impairment tongue movement, n. (%)	33	13	20	1.0
Pharyngeal & esophageal phase	Cough during meal, n. (%)	13	6	7	0.74
	Retching, n. (%)	31	15	16	0.20
	GER, n. (%)	35	14	21	1.0
	Bubbly voice/crying @meal, n. (%)	11	6	5	0.059
Feeding level	Pre-SLP therapy, mean (IQR)	2 (1-3)	1 (1-1)	2 (2-3)	0.0004
	Post-SLP therapy; mean (IQR)	4 (4-4)	4 (2-4)	4 (4-4)	0.40

[P012] Dysphagia in children with repaired oesophageal atresia

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Introduction: Dysphagia is a common problem in children with repaired oesophageal atresia (OA). The knowledge of dysphagia in this population is limited by the use of various definitions and lack of clarity regarding the swallowing phase in which it occurs. Abnormalities in the oropharyngeal and oesophageal phase have hardly been studied, even though they require different treatment approaches.

Aim: The aims of this study were to assess the prevalence of dysphagia in children with repaired OA, and to identify and differentiate oral and pharyngeal dysphagia based on videofluoroscopic swallow study (VFSS) findings in a limited number of children in this cohort.

Method: Medical records of 111 patients, born between January 1996 and July 2013 and treated at the Radboudumc Amalia Children's Hospital, were retrospectively reviewed. The prevalence of dysphagia was determined by the objective and modified Functional Oral Intake Scale (FOIS) in four age groups. In addition, the first performed VFSS of 13 children was reviewed according to a structural procedure.

Results: The prevalence of dysphagia was 55% (61/111) in age group <1 year. In age group 1-4, 5-11 and 12-18 years the prevalence of dysphagia decreased from 51% (54/106) to 17% (11/64) and 21% (5/24). The percentage of tube feeding dependent patients decreased from 42% (47/111) in age group <1 year to 4% (1/24) in age group 12-18 years. The review of the 13 VFSS's revealed oral dysphagia in 33% and pharyngeal dysphagia in 77% of the children.

Conclusion: This study highlights dysphagia as an important problem in children with repaired OA, with a high prevalence of oropharyngeal dysphagia. The use of an objective dysphagia scale is needed to give tailor made advices for feeding and swallowing in this patient group. In addition, VFSS imaging should be considered when dysphagia is present.

[P013] Sham Feeding in Children with Long Gap Esophageal Atresia: A Controlled Study

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Introduction: Children born with long gap esophageal atresia (LGEA) are prone to develop feeding disorders. After delayed primary anastomosis, most of the patients have impairment of oral skills that prolong nutrition via gastrostomy. Sham feeding has been proposed to prevent feeding aversion and to promote appropriate development of oral skills in these children. Since 2013, a sham feeding protocol is proposed in our centre to all children with LGEA without severe cardiac or respiratory comorbidities, born ≥ 32 weeks and > 1500 g and for whom a delayed anastomosis > 2 months is needed. This study aims to evaluate the nutritional outcomes of these children as compared to an historical control group.

Methods: After parental consent, children meeting inclusion criteria born between 2013 and 2015 were included in a bottle or breast sham feeding program initiated and monitored throughout hospitalization until anastomosis by a nutritionist and an occupational therapist. Nurses ensured adequate functioning and safety of the repleg tube and noted the daily tolerance and side effects. Patients were compared to a control group of term- and weight-matched children born between 2008 and 2012 with LGEA with a delayed anastomosis > 2 months who benefited from standard oral stimulation with a pacifier and occasional drops of milk.

Results: are presented in the table. Data are presented as median (range). There was no complication or side effect reported in the sham feeding group.

Patients characteristics	Oral stimulation Pacifier & drops of milk 2008-2012 (n= 4)	Sham Feeding Bottle / Breast 2013-2015 (n=6)
Age at anastomosis (days)	87 (81-115)	133 (109-170)
Duration of hospitalization (days)	130 (117-141)	195 (156-422)
Age at 100% per os (months)	12.5 (4-48)	9.5 (6-16)
100% per os-# months post-hospitalization	8.2 (0.1-43.3)	2.2 (0.5-6.4)
100% per os-# months post-anastomosis	9.1 (1.1-45.3)	4.9 (1.3-12.4)

Conclusion: A sham feeding program demonstrates positive effects on infants' oral skills allowing earlier weaning of enteral nutrition. Additionally, we believe this program promotes daily child-parent attachment and involvement which permits better skills development during hospitalization and after discharge.

[P014] Clinical outcomes and predictive factors for hospitalisation and mortality rates in Oesophageal Atresia: a ten-year multi-centre Australian experience

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Aim: To characterise childhood hospitalisations and mortality after OA/TOF repair.

Methods: Linked perinatal, neonatal, hospitalisation and death records for all births in News South Wales, Australia, between 2000-2011 were obtained. Infants with a diagnosis of OA/TOF (n=200) during this period were identified and compared to gender and gestational age matched controls (n=400). OA/TOF children were identified using the International Statistical Classification of Diseases and Related Health Problems, Australian Modification (ICD-10-AM). Details regarding neonatal characteristics, associated anomalies, neonatal and subsequent hospital admissions and deaths were collated and analysed.

Results: Neonatal length of stay (LOS) for OA/TOF infants was significantly longer than controls (median 20 vs 4 days, $p < 0.0001$, RR, 95% CI: 2.56, 2.42-2.70), but decreased over the study period. Almost half (46.9%) of OA/TOF children required post-neonatal readmission for lengthy durations, especially up to 2.5 years of age (median 6 days, 0-276 days). The presence of VACTERL association, low 5-minute APGAR scores and increasing age within OA/TOF children was associated with increased length of stays for subsequent hospitalisations (RR 4 days; RR 1.33 days; 1.21 days).

The majority (88%) of readmissions were for gastrointestinal (42%), respiratory (24%) and otolaryngological (22%) conditions. Specifically, the readmissions were for gastrointestinal reflux disease (16%), tracheomalacia (12%) and oesophageal strictures (8%).

10% of OA/TOF children died. The death rate of OA/TOF children was 6.2 times higher than controls ($p < 0.0001$, 95%CI 2.51-15.53, RR 6.24). Factors that were significantly associated with death on multivariate analysis included male gender, rural residence, abnormal APGAR scores and concomitant congenital malformations. OA/TOF children residing in rural areas were 3.7 times more likely to die ($p = 0.018$, 95%-CI-1.26-11.74) than their urban counterparts.

Conclusion: OA/TOF children have a significantly longer neonatal LOS, and are more likely to die than controls. They are also frequently hospitalised in infancy and childhood, especially with gastroenterological issues.

[P015] Prevalence and risk factors of sagittal posture abnormalities in oesophageal atresia

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Introduction: Scoliosis has been described in oesophageal atresia (OA) as a consequence of associated spine malformations and/or thoracotomy. However sagittal posture abnormalities have never been studied in OA.

Aim: to evaluate the prevalence and risk factors of sagittal posture abnormalities (SPA) at the age of 6 years in patients operated at birth for OA.

Method: A prospective cohort of 64 patients with OA was examined by the same rehabilitation doctor at the time of a multidisciplinary visit scheduled at the age of 6 years. Children presenting with scoliosis (n=2) or who missed the consultation (n=17) were excluded. The following factors potentially associated with SPA were analyzed: quality of healing, type of atresia (II, III, IV versus I), delayed surgical reparation of the atresia (<1 week versus >1 week), scapular dyskinesia, prematurity, spinal abnormality, history of laparotomy (open or coelioscopy), delayed psychomotor development, abnormal feeding behavior, lower limb orthopedic abnormality, complications after surgery (including reoperation, stenosis, infection, leaking, pneumothorax or fistula). Chi square test was used for analysis.

Conclusion: Prevalence of SPA was 22.7% [ranges: 16.9%-32.2%]. None of the factors studied were significantly associated with SPA excepted for history of laparotomy (p<0.005). This study shows that SPA is frequent in children operated at birth for OA and should be screened and treated by postural physiotherapy. Initial pain (although not assessed here) and/or abdominal hypotonia following laparotomy could be the underlying mechanisms.

[P016] Is tracheal pepsin a marker of reflux aspiration in children with repaired oesophageal atresia/ tracheo-oesophal fistula?

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Introduction: Children with Oesophageal Atresia and Tracheo-Oesophageal Fistula (OA/TOF) may suffer from reflux aspiration secondary to gastroesophageal reflux disease (GORD). Tracheal pepsin has been reported in literature as a marker of reflux aspiration.

Aim: To measure pepsin in the tracheal aspirates (TA) of children with OA/TOF undergoing an oesophagogastroduodenoscopy (OGD) and to correlate the presence of pepsin with: (a) Gastrointestinal symptoms reported by parents/children (PedsQL), (b) OGD results (c) Gastrointestinal and respiratory symptoms.

Method: Children with OA/TOF attending a multidisciplinary clinic at Sydney Children's Hospital between 2015 to 2016 were recruited. Tracheal aspirates collected during OGD were analyzed using two specific monoclonal antibodies against human pepsin A (Peptest). The parents and children > 5 years old completed a validated gastrointestinal symptom questionnaire (PedsQL). Symptoms were also recorded by the clinician during the clinic visit.

Results: Twenty-eight patients were recruited. Thirteen patients were males (46%), the median age was 5.2 years (IQR, 1.88-6.85), 20 (71%) had a type C OA/TOF. In addition, 31 (91%) patients were on PPI, 5 (18%) had prior fundoplication and 7 (25%) had a gastrostomy in use. Five patients had >1 OGD. Pepsin was detected (>16ng/l) in 42% (14) of the 34 samples. There were no significant differences between the groups with and without detected pepsin regarding: age at measurement, gender, type of OA/TOF, PPI use, prior fundoplication or presence of gastrostomy. Presence of pepsin also did not significantly correlate with PedsQL scores (overall and subsets), reflux oesophagitis on biopsy or presence of gastrointestinal/respiratory symptoms. (Table 1)

Conclusion: Pepsin was detected in nearly half (42%) of children with OA/TOF in this study. However, the presence of pepsin in tracheal aspirate did not significantly correlate with quality of life scores, clinical symptoms or reflux oesophagitis on EGD.

Table 1. Measurement of tracheal pepsin (Peptest)

	Negative (n = 20)	Positive (n =14)	p - value
(a) Quality of life questionnaire : PedsQL ^{1,*}			
Parent : "Overall", median (IQR)	78.1 (69.0-89.9)	86.8 (74.2-94.4)	0.48

Parent : "Heartburn/Reflux", median (IQR)	75 (56.3-93.8)	87.5 (75.1-89.6)	0.49
Parent : "Nausea/Vomiting", median (IQR)	100 (65.5-100)	100 (93.8-100)	0.32
Child : "Overall", median (IQR)	75 (58.2-76.8)	89.6 (80.6-92.2)	0.05
Child : "Heartburn/Reflux", median (IQR)	53.2 (50-71.9)	75 (50-93.8)	0.18
Child : "Nausea/Vomiting", median (IQR)	75 (62.5-100)	75 (87.5-100)	0.39
(b) Histologic findings of reflux esophagitis ^{2,§}			0.25
Normal, n (%)	7 (44)	9 (56)	
Reflux Oesophagitis, n (%)	7 (70)	3 (30)	
(c.1) Gastro-intestinal symptoms reported by clinician ^{3,§}			0.12
No, n (%)	9 (50)	9 (50)	
Yes, n (%)	10 (83)	2 (17)	
(c.2) Respiratory symptoms reported by clinician ^{4,§}			0.37
No, n (%)	14 (58)	10 (42)	
Yes, n (%)	5 (83)	1 (17)	

*Mann-Whitney test, § : Fisher's exact test

1: Scale of 0-100, with higher score indicating a better quality of life

2: Patients with active eosinophilic oesophagitis were excluded from the analysis.

3: Regurgitation, vomiting, dental enamel erosion, chest pain, heartburn

4: Cough, recurrent chest infections, hoarse voice, sore throat

[P017] Combined treatment in refractory severe pediatric esophageal anastomotic strictures

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Introduction: Mitomycin C (MMC) is an antineoplastic and antiproliferative agent. Dexamethasone has anti-inflammatory action, with not established role after esophageal dilations.

Aim: This study evaluates the efficacy of combined treatment in refractory severe esophageal anastomotic strictures (RSEAS).

Methods: From 2013 to 2016, 5 children with RSEAS were enrolled in a prospective open trial. Age, type of esophageal atresia, dysphagia score (DS), previous endoscopic dilations, stent placement and surgery pre-post treatment were recorded. All patients underwent sessions of combined treatment (within interval from 2 to 4 weeks), consisted of esophageal dilations with Savary-Gilliard bougie, topic MMC (0.1 mg/dl; 5 minutes) applied at dilated stricture and intravenous/oral dexamethasone (0,5 mg/kg bid, 3 days). All patients received proton pump inhibitors. Outcomes were to measure DS (0 no dysphagia-4 severe dysphagia) and dilation requirement.

Results: 5 children (mean age 25 months, range 13-40 months) with RSEAS, before combined treatment, underwent endoscopic dilations (mean 7; range 2-10) and Dynamic Stent[®] placement. Because of persistent dysphagia (DS 4), all patients received combined treatment (mean 6,4; range 3-10). After treatment, 3 patients improved the DS (from 4 to 1) and required a mean of additional 7 dilatations (range 4-11). In 2 patients, treatment failed; they had a second esophageal Dynamic Stent[®], and one of them also an indwelling balloon treatment. Mean follow-up was 4 years (range 13 months - 5 years).

Conclusions: Combined treatment with dilations, MMC and Dexamethasone in our experience shows dysphagia improvement, reducing number of dilations in RSEAS, but not resolution of the stricture.

[P018] Prevalence and natural history of scoliosis and associated congenital vertebral anomalies in patients operated for esophageal atresia

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Introduction: Scoliosis reported after esophageal atresia (EA) repair may be secondary to thoracotomy or due to associated congenital vertebral anomaly. However, the prevalence and natural history of scoliosis in patients with EA are not clear.

Aim: To review the prevalence and natural history of thoracogenic scoliosis and associated congenital vertebral anomalies in 190 children who underwent EA repair.

Method: A retrospective review of X-ray imaging and chart of 190 patients operated and followed at the Ste-Justine EA Clinic between 1990 and 2015. We excluded patients who only had radiological images younger than 5-year-old due to insufficient follow-up for scoliosis believed to be of thoracogenic origin.

Results: Radiological examination for presence of scoliosis or associated spine congenital anomalies was done in 108 patients (62 boys) aged 5 to 19 years. Scoliosis was found in 53 patients (49%) of which 46 (87%) were in the thoracic region and 33 (72%) with a right thoracic curve. After a mean follow-up of 7 years (range 2-14), 4 patients (7 %) were operated for scoliosis after failure of bracing to prevent progression. Right-sided thoracotomy (RST) was the identifiable risk factor for scoliosis development; 50 patients (94%) with scoliosis had their EA repaired through RST versus 24 out of the 55 patients without scoliosis (43%, OR 21.5 95% CI 6–77, P<0.001). Congenital vertebral anomalies were found in 8 patients (7%). After a mean follow-up of 7.8 years (range 3-13), no patient with associated congenital anomalies progressed to the indication of surgery.

Conclusion: Scoliosis affects one of every two patients operated for EA; the course is usually benign but could progress to the indication of surgery and need extended follow-up. Associated vertebral congenital anomalies is another reason to follow these patient's despite in the present study these anomalies did not increase the need for surgery.

[P019] Does my baby have a feeding problem? It depends on who you ask.

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Introduction: Feeding and the incidence of feeding problems in babies with oesophageal atresia (OA) is well described in the literature. However, there is little reference to the bio-psycho-social factors that influence feeding outcomes in this vulnerable population. We present a prospective study conducted at The Royal Children's Hospital in which a range of factors having the potential to impact on feeding development were analysed.

Aim: To describe the interactions between biomedical, psychological and social factors associated with the development of oral feeding in babies with OA.

Method: This study was a prospective, longitudinal study of 30 infants with OA and their parents. Subjects were recruited whilst inpatients on the NICU. Data were collected at four time points (4 weeks, 3 months, 6 months and 9 months). The study used mixed methodology with a qualitative component using phenomenological analysis of semi-structured interviews to explore parental experience of feeding a baby with OA. Feeding outcome measures included feeding success, feeding efficiency and growth. Qualitative analysis identified key themes.

Results: 30 babies and their families were recruited. Mean GA at birth was 37 weeks and mean birthweight was 2681grams. Mean inpatient length of stay was 55 days. Mean time to first suck feeds was 39.42 days. There was a weak negative correlation between time to first suck feeds and weight at 9 months. A range of factors operate alongside the development of feeding and these infants were slow to feed, which had an impact on their growth. Qualitative analysis revealed a number of themes, including parental stress, satisfaction with support available and negative and positive accounts of feeding.

Conclusion: Many factors interact to result in delayed establishment of suck feeds in infants with OA. Families need multi-disciplinary support if outcomes for infants are to be optimised.

[P020] Thoracoscopy and Thoracotomy: Choice of Surgical Approach and Outcomes in Oesophageal Atresia-Tracheoesophageal Fistula

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Introduction: Thoracotomy continues to be considered the gold standard surgical approach for Oesophageal Atresia-Tracheoesophageal Fistula (OA-TOF) despite the increasing popularity of thoracoscopy. As thoracoscopy has gained wider utilization over the last decade reports emerged regarding surgical complications not previously associated with OA-TOF repair including hypercapnia and vocal cord paralysis. This study aims to compare outcomes and complications between thoracotomy vs thoracoscopic repair of OA-TOF.

Methods: A Retrospective chart review was conducted. Infants undergoing surgical repair of OA-TOF from the years 2005 - 2016 for whom data were available were included. Exclusion criteria were death before surgical repair and tracheoesophageal fistula without oesophageal atresia. Primary data points were survival, anastomotic leak/stricture, vocal cord paralysis, and intraoperative hypercapnia. Statistical analysis was performed using chi-squared analysis and student's T test.

Results: 55 patients were included, 38 underwent thoracotomy, 17 underwent thoracoscopy. Survival did not differ significantly between groups at 93%. Anastomotic leak and stricture did not differ between groups at 14% and 28% respectively. The incidence of hypercapnia intraoperatively was not significantly different between groups: maximum PaCO₂ was 58mm Hg (+/- 48) in the thoracoscopy vs 69 (+/- 19) in thoracotomy. Vocal cord paresis (transient or permanent) was documented in 4 infants, 2 had thoracoscopy repairs and 2 thoracotomy. Mean operative time was longer for thoracoscopy vs thoracotomy: 148 min (+/- 62 min) vs 109 minutes (+/- 35 min) (p<0.05). Thoracotomy patients had more preoperative comorbidities (p<0.05) and more preoperative cardiopulmonary instability events (p<0.05) than thoracoscopy patients.

Conclusion: Thoracoscopy is equivalent to thoracotomy in terms of survival and anastomotic leak/stricture. Importantly, vocal cord paralysis and intraoperative hypercapnia rates were not higher in our series of thoracoscopic repairs as compared to thoracotomy. These data improve understanding of outcomes can be applied develop strategies for selecting the best surgical approach for each infant.

[P021] Post operative non-invasive ventilation and complications in oesophageal atresia-tracheoesophageal fistula

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Introduction: Advancements in critical care have been instrumental in the observed improvement in survival and complication rates in Oesophageal atresia-Tracheo-Oesophageal Fistula (OA-TOF). Nearly all neonates with OA-TOF undergo assisted ventilation at some point in the course of their post operative care. Post operative assisted ventilation strategies are heterogenous from one center to another and to date limited data are available to guide current practices of assisted ventilation in cases of OA-TOF. The present study aims to examine the impact of post operative assisted ventilation strategies on clinically relevant outcomes in a retrospective series of OA-TOF patients.

Methods: A single center retrospective chart review was conducted including all neonates born with OA-TOF 1986-2016 for whom complete ventilatory data were available. Exclusion criteria: death prior to surgical repair, presence of pulmonary disease, cardiac malformation resulting in severe pulmonary hypertension. Primary data points evaluated were: Post-operative ventilation strategy, Survival, Anastomotic Leakage, Stricture, Pneumothorax and Mediastinitis. Statistical significance was determined using Chi-square test for p less than 0.05.

Results: 28 infants were included. Assisted ventilation was used in all infants. 1(3%) of infants required HFOV postoperatively, 28 infants required conventional ventilation for some period of time and 13 were bridged with postextubation non-invasive ventilation. Survival was 27 (97%), incidence of anastomotic leak was 7 (25%), Stricture 7 (25%), Pneumothorax 4 (14%) and Mediastinitis 2 (7%). The relationship between non-invasive ventilation strategy and complications is expressed in the table below.

Conclusions: The most important finding of the present study is that High Flow Nasal Cannula assisted ventilation is associated with a significantly higher rate of anastomotic leakage and stricture following repair of OA-TOF. We hope to use these findings to develop guidelines for ventilation strategies in the care of babies born with OA-TEF.

Table 1. Complications Associated with Non-Invasive Ventilation in OA-TOF

Ventilation Strategy	Leak	Stricture	Mediastinitis	Pneumothorax
	n (%)			x
CPAP	2 (33) NS	0 (0) NS	1 (17) NS	0 (0) NS
NIPPV	0 (0)	0(0) NS	0 (0) *	0(0) NS
HFNC	4 (67) * (p = 0.01)	2 (33) NS	1(17) * (p<0.001)	1(17) NS

CPAP: Continuous Positive Pressure Ventilation; NIPPV: Non-invasive positive pressure ventilation; HFNC: High Flow Nasal Cannula. NS = not statistically significant. * = statistically significant for p <0.05 via Chi-square Test.

[P022] Transnasal Esophagoscopy (TNE) is Successful for Monitoring and Evaluating Esophageal Mucosa at TEF/EA Repair Site in a 10-year Pediatric Patient with Dysphagia

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Introduction: A child with esophageal atresia and/or tracheal esophageal fistula may be subjected to multiple radiographic studies or endoscopic procedures to evaluate the post-operative site in their lifetime due to complaints of dysphagia and risk of esophagitis. These studies offer increased risk of radiation and anesthesia exposure to children who have rapidly developing gastroenterological and neurological systems. Our center recently introduced the use of unsedated transnasal esophagoscopy (TNE) for monitoring eosinophilic esophagitis in pediatrics and has now been performed on children as young as 5 years of age¹. We hypothesized that this technique would be successful in a patient who needed esophageal mucosal monitoring due to dysphagia and whose family requested to not use fluoroscopy or anesthesia to assess his esophagus.

Aim: To evaluate the success and safety of using TNE for monitoring the esophageal repair site in a 10-year child with complaints of dysphagia.

Method: A 10-year old otherwise healthy child with known history of tracheal esophageal fistula with long gap esophageal atresia (TEF/EA) underwent unsedated TNE as previously described at our center. Adverse events, visual, and histological results were recorded.

Results: Procedure was tolerated by the patient with no significant adverse events occurring. Sore throat was noted. Total time in the office was approximately 1 hour with child returning to sports practice after procedure. Anastomosis site was found to be visually patent without shelf or narrowing. Biopsy found evidence of ongoing mild esophagitis <15 eos/hpf both distally and at the anastomosis site.

Conclusion: TNE with biopsies appears to be a safe and effective alternative to sedated esophagoscopy or a fluoroscopic esophagram for monitoring the anastomosis site and esophageal mucosa in TEF/EA, further study is needed.

[P023] Adults operated on for esophageal atresia – the need for a follow-up program regardless of symptoms

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Introduction: Gastro-esophageal reflux (GER), pulmonary symptoms and functional impact (esophageal and pulmonary) are common among adults operated for esophageal atresia (EA). However, there are no guide-lines concerning follow-up.

Aim: We wanted to investigate if symptoms could point out those in need of further follow-up in adulthood.

Method: Between 1968-1983 110 patients were operated for EA in Gothenburg, Sweden. 79/80 survivors were localized and 69/79 with the most common form of EA, proximal atresia with distal tracheo-esophageal fistula (TEF), were chosen for further investigations. In 2011 twenty-eight patients accepted to undergo additional studies concerning their esophageal and pulmonary function. A drop-out analysis showed no differences between participators and non-participators. Fifteen patients completed both the esophageal investigations, including gastroscopy and pH multichannel intraluminal impedance (pH-MII), as well as the pulmonary function tests, including multiple breath wash-out, spirometry and whole-body plethysmography.

Seven women and 8 men participated (mean age 35 years). Ten had respiratory symptoms; 8/10 had long-standing cough, 6/10 had wheezing during the last 12 months and 4 had a doctor-diagnosed asthma. Seven patients complained of GER; 5/7 had heartburn and 5/7 had regurgitation. Eleven patients had a pathological pulmonary function test while 11/15 had pathological pH-MII findings. Six patients had histological esophagitis of whom two had intestinal metaplasia.

No correlation was seen between symptoms on one hand and pulmonary function test, pH-MII or histology on the other. However, the number of acid and weakly acidic reflux and also the reflux index (RI) showed a strong correlation to reduced FEV1 (not number of weakly acidic though) and FEV1/FVC ($p < 0.05$ for all).

Conclusion: Even though pH-MII verified GER seems to affect pulmonary function, and respiratory and GER symptoms are common, there is no correlation between function tests and these symptoms. This stresses the need for a continued follow-up in adulthood regardless of symptoms.

[P024] Review of tracheomalacia in tof/oa patients from an Australian tertiary centre

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Introduction: Tracheoesophageal fistula (TOF) is a common congenital anomaly occurring in conjunction with oesophageal atresia (OA) in approximately 1:3500 live births¹. The reported association with tracheomalacia (TM) is variable, a large series reporting a 15% association, of which 40% required surgical intervention². Other studies have shown an association as high as 86%.³

Aim: To review the clinical characteristics, investigations and management of TM in the TOF-OA population at an Australian tertiary referral centre

Methods: A retrospective case note review was performed of all patients who underwent primary or delayed TOF and/or OA repair at the Sydney Children's Hospital between May 2010 and June 2016. All patients underwent a laryngo-bronchoscopy at the time of the repair to evaluate the site of the abnormality and to assess for other airway pathology. Patients with confirmed tracheomalacia on endoscopy were specifically identified and data was collected on this group regarding demographics, clinical features, operative findings, adjunctive investigations and further management.

Results: A total of 32 patients were identified with TOF-OA (Type A - 12.5%, Type B - 3.1%, Type C - 81.2%, Type H - 3.1%). Exactly 50% were male. TM was reported in 18/32 patients (56.2%) graded as mild, moderate and severe in 27.7%, 44.4%, and 27.7% respectively. The commonest site of malacia was the distal trachea in 58% of cases. Laryngeal clefts were identified in 3 patients. Airway noise, cough and work of breathing were frequent presentations. 6/18 (33.3%) patients with TM were further investigated with CT pulmonary angiogram to exclude a vascular anomaly. Only 2/18 (11.2%) proceeded to aortopexy.

Conclusion: TM is a common airway anomaly associated with TOF-OA in this cohort. It was frequently graded as moderate and in the distal trachea, corresponding to the TOF site. Many patients had significant symptoms however only a small percentage required further investigation or intervention.

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[P025] The handling of gastro-esophageal reflux following congenital esophageal atresia (ea) – a patient survey

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Introduction: Patients with EA often suffer from gastroesophageal reflux disease (GERD).

Aim: We aimed to study the patients' understanding for GERD pathophysiology, individual symptoms, adherence to follow-up, health-care satisfaction and need for professional information.

Method: An international web-based survey was conducted between December 2014 and June 2015. 101 responses were evaluated and the survey was augmented by personal interviews with 25 patients and 5 consultants.

Results: The participants (33%: age <9 (answered by parents), 67%: age 9-57years, 49% male) were predominantly recruited in USA 22%, Switzerland 20%, England 17%, Germany 16% and Australia 7%. The majority (80/101) of participants understood GERD pathophysiology. Indeed, 96 of 101 patients reported a minimum of one GERD symptom and more than 50% suffered from multiple symptoms. Acidic burp is the most frequent symptom (n=59), followed by heartburn and iron deficiency. 60 (N=67) patients ever had an endoscopy, 27 ph-metry, 37 x-ray. Spicy food and stress are the most frequent causes for a reflux episode. Sparkling soda-waters, alcohol, chocolate and meat are the most frequent foods that trigger reflux. 18 patients reported having no follow-up care at all, while only 11 patients have yearly follow-up care. 67 wished to consult a GERD/EA specialist. In total 70 patients felt under-informed about GERD.

Conclusion: Many EA patients lack regular consultation with a GERD specialist and fear that their after-care may be negatively impacted by doctors who lack experience with rare conditions like EA. Study participants report a need for professional information about GERD, also to improve patient-doctor-communication. Following these results a comprehensible, educational brochure about GERD for OA-patients has been published.

Key Words: Esophageal Atresia, Gastro-esophageal Reflux Disease (GERD), Patient-Survey, after-care

[P027] Is the Foker procedure for long gap oesophageal atresia superior to traditional techniques?

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Introduction: Foker procedure (FP) has recently been used for repair of long gap oesophageal atresia (LGOA). We report our experience using this procedure.

Aim: The aim of this study was to compare the outcomes in LGOA following Foker procedure with traditional techniques.

Methods: A retrospective chart review was performed of all LGOA children repaired using the Foker procedure at Sydney Children's Hospital with those repaired using traditional techniques.

Results: Nine children have undergone the Foker procedure (FP) at our hospital since 2007. In the FP group the median age at follow up (F/U) 5.3years (2.2-9.1), and there were 5 (55.5%) males, In the Non Foker procedure group of 9 children there were 3 males and median follow up was 7.1years (3.1-12.9). There was no significant difference between both groups in these demographics. All children in both groups have kept their native oesophagus. Other demographic details including associated co morbidities, type of OA, time taken to join, and short and long term clinical outcome measures including post operative complications, number on oral feeds at discharge, number on full oral feeds at F/U, gastrostomy in use at F/U, strictures needing dilatation/patient, anti reflux medication (PPI) use, reflux and eosinophilic esophagitis on biopsy, need for fundoplication, cyanotic spells, recurrent fistula, dysphagia, reflux symptoms, height and weight z scores at F/U are shown in Table 1. Although the Foker group had significantly more pure atresia patients and associated comorbidities, there were no significant differences in clinical outcome measures between both groups apart from the significantly higher incidence of strictures needing dilation in the Foker group.

Conclusion: Clinical outcome measures in children with LGOA repaired using the Foker technique were similar to those repaired using traditional techniques apart from the significantly higher incidence of strictures needing dilation in the Foker group.

Table 1: Comparison of Foker Group with Non Foker Group

Demographic And Outcome Variables	Foker Procedure (FP)	Non Foker Procedure (NFP)	P Value
Type of OA/TOF	7 Type A ^a 1 Type B 1 Type C	1 Type A ^a 7 Type C 1 Type D	P ^a = 0.01*
Associated Comorbidity	8 (89%)	2 (22%)	P = 0.00*
Median age at joining	59 days (15-117)	2 (1-330)	P = NS
Post operative leak	3 (33%)	2 (22%)	P = NS
Number on oral feeds at discharge	7 (78%)	2 (22%)	P = 0.05
Full oral feeds at F/U	8 (89%)	8 (89%)	P = NS

Gastrostomy in use at F/U	1 (11%)	1 (11%)	P = NS
Strictures needing dilation/patient	6 (2-15)	2 (0-8)	P = 0.02*
Reflux Symptoms at F/U	8 (89%)	8 (89%)	P = NS
PPI Use at F/U	7 (78%)	8 (89%)	P = NS
Reflux Oesophagitis	1/5 (20%)	1/9 (11%)	P = 0.02*
Eosinophilic Esophagitis	3/5 (60%)	3/9 (33%)	P = NS
Fundoplication	2 (22%)	3 (33%)	P = NS
Cyanotic Spells	2 (22%)	1 (11%)	P = NS
Recurrent Fistula	0	0	
Dysphagia at F/U	4 (44%)	5 (55%)	P = NS
Height "z" score at F/U	-1.245 (-2.46-0.88)	-1.27 (-1.92-1.26)	P = NS
Weight "z" score at F/U	-1.5 (-1.77- -0.34)	-1.64 (-2.81-0.84)	P = NS

[P029] Is fundoplication required after the foker procedure for long gap oesophageal atresia?

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Introduction: Fundoplication has been almost universally performed in children treated with the Foker procedure (FP) for long gap oesophageal atresia. We report our experience of not routinely performing fundoplication in children treated with this technique.

Aim: The aim of this study is to demonstrate that not all children require early fundoplication following the Foker procedure.

Method: A retrospective chart review was performed of all children treated with the Foker procedure at our institution.

Results: Nine children have undergone the FP at our hospital since 2007. Follow-up data was available for six children. Seven children had traction sutures placed thoracoscopically and two also had their definitive anastomoses performed thoracoscopically. The median time between FP and definitive anastomosis was 22 days (range 14-114). All nine children kept their native oesophagus. There were three anastomotic leaks all treated non-operatively. All children required dilatation of anastomotic strictures with a mean number of 7 dilatations (range 2-15). All have been treated with proton pump inhibitors (PPI). Three children had eosinophilic oesophagitis and one had Barrett's oesophagus; all treated pharmacologically. Only two children in this series have undergone fundoplication, which were done for symptomatic, erosive oesophagitis despite optimal PPI therapy. Eight children have had their gastrostomies closed and are fed orally.

Conclusion: The question of immediate versus delayed fundoplication in long gap oesophageal atresia managed with the Foker procedure remains unanswered. Our series demonstrates that it is possible to achieve good long-term outcomes when the operation is reserved only for complicated gastro-oesophageal reflux disease resistant to maximal medical therapy.

[P031] Prevalence and risk factors of sagittal posture abnormalities in oesophageal atresia

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Introduction: Scoliosis has been described in oesophageal atresia (OA) as a consequence of associated spine malformations and/or thoracotomy. However sagittal posture abnormalities have never been studied in OA.

Aim: to evaluate the prevalence and risk factors of sagittal posture abnormalities (SPA) at the age of 6 years in patients operated at birth for OA.

Method: A prospective cohort of 64 patients with OA was examined by the same rehabilitation doctor at the time of a multidisciplinary visit scheduled at the age of 6 years. Children presenting with scoliosis (n=2) or who missed the consultation (n=17) were excluded. The following factors potentially associated with SPA were analyzed: quality of healing, type of atresia (II, III, IV versus I), delayed surgical reparation of the atresia (<1 week versus >1 week), scapular dyskinesia, prematurity, spinal abnormality, history of laparotomy (open or coelioscopy), delayed psychomotor development, abnormal feeding behavior, lower limb orthopedic abnormality, complications after surgery (including reoperation, stenosis, infection, leaking, pneumothorax or fistula). Chi square test was used for analysis.

Conclusion: Prevalence of SPA was 22.7% [ranges: 16.9%-32.2%]. None of the factors studied were significantly associated with SPA excepted for history of laparotomy (p<0.005). This study shows that SPA is frequent in children operated at birth for OA and should be screened and treated by postural physiotherapy. Initial pain (although not assessed here) and/or abdominal hypotonia following laparotomy could be the underlying mechanisms.

[P032] Evaluation of motor function in children with esophageal atresia.

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Introduction: Short-term motor outcome in esophageal atresia (EA) survivors are scarce and the results are ambiguous and based on small samples.

Aim: The aims of the study are to measure motor skills in EA children at 12 and 24 months of age and to examine if motor skills change from 12 to 24 months of age.

Method: 21 EA children (GA 36.8, range 30-41) born 2012- 2013 were included. Children with genetic syndromes were excluded. Eleven were premature. Muscle preserving right thoracotomy in all. Cardiac anomaly in 9 (43%) and VACTERL association in 5 (24%). At one year of age, the children were examined with the Alberta Infant Motor Scale, and at two years of age, the Peabody Developmental Motor Scales was used. Both tests are validated and well correlated.

Conclusion: At 12.5 months (range 11-16), 4 (19%) children (2 premature) were classified with motor delay, 2 (10%) at risk of motor delay (1 premature) and 15 (71%) had motor skills within normal range. At 25 months, (range 23-32), 2 (10%) were classified with motor delay, 2 (10%) at risk and 17 (81%) within normal range. Among 4 children with motor delay at one year, one was categorized as normal at two years, one infant was still delayed and 2 identified at risk. The two classified at risk at one year, had normal motor skills at two years. One child changed from normal to definite motor delay. Among the 6 patients with delay at one year, 5 had physical therapy at home and all those patients improved after one year. 29 % had or were at risk of motor delay at 12 months. From one to two years the overall motor skill score changed in 6/21 of the patients, in 5 of them to the better following home physical therapy.

[P033] Admissions and outcomes of infants with oesophageal atresia/tracheo-oesophageal fistula in new south wales and the australian capital territory from 1992 to 2010

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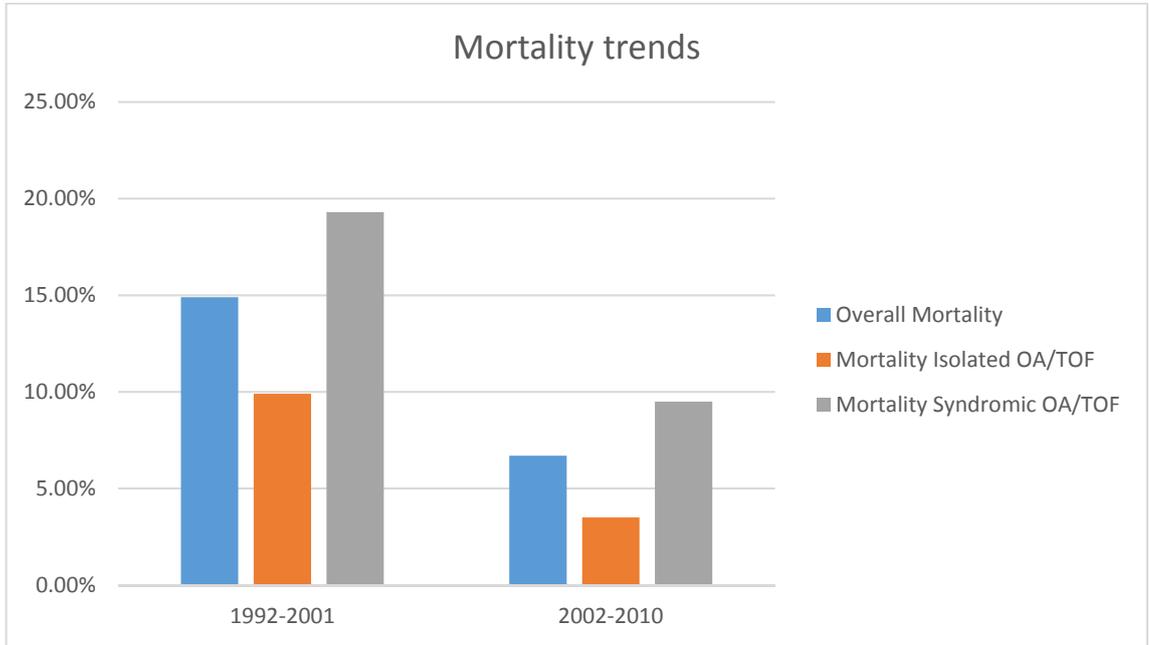
⁶ *NSW Pregnancy and newborn Services Network*

Introduction: In recent years, significant advances have been made in the early diagnosis of fetal malformations and management of infants with congenital conditions.

Aim: To report on the trends in antenatal diagnosis, admissions and outcomes of liveborn infants with OA/TOF in New South Wales (NSW) and the Australian Capital Territory (ACT).

Method: A regional retrospective cohort study using prospectively collected (NICUS) database on all live born infants with OA/TOF admitted to any of the 10 Newborn Intensive Care Units (NICUs) between 1992 and 2010. The cohort was divided into 2 epochs. Epoch 1 included infants born 1992-2001 and Epoch 2 included infants born 2002-2010.

Conclusion: 334 cases were admitted (1.9 per 10,000 livebirths). Antenatal diagnosis increased from 5% in 1992 to 35% in 2010. OA with distal TOF was the most common variety (90.1%). Median birth weight and gestation were 2647g and 37.5 weeks. 38% were preterm (<37 weeks) and 31% were growth restricted (<10th percentile weight for gestation). Approximately half (156, 47%) were isolated OA/TOF while 178 (53%) were associated with other anomalies (syndromic OA/TOF), most common being cardiac (ASD, VSD and PDA) and renal anomalies. VACTERL association was found in 5.6% cases and chromosomal anomalies were detected in 4.5% cases. Overall mortality rate decreased significantly from 14.9% in epoch 1 to 6.7% in epoch 2 (p 0.02). There was a non-significant decrease in mortality in infants with isolated (3.5% vs 9.9%, p 0.2) and syndromic OA/TOF in Epoch 2 (9.5% vs 19.3%, p 0.1). Preterm infants carried significantly higher mortality compared to term infants (48.7% in 23-31 wk GA; 13.5% in 32-36 wk GA; 2.9% in ≥37 wk). In multivariate analysis, only lower gestation (OR 1.29, 95% CI 1.05, 1.60) and presence of associated anomalies (OR 3.48, 95% CI 1.34, 9.01) were significant predictors of increased mortality.



[P034] Complex care of esophageal atresia +/- tracheoesophageal fistula (ea/tef) patients - a population-based healthcare utilization analysis.

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Introduction: Children with esophageal atresia +/- tracheoesophageal fistula (EA/TEF) can experience significant morbidities following surgical repair. Additionally, up to half of the EA/TEF patients have other anomalies (e.g. VACTERL association, duodenal atresia and Trisomy 21). However, the burden of care required for EA/TEF patients has not yet been studied.

Aim: The objective of this study is to determine the population-based incidence of EA/TEF in the province of Ontario and examine overall healthcare utilization of EA/TEF patients.

Methods: Inpatient and outpatient health services data for patients born with EA/TEF in ON from 1998 to 2011 were obtained from provincial administrative datasets at the Institute for Clinical Evaluative Sciences. Matching data for non-EA/TEF control patients (n=6885) selected randomly from ON births during the study period were also obtained.

Results: There were 345 EA/TEF births in ON (1 in 5,446 live births) during the study period: 196 (56.8%) were males, and 296 (85.8%) resided in urban centres. Patients were evenly distributed across income quintiles (measure of socioeconomic status) and geographic regions. In total, 197 (57%) had coexisting cardiac defects. Overall, 21 (6.1%) died with 16 (76.2%) deaths occurring during the initial hospitalization. During the first year of life, EA/TEF patients averaged 2.4 hospital admissions (control 1.1), and spent 60 days in hospital (control 4), with 48 days for the initial hospital stay (control 3). Outpatient utilization by patients for pediatricians, medical specialists and surgeons was substantially greater compared to controls (Table-1). Post discharge, 52 home care nursing services were delivered over 428 hours per EA/TEF patient (control 1 service over 7 hours).

Conclusion: EA/TEF is a rare birth defect. Patients with EA/TEF require substantial hospital-based and community care. These data will be useful in planning and providing multi-disciplinary, coordinated care for EA/TEF patients.

Table 1 – Fold-Difference in the Average Frequency of Outpatient Consultations & Assessments Utilized by EA/TEF Patients Compared to Controls

Outpatient Services	Average Frequency per EAT/TEF Patient	Average Frequency Per Control Patient	Fold-Difference
Primary Care			
Pediatrics	21.25	3.89	5.46
Emergency Medicine	7.92	4.59	1.73
Family Practice	29.36	26.94	1.09
Medical Specialties			
Genetics	0.43	0.018	23.89

Cardiology	0.68	0.061	11.15
Respirology	0.35	0.030	11.67
Gastroenterology	0.28	0.032	8.75
Surgical Specialties			
General Surgery	7.30	0.12	60.83
Neurosurgery	0.51	0.023	22.17
Thoracic Surgery	0.0058	0.00058	10
Urology	0.68	0.13	5.23
Plastic Surgery	0.40	0.095	4.21

[P035] Assessment of Ventilation Inhomogeneity in Children with Oesophageal Atresia – Tracheo-Oesophageal Fistula

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Introduction/Aim: Children with oesophageal atresia-tracheo-oesophageal fistula (OA-TOF) experience lifelong respiratory morbidity. Spirometry is used to evaluate the presence, severity and progression of lung disease. However spirometry mainly assesses large airway function. The degree to which the peripheral airways are affected, particularly as a consequence of recurrent infections, atelectasis and aspiration, in the years following surgical repair, has not been clearly determined. We aimed to evaluate the clinical utility of multiple breath inert gas washout (MBW), in particular the Lung Clearance Index, to detect peripheral airway disease in children with OA-TOF. We hypothesised that MBW testing would be more sensitive than spirometry, to identify early lung function impairment.

Method: Prospective cross-sectional study evaluating children attending a multidisciplinary OA-TOF clinic. Spirometry was reported according to ATS/ERS criteria. Multiple breath nitrogen washout (MBW) was performed using a commercial device (Exhalyzer D, Eco Medics, Switzerland). Results were compared to published normative values.

Results: 10 children were assessed; 5 (50%) had abnormal spirometry (1 mild obstructive, 2 mild restrictive and 2 moderate mixed impairment). LCI was elevated in 5 children, four with abnormal spirometry. One child had a mildly restrictive defect but normal LCI. Abnormal LCI z-scores ranged from 2.1 to 10. There was a moderate negative correlation between LCI and FEV₁, ($r = -0.51$).

	Children with OA/TOF (n =10)	Healthy reference population (n=44)	P value
Mean Age (yrs)	11.59 (7.89-15.62)	13 (7 - 19)	<0.05
FEV ₁ % pred	76.70 ± 20.16	101 ± 10	<0.01
LCI	7.44 ± 1.30	6.54 ± 0.28	<0.01
Scond	0.028 ± 0.01	0.021 ± 0.004	Ns
Sacin	0.0967 ± 0.05	0.051 ± 0.012	Ns

Conclusion: We found abnormal peripheral airway function in a substantial proportion of children with OA-TOF assessed by MBW. Both spirometry and MBW testing are simple, useful, repeatable lung function assessments which together may provide a comprehensive evaluation of large and small airway dysfunction in children with OA-TOF.

[P036] Prevalence of acidic gastro esophageal reflux after the age of 18 months in infants with esophageal atresia.

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Introduction: The high incidence of gastro esophageal reflux disease (GERD) in children operated for esophageal atresia (EA) led to the recommendation for a systematic treatment by proton-pump inhibitor (PPI) until the age of 1 year.

Aim: The aim of our study was to determine the prevalence of GERD after the age of 18 months in children who were operated for EA and to identify associated risk factors.

Methods: All patients with EA type III or IV who were born between January 2007 and December 2012 were prospectively included. Patients were *a priori* classified into 3 groups: 1) Clinical reflux: digestive or respiratory symptoms compatible with GERD after PPI discontinuation which resolved or improved when PPI was started again; 2) Definite GERD: positive pH-monitoring and/or esophagitis and/or anti-reflux surgery; 3) No GERD (normal pH-monitoring). Patients who still had GERD at the age of 18-30 months were re-evaluated during follow-up. Potential factors associated with the persistence of GERD were recorded.

Conclusion: 57 patients were studied. Among them, 82% could be evaluated for GERD at a median age of 24 months (18-30): 82% had GERD (27% clinical reflux, 40% positive pH-monitoring, 9% esophagitis, 6% anti-reflux surgery); only 18% had no GERD. At last follow-up (median age: 3.7 years (3.0-6.5)), 27 patients who previously had GERD were re-evaluated: 48% still presented GERD including 4% clinical GERD, 18% positive pH-monitoring and 26% anti-reflux surgery. No significant association was found between GERD after the age of 18 months and prematurity, associated malformation, undernutrition, or tension of the esophageal anastomosis.

The prevalence of acidic GERD after 18 months of age is high in patients with EA, and decreased to 50% after the age of 3 years. This supports systematic PPI treatment until the age of 18 months and pH-monitoring thereafter.

[P037] Growth in adolescents with esophageal atresia

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Introduction: Data on growth in adolescents with esophageal atresia (EA) are limited.

Aim: To study growth in Dutch and Norwegian adolescents with EA.

Method: EA patients, born 1996-2003, who attended follow-up in one of two participating academic centers were included. Children with genetic syndromes associated with growth disorders were excluded. Data were collected from medical records and at follow-up. Standard deviation scores (SDS) for Body Mass Index (BMI), height-for-age (HFA), distance-to-target-height (DTH) and weight-for-age (WFA) were calculated. Differences in SDS and demographics in relation to growth between both centers were evaluated. Eighty-four EA patients (53/31 Dutch/Norwegian, 55% male) were followed up at a median age of 16.5 (range 11-20) years. Gross type C EA was present in 84.5%, cardiac anomalies in 26% and VACTERL association in 33%. First hospital stay was median 34 (range 9-264) days. In forty-nine (58%) patients a median of 5 (range 1-108) esophageal dilatations had been performed. At last follow-up, nineteen (23%) patients had symptoms of gastroesophageal reflux and fifty-six (68%) had complaints of dysphagia. Twenty-six (31%) patients underwent Nissen's fundoplication. Median BMI was 19.4 (range 13.7- 34.4) kg/m². Median SDS-HFA and SDS-WFA were -0.67 (range -4.56 – 1.94) and -0.23 (range -6.03 – 3.60), respectively. Median SDS-DTH was -0.29 (range -2.06 – 1.02). Stunting (SDS-HFA ≤ -2) was present in thirteen (15.5%) patients. In thirty-three (39%) and seven (8%) patients BMI was <18.5 kg/m² and >25 kg/m², respectively. Median BMI in the Dutch and the Norwegian population was 18.5 (range 13.9-34.5) and 20.8 (range 13.7-29.3) (p<0.004), respectively. There were no significant differences in SDS for HFA, BMI, WFA or DTH between the two populations.

Conclusion: Among adolescents with EA 39% were underweight and 15.5% had stunting. Factors predicting growth in EA patients need to be identified and addressed.

[P038] Delayed primary repair with or without esophagostomy is an ideal option for long gap esophageal atresia. Report of two cases and review of literature.

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Introduction: Eventhough various procedure have been tried for management of long gap esophageal atresia, most paediatric surgeon agree that native esophagus is the best.

Aim: This study is to illustrate that avoiding esophagostomy is preferable; and primary anastomosis is possible even in those who had underwent esophagostomy during initial management.

Method: From 2012 till 2015,twelve babies with esophageal atresia were treated in Paediatric Surgical Department, Sabah Women And Children Hospital. Among those, four had long gap more than 4cm. One died at three months old due to severe cardiac disease and one had gastric pull up. Another two had primary delayed repair were analyzed retrospectively. First baby was borderline premature baby with weight 2.2kg had esophageal atresia with distal tracheoesophageal fistula, VACTERL association and duodenal stenosis. Thoracotomy done on day two found to have fistula at the carina which was ligated. Attempted primary anastomosis failed, hence distal esophagus was anchored to the T3 vertebra fascia. Upper pouch was brought out as cervical esophagostomy. Duodenoduodenostomy, gastrostomy, and colostomy was performed. Posterior sagittal anorectoplasty and colostomy closure was performed later. Delayed primary repair of esophagus was delayed up to 4 years of age due to mother delivered another baby. The second child was one of the twins who delivered at 34 weeks with extreme low birth weight of 850g had pure esophageal atresia. Gastrostomy was done at fifth day. Child was managed in neonatal ward with gastrostomy feeding and upper pouch suction. Primary esophageal anastomosis was performed at 6 months later with the weight 3.6kg. Both patients had uneventful recovery and tolerating oral feeds.

Conclusion: Esophagostomy can be avoided if the baby can be managed in the ideal environment. Delayed primary anastomosis is possible even after esophagostomy. Mobilizing and anchoring of the lower pouch helps in successful anastomosis.

[P039] Simultaneous bronchoscopy and oesophagoscopy for recurrent tracheoesophageal fistula

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Introduction: The incidence of recurrent tracheoesophageal fistula (TOF) is reported to be 5-15%.^{1,2} It can be diagnosed radiologically or by endoscopic evaluation. Endoscopic repair techniques have been described in recent times as an alternative to revision thoracoscopy or thoracotomy.

Aim: To describe a novel technique for visualisation of the tracheal and oesophageal openings in recurrent TOF using simultaneous rigid bronchoscopy and flexible gastroscopy.

Method: A case presentation of a 6 year old male with a recurrent TOF that was refractory to 2 previous endoscopic attempts at cauterisation. Standard laryngobronchoscopy was performed under spontaneous inhalational general anaesthesia initially then an endotracheal tube (ETT) was placed. A flexible 2.8mm gastroscope was introduced into the oesophageal lumen. A Parson's laryngoscope was positioned whilst the gastroscope remained in situ and the ETT was exchanged for a size 5 ventilating bronchoscope. This allowed ongoing delivery of anaesthesia and simultaneous visualisation of the tracheal lumen with a 4mm Hopkins rod. The recurrent TOF was identified, probed, measured and re-cauterised in its entirety with a Bugby diathermy under direct visualisation from both lumens, safely avoiding damage to surrounding structures.

Results: The procedure was performed uneventfully. It required good communication and coordination between the Anaesthetists, ENT and Gastroenterology teams. The patient tolerated the procedure well without any adverse effects from the dual endoscopic assessments. He had significant improvement in his symptoms and signs of aspiration, reduced requirement for antibiotic treatments and near complete closure of the fistula lumen several months later. His parents however elected for revision thoracoscopic repair.

Conclusion: Simultaneous bronchoscopy and oesophagoscopy can allow direct visualisation of a recurrent TOF tract and the safe passage of catheters, diathermy or other treatment modalities. This can avoid trauma to the surrounding anatomy and allow application of the therapy to full length of the tract.

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[P040] Standardised clinical reporting in long term otolaryngology follow-up of tracheoesophageal fistula patients

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Introduction: Patients with tracheoesophageal fistula (TOF) and/or oesophageal atresia (OA) may have a spectrum of comorbidities. The initial repair of their TOF-OA in infancy is often only a small part of their overall medical journey. A multidisciplinary approach for follow up throughout their paediatric development and beyond is essential to understand the natural history of the condition. This allows for early recognition of clinical issues, need for investigation/intervention, improved symptom control and quality of life.

Aim: To audit the clinical profile and recommended management of TOF-OA patients referred for Otolaryngology assessment in a multidisciplinary outpatient setting.

Method: A retrospective review was performed from January 2014 to June 2016 of patients attending a TOF-OA clinic referred for Otolaryngology review. Data was obtained from a standardised Otolaryngology template completed by the ENT team for each patient during clinic attendance. Information was gathered on demographics, airway, breathing, swallowing and other general ENT symptoms, clinical signs and management plan.

Results: The template was completed for 66 patients during the study period. The average age at review was 6 years (2 months to 17 years) and 42 % were male. The most common airway symptoms were TOF cough (60.6%) and airway noise (22.7%). 21.2% suffered from recurrent respiratory tract illness and 13.6% had exercise limitation. Ongoing gastro-oesophageal reflux symptoms were reported in 34.8%, with 19.6% describing choking/gagging, 16.6% aspiration, and 7.5% bolus obstruction. 12.1% had to modify dietary textures due to enduring swallowing difficulties. Other positive ENT symptoms and signs were noted (snoring 24.2%, hearing loss 4.5%, hayfever/environmental allergies 15.1%). 12.1% were recommended for laryngobronchoscopy, the main indicator being TOF cough (100%) associated with recurrent lower respiratory tract infections (62%), aspiration symptoms (50%) and reflux (50%).

Conclusion: The standardised reporting template for Otolaryngology assessment facilitates comparative data collection and improves our knowledge of the natural progress of the condition.

[P041] The refractory oesophageal stricture: a tale of multiple dilatations and unexpected resolution

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While oesophageal dilatation is a widely accepted method of dealing with post anastomotic strictures, this procedure is not without risk and can be complicated by occult or life-threatening oesophageal perforation.

We present a case of a Caucasian boy who developed a refractory anastomotic stricture shortly after an uncomplicated repair of oesophageal atresia with distal trachea-oesophageal fistula. At surgery, there was a gap of 3 vertebral bodies after full mobilization and the anastomosis was under some tension. A contrast study on the tenth post-operative day showed minimal waisting and the absence of leak. He was discharged well on anti-reflux medications.

He returned 2 weeks after discharge with a diagnosis of bronchiolitis. A repeat contrast study showed a short, tight anastomotic stricture. A schedule of dilatations was started. However, the stricture proved particularly refractory, resulting in almost total oesophageal occlusion at intervals of only 2 to 3 weeks. A Nissen fundoplication was done to address gastro-oesophageal reflux and oesophageal biopsies were negative for eosinophilic oesophagitis. Parents were offered excision of the stricture with re-anastomosis but declined. In all, a total of 38 dilatations over a 24 month period were performed, with adjuncts of topical mitomycin and intra-lesional corticosteroid injections.

Unexpectedly, at about 25 months of age, the stricture ceased to be a problem. He managed 10 months between dilatations and his final procedure was at the age of 33 months. He has been well since. Review of early dilatations showed evidence of localized perforation which likely contributed to the refractory nature of the stricture.

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We present a case of a child with multiple complex congenital anomalies and long gap oesophageal atresia without fistula. A feeding gastrostomy was done on day 1 of life and serial stretching of oesophageal ends was done at intervals under fluoroscopic guidance. Thoracotomy and anastomosis of the oesophagus was done at 5 months of age. The resultant anastomosis was under considerable tension and a contrast study showed a contained leak which resolved with conservative management, leaving a friable 1.5 cm-long oesophageal stricture.

She was discharged from hospital and returned electively for oesophageal dilatation. The first was uneventful, but the next was complicated by a delayed perforation with tension pneumothorax and collapse. The ensuing 3 months were marked by multiple episodes of pneumonia and severe sepsis with long periods of stay in the Children's ICU.

Parents subsequently declined all forms of intervention offered for the oesophageal stricture except for change of nasogastric tube under fluoroscopic guidance. Eventually, they opted to remove the nasogastric tube with full understanding that the oesophageal ends would seal up. She was sent home on gastrostomy feeds and regular upper pouch suctioning.

In the year since discharge, she has made significant developmental progress. Recent assessment of the oesophagus showed absence of continuity of both oesophageal ends. Significant lengthening of the lower oesophagus has occurred due to reflux through a wide-open oesophageal hiatus, with both ends of the oesophagus now abutting each other. This will greatly facilitate reconstruction of the oesophagus should parents decide to proceed.

[P043] Anastomotic stricture post repair of oesophageal atresia – our experience

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Introduction: Oesophageal Atresia (OA) and Tracheo-oesophageal Fistula (TOF) is a group of congenital anomalies which comprise of an interruption of the continuity of the oesophagus with or without a persistent communication with the trachea. Oesophageal strictures are the most common complication following OA/TOF surgery, and convey a significant morbidity to the care and progress of a child.

Aim: To evaluate the incidence of anastomotic stricture post OA/TOF surgery.

Method: A retrospective analysis was conducted of 116 patients with OA/TOF, surgically treated during the time period from 1998 – 2010, at the Sydney Children's Hospital, Randwick, Australia. Eleven patients were excluded, three due to multiple congenital anomalies and 8 due to lack of case notes.

Conclusion: The incidence of stricture overall was 56.2%. Long gap OA/TOF and gastroesophageal reflux disease were both found to be statistically significant independent risk factors of stricture formation. Long gap OA/TOF was also shown to be a statistically significant independent risk factor for long term stricture development. Presence of a leak on the first contrast suggested an association with stricture formation; however the sample size was too small to demonstrate a statistically significant difference. Gender, presence of stricture on first contrast and a VACTERL association were demonstrated to be non-risk factors for stricture development.

[P044] Thoracoscopic multistage repair of long-gap esophageal atresia using internal traction sutures - what time between stages is optimal?

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Introduction: Treatment of long-gap esophageal atresia poses a great challenge for a surgeon. Commonly utilized techniques require multiple surgeries carrying negative complications of open thoracotomy and often do not allow to preserve native esophagus. In our Department a new multistage thoracoscopic technique of long-gap esophageal atresia treatment using internal traction sutures was developed.

Aim: The aim of the study is a retrospective analysis of performed procedures aimed at finding optimal time interval between following surgeries in a multistage treatment.

Methods: 23 cases of newborns with long-gap esophageal atresia (type A and B) were operated thoracoscopically using internal traction sutures between 2008 and 2015 in different hospitals with first author involved personally. Originally time interval between stages was set to be 4 weeks.

Results: Of 23 newborns operated on using internal traction 1 patient died before finishing treatment and was excluded from the analysis. Overall 57 procedures were performed. 13 newborns were operated in two stages, 7 in three stages and 2 in five stages. Time interval between procedures was longer than 4 weeks almost exclusively due to logistical complications and only in two cases because of patient's condition. In two cases time intervals were purposefully shortened to 8 and 15 days. The mean time interval between surgeries was 41,42 days (minimal 8 days and maximal 101 days). For newborns treated in two stages mean time interval was 31,23 days and after exclusion of 2 last patients with aforementioned shortened time interval the mean was 34,81 days. In newborns with esophageal anastomosis performed in third stage mean time interval between 1st and 2nd stage was 51,85 days and between 2nd and 3rd 52,28 days.

Conclusion: Results of the study may indicate that prolonged time interval between procedures does not increase the chance of performing faster esophageal anastomosis.

[P045] Diagnostic value of upper digestive endoscopy in symptomatic children with esophageal atresia

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Introduction: Upper endoscopy (EGD) with esophageal biopsies are recommended to evaluate symptomatic patients with esophageal atresia (EA). However, the diagnostic yield of EGD is unknown in this population. We therefore aimed to report the diagnostic value of EGD in a cohort of EA patients prospectively followed in a EA Clinic.

Patients and methods: All children born with EA from September 2005 to December 2014 and followed at the Ste-Justine EA Clinic were included. Digestive and/or respiratory symptoms systematically collected and endoscopic and pathological findings were analyzed.

Results: 84 patients (33 girls, 8 type A, 65 type C, 3 type D) with a median age at last follow-up of 57 months (range 17-130) were included. Eighteen patients (12%) were found asymptomatic. Nine patients, (8 < 4 years, asymptomatic), did not undergo EGD during the study period.

67 patients underwent 92 EGD. They all presented at least 1 digestive or respiratory symptom. The diagnostic of anastomotic stricture leading to further dilations (median number 4 (range 1-11)) was performed in 32 patients. Endoscopic findings in the 35 remaining EGD were as follows: peptic esophagitis (10), eosinophilic esophagitis (6), congenital stenosis (4), recurrent fistula (1), gastric metaplasia (4), duodenal web (1), candida infection (1), hiatal hernia (1) and 5 recurrence of stricture. Normal macroscopic and microscopic findings were reported in 25 (27%) symptomatic patients. Overall, 72 % of the endoscopic procedures performed in symptomatic patients yielded to a diagnosis.

Conclusion: The diagnostic value of the EGD with esophageal biopsies is high in children with EA presenting with digestive and/or respiratory symptoms. EGD with biopsies should therefore be systematically indicated in this context.

[P046] How often do we need to evaluate exercise capacity in children with oesophageal atresia-tracheo-oesophageal fistula (oa-tof)?

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Introduction: Children with OA-TOF can experience significant long-term respiratory morbidity, limiting exercise capacity and affecting pulmonary function. Few studies have assessed aerobic capacity (VO_2 peak), ventilatory limitation, and other Cardiopulmonary Exercise Testing (CPET) measures, in children with OA-TOF. No current studies have reviewed longitudinal exercise limitations in children with OA-TOF.

Aim: This study prospectively investigated the longitudinal response to exercise in children with OA-TOF, to determine if children with OA-TOF maintain exercise capacity and, if short-term longitudinal functional assessment (CPET) provides information aiding management.

Method: Children aged ≥ 7 years attending a multidisciplinary OA-TOF clinic were evaluated prospectively. CPET and Spirometry follow-up testing were performed within 2.5 years of baseline testing. CPETs were performed on a treadmill ergometer, using a standardised incremental protocol (Bruce). Peak exercise capacity was defined as achieving 2 or more of the following: respiratory quotient >1.0 , a modified Borg rating ≥ 8 and/or a heart rate $>80\%$ of predicted maximum. Spirometry was undertaken according to ATS guidelines. Paired sample t-tests were utilised to analyse data.

Results: Thirteen children (6 male), aged 7.34-15.99 years, performed both a peak CPET and Spirometry, on two separate occasions (mean time interval 1.7 ± 0.4 (SD) years).

	Baseline (n=13)	testing	Follow-up (n=13)	testing	P Value
Age (years)	10.7 \pm 2.3		12.4 \pm 2.3		
FEV ₁ (%pred)	74.5 \pm 18.4		77.3 \pm 17.9		0.336
VO ₂ peak (mL/min/kg)	41.3 \pm 7.5		45.4 \pm 7.3		0.089
O ₂ Pulse (mL/beat)	7.1 \pm 1.6		9.2 \pm 2.6		0.002
VE/VCO ₂ peak	27.7 \pm 2.5		28.2 \pm 3.7		0.563
VE/VO ₂ peak	31.3 \pm 3.9		30.1 \pm 4.5		0.303
Tidal Volume Increase	2.3 \pm 0.6		2.7 \pm 0.9		0.088
Breathing Reserve (%)	19.2 \pm 11.5		18.3 \pm 17.1		0.767

Values are Mean \pm SD

Conclusion: This pilot study indicates that children with clinically stable OA-TOF maintain exercise capacity, within the time frame observed (12-30 months). Thus, short-term repeat CPET is unlikely to provide additional information which would alter clinical management. This study may help inform future management guidelines for the longitudinal evaluation of respiratory function in children with OA-TOF.

[P047] Skeletal thoracic abnormalities after oesophageal atresia cure. A national register CRACMO study

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Introduction: Thoracotomy performed during the cure of oesophageal atresia (EA) leads to thoracic deformation (20 à 50% of adults), with 14% of thoracotomy-induced scoliosis. Recent progress with mini-invasive approach could reduce skeletal/costal sequelae, but this has never been assessed in large series of EA.

Aim: to assess incidence of these abnormalities in a population-based national cohort and look for associated factors.

Method: Paediatric surgery departments from the CRACMO network were asked to send the more recent thoracic X-ray of their patients born between 2008 and 2010 and included in the national register. Thirty-two out of the 37 centres from the national network included 326 patients. All X-ray were blindly and centrally reviewed by a radiologist and a paediatric surgeon.

Results: Two hundred and twelve X-ray (65%) showed skeletal thoracic abnormalities, including 25 (7.7%) thoracic congenital malformations and 187 (57.4%) costal or intercostal acquired abnormalities (129 (39.6%) costal and 58 (17.8%) intercostal). Acquired abnormalities were not influenced by age at procedure, birth weight or atresia type. However, these abnormalities were more frequent when X-ray was performed more than one year after surgery compared to early X-ray (60.4% versus 43.4%; $p < 0.01$) and mostly consisted in costal hypoplasia (69.3% versus 42.9%; $p < 0.05$). The rate of sequelae was lower for mini-invasive approaches - thoracoscopy (n=9) or axillar Bianchi thoracotomy (n=11) - compared to classical posterior-lateral thoracotomy (4/20 (20%) versus 142/239 (59.4%); $p < 0.001$).

Conclusion: In addition to 8% of congenital thoracic bone malformations associated with oesophageal atresia, more than 50% of patients have thoracotomy-induced costal abnormalities than could be reduced by use of mini-invasive approach. The long term morbidity induced by those abnormalities remains to be assessed.

[P048] Respiratory problems in children with repaired esophageal atresia with tracheoesophageal fistula.

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Introduction: Children with congenital esophageal atresia (EA) and tracheoesophageal fistula (TEF) have chronic respiratory symptoms including recurrent pneumonia, wheezing and persistent cough. Aim: The aim of this study is to describe the clinical findings of patients with EA and TEF surgically corrected and the instrumental investigation to which they have undergone during follow-up evaluation in order to better define a standardized algorithm for their long term management.

Methods: a retrospective data collection was performed on 105 children with EA and TEF followed at Department of Pediatric Medicine of Bambino Gesù Children Hospital (Rome, Italy) between 2010 and 2015.

Results: 64 (61%) children were treated at Bambino Gesù Children's Hospital for surgical repair of EA with TEF. 70 (66,6%) children reported lower respiratory symptoms with a mean age onset of 2.2 ± 2.5 years and only 63 (60%) performed specialist assessment at Respiratory Unit. The first pneumological evaluation was performed at mean age of 3.9 ± 4.2 years. 29 patients have undergone to chest CT with contrast enhancement detecting localized atelectasis (43%), residual tracheal diverticulum (35,7%), bronchiectasis (32,1%), tracheal vascular compression (21,4%) or without other causes (17,8%) and esophageal diverticulum (14,3%). 53 patients have undergone to airways endoscopic evaluation detecting tracheomalacia (74%), residual tracheal diverticulum (29,8%), tracheoesophageal fistula recurrence (21,2%) and vocal cord paralysis (12,7%). Of the remaining 35 patients, 13 (12,4%) referred only upper respiratory tract infections and 22 were asymptomatic for respiratory disturbances. None of them performed pneumological assessment.

Conclusion: our study underlines that respiratory symptoms often complicate AE and TEF; their persistence despite surgical treatment of gastroesophageal reflux means that other etiological hypothesis must be examined. Associated cardiopathy and atopy may contribute to the early onset of symptoms. On the basis of above considerations, due to patients' complexity and comorbidity, a delayed pneumological assessment is unjustified.

[P049] Spirometry: a useful and reliable tool for monitoring tracheomalacia in patients with esophageal atresia.

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Introduction: airway endoscopy is the gold standard for the diagnosis of tracheomalacia (TM). The need of sedation and invasiveness limit its use in specialized centers. The role of spirometry in patients affected by TM is rarely described. Due to non invasiveness and easy execution, it is particularly interesting in clinical practice.

Aim: to define sensitivity and diagnostic predictive value of spirometry in detecting tracheal collapse in patients with TM endoscopically detected.

Methods: 53 patients with esophageal atresia surgically corrected and tracheomalacia in follow-up at Department of Pediatric Medicine of Bambino Gesù Children's Hospital, underwent to laryngotracheobronchoscopy. The morphology of the volume-flow curve during expiratory phase was examined in patients taking a spirometry test.

Results: 12 patients with intrathoracic tracheomalacia and 4 patients with normal airway endoscopy performed spirometry at mean age of 9.3 ± 3.1 years. A sudden and temporary drop during expiratory phase was observed respectively in 8 and 1 patients. FEV1/PEF ratio was 8.6 mL/L/min (range 4.29 and 12.5) and 7.6 mL/L/min (range 7.1 e 8.1) respectively in the first and second group. If compared with endoscopy, spirometry detected tracheal collapse during expiratory phase with diagnostic sensitivity of 67%, specificity of 75% and positive and negative predictive value respectively of 89% and 42%. FEV1/PEF ratio, that was expressive of intrathoracic obstruction, was higher in patients with TM detected by endoscopy.

Conclusions: even if laryngotracheobronchoscopy is the gold standard for the diagnosis of TM, spirometry is a useful and reliable tool for monitoring intrathoracic tracheomalacia in symptomatic patients and/or in patients for which sedation is not indicated.

[P050] Esophageal stents for post-operative complications after tracheoesophageal fistula/esophageal atresia (tef/ea) repair.

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Introduction: Anastomotic stricture or leak may occur after TEF/EA repair. Most strictures ultimately respond to dilatation, and conservative management of a leak may be effective, whereas recalcitrant problems may require surgery. An esophageal stent could facilitate non-operative management, but until recently their use has been limited by the availability of appropriately-sized stents for small children and experience in placing them.

Aim and Method: Retrospective chart review of our experience with esophageal stents.

Case Series: 5 children with a history of repaired TEF/EA (ages 26 months-16 years) underwent a total of 11 esophageal stent placements since 2014. 7 of the stents were re-purposed partially-covered self-expanding metal biliary stents (8mm or 10mm x 6cm) and 4 were fully covered self-expanding metal esophageal stents (FCSEMS; 12mm x 6cm to 16mm x 10cm). All were placed under general anesthesia using endoscopic and fluoroscopic guidance. 3 stents were placed for anastomotic leaks and were left in place 7 days, 18 days, and 41 days, respectively, with no recurrence of the leak upon removal. 1 FCSEMS was in place for 56 days to cover a TEF after endoscopic cautery, with no leak upon removal. 7 stents were placed to manage recalcitrant anastomotic strictures, none of which resulted in sustained luminal patency after removal, although 2 migrated distally and one is still in place. One patient complained of transient chest pain and one had mild dysphagia from ingrowth of granulation tissue prior to stent removal. There was no occurrence of obstruction, bleeding, or perforation.

Conclusion: Stenting the esophagus to cover an anastomotic leak is feasible with available products, although they are not approved for such use in children in the United States. Dysphagia responded to stent placement across strictures, but long term anastomotic patency was not achieved without subsequent surgical intervention or ongoing dilatation.

[P051] Esophageal substitution for atresia: experience in a tertiary care centre with resource constraints

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Introduction: Esophageal substitution for pure esophageal atresia (PEA) or after complications following long gap esophageal atresia with tracheo-esophageal fistula (LG-EATEF) repair at birth is a major procedure in infants and children.

Aim: Experience with two operative procedures, where major portion of stomach is retained in the abdomen for storage function, is presented from a resource constraint set up. (a) Reverse Gastric Tube Esophagoplasty (RGTE) in children (n=16) with gastro-epiploic arterial arcade on the greater curvature of stomach and (b) Fundal Tube Esophagoplasty (FTE) where gastro-epiploic arcade was not available (n=34).

Methods: Fifty children [group A: PEA (n= 25); group B: LG-EATEF (n= 25)] who underwent surgery from 2001 to 2015 were included. Children with PEA underwent a gastrostomy & cervical esophagostomy at birth. Esophageal substitution was performed between the ages of 6 to 36 months (mean 12 months). Patients with LG-EATEF underwent thoracotomy, ligation of fistula and esophageal anastomosis at birth. Consequent to various complications, the anastomosis was taken down, converted to gastrostomy & cervical esophagostomy and followed by esophageal substitution between 9 to 84 months (mean 18 months). Follow up ranged from 1 – 15 years.

Results: Serious associated congenital anomalies were present in 18 (36%) children. There were 2 (4%) mortalities, one each in PEA & LG-EATEF groups. Eight out of 48 (16.7%) children required revision of esophageal anastomosis (3 in group A and 5 in group B). Eighteen (37.5%) out of 48 required 2 to 8 esophageal dilatations (12/24 in group A and 6/24 in group B). Two children in LG-EATEF group required a second procedure of colonic conduit. Forty-seven children are eating normal diet orally and maintained satisfactory somatic growth.

Conclusions: Complicated LG-EATEF patients with previous thoracotomy at birth had more complications. Children undergoing esophageal substitution require prolonged and constant surgical care for satisfactory results.

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Aim: To achieve extubation on table after the procedure, to reduce- cardiopulmonary morbidity, anastomotic leak and financial burden.

Introduction: Oesophageal replacement surgery in pediatric age group is mostly done for benign conditions. For safe & successful outcome, it involves distinct and demanding perioperative care. Different centers follow their own pattern of management with or without intensive care team, but there is no well described protocol of care. However, for optimal results it is critical to have goal directed guidelines of management. Our Evolving Protocol (EP) is an attempt in this direction. It is more pertinent in resource limited situations.

Methods: Records of 69 out of 102 patients of oesophageal replacement done over past twenty years were shortlisted for this study. Stomach (tube or pull up) is used as a conduit in all the patients by transhiatal approach. Fifty patients of OA requiring oesophageal substitution were managed according to Conventional Protocol (CP) while our Evolving Protocol (EP) was used in 19 patients. This protocol involves modifications in, anesthesia techniques - minimum necessary intra op iv fluids, extubation on table etc; surgical techniques - gastric pullup, pyloroplasty in all, two layered intussuscepting oesophago - gastric cervical anastomosis and modified early post operative care viz..restricted as opposed to liberal intravenous fluids, use of anti-sialogogue, hydrocortisone, diuretics & mild sedation with midazolam drip, diclofenac suppository for analgesia etc. Tests of significance were applied for validation.

Conclusions: Statistically significant reduction in pulmonary complications were noted hence requiring less use of PICU facilities leading to shorter hospital stay, less anxiety moments, substantial decrease in overall cost of management without compromising on final outcome. Though EP shows encouraging results multicentric studies & randomized control trials would be pragmatic.

[P054] Risk factors for early morbidity and mortality in esophageal atresia type C: data from a population based register

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Introduction: Prognosis of Esophageal atresia (EA) dramatically improved over the last decades. However, there remains a high morbidity especially during the first year of life. Data about factors associated with morbidity in large population is lacking and should help defining risk group.

Aims: to assess the one-year mortality and morbidity in EA type C on a population based national register.

Methods: Data from all the 562 new cases of EA type C born from 2008 to 2011 in France were analysed. Mortality and morbidity (length of stay (LOS) and full oral rate at one year of age) were studied within the first year of life. Multivariable analyses were performed using logistic or linear regression.

Conclusions: Early mortality occurred in 31 cases (5%). Multivariable analysis showed that birth weight ≤ 2600 g (OR, 5.34; 95%CI, 1.81–15.81; $p=0.003$), associated malformation (cardiac malformation: OR, 9.22; 95%CI, 2.62–32.41; $p=0.001$, other malformation: OR, 4.59; 95%CI, 1.18–17.89; $p=0.028$, using no malformation as reference) and prenatal diagnosis (OR, 2.66; 95%CI, 1.14–6.192, $p=0.024$) were significantly associated to early mortality. LOS during the first year of life was related to birth weight ≤ 2600 g ($p<0.0001$) and associated malformation ($p<0.0001$) in multivariate analysis. Full oral feeding at one year of age was related only to associated malformation ($p=0.018$). This study confirms that, still in 2016, birth weight and malformation are associated with mortality and identifies new risk factors for mortality and morbidity in EA type C.

Characteristics of the study population	N=562
Prenatal diagnosis	16%
Sex ratio (M/F)	1.34
Primary anastomosis	93%
Associated abnormalities	53%
Vacterl association	23%
Anastomotic stricture	20%
Fundoplication	6%
Need of respiratory medications at one year of age	30%

Survival	95%
Fully orally fed at one year of age	91%
LOS during the first year of life, days (median, ranges)	26 (1–730)

Dr Parthiv Shah

Introduction: Type H trachea-esophageal fistula accounts for 4-5 % of congenital esophageal atresia / trachea-esophageal fistula cases. The symptoms are variable and sometimes subtle causing delayed diagnosis. We present our 16 years' experience of treating this rare congenital anomaly.

Patients and Methods: This is a retrospective analysis of all cases operated between 2001 and 2016 by a single surgeon. All patients who had clinical suspicion based on their symptoms and signs underwent esophagogram and rigid video bronchoscopy. All patients were operated and esophagogram was performed on 7th post operative day.

Results: Twelve patients, age ranging from 7 days to 10 years, were operated during this period. Follow up of these children ranged from 15 years to 15 days. Five neonates (weight 1.8 kg to 3.2 kg), five infants and two older children underwent repair soon after diagnosis. Fistula was located with rigid video bronchoscopy in all cases and all but one case was operated through cervical approach. Post operatively 4 newborns and one infant required ventilator support for 2-3 days and total hospital stay was 10 to 24 days. One leak (closed spontaneously) and one recurrent extubation failure (due to congenital distal tracheal stenosis) were complications.

Conclusion: A high index of suspicion and early bronchoscopy are essential for diagnosis of this rare anomaly. Surgical outcome is excellent with minimal morbidity.

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Introduction: Evaluation of quality of life (QoL) after surgical repair of esophageal atresia (EA) is an important factor for effective treatment and proper care of the child.

Aim: The aim of this study was to assess QoL of children with isolated or syndromic EA.

Method: The survey was conducted from July 2015 to January 2016. The data of 73 patients were analyzed. Quality of life was evaluated by means of PedsQL 4.0 questionnaire. *Mann-Whitney U* test was applied and p-values <0.05 were considered statistically significant.

Conclusions: Out of 73 patients (31 girls, 42 boys) that have participated in the study, 61 (84%) had EA with tracheoesophageal fistula (TEF) and 12 (16%) had EA without TEF. Gestation (GA) 27-42 weeks, 37 (51%) premature infants were identified. 41 (56%) had associated malformations, isolated (32 patients, 44%). 26% with cardiac, 23% with skeletal, 14% with respiratory abnormalities. In the surveyed group the mean QoL score in emotional functioning were significantly lower 57 [range 30-100] then in social functioning 72 [range 0 – 100], whereas mean physical functioning score was 69 [range 25-100]. The comparison between the QoL of children with TEF and those without TEF revealed no statistical differences ($p > 0.05$). The analysis showed that there is a significant correlation ($p < 0,034$) between gestation age (whether the child was born preterm or in term) and QoL in the domain of social functioning. QoL improves with age in physical functioning. The presence of concomitant anomalies does affect the overall generic QoL. Children with associated central nervous system anomalies has the lowest mean score ($M=49$) in emotional functioning. QoL of premature and syndromic with EA appears to be low. It seems to be needed to extend psychological care of premature babies with EA.

[P057] Alternations in genes expression microarray in esophageal atresia tissues – the role of cytokine-cytokine receptor interaction and rho cell motility signaling pathways.

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Introduction: Esophageal atresia (EA) with or without tracheoesophageal fistula belongs to common congenital anomalies. EA may occur as isolated (IEA) or syndromic (SEA). EA etiology is complex and still unexplained. It is suggested that multifactorial mechanism combined with epigenetic factors play role in EA etiology.

Aim: The aim of study was to assess the level of gene expression directly in the esophageal tissue and compare these results with control esophageal tissue.

Method: The material for study was RNA extracted from esophageal tissue acquired from newborns with esophageal atresia. The control group was RNA extracted from esophageal tissues taken from aborted fetuses and stillborn neonates without congenital defects. In first step the gene expression profiling in IEA and SEA vs control was done. The expression analysis was performed by microarray methods (Agilent SurePrint G3 Human GE).

Conclusion: After the functional analysis of signaling pathways (by KEGG database and DAVID) two pathways were chosen - *cytokine-cytokine receptor interaction* and *Rho cell motility signaling pathway* and nine genes (*CCL2*, *LIF*, *TNF*, *TNFRSF6B*, *LIMK1*, *ARHGEF1*, *ARHGEF11*, *TLN1*, *VCL*) were analyzed by real-time PCR (The LightCycler® 480 System, Roche). Microarray examination was performed on 26 tissues, real-time PCR was done in group of 20 tissues (10 of IEA and 10 of SEA). Statistical analysis after Real-time PCR was done by Rest2009 Software, Qiagen GmbH. In microarray we identified about 4800 up and down regulated probes between IEA and controls and about 5000 up and down regulated probes between SEA and controls. About 2300 probes occurred both in IEA and SEA groups. Analysis after real-time PCR showed down-regulation for *ARHGEF1* gene in IEA vs. control and for *LIF* gene in SEA vs. control.

[P058] Perspection of caregivers concerning waiting period for delayed primary esopgaheal repair: at home or at hospital?

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Introduction: Patients with long gap esophageal atresia (LGEA) may require delayed primary esophageal repair (DPER) and wait at home or at hospital until esophageal anastomosis become possible.

Aim: To evaluate the perception of parents' concerning the waiting period.

Methods: Caregivers responded a questionnaire including statements about feeding, aspiration and care-giving problems with a 5-point Likert scale. The effect of waiting period on parents' social and economical status and their child's health was also surveyed. Patients waited at home (HMG) and at hospital (HSG) were enrolled in 2 groups and results were compared with non-parametric test.

Conclusions: Twenty mothers (n=13-65% HSG/n=7-%35 HMG) responded the questionnaire. There was no difference between parent's age and educational status ($p>0.05$). Caregivers' male-to-female ratio was 7:6 in HSG and 2:5 in HMG. Birth weights and gestational week of the groups were similar ($p>0.05$). Nine of the cases were isolated atresia (76.9%) and four of them were EA-with fistula in HSG. All of the patients in HMG were isolated atresia. The mean operative time was 120 days (77-143). When statements were compared, parents in HSG had better and easy information about their baby form health-care professions ($p<0.05$). Parents in HMG felt themselves happier then parents at hospital ($p<0.05$). Most important problems were unable to understand a serious health problem, aspiration and respiratory problems respectively. Feeding difficulties and problems with medical devices were least important issues. HMG need hospital visits because of emergency in 1 case (14.3%) and for control purposes (n=6, 85.7%). Although rate of complications were similar in groups, HSG had higher infectious complications. Perceptions of caregiver's reveal that they feel and care themselves better at home, however they had doubts about ability to understand severe health problems.

[P059] The effects of swallowing rehabilitation programme in patients with esophageal atresia and tracheoesophageal fistula

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Introduction: Dysphagia is a common clinical problem in patients with esophageal atresia and tracheoesophageal fistula (EA-TEF).

Aim: To propose a Swallowing Rehabilitation Protocol (SRP) for EA-TEF patients with pharyngeal swallowing disorder (SD) and to evaluate the results of SRP on swallowing functions (SF).

Methods: Patients were evaluated for age, sex, type of atresia, repair time and time of oral intake. Patients with SD and airway aspiration during deglutition in videofluoroscopic swallowing study (VFSS) were included. SRP including thermal tactile stimulation, laryngeal mobilization and neuromuscular electrical stimulation was performed. SF was evaluated with 3 ml liquid and pudding barium by VFSS before and after 20 sessions of SRP. Parameters of oral phase dysfunction, delay in swallowing reflex, nasal regurgitation, laryngeal penetration, aspiration, abnormal esophageal body function, reflux, residue were coded as either 'absent' or 'present'. The penetration-aspiration scale (PAS) was also used to quantify a patient's ability to protect the airway during VFSS.

Conclusions: Nine children were included. The median age was 10 months (4-31 months) and male-female ratio was 5:4. Six (66.7%) of the patients had isolated-EA and three (33.3%) of them had EA+TEF. The median repair time was 12 weeks (1-20 weeks) and oral feeding started with a median of 5 months (1-8 months). The VFSS results before SRP were indicated that non-oral feeding was essential in 44.4% (n=4) of the patients and liquid restricted diet was essential in 55.6% (n=5). A significant improvement in terms of oral phase dysfunction, delay in swallowing reflex, PAS, residue in valleculae, pharyngeal wall and pyriform sinuses was found after SRP ($p<0.05$). After 20 sessions, full oral feeding was recommended in 55.6% (n=5) of the patients and liquid restricted diet was continued in 44.4% (n=4) patients. SRP improves swallowing function in children with SD. Liquid restricted cases should require longer swallowing rehabilitation sessions.

[P060] A questionnaire-based survey on feeding and swallowing problems in children with esophageal atresia

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Introduction: Feeding and swallowing problems are most common complaints in children with esophageal atresia and/or tracheoesophageal fistula (EA-TEF).

Aim: A questionnaire-based survey was performed to determine feeding and swallowing problems of children with EA-TEF.

Methods: Data was obtained from families of children with operated EA between August-November 2014 by an online survey. Among 44 responses, completely answered (n=35) questionnaires were evaluated.

Conclusions: Mean age was 3.3 ± 2.6 years and male:female ratio was 16:19. Forty-five percentage of children had isolated-EA, 49% had EA-TEF and 6% had isolated-TEF. Except one child with gastrostomy, all children fed orally. Esophageal dilatation was needed in 60% while 3 or more dilatations were made in 67% of all children. Nutrition consultancy was given before hospital discharge in 77% of patients by doctors (92%), nurses (28%) and dietitians (16%). The most common recommendations were serving small and frequent meals (37%), accurate feeding posture (26%) and mechanical food process before serving meals as mashing or cutting into small pieces (22%). Only 76% of children over 6 months of age (n=33) were able to start eating solid foods (beginning at 18.6 ± 10.5 months). In children who were able to eat solid foods, 24% were having difficulty in chewing and 82% were having problems with certain foods (e.g. meat/bread/crusty-hard fruits). We also found that 69% of children had swallowing difficulty, 67% had reflux and 46% were admitted to emergency at least once for food impaction. More than 80% of families reported that the disease of their children negatively affected their social life and psychology. According to weight-for-age and height-for-age z-scores, 22.8% of children were classified as both underweight and stunted. Feeding and swallowing problems are very common in children with EA/TEF. Early detection and treatment of these problems will positively affect families' quality of life and growth & development of these children.

[P061] Comparison of growth and feeding problems in children with esophageal atresia with early or delayed oral feeding after esophageal repair

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Introduction: Feeding problems are common in patients with esophageal atresia-tracheoesophageal fistula (EA-TEF) and may cause growth problems in children with delayed oral feeding.

Aim: A cross-sectional study was planned to compare growth and feeding problems in children with EA with early (<1 month) or delayed (>1 month) oral feeding.

Methods: Data was obtained between August 14th- June 15th from families of patients operated for EA, by an online survey with a total of 79 questions. From 45 survey responses, completely answered questionnaires (n=36) were evaluated. Patients were divided into two groups as early (EF) or delayed oral feeding (DF).

Conclusions: Thirty-six children (44.4% boys) with a median age of 25.5 months (1-130 months) were included. Isolated-atresia was present in 47.2% of children while 52.8% had EA-TEF. Primary repair was performed in 2 days (median; 1-5 days) and oral feeding was started in 12 days (median; 7-27 days) after birth in EF group (n=17; 47.2%) while they were 90 days (1-540 days) and 150 days (30-900 days) respectively in DF. Mean birth-weight, weight-for-age and height-for-age z-scores were significantly lower in children with DF (p<0.05). Longer meal times, food refusal, choking and vomiting at meals were significantly more common in children with DF in milk-based feeding period (p<0.05). The age at onset of lumpy and solid complementary foods, the age at which the feeding problems were resolved and duration of meal times were higher, but total breastfeeding time was lower in children with DF (p>0.05). Feeding difficulties (particularly with solid foods), swallowing and chewing problems, strictures and reflux were more prevalent in DF without any significance. The age at primary repair and the age at onset of oral feeding may effect growth and may lead to more frequent feeding problems particularly in infancy and early childhood in children with EA.

[P062] Validity of pediatric eating assessment tool-10 to predict aspiration in children with esophageal atresia

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Introduction: Aspiration is a common problem in children with esophageal atresia (EA) and may cause severe respiratory problems. Pediatric eating assessment tool-10 (pEAT-10) is a self-administered questionnaire to evaluate dysphagia symptoms in children and may predict aspiration.

Aim: To evaluate the validity of pEAT-10 for prediction of aspiration in children with EA.

Methods: Patients operated for EA were evaluated for age, sex, type of atresia, presence of associated anomalies, type of esophageal repair, time of definitive treatment and onset of oral feeding. Penetration aspiration score (PAS) was evaluated with videofluoroscopy (VFS) and parents were surveyed for pEAT-10, dysphagia score (DS) and functional oral intake scale (FOIS). PAS>7 was considered as risk of aspiration, and EAT-10>3 was abnormal.

Conclusion: Forty patients were included in the study. Children with penetration-aspiration in VFS (PAS>7) were assessed as PAS+ group, and the ones with PAS<7 were PAS- group. Demographic features, results of surgical treatment, and median levels of PAS, pEAT-10, DS and FOIS are listed in Table 1. The sensitivity and specificity of pEAT-10 to predict aspiration were 88% and 77%, positive and negative predictive values were 22% and 11%, respectively. Type-C cases had better pEAT-10 and FOIS than type-A cases, and both scores were statistically better in primary repair than delayed repair ($p<0.05$). Onset of oral feeding did not differ for each score. Among the postoperative complications, only leakage had impact on DS, pEAT-10, PAS and FOIS scores ($p<0.05$). Aspiration is a common problem in patients with EA. pEAT-10 is a simple and reliable tool to predict airway aspiration in children. Patients with higher pEAT-10 scores should undergo detailed evaluation of deglutitive functions and risk of aspiration to promote safer feeding strategies

[P063] Evaluation of the first year data of Turkish esophageal atresia registry

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Introduction/Aim: To evaluate the first year data of Turkish Esophageal Atresia Registry (TEAR).

Methods: Patients registered to TEAR between March-2015/April-2016 were evaluated for age, sex, demographic features, type of atresia, diagnostic methods and results of surgical treatment.

Results: Hundred cases from 15 centers were included. Male-female ratio was 54:46 and mean birth weight was 2256,28 g (870-3920 g) and height was 45 cm (33-52 cm). Mean gestational week was 35,4 weeks (28-41 weeks) and mother age was 28.7 years (18-44 year). The types of atresia according to Gross classification were 14%-A, 4%-B, %80-C and 2%-D. 91% of cases were born after spontaneous pregnancy and 3 of them were twins. Prenatal diagnosis was obtained in 34% of cases. Most common prenatal findings were polyhydramnios (45%) and absence of stomach gas (13%). Family history of VATER was seen in 3 cases. The incidence of associated anomalies was 75% and most common anomalies were cardiac (65%), renal (13%) and anorectal (12%). Primary anastomosis was obtained (n:82) with thoracotomy (n:78) and thoracoscopy (n:4). Standard repair was performed in 64 cases whereas 39% of them were tensioned. The mean gap was 2.26 vertebral body (1-5). Foker's technique and Livaditis was performed in one case. Gastric pull-up, colonic interposition and cervical esophagotomy were performed in one patient each. Mean endotracheal intubation and non-invasive ventilation time were 15.16 (0-210) and 67.22 (0-200) days respectively. After a mean time of 109 (9-200) days patients were free of oxygen. Gastrostomy (n:27), antireflux surgery (n:1) and aortopexy (n:1) was performed. 60% of patients discharged from hospital with full oral feeding and 25% with enteral feeding. The survival rate was 78% and mortality was 22% at the end of first month. Seven cases waiting for delayed primary repair have been following up at home (n:4) and at hospital (n:3). The data obtained from TEAR is the first national

data about EA in our country. If more centers will register, it will be possible to define the current outcome of EA in Turkey and compare the results with the international data.

[P064] Correlation of respiratory problems with results of surgical treatment in patients with esophageal atresia and tracheoesophageal fistula

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Introduction/Aim: To evaluate the correlation between respiratory symptoms (RS) and results of surgical treatment in esophageal atresia-tracheoesophageal fistula (EA-TEF).

Methods: Patients operated for EA-TEF were evaluated for age, sex, type of atresia, time and type of esophageal repair (ER) and surgical complications. Respiratory functions were correlated with type of atresia, time of ER and oral feeding with Chi-square test.

Results: Fifteen patients operated for EA-TEF with RS were included. The mean age of the patients was 36.9 months (4-8). Male to female ratio was 8:7. Types of atresia were isolated-EA (n=10, 66.6%), EA-distal-TEF (n=4, 26.6%) and EA-proximal-TEF (n=1, 6.6%). 73% of cases had associated anomalies. Primary ER (n=10), delayed ER (n=4) and colonic replacement (n=1) were performed. Anastomotic leak was seen in 3 patients (20%) whereas stenosis and recurrent fistula were seen in 1 patient. Oral feeding started early (<1 week) in 5 patients (33.3%), less than 1 month in 6 patients (40%) and delayed (>1 month) in 4 (26.6%) patients. Gastroesophageal reflux (GER) encountered in upper GI (n=5), and pH studies (n=1). In videofluoroscopic evaluation, aspiration (n=3) and airway penetration (n=2) was seen during deglutition. Five cases had a history of lower respiratory tract infection (LRTI). When type of atresia was correlated with RS, isolated cases had more LRTI, aspiration, atelectasis, mechanical ventilation and postoperative wheezing (p<0.05). Patients with primary ER had less aspiration and consolidation than delayed repair (p<0.05). Patients with late-oral-feeding had more mechanical ventilation, GER, need for inhaled medication and peribronchial thickening than patients with early-feeding (p<0.05). Also, early-fed cases had less atelectasis, and LTRI than delayed-oral-feeding (p<0.05). RS are common in patients with EA-TEF and may be correlated with type of atresia especially in isolated cases. Delayed ER and oral-feeding one month after ER had more associated RS than primary cases and early-fed patients.

[P066] Postoperative infections in children with esophageal atresia

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Introduction: Postoperative infections (PI) are common cause of mortality and morbidity in children with esophageal atresia and tracheoesophageal fistula (EA-TEF).

Aim: A retrospective study was performed to evaluate the incidence and clinical features of PI in children EA-TEF.

Methods: Children who underwent primary esophageal repair were evaluated for age, sex, type of atresia, birth weight and gestational age, time of repair and oral intake, clinical findings and microbiologic evaluations for PI. The results of isolated cases and cases with TEF were compared to evaluate the effect of atresia type on PI.

Conclusions: Forty-four cases with a median age of 4.9 months (0-120 months) were included. Male female ratio was 21:23 and 9 (20.5%) of the patients had isolated EA. Mean gestational age was 36.97 ± 3.6 weeks (27-42 weeks) and mean birth weight was 2540 ± 694 grams (1200-3500 g). Time of repair and oral intake were less than one month in cases of TEF and more than one month in isolated cases. The incidence of PI was 22.7%. The most common infections were wound infections (n=4 cases, %40), blood stream infections (n=3, %30), pneumonia (n=1), peritonitis (n=1) and urinary tract infections (n=1). Microbiologic evaluations revealed that 3 cases had *staphylococcus spp*, 2 cases had *streptococcus spp*, 2 cases had mixed bacteria. *Candida albicans*, *Rothia mucilaginosa* and *Acinetobacter baumannii* were isolated in one case each. Gram (+) bacteria were detected in 60% of cases. One of the cases was died bacteremia caused by *Rothia mucilaginosa*. There was no difference between isolated and cases with TEF related with incidence and risk of PI for all parameters (p>0.05). In conclusion, PI can be seen in 22% of cases after EA repair. The incidence and risk of PI is found similar in both isolated and cases with TEF.

[P067] Growth impairment in esophageal atresia patients improves at school age

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Introduction: Esophageal atresia (EA) patients are at risk for respiratory and gastro-intestinal morbidity. Previous studies reported impaired growth in EA patients, but long-term follow-up data on growth are scarce.

Aim: To evaluate growth longitudinally from infancy up till school age.

Method: EA patients, born 1999-2013, who participated in a longitudinal follow-up program were included. Children with genetic syndromes known with associated growth disorders were excluded. Growth status and possible determinants were retrieved until 12-2015. Standard deviation scores (SDS) for height-for-age (HFA), weight-for-height (WFH) and distance-to-target-height (DTH) were calculated for each routine outpatient visit at 0.5, 1, 2, 5, 8 and 12 years of age. Linear mixed models were used to evaluate explanatory variables for growth.

Results: We included 126/155 children (61.9% male, 31.7% prematures, 19.8% small for gestational age). Anastomotic stricturing occurred in 59.5%(n=75; median of 2 (range 1-15) dilatations) and fundoplication surgery was performed in 23.8%(n=30; median age of 175 (range 54-1343) days). SDS-HFA was below the population norm in the first 8 years and improved over the years. After correction for target height (SDS-DTH) scores improved. SDS-WFH was below the norm in the first 5 years and normalized at 12 years. Stunting (SDS-HFA \leq -2) was present in 11/8/2% of the children aged 0.5-1 years, 2-5 years, and 8-12 years, respectively. Wasting (SDS-WFH \leq -2) was present in 11/14/2%, respectively. Risk factors for growth impairment were fundoplication surgery and low birth weight.

Conclusion: Impaired growth in EA patients was present during the first years of life, but normalized at 12 years. Further large longitudinal cohort studies are needed to evaluate if catch-up growth into adolescence continues. After fundoplication surgery lower scores were reported for all growth parameters. Earlier involvement of a dietician, who closely monitors growth and starts nutritional intervention when needed, should be considered in these patients.

Table: Biometric results per age group

Age group (n)	SDS-HFA; mean (SE)	p-value	SDS-DTH; mean (SE)	p-value	SDS-WFH; mean (SE)	p-value
0.5 year (126)	-0.480 (0.089)	<0.001	-0.291 (0.101)	0.005	-0.169 (0.099)	0.091
1 year (126)	-0.461 (0.091)	<0.001	-0.289 (0.098)	0.004	-0.521 (0.094)	<0.001
2 years (123)	-0.321 (0.094)	0.001	-0.144 (0.100)	0.151	-0.642 (0.095)	<0.001
5 years (97)	-0.414 (0.092)	<0.001	-0.275 (0.097)	0.006	-0.228 (0.094)	0.017

8 years (66)	-0.315 (0.089)	0.001	-0.179 (0.092)	0.055	-0.167 (0.109)	0.127
12 years (32)	-0.179 (0.132)	0.180	-0.076 (0.134)	0.572	0.036 (0.146)	0.806

The estimated marginal means, based on linear mixed model analysis with adjustment for center and age, were used to compare the mean SDS with the norm in the general population (SDS = 0).

[P068] One-step simple traction or staged traction-and-growth repair: impact on early post-operative outcome in long-gap esophageal atresia

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Aim: Many techniques were described in order to preserve native esophagus in long-gap esophageal atresia (LGEA). Aim of this study is to assess the impact of esophageal one-step primary surgery versus staged traction-and-growth techniques in LGEA.

Method: Patients with LGEA (gap wider than 3 vertebral bodies-vb and/or absence of distal TEF) were treated from 2010 to 2016. Traction-and-growth techniques were used to induce esophageal growth before the final esophageal anastomosis, when gap was wider than 3.5 vb. Patients were categorized on: STAGED traction-and-growth group and ONE-STEP simple-traction group.

Results: During the study period, 39 patients with LGEA were managed. Staged strategy was applied in 16 patients, who underwent 1 to 7 Kimura advancements in 12 cases and Foker Technique in 4 cases. Two of these patients finally underwent esophageal replacement. Primary immediate or delayed anastomosis was achieved in 23 patients using one-step strategy.

	STAGED	ONE-STEP	p
GA (weeks)	37 (33.25-38)	37 (33.75-38.25)	0.83
BW (grams)	2355(1770-2818)	2563 (1945-2883)	0.50
Type I-II vs III-IV	7/9	13/9	0.5118
Referred	12	4	0.0007
Esophagostomy	12	2	<0.0001
Gap (vb) (first measurement)	4 (3.5-6)	3 (1.8-3.5)	0.0008
GAP (vb) at surgery	1 (0.5-2)	2.75 (1.375-3.125)	0.0041
Age at surgery	254 (179-436)	62 (26-130)	<0.0001
Dilatation 3 months	0 (0-2)	1,5 (1-3)	0.015
Dilatation 6 months	1 (0.75-2)	1(0-2)	0.56
Dilatation 12 months	0.5 (0-3.2)	0 (0-3.2)	0.77

Conclusion: Data confirm that preservation of native esophagus is achievable also in LGEA and should be considered the main goal in all patients, including those referred with esophagostomy and extremely LGEA. Moreover, esophageal traction-and-growth procedure may enhance the possibilities to succeed in direct esophageal anastomosis, reducing early anastomotic stricture and the need for endoscopic dilations. Consequently this approach seems to be a feasible, effective and safe option in all cases of LGEA, not only as rescue strategy.

[P069] Is a selective approach to preoperative echocardiography in oesophageal atresia the future standard of care?

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Introduction: Oesophageal atresia (OA) is associated with cardiac defects that may have implications for perioperative care. Gold standard for preoperative care includes echocardiography, to identify major and moderate cardiac defects, including right-sided aortic arch (RAA) or double aortic arch (DAA). Nasr *et al* (2010) challenged this dogma and proposed selective preoperative echocardiography, albeit based on a small sample. Whilst local and international practice does not reflect its adoption, a selective approach may enhance efficiency of OA preoperative care.

Aim: We aim to assess selective preoperative echocardiography for pragmatism and accuracy in a larger cohort, to inform change in local practice.

Method: Single-centre retrospective review was performed of neonates with OA over 6-year period (2010-2015). Data recorded include preoperative clinical examination, chest x-ray, echocardiography. Endpoints were cardiovascular (cyanosis, murmur, tachycardia, abnormal 4-limb blood pressure, shock), respiratory (tachypnoea, intercostal recession, desaturations), radiological (cardiomegaly, abnormal pulmonary vasculature), echocardiography (major, moderate, minor cardiac defects). Performance of candidate-selective echocardiography strategies were assessed using sensitivity, specificity, positive predictive value (PPV), negative predictive value (NPV).

Conclusion: 115 neonates with OA were identified. All underwent preoperative echocardiography. 49/115 (43%) had cardiac defects (9/115 major, 4/115 moderate, 36/115 minor). Performance of candidate-selective strategies is summarised in Table 1. Interestingly, inclusion of chest x-ray findings did not enhance performance. However, further rationalisation of strategy to absence of murmur, cyanosis, and abnormal respiratory examination enhanced performance. Nasr *et al* concluded that normal clinical and radiological examination predicted absence of significant cardiac abnormalities in 100% of cases. In our experience, this missed 1 major cardiac defect – a RAA. Whilst missed asymptomatic RAA is a potential shortfall of selective strategies, RAAs are also missed with a non-selective echocardiogram strategy. Any of these selective strategies would have reduced echocardiograms performed by 22%, and reduced ‘number needed to scan’ for major and moderate cardiac defects from 9 to 7.

Table 1. Performance of selective strategies (with 95% confidence interval [CI]) based on clinical and radiological findings in detecting major (including RAA and DAA) and moderate cardiac defects

	Sensitivity (CI 95%)	Specificity (CI 95%)	PPV (CI 95%)	NPV (CI 95%)
Clinical + chest x-ray*	92% (0.62-1.00)	25% (0.17-0.34)	13% (0.07-0.23)	96% (0.78-1.00)
Clinical exam only	92% (0.62-1.00)	25% (0.18-0.35)	14% (0.08-0.23)	96% (0.79-1.00)
Murmur + cyanosis + respiratory	92% (0.60-1.00)	28% (0.20-0.38)	13% (0.07-0.22)	97% (0.81-1.00)

*Strategy proposed by Nasr *et al.* (2010), which had no cases of RAA or DAA

[P070] Rare variants of esophageal atresia: a case series

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Introduction: Rare variants of esophageal atresia (EA) exist in addition to the most commonly encountered forms.

Aim: This study aims to present a case series involving rarely seen variants.

Method: A retrospective evaluation of a 6-year period was done and Kluth's classification was used. Among 53 patients who underwent EA surgery, five had rare variants. Four were males and one was a female. The mean gestational age was 37 (29-40) weeks and the mean birth weight was 2042 (1310-3200) grams. The female patient who underwent a planned delayed primary repair surgery for isolated atresia, a 2.5-cm long solid cord was found to be connecting the two esophageal ends (Type II₃). A primary repair was done. Esophageal continuity was not disrupted in one patient having a high tracheal fistula and an oblique intraluminal membrane (Type IV₅). The membrane was excised with repair of esophageal wall and fistula. He died on the 10th postoperative day due to a major cardiac anomaly. The distal fistula was at the thoracic inlet level and the proximal end was extending about 2 cm below the distal fistula (overlapping ends) in one patient (Type IIIb₄). The esophageal ends had to be "resected" for a successful anastomosis. There were two patients with right main bronchus fistulae (Type VIb₁). One of them underwent primary repair. The other underwent colonic transposition surgery after an initial unsuccessful Foker approximation. The long-term follow-up is uneventful in the four surviving patients with a mean of 39 (30-48) months.

Conclusion: Studies concerning rare variants of esophageal atresia are limited and most are in the form of case reports. Interestingly, rare forms were present in one patient out of 10 in the presented series. Esophageal atresia is the index case for pediatric surgical practice. Intraoperative surprises do happen. Alternative "tailor-made" surgical approaches should be applied whenever necessary.

[P071] A novel technique for long gap esophageal atresia surgery: colonic transposition without cervical esophagostomy

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Introduction: An initial cervical esophagostomy is done before colonic replacement surgery in long gap esophageal atresia (EA) patients. Esophagostomy may itself cause complications before and after the definitive surgery.

Aim: Presentation of a clinical experience on esophageal colonic replacement surgery without cervical esophagostomy.

Methods: A retrospective analysis for the years 2010-2016 was done. Among the four male and one female patients, four had isolated EA and one had EA with proximal fistula. Their mean birth weight was 2200 (1420-3530) g. After an initial Stamm gastrostomy, the definitive surgery was done at a mean of 5.5 (3-10) months with a mean weight of 6 (4.5-8) kg. Through a right thoracotomy incision, the proximal pouch was dissected free. An exploration revealed no distal esophageal pouch in four. A left colonic segment in three and transverse colonic segment in the two were prepared and brought up to chest via transhiatal route in an isoperistaltic manner. The proximal esophagocolonic anastomosis was done within the thorax. The distal gastric anastomosis was done behind the stomach in four and the distal stump was used in one. The early postoperative course was uneventful. A proximal anastomotic stricture needed dilatations in one patient. The mean follow up period is 24 (1-65) months. The first three patients are fully and the two recently operated ones partially fed orally by age-appropriate diet.

Conclusion: Protection of proximal esophageal pouch intact eliminated the need of working in the cervical region. The proximal anastomosis was kept within the thorax preventing any compressive force that might act on esophagus at the level of thoracic inlet with the potential to impair vascularity. Moreover, the patients had the chance of using a much longer segment of their native esophagus. Long term clinical follow-up in three showed a quite satisfactory result in terms of swallowing and feeding.

[P072] Transhiatal isoperistaltic colonic interposition for esophageal replacement in long gap esophageal atresia

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Introduction: Isoperistaltic transhiatal colonic interposition (THCI) is a relatively less used method for esophageal replacement in children

Aim: Evaluation of the results with THCI.

Method: Records for the last ten years revealed 12 children who underwent THCI for esophageal atresia (EA). The primary diagnosis was isolated EA in seven, EA with distal fistula in four and EA with proximal fistula in one. Seven were males and five females. The mean age at operation was 13.84 ± 9 (4-18) months. The replacement was done after an esophagostomy in seven and without an esophagostomy in five. A thoracotomy at the time of interposition was done in nine (75%). Left colon was used in eight (67%) patients with left colonic artery pedicle and right colon (33%) was used in four with middle colic artery pedicle. The colon was interposed through the transhiatal route in an isoperistaltic manner in all. The proximal anastomosis was done in single layer by absorbable sutures and the cologastric anastomosis was done retrogastrically to the antrum in all but one. A pyloroplasty was added in all. There were two early postoperative deaths because of nonsurgical complications. Serial balloon dilatations were done for the stricture development in the proximal anastomosis in two patients with resultant success in both. The mean follow-up period is 43 ± 35 (2-109) months. All patients can be fed orally by age-appropriate diet; two recent cases are fed via gastrostomy as well. Colonic redundancy, clinically significant reflux or chronic respiratory difficulty was not observed in any patients on the long term.

Conclusion: Esophageal atresia was the most common indication for THCI in the presented series. The long-term functional results of are quite satisfactory. As there is no "perfect" replacement surgery, this is a favorable method and is a viable option for esophageal replacement surgery.

[P073] Prospective longitudinal evaluation of long-gap esophageal atresia patient at 2 years: A long-lasting morbidity

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Aim of the study: No data are available on longitudinal evaluation of long gap esophageal atresia (LGEA) patients. Aim of present study was to evaluate 1 and 2-year outcomes of patients treated for LGEA

Methods: a prospective, longitudinal multi-specialistic evaluation of all LGEA patients treated between 2005 and 2014, was performed at 1 and 2 years of age. LGEA was defined as gap wider than 3 vertebral bodies. Outcomes included auxological assessment, esophago-gastric morbidity, rate of respiratory tract infections. Paired t-test and Fisher's exact test were used; $p < 0.05$ was considered significant.

Main Results: During the study period 48 patients, with a minimum of 1-year follow up, were studied. Table summarizes main results.

Conclusions: LGEA patients continue to experience morbidity at 1 and 2-year follow-up, although progressively decreasing. Our data indicates the need for close multi-specialistic monitoring of those complex patients, well beyond surgical correction of esophageal atresia.

	1st year 48 pts	2nd year 42 pts	p
BMI; median (IQR)	14,8 (13,5-15,7)	15,2 (14-16)	0.14
N° dilatations/year; median (IQR)	3 (0,25-5,8)	1 (0-4)	0.05
GER; n° (%)	40 (95)	32 (76)	0.02
Nissen fundoplication; n° (%)	14 (29)	11 (26)	0.8
Oral aversion; n° (%)	30 (62)	17 (40)	0.04
Lower respiratory tract infections/year; n° (%)	28 (54)	24 (57)	0.8
Deaths, n° (%)	0	0	1.0

[P074] Prenatal diagnosis of esophageal atresia: a prospective study

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Aim: Prenatal suspicious of esophageal atresia (EA) with or without tracheo-esophageal fistula is rare and diagnosis possible only in a minority of cases. Polyhydramnios and small/absent stomach bubble are considered non-specific/indirect signs of potential EA. Aim of our study is to assess the role of upper pouch sign (UPS) in prenatal diagnosis of EA.

Method: Prospective collected data was performed. We evaluate all prenatal ultrasound scans of fetuses referred for suspected EA between January 2009 and December 2015. We analyzed the presence of small/absent stomach bubble, polyhydramnios and the presence of UPS. Fisher exact test was used as appropriate.

Results: Forty patients were referred for suspected EA during the study period. There were 3 terminations of pregnancy, and 3 expectant women were lost before delivery. Thirty-four fetuses were prospectively followed up during pregnancy and reevaluated after birth. Table summarized main results.

Conclusion: Our data suggest that repeated prenatal evaluation has crucial importance for prenatal diagnosis of EA. Polyhydramnios and UPS are frequently detected in EA patients confirmed postnatally. Temporary detection of polyhydramnios and/or absent/small stomach has no role in prenatally suspicious of EA, while the persistence of those signs are statistically encountered in postnatally confirmed EA patients. Strong prenatal suspicious of EA seems to correlate with high prevalence of LGEA and/or syndromic association.

	Postnatal Confirmed EA 12 pts	Postnatal Not EA 22 pts	p
Temporary polyhydramnios detection; n(%)	12 (100)	16 (73)	0.07
Persistent polyhydramnios; n (%)	12 (100)	6 (27)	0.0001
Temporary absent/small stomach; n (%)	12 (100)	22 (100)	1.0
Persistent absent/small stomach; n (%)	11 (91)	1 (5)	0.0001
UPS; n (%)	8 (67)	0 (0)	0.0001
POSTNATAL OUTCOMES			
LGEA; n (%)	9 (75)	-	
Syndromic patients; n (%)	8 (67)	3 (14)	0.005

Of the 34 infants followed after birth, prenatally we observed 18 polyhydramnios, 12 small/absent stomach bubbles, while UPS was evident in 8 cases. At birth, 12 newborn had diagnosis of EA (37%), 8 of which with pouch evidence prenatally. UPS was the only significant marker of EA (p 0.0002) with sensitivity 0.8 (CI95% 0.28 to 0.99) and specificity 1 (CI 95% 0.85 to 1). Polyhydramnios and small/absent stomach bubble were no significantly associated with postnatal diagnosis of EA (p 1.0 and p 0.3 respectively). Long-gap esophageal atresia were defined after birth in 9 children: 5 type I/II and 4 type III. The other 3 child were AE type III syndromic.

[P075] Growth impairment in patients born with esophageal atresia in rotterdam and oslo: 0-2 year olds

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Introduction: Long-term follow-up data on growth in esophageal atresia (EA) patients is scarce.

Aim: To evaluate growth and morbidity in Dutch and Norwegian EA patients up till the age of 2 years.

Method: EA patients, born 2011-2015, who attended follow-up in one of 2 participating academic centers were included. Children with H-fistulas or genetic syndromes known to be associated with growth disorders were excluded. Growth status and demographic data were retrieved at 0.5, 1 and 2 years of age. Standard deviation scores (SDS) for height-for-age (HFA), distance-to-target-height (DTH), weight-for-age (WFA), and weight-for-height (WFH) were calculated. Differences in SDS and demographics in relation to growth between both centers were evaluated.

Results: We included 87 children (40/47 Dutch/Norwegian, 59.8% male, 37.9% prematures, 14.9% small for gestational age, 93.1% Gross type C, initial hospitalization of median 21.5 (range 7-169)days). Anastomotic stricturing occurred in 66.7% (median of 3.5 (range 1-27)dilatations; first dilatation at median age of 66 (range 21-690)days) and fundoplication surgery was performed in 14.9% (median age of 180 (range 43-750)days). Thoracoscopic EA repair was not performed in Norway (62.5% in the Netherlands;p<0.001). SDS-HFA was below the population norm at all assessments and improved after correction for target height (SDS-DTH;Table). SDS-WFA and SDS-WFH were below the norm at 2 and 1 year, respectively. Stunting(SDS-HFA≤-2) was present in 24.0/9.5/9.5% of children aged 0.5/1/2 years, respectively. Wasting(SDS-WFH≤-2) was present in 4.0/8.1/6.3%, respectively. Norwegian children had higher SDS-WFH at 2-years than Dutch children (mean(SE) of 0.022(0.154) and -0.563(0.176);p=0.014). Other parameters did not differ significantly between centers.

Conclusion: Impaired growth in 2 cohorts of EA patients was present during the first 2 years of life. As a good nutritional status is crucial for normal brain- and immune system development, these results warrant early dietary management. Longitudinal multicenter studies are needed to study possible determinants of growth impairment.

Table: Biometric results per age group

Age group (n)	SDS-HFA; mean (SE)	p-value	SDS-DTH; mean (SE)	p-value	SDS-WFA; mean (SE)	p-value	SDS-WFH; mean (SE)	p-value
0.5 yr (50)	-0.887 (0.152)	<0.001	-0.659 (0.152)	<0.001	-0.976 (0.167)	<0.001	-0.282 (0.122)	0.024
1 yr (74)	-0.647 (0.124)	<0.001	-0.439 (0.131)	0.001	-0.849 (0.121)	<0.001	-0.366 (0.103)	0.001

2 yr (63)	-0.351 (0.135)	0.011	-0.110 (0.134)	0.416	-0.614 (0.132)	<0.00 1	-0.227 (0.122)	0.066
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The estimated marginal means, based on linear mixed model analysis with adjustment for center and age, were used to compare the mean SDS with the norm in the general population (SDS = 0).

[P076] Molecular profiling of eosinophilic esophagitis in children with esophageal atresia

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Introduction: Recently, the high prevalence of eosinophilic esophagitis (EoE) in esophageal atresia (EA) patients has been reported.

Aim: The aims of this study were to characterize the EoE transcriptome in EA patients, to compare the EoE transcriptome in EoE patients with and without EA and to identify any genes that could potentially predispose EA patients to EoE

Methods: This was a retrospective review of epidemiological data and esophageal biopsy slides collected at Sydney Children's Hospital (SCH) between 2000 to 2014. Patients were divided into 4 major cohorts: Healthy control (NL), EA patients with (EA+EoE+) and without EoE (EA+EoE-) and EoE patients without EA (EA-EoE+). The EoE+EA- and EA+EoE+ groups were also analyzed before and after EoE treatment. Molecular signature acquired by EoE diagnostic panel (EDP) was quantified by a 96-gene RNA expression signature array from formalin fixed esophageal biopsy sections at Cincinnati Children's Hospital Medical Centre (CCHMC),

Results: Out of a cohort of 110 EA patients treated at SCH, 20 patients (18%) were diagnosed with EoE. When compared to the reported prevalence of EoE in 1 in 2000 of the general pediatric population, there was a 364-fold increased risk of EoE development in EA patients (OR 444; 95% CI 59-3347; p <0.001). EA patients with EoE were 60% male, 70% atopic, and were diagnosed with EoE at median age of 26 months (35 +/- 27). The EDP molecular signature analyses was performed in 10 patients in each of the four groups, and EoE scoring did not reveal any major difference between EoE+EA+ and EoE+EA- cohorts (Table 1). Dysregulated Th2 genes were identified in EA+EoE+ cohort.

Conclusion: EoE patients with EA have a similar patho-molecular profile and remission characteristics post treatment compared with the EoE cohort without EA. Dysregulated Th2 genes may explain the higher EoE prevalence within the EA+EoE+ cohort.

Table 1: EDP Analysis

EDP Molecular Th2 Signature	EoE+EA+	EoE+EA-	EA+EoE-	NL	P Value*
EoE Mean Score at baseline time of diagnosis of EoE	543 (502-584, 95% CI)	532 (491-573, 95% CI)	632 (603-662, 95% CI)	655 (636-674, 95% CI)	NS
EoE Mean Score post treatment of EoE	662 (648-676, 95% CI)	654 (631-671, 95% CI)	N/A	N/A	NS

CCL26 mRNA level post treatment of EoE
85%± 27% 91% ± 12% N/A N/A NS
(mean ± SD (mean ± SD %)).

*Non-significant, EoE+EA+ vs. EoE+EA-, two-tailed student T-test, p < 0.05 deemed as significant

[P077] Evaluation of motor function of the esophagus with high-resolution manometry in adolescents with esophageal atresia.

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Introduction: The incidence of esophageal atresia is 1: 3000 live births, type 3 being the most common. The condition is frequently associated with motility disorders.

Objective: To evaluate segmental esophageal motility disorders and gastroesophageal reflux in patients with history of esophageal atresia.

Methods: Population: Children over 12 years of age with data of esophageal atresia evaluated at two hospitals, without jamming or dilations in the previous six months. The methods included high resolution esophageal manometry, conventional esophageal manometry, pH-impedance and video endoscopy with biopsy.

Results: The study included 10 children (mean age 15.4 years, range 12 to 18). Nine had presented esophageal atresia type III, and one type I. At endoscopy 30% showed C esophagitis (Los Angeles). Normal pH-impedanciometry was recorded in 70%. The conventional manometry showed:

Lower esophageal sphincter: 100% presented normal basal pressure, and 90% normal relaxation.

Esophageal body: 90% showed upper esophagus with half aperistalsis; lower esophagus: 3 children presented peristalsis in 30% of swallows. One child showed peristalsis all along the esophagus in 30% of swallows.

High resolution manometry:

Gastro-esophageal junction: the lower esophageal sphincter pressure was normal in all (100%), the relaxation was normal in 90%, and the integrated relaxation pressure was normal in 100%.

Esophagus: Lower esophagus: the peristalsis was present in 30-40% of swallows in 50% of children. Middle-lower esophagus: peristalsis present in 10%. Middle-upper esophagus: aperistalsis in 100% of the patients. Upper esophagus with peristalsis in 10%.

Integral pressure distal pathological contractility: abnormal in 100% (mean 68 mmHg/cm/second).

Upper esophageal sphincter, normal 90%, mean 104 mmHg. Pharyngeal pressure and coordination: normal in 100% .

Conclusions: Esophageal high-resolution manometry allowed evaluation of esophageal motility disorders and segmental esophagus fully and more accurately. The ph-impedance and video endoscopy with biopsies prove useful tools to assess gastroesophageal reflux in these paucisymptomatic children.

[P078] Our experience with management of H-type Fistula

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Background: H-type of trachea-oesophageal fistula (TOF) is rare & forms 4% of OA & TOF. (1:100,000 live births) Delay in diagnosis is common, often mistaken for gastro-oesophageal reflux. We report our experience with H-type(N-type) of fistula over a period of 20 years (1988 to 2008).

Method: 8 children (M:F – 6:2) aged between, 1 month to 6-years were treated. coughing & choking after liquids, was mistaken in 4 cases as G-O reflux. One had fundoplication & referred at 15 months. Another, presented with abdominal distension, increased during crying & mistaken for Hirschprung's disease (HD) & referred at 1-year. Unlike HD, this infant was passing out lot of flatus, Catheter test & oral nonionic contrast study clinched the diagnosis. A 6-year old was investigated by Ba enema to rule out HD. Two had associated problems, -cyanotic spells & was treated with morphine. Another had Leukemia. Clinical observation, Contrast study, in prone head down position & dye injection, Bronchoscopy & introducing a ureteric catheter or Fogarty catheter, helped in diagnosis. Identification of the fistula, with due care to recurrent laryngeal nerves through a right cervical approach & repair of fistula with 5/0 prolene & muscle interposition, was performed in all cases.

Results: Postoperative ventilation for a day or two was needed in 5 of them. All recovered well. 2 children died: one with cyanotic spells, & another with leukemia died. The rest are doing well.

Conclusion: N-type fistulas are rare. A high index of suspicion is essential to diagnose early. Although some have recommended MRI & CT scan, properly done non-ionic contrast study & bronchoscopy are the main modalities, we prefer. The right cervical approach is our choice, although thoracotomy or thoracoscopic repair may be needed rarely, in fistulas below T2 level. This type poses diagnostic challenges which leads to delay in management.

[P079] Uncommon Foregut anomalies in Neonates

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Introduction: Uncommon Foregut anomalies (UFG), in 4 infants, lead to diagnostic & therapeutic challenges. The pathology, associated anomalies & challenges in management are discussed.

Method: 3neonates had large diaphragmatic defects. Another had surgery for TOF. She had Bilateral VUR & High Vaginal atresia. In two infants while repairing the defect, several episodes of hypoxia & difficulty in maintaining the airway occurred. Case: 1. the contents were reduced, the left lobe of liver folded inside the chest & was delivered. Stomach could not be brought down. Inspection showed, the oesophagus was absent or very short. The dorsal & ventral pancreas was, attached to the right & left side of duodenum in the chest. Stomach was distending with air with ventilation. The bile ducts opened below the diaphragm. The defect was closed with tranverse abdominis muscle flap. To rule out a H-type fistula, bronchoscopy, rigid & flexible a few times did not show any fistula from trachea, Upper GI endoscopy showed, multiple areas of air leaks indicated multiple peripheral bronchiolar communication with stomach. He needed parenteral nutrition & ventilation. Barium meal showed, intrathoracic stomach & congenital absence of oesophagus or a mini oesophagus. Attempts to open the chest, led to respiratory compromise. Case 2, had tracheobronchial cleft type 4. This was diagnosed on table, the ETT was pushed into the right bronchus, & ventilated. The large diaphragmatic repair could not be repaired due to episodes of desaturation & the child died at 6months. Case 3, had tachypnea & needed ventilation. Bile was coming out in ETT & the tracheobiliary fistula, diagnosed by MRI, was repaired successfully. Case:4, The child who had repair of TOF, the lower oesophagus, remained narrow, in spite of several dilations. There was no resistance to dilations. Discussion with a visiting thoracic surgeon felt, it looks like, achalasia of the lower segment & a long myotomy & loose nissen's fundoplication, settled her problem.

Conclusion: Severe anomalies of foregut, may be associated with TOF or diaphragmatic hernias. Inability to maintain the airway, should alert the anesthetist & surgeon to look for major anomalies of the tracheobronchial tree.

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Introduction: The Lancet Commission on Global Surgery in 2015 concluded that the poor world suffers a serious shortfall in available surgery, leading to significant avoidable disability adjusted life years. (DALYs) At the same time the rich world has increasing numbers of medical specialists who often have small exposure to index cases. Tracheo-oesophageal fistula, particularly the long gap variant, is one such challenging index case, difficult to treat in poor countries, but requiring considerable experience to do well even in rich countries. We present one such case. The support of ROMAC in this and other cases is gratefully acknowledged.

Case report : TB was transferred at the age of 12 weeks from the Solomon Islands with a diagnosis of a long gap pure oesophageal atresia. There was no evidence of other VACTERL associations. The course had been complicated by pneumonia and nutritional insufficiency relating to gastrostomy feed problems. Weight on arrival was 2.3 kg.

Progress: After nutritional stabilisation, a Foker procedure was initiated. Progress was complicated by a mediastinal leak requiring drainage, and precluding straightforward Foker style anastomosis. Mobilisation of the stomach with proximal advancement was performed. The recovery was complicated by repeated stricturing at the anastomosis managed with multiple dilatations, and finally intra-strictural steroid injection. Total length of stay in Australia was 8 months, for a potential cost of ~\$300,000. Follow up in the Solomon Islands in 2016 by the senior author reveals good weight gain, no stricturing, and a healthy oesophagus.

Discussion: Long gap oesophageal atresia presents technical problems even in the rich world. Our choice of the Foker procedure as a first choice operation was dictated by the need for a procedure with the lowest possible likelihood of long term complications for a child who is to live his life in a remote area. Failure to achieve a Foker style anastomosis has highlighted the importance of follow up. Follow up for children with major malformations presents a challenge in poor countries. One of the important factors in care of this child has been the possibility of follow up locally both by a local paediatrician and by the Australian operating surgeon.

Conclusion: Co-operative health care between nations allows skills development on both sides, and is likely to be of benefit to both. Private initiatives like ROMAC are a great part of that. Young TB is likely to live a productive life with little or no disability in his country of birth.



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