Gastro-oesophageal reflux in case of esophageal atresia: surgical options

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Gastro-oesphageal reflux (GER) is one of the main problems after oesophageal atresia (OA) repair. Fundoplication is necessary in many of these children, probably around thirty percent of all the patients. There are many problems concerning this procedure. When is it necessary? what is the best time to perform this procedure (during the growth) and what is the best procedure ?. Finally, the last point will be: witch is the best strategy in case of failure?

GER is very usual after OA repair and many team used medical treatment during the first year of life, expecting amelioration after first walking. Interestingly, some study find that the number of children with significant GER associated with OA is increasing from 6 months to 1 year after primary repair (1).
During the first months of life, indications for fundoplication are mostly apnea and cyanotic spells. For these cases, correct evaluation is mandatory to exclude major tracheomalacia or arterial compression. Sometimes, appreciation of the responsibility of GER in these episodes is difficult and aortopexy can be performed before, after or at the same time of the fundoplication. The need for multiple balloon dilatation of the anastomosis can be an indirect sign of bad control of GER. In our team, we propose a Nissen procedure after failure of 3 dilatations.

In infant, indication for fundoplication is also a difficult decision. Two main important factors need to be discussed: first, the pulmonary status, and secondary the oesophagus motility. Pulmonary status must be appreciated during the first years. Recurrent aspirations lead to antireflux procedure in order to avoid important pulmonary destruction. The dysmotility of the oesophagus is well known in case of AO. Evaluation by simple manometry is not sufficient to appreciate the risk of dysfunction after fundoplication procedure. We know that during the growth, a better oesophageal function is noted.

Numerous type of antireflux procedure are available. The most commonly performed in the word is probably the Nissen procedure. In paediatric teams, this procedure is well known, easily performed with good results, by laparotomy or, better, by laparoscopy (3). However, there is some trouble with this procedure. The first is the risk of failure. During the previous 20 years, several publications state a percentage of failure from 20 to 30 % in OA (2). Recent publications are less pessimists, with a risk of failure not correlated with oesophageal atresia but with open surgery at first fundoplication and neurological impairment (4).

Collis Nissen could be an option in case of AO. Because this procedure authorizes a lengthening of the oesophagus, it is probably an interesting option (5). For long term follow up, the trouble is that the ascension of Z-line induces difficulty to evaluate GER and metaplasia.

Partial posterior and anterior wrap (6) are recommended by some team because they decrease the risk of post operative dysphagia. Some studies reported similar results in term of GER control in Thal or Nissen procedure. This fact is very difficult to establish. A randomized study is probably the only way in order to have an objective evaluation.

In case of failure of Nissen procedure or in some difficult cases of OA (as associated with diastema) a total oesogastric disconection (TED: Bianchi procedure) can be proposed (7). Long follow up is mandatory but efficiency is particularly good when the pulmonary function is altered by aspiration (severe persistent GER or swallowing aspiration after dysmotility post Nissen procedure)
For the long term, there is few study which indicate the necessity of long term follow up for these patients. The

risk of Barret oesophagus or the risk of oesophageal carcinoma is not fully established.. I f we consider these risks, Collis Nissen or partial gastroplasty must not be recommended. News studies are necessary to evaluate these problems.

1. Koivusalo A, Pakarinen MP, Rintala RJ.
The cumulative incidence of significant gastrooesophageal reflux in patients with oesophageal atresia with a distal fistula- a systematic clinical, Ph-metric, and endoscopic follow up study. J pediatr Surg 2007; 42:370-374.
2. Wheatley MJ, Coran AG, Wesley JR. Efficacy of the Nissen fundoplication in the management of gastroesophageal reflux following oesophageal atresia repair. J Pediatr Surg 1993; 28:53-55.
3. Esposito C, Langer JC, Scaarschmidt K and Co. Laparoscopic antireflux procedures in the management of gastroesophageal reflux following esophageal atresia repair. J Pediatr Gastroenterol Nutr 2005; 40 (3): 349-51.
4. Pacilli M, Eaton S, Maritsi D and Co Factors predicting failure of redo Nissen fundoplication in children.
Pediatr Surg Int 2007; 23: 499-503.
5. Kawahara H, Imura K, Yagi M and Co Collis Nissen procedure in patients with esophageal atresia: long term evaluation. Word J Surg 2002; 26: 1222-7.
6. Snyder BL, Ramachandran V, Kennedy AP and Co. Efficacity of partial wrap fundoplication for gastroesophageal reflux after repair of esophageal atresia. J Pediatr Surg 1997; 32 (7): 1089-92.
7. de lagausie P, Bonnard A, Schultz A and Co Reflux in esophageal atresia, tracheoesophageal cleft and esophagocoloplasty: Bianchi's procedure as an alternative approach. J Pediatr Surg. 2005 Apr;40(4):666-9.
8. Madre C, Serhal L, Michaud L and Co Prolonged enteral feeding is often required to ovoid long-term nutritional and metabolic complications after esophagogastric dissociation. J Pediatr Gastroenterol Nutr. 2010 Mar;50(3):280-6.

Extra-digestive complications

Abnormal pharyngo-esophageal function in infants and young children: diagnosis with high resolution manometry

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Dysphagia is common in the paediatric population and hinders the provision of adequate nutrition to infants and children within a wide range of morbidity 1,2. The clinical impact of dysphagia is significant, but the current understanding of pathophysiology is poor and the therapeutic interventions available are limited. Before treatment and management of paediatric dysphagia can commence, there is a need for a thorough assessment of swallowing function. The infant and the adult swallow are quite different and warrant individual discussions in their own right. One cannot assume that assessment techniques that are suitable for adults will also be suitable for babies, infants and children 3. There are also technical considerations of catheter size, sensor spacing and response time which preclude extrapolation of adult findings to infants and children.

As in any observational method, the quality of information from a clinical examination depends mainly on the examiner's perceptual skills. If applied properly, instrumental assessment has the potential to document oropharyngeal function objectively. Many different functional tests are available to assess oropharyngeal function during swallowing, each with advantages and disadvantages 3,4. Children with dysphagia are typically investigated by videofluoroscopic swallow study 5,6. Although helpful clinically, the limitations of the technique are well known and relate to radiological exposure and the lack of measurable objective parameters. Currently the outcomes of a videofluoroscopic swallow study are largely approximate and descriptive.

To make a videofluoroscopy diagnostically more useful, it can be combined with a novel high resolution manometry (HRM) technique to assess pressure changes in the pharynx and upper oesophageal sphincter during swallowing. The integration of both manometric and videofluoroscopic data, called high resolution videomanometry, represents a significant advance over a standard videofluoroscopic swallow study for the assessment of dysphagia. This is because it (1) allows quantification of the pharyngeal movements in relation to bolus passage and to the opening of the upper oesophageal sphincter (UOS) and (2) allows determining the pathological basis for swallowing disorders precisely in terms of abnormal pressure flow patterning across the pharyngo-oesophageal segment.

High resolution manometry (HRM) as individual technique, provides the ability to measure pressure across the length of the pharyngo-oesphageal segment using a chain of closely spaced point pressure sensors at high spatial resolution. This enables highly accurate spatiotemporal interpolation of dynamic pressure changes caused by luminal closure following contraction.

Recently, HRM has become more accessible and popular to study dysphagia. High-resolution manometry can be performed using miniature, multichannel, water-perfused catheters 7 or solid-state catheters 8. Both perfused and solid-state manometry acquisition systems are commercially available. Pressure dynamics of the pharynx and UOS are difficult to obtain with conventional manometry due to the rapid response time needed to measure the physiology of the striated muscle of the pharynx. Also, recordings with a single to a few sideholes are difficult to interpret because of the movement of the UOS during swallowing. Solid state HRM meets both requirements of rapid response rate and insensitivity for axial movement 9 and is therefore promising in the study of pharyngo-oesophageal motility.

Manometry is particularly helpful in instances where videofluoroscopic imaging demonstrates impaired or absent transit from the hypopharynx to the oesophagus 9-12. When this occurs, it may be difficult to distinguish between failure of the UOS to relax or its inability to open. In this case, failure of the upper oesophageal sphincter relaxation can be documented by manometry which can provide the differential diagnosis between both causes of dysphagia 9,11-13 .

HRM has been successfully applied to studies of the pharynx and upper esophageal sphincter in adults, either through perfusion 11 or through solid-state technology 9,14.

So far and mainly because of ethics, the biomechanics of oropharyngeal patterns in normal children have not been determined. 15. A series of manometrically confirmed UOS dysfunctions in children with VCFS have been reported recently 13 as well as the development of the pharyngeal phase of swallowing in healthy preterm infants as assessed with HRM 16. The outcome of these pediatric studies will be discussed during the talk.

When the likely underlying cause of pharyngeal dysphagia is considered in any given patient, Cook suggested four fundamental issues to consider during work-up 17. First, a correctable structural lesion should be identified if possible. Second, any underlying systemic condition that might be treatable in its own right should be identified. Third, the risk of aspiration should be established. Fourth, the mechanics of dysfunction should be determined as a precursor to swallow therapy 17. In the assessment of pediatric dysphagia, it is exactly the latter step is too often missing leading to the current trial and error in diagnosis experienced by clinicians. In order to overcome this lack of information on the biomechanics of the infant swallow, we started applying HRM for the study of paediatric dysphagia. We were clearly able to recognise a range of pharyngeal and UOS dysfunctions. The pathology varied from synchronous pharyngeal peristalsis to pharyngeal paralysis and from a normally relaxing UOS to a poorly relaxing UOS and a non-relaxing UOS (achalasia). In our opinion, HRM is an adequate diagnostic tool to distinguish the subtle differences between these different pathologies that may be treated by different means.

In summary, objective assessment should be the key point for investigation into the feeding and swallowing abilities of infants and children. HRM is a promising technique providing the ability to describe the (patho)-physiological mechanisms of dysphagia in pressure-flow mechanical terms. Current studies with HRM are characterizing the biomechanics of the infant swallow and will lead to better understanding of pharyngo-oesophageal dysmotility causing dysphagia. This type of objective diagnostics must continue to be developed in order to meet the specific needs of the pediatric population 18.

Reference List

(1) Rudolph CD, Link DT. Feeding disorders in infants and children. Pediatr Clin North Am 2002; 49(1):97-112, vi.
(2) Rommel N, Meyer AD, Feenstra L, Veereman-Wauters G. The complexity of feeding problems in 700 infants and young children presenting to a tertiary care institution. J Pediatr Gastroenterol Nutr 2003; 37(1):75-84.
(3) Rommel N. Assessment techniques appropriate for the use in babies and children. In: Murdoch B, Chicero J, editors. Dysphagia: Foundation Theory and Practise. London: Wiley Publishers, Ltd, 2006: 466-86.
(4) Arvedson JC. Assessment of pediatric dysphagia and feeding disorders: clinical and instrumental approaches. Dev Disabil Res Rev 2008; 14(2):118-27.
(5) Arvedson J, Lefton-Greif M. Videofluoroscopic swallow procedures in pediatrics. In: Arvedson J, Lefton-Greif M, editors. Pediatric videofluoroscopic swallow studies. San Antonio Texas: Communication Skill Builders, 1998: 72-116.
(6) Hiorns MP, Ryan MM. Current practice in paediatric videofluoroscopy. Pediatr Radiol 2006; 36(9):911-9.
(7) Omari T, Bakewell M, Fraser R, Malbert C, Davidson G, Dent J. Intraluminal micromanometry: an evaluation of the dynamic performance of micro-extrusions and sleeve sensors. Neurogastroenterol Motil 1996; 8(3):241-5.
(8) Pandolfino JE, Ghosh SK, Rice J, Clarke JO, Kwiatek MA, Kahrilas PJ. Classifying esophageal motility by pressure topography characteristics: a study of 400 patients and 75 controls. Am J Gastroenterol 2008; 103(1):27-37.
(9) Ghosh SK, Pandolfino JE, Zhang Q, Jarosz A, Kahrilas PJ. Deglutitive upper esophageal sphincter relaxation: a study of 75 volunteer subjects using solid-state high-resolution manometry. Am J Physiol Gastrointest Liver Physiol 2006; 291(3):G525-G531.
(10) Benson J, Tuchman D. Other diagnostic tests used for the evaluation of swallowing disorders. In: Tuchman D, R Walter., editors. Disorders of feeding and swallowing in infants and children. San Diego: Singular Publishing Group, Inc, 1994: 201-9.
(11) Williams R, Pal A, Brasseur J, Cook I. Space-time pressure structure of pharyngo-esophageal segment during swallowing. Am J Physiol 2001; 281:1290-300.
(12) Dejaeger E, Pelemans W, Ponette E, Joosten E. Mechanisms involved in postdeglutition retention in the elderly. Dysphagia 1997; 12(2):63-7.
(13) Rommel N, Davidson G, Cain T, Hebbard G, Omari T. Videomanometric evaluation of pharyngo-oesophageal dysmotility in children with velocardiofacial syndrome. J Pediatr Gastroenterol Nutr 2008; 46(1):87-91.
(14) Takasaki K, Umeki H, Enatsu K, Tanaka F, Sakihama N, Kumagami H et al. Investigation of Pharyngeal Swallowing Function Using High-Resolution Manometry. Laryngoscope 2008.
(15) Mathur NB, Banerjee S, Maria A, Bhatnagar V. Congenital cricopharyngeal achalasia. Indian Pediatr 2001; 38(7):783-8.
(16) Rommel N, Van Wijk M, Boets B, Hebbard G, Haslam R, Davidson GP et al. Development of pharyngo-oesophageal motility in healthy preterm infants. In review 2010.
(17) Cook IJ. Diagnostic evaluation of dysphagia. Nat Clin Pract Gastroenterol Hepatol 2008; 5(7):393-403.
(18) Rommel N, Omari T. The use of high resolution manometry for the assessment of swallowing in infants and young children. B-Ent 2008; 4:27-39.

Esophageal atresia : pulmonary outcome

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Children with esophageal atresia (EA) are now reaching their adulthood in large numbers. Many patients, however, continue having functional problems from their respiratory system after the initial postoperative period. There are now several series reporting long-term follow-up in adolescents or young adults after repair of EA (1-5) and recent reviews have been published (6, 7). The objectives of the present communication are as follows :
(a) to summarize the long-term pulmonary outcome of children with EA, in terms of symptoms and respiratory function
(b) to focus on the underlying mechanisms contributing to the occurrence of prolonged respiratory complications

Persisting respiratory symptoms are commonly observed in adolescents or adult patients. Around 40% of adolescents and adults report current respiratory symptoms (4). However, the prevalence of symptoms clearly decreased with age (2). Wheeze appears as the most frequent symptom beyond childhood. An obstructive defect is measured in 20 to 30% of subjects (4, 5). Airway hyperreactivity to histamine is increased in 40 to 78% of adolescents or adults, and is consistent with asthma in 15 to 26% of subjects (4, 5). Barking cough is still present in 40% of adults, and chronic cough is reported by 8 to 16% of adults (4, 6). A restrictive ventilatory defect can also be measured in up to 21% of adults, mainly related to thoracotomy-induced rib fusions (5). Although children wit EA are at higher risk of scoliosis, the general clinical course of spinal deformities is mild (8), and may not contribute significantly to restrictive pattern. Maximal exercise performance has not been studied in adults, but has been demonstrated impaired in children (9). As a whole, respiratory morbidity following EA is high, and the complication rate is comparable to that observed in the congenital diaphragmatic hernia (9). Complicated evolution is associated with long gap atresia (10). Respiratory symptom-related quality of life is significantly impaired in adults with EA (5).
Developmental abnormalities of tracheal structure and innervation may contribute to the persistence of respiratory symptoms beyond childhood. Congenital structural deformities, such deficiency of cartilage and increase in the length of the transverse muscle, are frequently observed, and are not restricted to the site of the fistula (11). This is associated to congenital abnormalities of the intrinsic nervous plexus (12). Experimental models demonstrated that these abnormalities are observed in the whole length of the respiratory tract (13). Persistent or worsening respiratory symptoms in childhood or adolescence are often associated to persistent tracheomalacia, potentially aggravated by a tracheal diverticula at the fistula repair site (14). Acquired bronchial inflammatory remodeling processes are unlikely to be responsible for persistent symptoms (4).

References:

1. Agrawal, L., C. S. Beardsmore, and U. M. MacFadyen. 1999. Respiratory function in childhood following repair of oesophageal atresia and tracheoesophageal fistula. Arch Dis Child 81(5):404-8.
2. Chetcuti, P., and P. D. Phelan. 1993. Respiratory morbidity after repair of oesophageal atresia and tracheo-oesophageal fistula. Arch Dis Child 68(2):167-70.
3. Little, D. C., F. J. Rescorla, J. L. Grosfeld, K. W. West, L. R. Scherer, and S. A. Engum. 2003. Long-term analysis of children with esophageal atresia and tracheoesophageal fistula. J Pediatr Surg 38(6):852-6.
4. Malmstrom, K., J. Lohi, H. Lindahl, A. Pelkonen, M. Kajosaari, S. Sarna, L. P. Malmberg, and M. J. Makela. 2008. Longitudinal follow-up of bronchial inflammation, respiratory symptoms, and pulmonary function in adolescents after repair of esophageal atresia with tracheoesophageal fistula. J Pediatr 153(3):396-401.
5. Sistonen, S., P. Malmberg, K. Malmstrom, T. Haahtela, S. Sarna, R. J. Rintala, and M. P. Pakarinen. 2010. Repaired esophageal atresia: Respiratory morbidity and pulmonary function in adults in a population-based long-term follow-up study. Eur Respir J.
6. Kovesi, T., and S. Rubin. 2004. Long-term complications of congenital esophageal atresia and/or tracheoesophageal fistula. Chest 126(3):915-25.
7. Rintala, R. J., S. Sistonen, and M. P. Pakarinen. 2009. Outcome of esophageal atresia beyond childhood. Semin Pediatr Surg 18(1):50-6.
8. Sistonen, S. J., I. Helenius, J. Peltonen, S. Sarna, R. J. Rintala, and M. P. Pakarinen. 2009. Natural history of spinal anomalies and scoliosis associated with esophageal atresia. Pediatrics 124(6):e1198-204.
9. Gischler, S. J., M. H. van der Cammen-van Zijp, P. Mazer, G. C. Madern, N. M. Bax, J. C. de Jongste, M. van Dijk, D. Tibboel, and H. Ijsselstijn. 2009. A prospective comparative evaluation of persistent respiratory morbidity in esophageal atresia and congenital diaphragmatic hernia survivors. J Pediatr Surg 44(9):1683-90.
10. Castilloux, J., A. J. Noble, and C. Faure. 2010. Risk factors for short- and long-term morbidity in children with esophageal atresia. J Pediatr 156(5):755-60.
11. Wailoo, M. P., and J. L. Emery. 1979. The trachea in children with tracheo-oesophageal fistula. Histopathology 3(4):329-38.
12. Nakazato, Y., T. R. Wells, and B. H. Landing. 1986. Abnormal tracheal innervation in patients with esophageal atresia and tracheoesophageal fistula: study of the intrinsic tracheal nerve plexuses by a microdissection technique. J Pediatr Surg 21(10):838-44.
13. Xia, H., C. Otten, L. Migliazza, J. A. Diez-Pardo, and J. A. Tovar. 1999. Tracheobronchial malformations in experimental esophageal atresia. J Pediatr Surg 34(4):536-9.
14. Shah, A. R., E. L. Lazar, and A. B. Atlas. 2009. Tracheal diverticula after tracheoesophageal fistula repair: case series and review of the literature. J Pediatr Surg 44(11):2107-11.

Management of severe tracheomalacia

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Tracheomalacia (TM) refers to a generalized or localized collapse of the tracheal lumen causing luminal obstruction during respiration and is present in 75% of patients with esophageal atresia (OA) with distal tracheoesophageal fistula (TOF). Severe TM often presents with acute life-threatening events in only 11 to 30% of patients. Tracheomalacia is one of the major causes of morbidity and late death in patient with EA/TEF. Death from airway problems may be preventable by its early recognition, adapted assessment, and aggressive management.

Physiopathology of TM in patients with EA/TEF remains unclear but histologic and experimental studies report deficiency of cartilage with a concomitant increase in the length of muscle in the membranous part of the tracheal ring. Moreover, the cartilaginous deformation is frequently associated with a bulging of the posterior wall related dilated former esophageal pouch that can lead to a total anteroposterior collapse of the trachea. In addition, TM is occasionally associated with laryngeal or tracheobronchial malformation that can participate to airways obstruction.

Mild to moderate TM may be asymptomatic apart from the characteristic barking cough or may present with stridor or recurrent chest infections. Severe TM present with dyspnea associated with recurrent acute life-threatening events, which are usually associated with feeds, vigorous crying, or coughing.
The current gold standard of diagnosing tracheomalacia is by visual assessment during bronchoscopy. Flexible bronchoscopy is superior to rigid bronchoscopy for evaluation of airway collapses. Flexible bronchoscopy provides the best assessment for the dynamic airway changes found in tracheo-bronchomalacia. Spiral CT gives high-resolution and advanced volumetric assessment but dynamic acquisitions inspiration and expiration spirals are impossible to obtain in young infant. Spiral CT reveals spatial relationship of airways and mediastinal structures.

The other methods of diagnosis are less reliable and/or have major pitfalls.
Based on the results of this functional and morphological assessment an appropriate surgical management of severe tracheomalacia can be proposed.

Signs of tracheomalacia responsible of acute life-threatening events include loss of consciousness, collapses followed by airway re-open, bradycardia and cardiac arrest (dying spells). These events must be rapidly identified and appropriate management proposed. esophageal stricture and upper segment dilatation must be ruled out as GER.

In these situations, medical management including anti-inflammatory medication, bronchodilators, antibiotics and oxygen therapy are not sufficient and anatomic opening of the malacic tracheal segment is mandatory. Open aortopexy is the oldest and the more frequent technique used and lately, thoracoscopic procedures were described. Stenting the tracheomalacic segment is another alternative for severe TM but results seems to be better in surgical procedures. The Cochrane review in 2008 didn't find any RCT study and include 13 papers in their analyses. Conclusions of the authors were that with the lack of evidence, recommendations for the routine use of any therapies for intrinsic tracheomalacia couldn't be made. But at the same time, it is unlikely that any RCT in surgically based management will be ever available for children with severe life threatening illness associated with tracheomalacia. Literature is richer with papers dealing with surgical options with satisfactory results. We present a series of 11 patients with aortopexy with a special focus on their pulmonary follow up.

References

Infants with repaired esophageal atresia and distal tracheoesophageal fistula with severe respiratory distress: is it tracheomalacia, reflux, or both? Nasr A, Ein SH, Gerstle JT. J Pediatr Surg. 2005;40(6):901-3.

Long-term complications of congenital esophageal atresia and/or tracheoesophageal fistula. Kovesi T, Rubin S Chest 2004;126(3):915-25.

Tracheomalacia with esophageal atresia and tracheoesophageal fistula in fetal rats. Qi BQ, Merei J, Farmer P, Hasthorpe S, Hutson JM, Myers NA, Beasley SW J Pediatr Surg. 1997;32(11):1575-9.

Analysis of morbidity and mortality in 227 cases of esophageal atresia and/or tracheoesophageal fistula over two decades. Engum SA, Grosfeld JL, West KW, Rescorla FJ, Scherer LR 3rd. Arch Surg. 1995;130(5):502-8

Tracheomalacia in esophageal atresia. Usefulness of preoperative imaging evaluation for tailored surgical correction. Briganti V, Oriolo L, Mangia G, Buffa V, Calisti A J Pediatr Surg. 2006;41(9):1624-8.

Thoracoscopic tracheoaortopexia for the treatment of life-threatening events in tracheomalacia. van der Zee DC, Bax NM. Surg Endosc. 2007 Nov; 21(11): 2024-5. Epub 2007 Mar

A decade of using intraluminal tracheal/bronchial stents in the management of tracheomalacia and/or bronchomalacia: is it better than aortopexy? Valerie EP, Durrant AC, Forte V, Wales P, Chait P, Kim PC. J Pediatr Surg. 2005;40 (6):904-7

Interventions for primary (intrinsic) tracheomalacia in children (Review) Masters IB, Chang AB 2008 The Cochrane Collaboration.

The role of aortopexy in severe tracheomalacia Sumit Dave, Bruce G. Currie J Pediatr Surg. 2006;41, 533–537

Outcome

Outcome of oesophageal atresia beyond childhood

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Backgound and aim

Survivors of oesophageal atresia are reaching their adulthood in large numbers for the first time thus allowing assessment of true long-term outcome among these patients. There are no previous population-based long-term follow-up on oesophageal atresia, therefore, the long-term sequelae are unclear.
In 2003 we initiated a study project to evaluate late outcome in adults patients who had undergone neonatal repair of oesophageal atresia. We aimed to study the incidence of oesophageal cancer, oesophageal function and morbidity, respiratory morbidity and clinical characteristics of musculoskeletal anomalies, especially spinal defects at adult age.

Patients and methods

The original study population consists of 588 patients treated for oesophageal atresia in the Children's Hospital, University of Helsinki, from 1947 to 1985. A total of 235 were alive with their native oe-sophagus. These were contacted and the first 101 (median age 36 years) who replied and agreed to par-ticipate made up the study group. The clinical and demographical characteristics of the study group were statistically similar to those of non-participants. The study group patients and also the patients not attending the clinical study responded to a symptom questionnaire including questions on oesophageal, respiratory, musculoskeletal symptoms and quality of life (SF-32, GIQLI, RSRQLI). Age and sex matched healthy controls (n=287) filled the same questionnaires. The study patients underwent upper gastrointestinal endoscopy with biopsies, oesophageal manometry, pulmonary function tests and full orthopaedic evaluation with radiographs. The incidence of cancer among oesophageal atresia patients was evaluated from population-based countrywide cancer registry that covers practically 100% of all cancer cases .

Results

Symptomatic gastro-oesophageal reflux occurred in 34% and dysphagia in 85% of the patients (8% and 2% in controls, $p<0.001$). The endoscopic findings included hiatal hernia (28%), Barrett's oesophagus (11%), oesophagitis (8%), and anastomotic stenosis (8%). Histology showed oesophagitis in 25% and epithelial metaplasia in 21%; the metaplasia was gastric in 15% and intestinal in 6%. Oesophageal ma-nometry demonstrated non-propagating peristalsis in most patients and ineffective distal oesophageal pressure in all. Manometrical abnormalities were significantly more common in those with epithelial metaplasia ($p<0.02$).
Fifty-six percent and 70% of the patients had a history of pneumonia and bronchitis (controls 20% and 50%, $p< 0.001$); 16% of the patients and 6% of the controls had diagnosed asthma ($p<0.001$). Respira-tory-related impaired quality of life was reported by 11% of the patients but only in 6% of the controls ($p< 0.001$). Pulmonary function tests showed obstruction in 21%, restriction in 21% and both in 36% of the patients.
Vertebral anomalies were detected in 45% of the patients, most commonly in the cervical spine (38% of the patients). Clinical and radiographical scoliosis was found in 56% of the patients; the risk of scoliosis was 13-fold when compared with healthy population. In most patients the clinical course of scoliosis was mild and did not require bracing or spinal surgery. Radial ray anomalies were found in 25% of the patients.
The patients had more frequently impaired gastrointestinal (GIQLI<105 in 23% of the patients vs. 8% of controls) and respiratory related (RSRQLI<45 in 12% of the patients vs. 2% of controls) quality of life in relation to controls ($P<0.001$). There was no difference in the global health related quality of life (SF-36) between the patients and controls.

Despite high incidence oesophageal metaplasia, none of the Finnish oesophageal atresia patients had oesophageal cancer. Three patients had had cancer in other organ systems but the overall cancer inci-dence was similar as in the general population.

Conclusions

Morbidity associated with oesophageal atresia is significant in adults. Oesophageal symptoms such as dysphagia and GER were common as well as abnormal oesophageal histology. There was no association between oesophageal symptoms and histological findings. Surgical complications, increasing age and impaired motility predicted the occurrence of epithelia metaplasia. Respiratory symptoms, asthma and infections were more common in oesophageal atresia patients than in the controls. Over half of the patients with repaired oesophageal atresia develop scoliosis. Vertebral abnormalities, especially cervical anomalies, and radial ray anomalies were also common; most of these had not been detected earlier. However, the overall quality of life of the patients was comparable with healthy controls.

Multidisciplinary Clinics : how to improve the follow-up of patients ?

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The concept of Multidisciplinary Specialty Clinics evolved in order to provide comprehensive coordinated care for patients with complex diseases. These models have repeatedly been shown to improve clinical outcomes in a variety of conditions.
The lifespan of children with Esophageal Atresia and Tracheo-Esophageal Fistua (EA-TEF) has greatly increased since 1941. Over the years, several related morbidities necessitating specific expertise have become obvious. A Multidisciplinary care approach appears optimally-suited to address the special needs of EA-TEF patients.

Key considerations include:

1. Who should be part of a collaborative multidisciplinary team for EA-TEF patients? What would be the "Perfect" team?

Based on the main expected co-morbidities, a core of specialists could be easily defined but other health services should optimally be integrated as well .

2. What are the prerequisites to a Successful team?

The mandate of the multidisciplinary team should be clear from the onset -- common goals are the key-- it is more than simply sharing complementary diagnostic and therapeutic skills.

Obviously, inter-professional collaboration, commitment, motivation, and being a good "team player" are all essential qualities of members in a multidisciplinary team (MDT). Appreciation for the role of others is also critical. In order to improve efficiency, role assignment and leadership should be clearly identified in order to avoid duplication, overlap or omissions.

3. The Quebec/Montreal EA-TEF model?

Different levels of Networking and co-management currently exist in Quebec. The RQAO (Réseau Québécois interuniversitaire d'Étude de l'Oesophage) is a provincial network regrouping the four University Centres of Quebec, both pediatric and adult. A transition program incorporating transition clinics has also bee integrated into this network model. Two MDT exist in Montreal: the Ste-Justine Hospital Team and the Montreal Children's Hospital (MCH) Team. The team of the MCH will hereby be presented.

4. What are the Advantages to a MDT for the EA-TEF population?

Advantages of this collaborative multidisciplinary care team will be reviewed from the patient, parents, community and physicians perpectives. Quality of life, Quality of care, ongoing education of patients and their families, and research are all domains where benefits appear obvious. Over time, we will gain knowledge on the real impact of this new ongoing multidisciplinary expert care approach on the global outcome and natural history of EA-TEF population. We are confident that these clinics will be a success from the clinical, scientific and human points of view.

1. Interprofessional collaboration :effects of practice-based interventions on professional practice & healthcare outcomes(Review); Cochrane database of system reviews(3): CD 000072,2009
2. Paediatrir.Child Health 11(7) Sept 2006
3. Pottie K. Research in social & administrative pharmacy 5(4):319-26,2009 dec
4. Lewis PJ. Journal of the Royal Society of medecine 102(11): 481-8
5. Seghal NL. Journal of General internal medecine 23(12) 2053-7, 2008 dec
6. Jain AK. Journal of surgical education 65(2) 145-50, 2008 Mar-Apr
7. Grosse S. Pediatrics 123(1)407-12, 2009 Jan
8. Doyle J .Paediatric nursing 20(2):26-29

P001 - Copy number variations in esophageal atresia / tracheo-esophageal fistula.

E.M. DE JONG, M. VISSER, D.A. SCOTT, A. DE KLEIN, D. TIBBOEL
(Erasmus MC – Clinical Genetics, Rotterdam, The Netherlands)

Esophageal atresia with or without tracheo-esophageal fistula (EA/TEF) is a congenital malformation that occurs in 1/3500 newborns. In about half of the cases additional malformations are present and in some patients a recognizable syndrome as CHARGE (CDH7 gene), Feingold (NMYC) or AEG (SOX2) can be confirmed. In approximately 10% of the patients with EA/TEF the VACTERL association (Vertebral anomalies, Anal atresia, Cardiac malformations, Tracheo-Esophageal fistula, Renal and Limb malformations) is seen. Clinical data of >400 patients, from 1988 onwards, is stored in our database. We have collected DNA, cell lines and pouch tissue (n=50) of over 200 patients with EA/TEF. This includes a set of 60 well characterized VACTERL associated patients. Conventional karyotyping and screening with subtelomeric MLPA are done routinely in all patients. A review of all published cases revealed 11 chromosomal regions that are recurrently affected in EA/TEF.

We are using a positional candidate approach based on chromosome aberrations and array data to localize and identify these genes that cause or predispose to the development of EA/TEF. To identify new chromosomal regions – and refine those previously reported – we have screened >100 patients with EA/TEF and/or VACTERL association for chromosomal deletions/duplications in affected individuals using a combination of Agilent and SNP Arrays. Although the majority of deletions/duplication had been identified previously in normal controls, over 100 rare genomic variants were identified. Q-PCR and FISH was used to confirm the copy number variation (CNV) and parents were included to determine the inheritance pattern of these clinically relevant CNVs within families. Although de novo changes are considered more likely to be causative, the 1 to 2 % recurrence risk in EA/TEF and VACTERL suggests that most cases may result from a combination of inherited changes affecting important developmental pathways combined with environmental stressors such as maternal diabetes.

P002 - Esophageal atresia – epidemiology in europe.

R. NEESS PEDERSEN, S. HUSBY, E. GARNE
(Hans Christian Andersen Childrens Hospital, Odense, Denmark)

Obective
The objective of this study is to describe prevalence, prevalence of prenatal diagnosis and epidemiologic data on esophageal atresia (EA) from twenty three well-defined European regions and compare total prevalence and prevalence of prenatal diagnosed between these regions. We wanted to characterize the cases as syndromic, multiple malformed or with isolated anomaly. Further more we wanted to characterize patients born with EA with regards to sex, the type of birth, gestational age (GA), birth weight (BW) and one week survival.
Material and Methods
The study is based on routinely collected data from 23 European registries of congenital malformations (EUROCAT). For further details on population coverage ad registration methods see www.eurocat.ulster.ac.uk/memberreg/.
All cases with ICD9 and ICD10 code with EA born in the period 1987-2006 were included in this study. Total number of births covered by the 23 registries in the years included in the study was 5,027,181.
Results
During the study period there were 1222 cases of EA from the 23 registries giving an overall prevalence at 2.43 per 10,000 births. There where large regional differences in prevalence. Also the prenatal detection rates showed large regional differences.
The majority of cases were boys (57.3%). The majority of cases (70.5%) had a tracheo-esophageal fistula along with the atresia. 88.7% of cases were live births (LB), 3.5% were still births (SB) and 7.8% were termination of pregnancy (TOP). Mean BW in the LB cases was 2462±766 grams and median GA was 38 (35-39) weeks.
The cases were classified in four groups: 44.7% had isolated esophageal malformation, 31.6% were multiple malformed, 23.7% had an association or syndrome.
The most common associated malformations were congenital heart defects, other gastrointestinal malformations, limb and urinary tract malformations.
During the study period the prenatal detection rates showed a statistical significant increase.
Discussion - Conclusion
There are large regional differences in prevalence and prevalence of prenatal diagnosis.
After pre or postnatal diagnosis of EA, it is important to evaluate the infant for associated malformations or syndromes, as these are present in more than half of all cases.

P003 - Magnetic compression anastomosis a nonsurgical treatment for esophageal atresia.

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Purpose
To communicate feasibility of performing an esophageal anastomosis with magnets, avoiding surgery and their results after more than 2- year follow- up.
Method and materials
Between 2001 and 2008, 8 children were admitted and evaluated. Seven were selected to be treated. Five of the patients had esophageal artesia without fistula and the other three had different types of artesia with fistula, which were surgically converted into type A.
Neodymium-iron-boron (NdFeB) magnets were used, mounted on catheters for the superior and inferior ends. Biplane chest radiographs were taken until the magnets were united. Then the magnet catheters were replaced with an orogastric tube. The patients started oral feeding 48 hours later.
Results
Anastomosis was achieved in all patients in 4,8 days average. All patients but one developed esophageal stenosis. All patients are currently free of treatment and with oral diet. Two patients require periodic balloon dilations. One patient underwent surgery because of recurrent esophageal stenosis not amenable to balloon dilatations. One patient, developed sepsis that was successfully treated with antibiotics.
Conclusions
Magnetic compression anastomosis is a feasible method in selected patients with esophageal atresia.
Esophageal anastomoses were achieved in all cases.
The only observed complication of significance was esophageal stenosis.

P004 - Post operative oesophageal atresia repair complications during the first year of life in a tertiary care center in paris, france : impact of the early post operative care.

J. NAUDIN, S. KATSAHIAN, S. AIZENFISZ, C. VITOUX, N. BEYDON, N. BELARBI, Y. NIVOCHE, S. DAUGER, A. BONNARD
(Hôpital Robert Debré, Paris, France)

Background
The prevention of long term morbidity is the current care challenge of newborn with an oesophageal atresia (OA). Available data concerning the impact of immediate post operative care on short and mild term oesophageal complications are limited. The aim of this study was to investigate immediate post operative follow up to determine if whether or not there were predictive factors of late oesophageal complications.
Methods
All consecutive case of type 3 OA (Ladd classification) with primary oesophageal anastomosis, between january 1997 and june 2008, admited in pediatric intensive care unit (PICU) at Robert Debré Hospital were included in a monocentric, retrospective study.
Clinical, surgical and post operative findings during the first post operative week in PICU were analysed. Children with or without oesophageal complications during the first post operative year of life were studied in an uni and multi-variate statistical analysis
Results
110 cases were included. 22 children developped one or two oesophageal complications (n=24) including 16 anastomotic strictures requiring dilatation, 4 fistula repermeabilisations, and 2 anastomotic leakage, 1 oesophagoplasty and 1 oesogastric disconnection. The duration of the surgical primary anastomosis was a risk factor of oesophageal complication only in univariate analysis (OR = 1,01 [1 – 1,03] p = 0.0496).
Surprisingly, an adequate post operative fluid resuscitation was a protective factor of oesphageal complication in uni (OR = 0,09 [0,01 – 0,68] p = 0,0201) and multi variate analysis (OR = 0,09 [0,01 -0,73] p = 0,0243).
Conclusion
Fluid resuscitation and more globally, the post operative hemodynamic status, could be an important post operative factor to prevent the occurrence of oesophageal complications especially the anastomotic site stictures. Improvement of oesophageal tissue healing could be explained by an early impact on the oesophageal microcirculation.

P005 - Outcomes of tracheomalacia as a complication of tracheo-oesophageal fistula and oesophageal atresia.

A. HAWLEY, K. GUY, E. J. MCLEOD, R. W. HUNT
(Royal Children's Hospital, Parkville, Australia)

Background
Tracheomalacia is a common complication following TOF/OA. Symptoms range from a brassy cough to acute life threatening events. The aim of this study was to describe the severity and outcomes of tracheomalacia in patients with TOF/OA at The Royal Children's Hospital (RCH).
Method
A retrospective review was performed of all infants treated for TOF/OA at RCH over 15 years (May 1993 – May 2008). We searched our TOF/OA database and referred to case notes where necessary. Tracheomalacia was defined as mild if the only feature was a 'TOF cough' or stridor; moderate if there was significant respiratory compromise, mechanical ventilation was required, or evidence on bronchoscopy/bronchogram; severe tracheomalacia was defined as an acute life threatening event requiring resuscitation, requiring prolonged ventilation >14 days, the need for surgical intervention (tracheostomy or aortopexy) or bronchoscope or bronchogram evidence (with airway collapse at >20cmH20) .
Results
238 eligible subjects were treated for TOF/OA at RCH during the study period. 92 (38.6%) had a recorded diagnosis of tracheomalacia. 65 (27.3%) had mild, 8 (3.3%) had moderate and 19 (7.9%) had severe tracheomalacia. 4 (1.7%) had severe tracheobronchomalacia, 2 (0.8%) had severe bronchomalacia, and 1 (0.4%) had mild tracheobronchomalacia. 11 (4.6%) required surgical intervention for their tracheomalacia (4 - aortopexy, 4 - tracheostomy and 3 both). 1 patient with moderate tracheomalacia required tracheostomy for management of subglottic stenosis. 7 (2.9%) had a Nissen fundoplication for gastro-oesophageal reflux thought to be contributing to respiratory compromise. Tracheomalacia contributed to the death of one subject that was unable to be resuscitated following respiratory arrest. Tracheobronchomalcia resulted in 3 deaths and bronchomalacia contributed to 2 deaths (aged 3 mo and 9 yrs).
Conclusions
The prevalence of tracheomalacia at RCH is consistent with reports in the literature. It should always be considered as a differential for respiratory morbidity in TOF/OA survivors, and its potential significance should not be underestimated.

MAY 27th

10:30-11:00 Epidemiology, Genetics posters

A001 - Oesophageal atresia and national french web database for children congenital pulmonary malformation.

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Introduction
The endorsement of a centre of rare respiratory diseases by the French ministry of Health in 2006 gave the opportunity to focus on congenital pulmonary malformation (CPM). In this context we have created a national database approved by the institutional review board and the Commission Nationale Informatique et Liberte (CNIL) for children CPM including oesophageal atresia.
Methods
this CPM database includes clinical, biological, functional, radiological, and pathological data which are updated at every patient's visit and daily use by the medical team in charge of the patients.
Results
In Armand Trousseau pulmonary department, all patients with CPM were collected. In February 2010 we had fulfilled the data for 225 CPM patients and among them, 43 had oesophageal atresia. These patients were included in the congenital pulmonary malformation group of the Reference Center for Rare Lung Diseases because of tracheomalacia consistently associated with oesophageal atresia. Diagnosis of oesophageal atresia was made during antenatal period for only 13 % of patients. The repartition of oesophageal atresia type was 10 % for the type I, 2 % for the type II, 75% for the type III and 13% for the type IV. All children underwent surgery. The mean age of patients in our database was 4.6 (range 0.24-19) years with a sex ratio of 1. The median of follow up was 7 (range 0-19) years. All patients had gastroesophageal reflux. Respiratory symptoms were noticed in 86 % and included tracheomalacia (98 %), chronic cough (65 %), dyspnea on exercise (14 %), chronic bronchial obstruction (70 %) and bronchial hypereactivity (30 %). The chest auscultation was abnormal in 46.5 % of the patients. Abnormal chest X ray was found in 56 % of the case. Thirty two patients (74 %) received treatments for respiratory symptoms including continuous antibiotherapy (44 %), physiotherapy (56 %) and inhaled corticosteroid (37 %).
Conclusion
The collected data provide new insight in the management of respiratory symptoms associated with oesophageal atresia and represent a useful tool for the survey of children after surgery.

A002 - Twelve years of esophageal malformations in Alsace (France): epidemiologic, genetic and clinical study and evaluation of prenatal diagnosis.

B. DORAY, D. BADILA-TIMBOLSCHI, C. CORDIER, B. DOTT, F. BECMEUR, M. FRADIN, E. LE BOETTE, Y. ALEMBIK, R. FAVRE, I. NISAND, B. VIVILLE, D. ASTRUC, B. GASSER, V. LINDNER, E. FLORI , F. GIRARD-LEMAIRE, H. DOLLFUS
(Hôpital de Hautepierre, Strasbourg, France)

Introduction
The aim of this study was to review clinical, genetic and epidemiologic data on esophageal malformations and to evaluate the efficiency and the impact of prenatal diagnosis.
Material and Methods
A retrospective study was carried out among data of the Registry of Congenital Malformations of Alsace (department of Bas-Rhin) in France between 1995 and 2006.
Results
Fifty-two esophageal malformations were recorded (overall prevalence: 0.34 per 1000, approximately 1 per 3000), subdivided into 49 cases of esophageal atresia with tracheo/broncho-esophageal fistula, 2 cases of atresia without fistula, and 1 case of fistula without atresia. The global male/female ratio was 1.36 (30/22).
There was no case of parental consanguinity. No case of familial recurrence was reported. Fertilization was spontaneous for all cases. Three gemellary pregnancies with CDH in one twin only were described.
Among these 52 cases, 41(79 %) were liveborn, 1 case (2 %) was stillbirth and 10 cases (19 %) resulted in termination of pregnancy.
Associated malformations and/or chromosomal abnormalities were observed in 25 out of the 52 cases (48 %). A chromosomal abnormality was identified in 5 cases (with 3 cases of trisomy 18). A malformative spectrum compatible with VACTERL association was discussed in 10 cases.
Only 12 cases (23 %) were prenatally suspected by sonographic examination considering the association hydramnios and abnormal stomach. Termination of pregnancy was performed only when associated or syndromic cases.
The postnatal prognosis of diaphragmatic hernias was good with 39 out of the 41 liveborns alive at 2 years of life (95 %) and 38 alive at 5 years (93 %). The three cases of postnatal death concerned associated or syndromic cases. All isolated cases were alive at 5 years of life.
Discussion - Conclusion
Esophageal atresia is an unfrequent congenital malformation, with a prevalence of 0.34 per 1000 total births. Ultrasound prenatal diagnosis of diaphragmatic malformations remains difficult with a sonographic detection in only 25 % of the cases. Associated or syndromic cases are frequent and must be carefully looked for improving the diagnostic and prognosis evaluation, isolated forms having a good survival after surgery and multidisciplinary follow-up.

A003 – Study of sonic hedgehog pathway genes in 80 patients with isolated and syndromic esophageal atresia – preliminary results.

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(Wroclaw Medical University, Wroclaw, Poland)

Esophageal atresia and tracheo-esophageal fistula are relatively frequently occurring foregut malformations of which etiology and pathogenesis are heterogeneous and not clearly understood. Chromosomal anomalies have been reported in up to 10% of cases of esophageal atresia, but no single specific chromosomal defect has been confirmed as a main etiological factor. Four genes MYCN, CHD7, MID1 and SOX2 have been implicated in rare cases of syndromic esophageal atresia. It is thought that a combination of genetic and environmental factors play a role in the etiology of foregut anomalies and it is the most likely that pathogenesis of esophageal atresia is also multifactorial. However, recent results of molecular genetic studies in esophageal atresia have yielded in the understanding of the molecular mechanism involved foregut morphogenesis.

This study focuses on the genes of Sonic Hedgehog pathway (SHH–PTCH–GLI) involved in the very early stages foregut embryology and which play important role in these processes.
The aim of study was to assess deletions and mutations in SHH and GLI genes and to analyze of the expression patterns of these chosen genes.

The study was carried out on a group of 80 patients with esophageal atresia. Patients with oesophageal atresia were classified as isolated and syndromic cases. Clinical aspects of associated malformations and diagnosed dysmorphic syndromes in patients with esophageal atresia were also presented in this study.
Screening mutations in chosen genes of SHH pathway were performed using TTGE method. After that, to identify point mutations, sequencing analysis of critical region were done. Simultaneously, MLPA study was performed to detect whole allele deletion in chosen genes. To analyze expression of panel genes, Real–Time PCR was used.

Moreover, we compare the results of SHH pathway genes expression in blood cells with expression in cells obtained from esophageal tissues taken out during surgery in thirty five patients with esophageal atresia.
To gain more insight of molecular changes and expression in SHH genes, we performed the genotype-phenotype correlation.

A004 – Esophageal atresia national registry results for 2008.

R. SFEIR, A. BONNARD, T. GELAS, N. KHEN-DUNLOP, F. AUBER, F. BECMEUR, A. BRETON, G. PODEVIN, F. LAVRAND, L. MICHAUD, M. MORINEAU, T. PETIT, F. SABIANI, V. FOUQUET, E. HABONIMANA, A. HOSSEIN, C. JAQUIER, J.L. LEMELLE, A. MAUREL, M.L. POLIMEROL, P. BUISSON, H. LARDY, M. LOPEZ, D. AUBERT, P. DELAGAUSIE, P. DE VRIES, J.L. GAUDIN, C. BORDERON, A. ECHAIEB, F. ELBAZ, L. FOURCADE, O. JABY, E. SAPIN, M. ARNOULD-POUZAC, J.L. BRÉAUD, S. GEISS, C. LAPLACE, C. PELATAN, D. WEIL, B DAGRY, F. GOTTRAND

(Hôpital Jeanne de Flandre, Lille, France)

Introduction
Incidence of esophageal atresia (EA) varies between 1/ 3000 and 1/5000 live births. The aim of this study is to present to results of the OA National Registry for the year 2008. (France and French overseas department and territory included)
Methods
Inclusions shafts for data collection were distributed all over the territory and analyzed by the reference centre. The medical information department (DIM) of every hospital was contacted as well as 4 regional registers of deformations to increase the number and the exhaustiveness of sources
Results
One hundred and forty eight cases were declared for year 2008 with a calculated incidence at 1/5597 birth (national birth rate in 2008 was at 828404 live births). There was boys' ascendancy (n=88), the average born weight was of 2529 g with a term at 37 ga. Prenatal diagnosis was possible in 17 % of the cases. There were 16 cases of type I in Ladd's classification and 128 type III. The diagnosis was made within 24 hours in 89 % of the cases and the initial surgery realized before day 3 in 94 % for the type III. The rate of the associated malformations was 53 % and the global mortality of 4,7%.
Conclusions
These first results show an incidence of the AO similar to that observed by the European register of the deformations. The prenatal diagnosis remains rare but with early diagnosis of the EA, mortality rate is comparable to the literature in western countries.

A005 – Use of collis gastroplasty in the repair of long-gap esophageal atresia.

A. SCHNEIDER, C. GOMES FERREIRA, I. KAUFFMANN, I. LACREUSE, F. BECMEUR
(University Hospital, Strasbourg, France)

Objective
To report our local experience of the Collis gastroplasty in the repair of long-gap esophageal atresia (EA).
Patients and Methods
The authors reviewed 5 cases of children with EA and long gap (5 to 6 vertebral bodies). The repair was carried out after 3 months of enteral nutrition and permanent aspiration of the proximal esophagus. The Collis gastroplasty was performed to create a gastric tube using an Endo-GIA linear stapler. The gastric tube can be easily mobilized into the thorax and anastomosed to the proximal esophageal segment.
Results
No intraoperative complication occurred. The average postoperative hospital stay was 40 days (range: 30-60) for an average follow-up of 16.8 months (range: 9-26). During early follow-up, a digestive occlusion occurred in one patient, and several months later, she suffered from a hiatal herniation. Other postoperative complications occurred: anastomotic stricture (n=2) requiring one or several dilatations, gastroesophageal reflux (GER) (n=3) and severe weight delay (n=4) requiring prolonged enteral nutrition in 2 cases.
Conclusions
We believe that the Collis gastroplasty is an efficient and easy esophageal lengthening procedure for EA in children with long gap. The complications seen are common complications of EA surgical repair such as GER, anastomotic stricture and ponderal

A006 – Comparison of post-operative course of end-to-end esophageal anastomosis and esophagoplasty in type 1 esophageal atresia.

A. DARIEL, N. KHEN-DUNLOP, M. MORINEAU, V. ROUSSEAU, C. NIHOUL-FÉKÉTÉ, Y. RÉVILLON, S. SARNACKI
(Hôpital Necker-Enfants Malades, Paris, France)

Aim
Type 1 esophageal atresia (EA) is mainly characterized by a long-gap and surgical repair is thus challenging. End-to-end esophageal anastomosis (EEA) or esophagoplasty (EP) are classically used. Our aim was to compare the post-operative course of these two procedures.
Methods
Between 1984 and 2008, 31 patients with type 1 EA were managed in our institution. Twenty-five cases had prenatal diagnosis (81%). Median gestational age at birth was 37 weeks with median birth weight of 2400g. Associated malformations were present in 16 cases (52%). Primary gastrostomia was performed in all cases at birth. EEA was realized in 16 and EP in 15 (13 colon and 2 stomach interpositions). Follow up was 29 months [12-91] for EEA and 86 months [34-233] in EP.
Results
EEA and EP groups were comparable for prenatal diagnosis, sex, gestational age, birth weight and associated malformations. Median esophageal gap was 4 vertebral bodies in EEA [2-5] and 5 in EP [2-6] (p=0.008). Procedures were performed at median age of 86 days [1-210] and 118 days [2-627] (NS) for EEA and EP respectively. Anastomotic strictures were not statistically different (7 versus 5) whereas anastomotic leaks were higher in EEA group although not significant (5 versus 1). Median initial hospital length of stay was 161 days [82-237] in EEA and 345 days [115 - 765] in EP (p=0.04).
Gastrostomia was used for complementary enteral support during 13 months [2-80] in EEA and 34 months [3-127] in EP (p=0.04). No difference in growth was noted between the 2 groups either at 1 year (-2.5 SD and -2 SD) or at the end of follow-up (-2 SD and -1.5 SD).
Conclusion
Despite the retrospective nature of this study, our series demonstrated as good if not better results following EEA as compare to EP for type 1 EA.

A007 – Thoracoscopic repair of tracheoesophageal fistula

D. PATKOWSKI, A. ANTCZAK, R. MIGIEL, A. POGORZELSKI, M. BAGŁAJ
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Increasing experience in minimal access techniques and growing evidences of benefits allowed to perform more complex procedures. Thoracoscopic approach seems to be a safe and effective alternative to the thoracotomy with possible avoidance of its long-term complications.

Purpose
The analysis of performed thoracoscopic repair of tracheoesophageal fistula (TEF).
Methods
During the last two years we have operated four cases aged 6 months to 7 years with TEF by thoracoscopic approach. There were two cases after open repair of esophageal atresia (EA) and two cases of H type TEF. All TEF were diagnosed by bronchoscopy.
Results
There were no conversion. Operative time was between 100 and 160minutes. There were no operative complications. Previous thoracotomy didn't make any problem to achieve operative space. In children after EA repair in one case there was a recurrent TEF, in other case it was probably missed upper TEF. All TEF were closed by 5mm titanic clips. In two cases it was possible to separate the TEF tract completely by division. The postoperative course was uneventful. The follow up was between 4 months and 2 years. The preoperative complaints resolved completely.
Conclusion
The thoracoscopic repair of TEF is effective method and based on our experience it is the procedure of choice if performed by experienced endoscopic pediatric surgeon.

A008 – Esophageal atresia; growing experience in thoracoscopic correction.

C. MEEUSSEN, CEJ SLOOTS, MC STRUIJS , CP VAN DE VEN, D. TIBBOEL
(Sophia Children's Hospital, Rotterdam, The Netherlands)

Background
Treatment of esophageal atresia is challenging. Thoracoscopic minimal invasive correction was first reported in 2000 by Rothenberg. In our institution we started this technique in 2006. Aim: to review our results regarding thoracoscopic procedure and complications.
Methods
Retrospective chart review was performed of patients undergoing thoracoscopic repair of esophageal atresia, admitted to our centre between July 2006 and February 2010. Patient characteristics, associated malformations, surgical procedure and complications, time on mechanical ventilator, time to start enteral and oral feeding and length of stay were reviewed. Data were presented as medians and range. Results: In the study period 20 patients (11 boys) underwent thoracoscopic correction. Gestational age was 38 weeks (28-40), birth weight was 2800 gram (1080-3600). Diagnosis was suspected on prenatal ultrasound in 4 patients. Associated anomalies were present in 4 patients (VACTERL). All but one had a type C esophageal atresia according to Gross. Age at surgery was 2 days (0-67). Operative time was 195 minutes (135-347). Early postoperative complications were leakage of anastomosis in 2 patients, treated conservatively. One of them, with multiple congenital anomalies and complex cardiac malformation, died at the age of 17 days, due to sepsis and cardiac failure. Three patients had recurrence of distal fistula, 2 of them were treated thoracoscopically. Postoperative mechanical ventilation was necessary for 1 day (0-4). Time to enteral feeding was 2 days (1-4), time to first oral feeding was 5,5 days (1-123). Median hospital stay was 18,5 days (5-161). Minimal invasive aortapexy and laparoscopic Nissen fundoplication were performed in 2 patients with life threatening events, due to tracheomalacia and gastro-esophageal reflux. Esophageal dilatations were performed, based on clinical symptoms in 8 patients. Six patients needed 2 or more dilatations (0-15).
Conclusion
Thoracoscopic repair of esophageal atresia is a feasible technique with an acceptable complication rate. This technique has become standard of care in our centre. Long term evaluation with respect to esophageal function, scoliosis and cosmesis will be performed.

A009 – Thoracoscopic operations for treatment of esophageal atresia.

A.Y. RAZUMOVSKY, S-KH.M. BATAEV, A.B. ALKHASOV, V.E. RACHKOV, Z.B. MITUPOV, R.O. IGNATYEV, R.M. KHANVERDIEV
(Filatov Children's Hospital, Moscow, Russia)

The aim of this study is to introduce the recent development of thoracoscopic operations in our clinic for the one-stage esophageal anastomosis to repair esophageal atresia in infants.

Patients and Methods
Since March 2008 we have performed 27 thoracoscopic operations for EA in infants weighing ranged from 1110 to 4065 g. 9 (33,3%) patients had associated VATER malformations. In 24 children there were EA with distal tracheoesophageal fistula (TEF), in 1 case - EA with proximal and distal TEF and in 2 cases - isolated EA without TEF. We used 3 troacars approach. A pneumothorax was maintained with 4-6 mm Hg and flow not more than 1 l/min. We prefer extracorporeal sutures for performing esophageal anastomosis. Chest tube was left in all cases. We have started feeding through nasogastric tube on 2-5 postoperative day.
Results
24 procedures were successfully completed without conversion. Average operative time was 90+/-32 min. The conversions were in 3 cases: in 2 cases due to large distance between esophageal porches in infants with EA without TEF, in 1 case due to lung rigidness which was caused by severe pneumonia. In these 3 infants the esophago- and gastrostoma were performed. The mean time to extubation was 10,5 days (ranged from 2 to 25 days).
There were 4 anastomotic leaks: 1 was managed conservatively and 3 required thoracoscopic closure. Reappearance of TEF appeared in 2 infants and was successfully managed thoracoscopically. Esophageal stenosis was founded in 11 (40,7%) patients and was treated by dilatation.
We had to performe laparoscopic Nissen fundoplication in one infant with gastroesophageal reflux. There were 2 deaths (7,4%) of causes not related with performed operation.
Conclusion
The thoracoscopic approach for repair of EA represents a natural evolution in the operative correction of this complicated congenital anomaly. The intraoperative complications observed if properly managed have a good prognosis.

A010 – Long gap esophageal atresia. a single center experience.

A. CONFORTI, B. D. IACOBELLI, A. CROCOLI, P. DE ANGELIS, A. BRAGUGLIA, F. MORINI, P. BAGOLAN

Aim
To describe a single centre experience in the treatment of long gap esophageal atresia (LGEA) and to define the differences between LGEA with and without distal tracheo-esophageal fistula (dTEF).
Methods
Retrospective review of all consecutive patients (pts) treated for EA between 1995 and 2009. LGEA was defined as a gap 3 cm or vertebral bodies. Pts with and without dTEF were compared for several variables (table) by Mann-Whitney or Fisher's exact test, as appropriate.
Results
173 consecutive pts with EA were treated. Overall mortality rate was 7.5%. 46 (27%) pts had LGEA (20 referrals). Native esophagus was preserved in all pts but one (referred), who required jejunal substitution at five months of age. Table shows findings.
Conclusions
Treatment of pts with LGEA is demanding, regardless the type of EA, but in most of them preservation of own esophagus is possible and effective. LGEA associated with dTEF had earlier surgical repair and a trend towards higher prevalence of associated anomalies. Pts with LGEA need long term follow-up for the high risk of complications that may require multiple operations.
Table
Results are medians (interquartile ranges) or prevalence

	dTEF+ (21pts)	dTEF- (25 pts)	p	All LGEA
Gest. age (wks)	38 (34-38)	36 (34-38)	0.34	37 (34-38)
Birth weight (kgs)	2.5 (1.8-2.7)	2.3 (1.9-2.8)	0.94	2.4 (1.9-2.8)
Age at surg. (d)	3 (2-83)	70 (40-158)	<0.01	34 (3-73)
Intraop. gap (cm)	3 (3-4)	4 (3-5)	0.06	3 (3-4)
Referrals (pts)	8	12	0.56	20
Anast. leak	10%	20%	0.43	15%
Anast. dysruption	0%	0%	1.00	0%
Anast. stricture	71%	76%	0.75	74%
Dilatations (#)	3 (2-5)	5 (3-7)	0.11	4 (3-6)
Vocal cord paral.	5%	16%	0.36	11%
Anti-reflux surg.	33%	44%	0.55	39%
Other anomalies	85%	56%	0.05	69%
Mortality rate	33%	12%	0.15	21%

A011 – Lessons we've learned in the treatment of long gap esophageal atresias.

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Background
A gap greater than 3 cm between both esophageal pouches is observed in 1 of 20 cases of esophageal atresia (EA). Our goal was to critically review our experience in the management of these patients.
Patients and Methods
From 86 EA treated at our institution during the last 11 years, there were 18 with a long gap. Of these, 11 were pure EA (8 treated with a Schärli reconstruction, 2 with esophageal flaps and 1 esophagocoloplasty). The remaining 7 were EA with distal fistula, treated as: 3 end-to-end anastomosis (1 Livaditis), 1 esophageal flap, 1 neonatal Schärli, and 2 awaiting definitive reconstruction.
Results
From 9 Schärli there have been 2 reinterventions for anastomotic leak and 1 parahiatal hernia that needed surgery after 8 years. Among the 3 flaps patients, 2 underwent reintervention for persistent stricture. The 3 patients with end-to-end anastomosis needed a second intervention for persistent stricture, and 2 and anti-reflux procedure. The esophagocoloplasty patient didn't developed any complications. In general, 8 patients achieved normal growth and development (4/9 Schärli, 3/3 Flaps and 1/3 E-E), and 10 were growth retarded (5/8 with associated malformations, 1/7 who developed an eosinophilic esophagitis, 3/7 preterm babies and 2/2 awaiting definitive treatment).
Some lessons we've learned…

1.- The own secretions could dilate the upper esophageal pouch.
2.- To feed the babies with bolus through the gastrostomy tube, because the gastro-esophageal reflux helps in growing the inferior pouch.
3.- Thechnical items to consider when performing an Schärli: reinforcement of the suture; to tailor the lesser curbature as a tube; to close the esophageal hiatus; and to fix the stomach.
4.- It's advisable to keep patient under mechanical ventilation and paralysis for a week to avoid disruptive forces and leakage from a tension esophageal anastomosis.
5.- Aspirative drains can allow a leak to persist.
6.- To treat all patients for gastroesophageal reflux, because the gastroesophageal union is always disarrayed. Occasionally, GER collaborates in stricture's development.
7.- Although sometimes 2 major procedures are required, the best esophageal substitute is the own esophagus.
8.- Despite difficulties, good results can be achieved.

A012 – Unconventional management of an uncommon form of esophageal atresia.

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Case report
A male infant presents with long gap esophageal atresia. The primary distance measures 5 cm.The upper stump reaches well beyond the tracheal bifurcation while the lower stump is extremely short ending below the diaphragm. A feeding gastrostomy is fashioned on Day 3. Bottle feeding is initiated soon and raised to up to 6 x 120 ml on continuous suction. Bedside bouginage of the upper stump is performed daily and tolerated well. At the age of 3 months, transabdominal end-to-end anastomosis is achieved, with gastrostomy closure in the same session. Postoperative recovery is uneventful. Following four esophageal dilatations within the subsequent two months, the boy has developed a normal eating behaviour and thrives well.
Discussion
Embryology and pathophysiology of this unusual type of esophageal atresia shall be discussed. The importance the authors ascribe to sham feeding for esophageal growth, comfort and development of the child and psycho-social well-being of the family is emphasized. Last but not least, the exclusively transabdominal access für primary esophageal atresia repair has not been found reported on pub med literature search.

A013 – Aortopexie in severe tracheal instability after esophageal atresia: short-term and long-term outcome in 26 infants and children.

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Background
Between 10-33% of neonates that undergo correction of esophageal atresia with a distal tracheoesophageal fistula develop a significant tracheomalacia. The symptoms due to malacia of the large airways occur directly or a few weeks to months after repair of the esophageal atresia and vary from stridor, wheezing, and recurrent lower airway infections to severe dyspnea or acute life-threatening episodes of cyanosis and/or apnea. In cases with severe tracheal instability leading to life-threatening apneas, aortopexie has been shown to be a therapeutic option. To asses the efficacy of this therapy, short-term and long-term outcome of our patients was analyzed retrospectively.
Methods
Between May 1994 and March 2009, we operated on 26 patients (13 males and 13 females; age 49 days-10 years) with tracheomalacia associated with esophageal atresia. In 23 children, there was a more than 90% collapse of the distal trachea. The major presenting symptoms were stridor in 22 patients and life-threatening apneic episodes in 24. All of the patients underwent aortopexy for the removal of the dynamic stenosis. The ascending aorta and aortic arch were elevated and fixated to the sternum and the ventral thoracic wall. The optimal positioning of the sutures was bronchoscopically monitored in 15 cases. The procedure was carried out for 14 patients via a left posterolateral or anterolateral thoracotomy, and 12 children underwent median sternotomy.
Results
There were no intraoperative complications. 25 children survived in short-term and long-term follow-up. One child died four months after the operation because of a severe bone metabolism disorder. 25 patients obtained full relief of airway obstruction, which has persisted in long-term follow-up from 3 month to 14 years.
Conclusion
Aortopexy is a safe and efficient method in the surgical treatment of tracheomalacia after correction of esophageal atresia. It helps to prevent recurrent airway tract infections and enables normal child development. There is no operation-related mortality, and the morbidity is low.

A014 – Preoperative evaluation of esophageal gap in esophageal atresia newborns.

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Purpose
Preoperative assessment of gap length in esophageal atresia (EA) is still not well established. Nonetheless, it is ascertain that the distance between the two pouches in EA plays a role in surgical strategies. Our aim was to describe a reproducible and reliable method for gap measurement in all types of EA.
Methods
TYPE A/B - Correction of type A/B EA is commonly delayed, performing tracheoscopy (even to exclude type B) and gastrostomy as first step. Gap is measured fifteen days afterwards under flouroscopy: 1) a 10F feeding catheter is inserted in upper pouch generating a loop; 2) a 4.5 Hegar dilator is then introduced through the gastrostomy into the lower pouch; 3) a dynamometer is connected to the Hegar (to define and standardize the pressure applied). Gap length is then measured at chest X-ray. TYPE C/D – Just before correction of EA, a radio-opaque 10F feeding tube is inserted in the upper pouch creating a loop. Tracheoscopy, to define numbers and level of the fistulas, is routinely performed. The bronchoscope tip is placed at the tracheal fistula opening, the gap is then measured at chest X-ray calculating the distance between the upper pouch and the lower fistula marked with bronchoscope's tip.
T-test was used to compare preoperative and intra-operative gap measurements.
Main results
We evaluate 5 patients (4 type C EA, 1 type A EA) from July to December 2009. Preoperative and intra-operative gap measurements were extremely similar (mean 9.2 mm, range 1-35mm vs. mean 9.8 mm, range 0-40 mm; $p = 0.65$)
Conclusions
The described technique consents accurate pre-operative evaluation of gap length, obtaining reliable data for prognostic and therapeutic purposes. In addition an objective measurement of the gap will consent to compare different series of patients with EA.

A015 – Our experience on postoperative complications of esophageal atresia.

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Aims
Esophageal atresia (EA) is one of the commonest congenital malformation. This study evaluated postoperative complications of esophageal atresia (EA).
Methods
We reviewed retrospectively reports of 21 patients (8 female, 13 male; 38 ± 1 WG ; 2751 ± 62 g) with EA operated in 2 differents units between October 1991 and december 2008. Data collected included : patients characteristics, associated anomalies, perioperative management parameters and postoperative complications.
Results
EA was classified type II (n = 1) or III (n = 20) according to Gross classification. The length of gap between esophageal pouches (LG) corresponded to 2 vertebral bodies or less in 18 children. Illness severity was classified type I (n = 17) or type II (n = 4) according to Spitz classification. In one case, a bronchogenic cyst was associated to EA. The others associated anomalies were cardiovascular (n = 6), skeletal (n = 4), genitourinary (n = 3), gastrointestinal (n = 2), and chromosomal (n = 2). Mean age of surgery , length of postoperative ventilation and hospital stay were respectively 2 ± 1, 6 ± 1, and 27 ± 2 days. A gastrostomy tube was placed before (n = 3) or after (n = 4) EA repair. All patients survived after a primary anastomosis. Postoperative complications were anastomotic leak (n = 1), recurrent fistula (n = 1), atelectasis (n = 5), tracheomalacia (n = 10), and esophageal stricture (n = 10). pH-metry monitoring done in 4 cases was positive. Rate of tracheomalacia was higher (61 vs 25 %) in case of associated anomaly. Esophageal strictures required 2 ± 0.2 dilation procedures 8 ± 5 months after surgery and were related (table 1) to LG, post operative ventilator dependency and nutritional status.
Conclusion
Tracheomalacia and esophageal strictures were the most common complications. Related risk factors of these complications suggested by our data need confirmation. Uncommon association of EA with a bronchogenic cyst is reported.

Table 1 : factors associated to esophageal stricture

	Stricture +	Stricture -
LG more than 2 vertebral bodies (%)	30	0
Post operative ventilation (days)	8 ± 1.9	3 ± 0.5
Weight/height after 430 ± 40 days (z-score)	-1.5 ± 0.5	+0.3 ± 0.4

A016 – Therapeutically aspects in esophageal atresia.

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The esophageal atresia (E.A.) treatment still represents a challenge, concerning both the maintaining of the native esophagus and the neonatal intensive care efficiency.
The aim of this paper is to present a single team experience in E.A. treatment in the last five years.
28 consecutive cases, treated in the last five years (between 2005 and 2009) were retrospectively analyzed. We studied the type of the malformation, associated diseases, results regarding the saving of the native esophagus, and also the value of colon esophagoplasty.
In our series 23 patients (82%) had E.A. with distal tracheo-esophageal fistula (TEF), one case (3.6%) – EA with both proximal and distal TEF, one case (3.6%) – isolated TEF, 2 cases (7.2%) – isolated EA, and in one case (3.6%) – congenital esophageal stenosis. 6 patients were initially treated in other surgical units (gastrostomy, TEF ligation and cervical esophagostomy), one of them subsequently suffering multiple failed interventions, in which it was attempted the saving of the native esophagus, by the traction of the esophageal ends. We noted different associated anomalies in 9 patients (cardiac malformations, duodenal atresia, imperforate anus, tracheomalacia, skeletal deformities, paraesophageal hernia and pyloric stenosis). Among the 22 patients treated from the beginning in our unit, in 19 cases we performed primary repair of the EA, in one case - isolated TEF ligation, in one case – removal of the congenital stenotic segment and esophageal repair and in one case we renounced at the native esophagus after a failed traction procedure. 2 patients developing esophageal anastomotic tight stricture and 2 patients with significant anastomotic leak required the revision of the anastomosis, which was performed successfully. In one case a recurrent fistula occured. All the 6 patients, initially having gastrostomy, TEF ligation and cervical esophagostomy, suffered colon esophagoplasty.
We encountered 4 deaths (14.3%), 3 of them weighting less than 1.500 g. at birth.
The developing of the neonatal intensive care allows us to increase the percentage of cases with preserving the native esophagus. In long gap cases, the esophageal traction may represent a good instrument for primary anastomosis achievement. However, the reconversion of the EA patients initially having esophagostomy and cervical esophagostomy, in order to elongate and save the native esophagus leads to a serious morbidity. Colon esophagoplasty in failed esophageal repair is a safe and functional alternative.

A017 – Interest of the ph-impedance monitoring in patients undergoing surgery for esophageal atresia.

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Objective
The gastro-esophageal reflux disease (GERD) is a common complication in patients (P) with surgery esophageal atresia. The intraluminal impedance (pH-IMP) detect the passage of bolus into an organ to determine the nature of the reflux fluid (LR), gas (GR) and mixed (MR). If it is combined with the recording of esophageal pH, this technique allows the study of acid reflux (AR), weakly acidic (WAR) and non-acid (NAR). This study aims to analyze the frequency of reflux using the pH-IMP in these P.
Patients and Methods
Seven P with an AO type III were investigated by pH-IMP between February and August 2008. The analysis includes: the number of reflux characterized by their pH (AR, WAR and NAR) and composition (LR, MR and GR).
Results
The total number of reflux disease is to high for 6P/7. The number and index of AR are normal. The number of WAR is always pathological even in 4P who were treated by proton.pump inhibitor The number of GR is abnormal for 6P/7.
Discussion
The absence of reference values in children, the absence of consensus on the definition of WAR is an obstacle to the interpretation of results of abnormal values of GR and WAR.
Conclusions
This review is one of the first that investigates. GERD by pH-IMP in the P with of AO type III. The results encourage us to obtain reference values in children and to advise the pH-IMP for the monitoring of these P.

A018 – Esophageal manometry : mandatory for ph probe placement in esophageal atresia paediatric patients.

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The pH probe study remains the gold standard to evaluate gastroesophageal reflux disease, which occurs up to 35% to 58% of children with esophageal atresia with or without tracheo-esophageal fistula (EA-TEF).Proper placement is essential. Many mathematical formulas have been developed to estimate the esophageal length (EL) in normal children.

Aim
To determine whether an equation can accurately calculate the esophageal length in paediatric patients with EA-TEF. Nonsurgical paediatric patients were also evaluated.
Sixteen patients with EA-TEF aged between 4 and 17 years old had an esophageal manometry (EM)(n=26) since 2000 at the Montreal Children's Hospital. 127 children without EA-TEF or any other esophageal surgery (total of 144 EM) have also been prospectively followed during the same period. For both groups we compared the measured esophageal length by manometry to the calculated EL determined by the Strobel, Song and Jolley formulas. Age, gender, history of surgery, indication of manometry, result of pH probe (if done) and manometry were also analysed.
All three equations were inaccurate to predict the esophageal lenght in EA-TEF population. The Strobel formula calculated an average EL 3.4±0.25cm too long compared to the one measured. The Song and Jolley formulas, calculated respectively an EL 3.3±0.25cm and 2.8 ±0.25cm shorter than the EM measurements ($P<0.001$). In the normal paediatric population, these formulas were also inaccurate ($P<0.001$). Of the 10 asymptomatic EA-TEF children, 30% of the pH probe studies done were positive. This study confirms that no mathematical formula should be used for the placement of pH catheters in EA-TEF patients as they are inaccurate to predict the esophageal length when compared to EM, the established gold standard for the location of the lower esophageal sphincter. The impact on pH study results could be non negligeable. Our proportion of positive ph studies in asymptomatic children emphasizes the necessity for an accurate positioning. As the high resolution manometry becomes more available and easier to perform, pH probe placement by esophageal manometry should become a standard clinical practice in the pediatric population especially in EA-TEF children and other surgical patients.

A019 – Tube feeding dependence in infants with repaired distal tracheo-oesophageal fistula and oesophageal atresia.

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Background
Tube feeding dependence (TFD) is often necessary at time of discharge from hospital following repair of TOF/OA. The aim of this study was to describe the frequency and associated factors of TFD in infants with TOF/OA at discharge.
Method
A retrospective review was performed of infants with repaired OA and distal TOF at The Royal Children's Hospital over a 10 year period (December 1995-December 2005). The Nate Myers Oesophageal Atresia Database was searched to identify cases and case notes were referred to.
Results
111 patients were discharged from our NICU following repair of OA and distal TOF during the study period. 7 (6.3%) were discharged on tube feeds alone (gastrostomy [5.4%]; nasogastric [1.8%]). 17 (15.3%) were discharged receiving a combination of oral and tube feeds. The remainder were entirely orally fed.
Factors associated with tube feeding on discharge included: (A) Surgical issues: 5 (20.8%) had delayed repair of oesophagus with prolonged time to introduction of oral feeds (mean time to feed = 46 d post-op [mean 125 d), 6 (25%) had cardiac surgery, 4 (16.6%) required other surgery (B) Postoperative complications related to TOF/OA delaying the introduction of oral feeds: 7 (29.1%) had anastomotic leak; 4 (16.7%) had oesophageal stricture requiring dilatation; 8 (33.3%) had significant GOR and 4 (16.7%) had GOR treated with Nissen fundoplication (C) Neonatal issues: 14 (58.3%) were premature (<37 weeks' GA); 16 (66.7%) had low BWt (<2500g); 8 (33.2%) had BWt <1500g. (D) Associated congenital abnormalities -12 (50%) cardiac, 8 (33.3%) VACTERL. (E) Respiratory issues: 8 (33.3%) had significant tracheomalacia, and 1 (4.1%) had severe tracheobronchomalacia. 2 (8.2%) had subglottic stenosis; 3 (12.5%) underwent tracheostomy; 8 patients required prolonged ventilation. These factors were not mutually exclusive as many infants had multiple issues.
Conclusions
Infants with TOF/OA have many factors that cause tube feeding dependence. Increased awareness of these factors is the first step in improving rates of oral feeding for infants with repaired TOF/OA.

A020 – High-resolution esophageal manometry : preliminary evaluation on patients with treated esophageal atresia.

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Esophageal manometry is used for various digestive issues without mechanical or mucous causes. Only few specialized centres can perform this technique among adult patients because of its complexity and interpretation toughness. Thanks to electronic sensors technology, high-resolution manometry allows to record precisely the esophageal motility, by defining a pressure spatiotemporal plot, with easier execution and better tolerance in adult patients.
The goal of this work is to describe the technical feasibility and datas of high-resolution esophageal manometry in children who had had an esophageal atresia surgical cure.
High-resolution esophageal manometry (ManoScan 360, Sierra Scientific, Los Angeles CA, USA) was performed in 4 patients aged 3.1 to 14.5. The catheter was an adult one (4.2mm diameter) with 36 circumferential channels spaced 1cm. All patients, followed for esophageal atresia, were complaining of upper digestive symptoms, without esophageal stenosis (X ray and/or endoscopic assessment). Collected datas were: indication, examination sequence (length, tolerance), qualitative and quantitative datas of interpretation.
The introduction of the catheter ran smoothly, even for the youngest. There wasn't any incident leading to a premature end of the examination, and the tolerance was excellent.
For all patients, the upper esophageal sphincter basal and relaxation pressures were normal, as well as the peristalsis of the striated muscle segment. On the other hand, under this portion, the motility was always abnormal, with 2 kinds of figures: a) a complete lack of peristalsis in 2 patients, with low basal pressure of the lower esophageal sphincter (LES) or, b) in the 2 others, a partial sporadic peristalsis under and away from the transition zone, with several propagated waves after the swallowings, associated with a normal LES basal pressure.
Compared to standard techniques, high-resolution esophageal manometry is easier to perform and to interpret, seems better tolerated and allows a better description of the motility disorders frequently seen in children with esophageal atresia. This preliminary series encourages us to collect more data in order to precise the link between manometric figures and symptoms.

A021 – Management of the barrett's esophagus in children.

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Patients and methods
345 patients with gastroesophageal reflux underwent Nissen fundoplication in our hospital from 1992 to 2009.
From this 52(14%) patients were diagnosed Barrett's esophagus by histological, endoscopic, and scintigrafic assessment. Multiple biopsies were taken systematically during endoscopy. The mean ages was 10,2 3,8 years (range 1 – 15). The boys were in 71% of cases.
Cardiac type of metaplasia were found in 21(40%) patients, fundal type in– 25(48%) and intestinal type in 6 patients 12%. In 75% patients had 2 or 3 types of metaplasia at the same patients.
Endoscopical signs of the Barrett's esophagus were found only in 21 (40%) patients. All patients with endoscopical signs of the Barrett's esophagus had esophageal strictures which dilatations due to the persistence of stenosis were necessary.
All 52 patients with Barrett's esophagus Nissen fundoplication was performed (in 76% using a laparoscopic approach). 13 (25%) patients with endoscopical positive signs of BE electro coagulations of metaplasia were performed (2-4 times after 4 weeks intervals), crio coagulations was performed in 1 patient.
Results
The median follow-up was 12 years (range 1–18).
In 9 patients (17%) with severe peptical strictures in whom Nissen fundoplication failed (2-4 times), exterpations of the esophageal was done and colonic replacement performed at the same time. In 43 (83%) cases fundoplication was successful, but complete disappearance of metaplasia not demonstrated in any case.
In 14 patients (27%) metaplasia of the esophagus was removing by electro and crio coagulations. In 29 patients (56%) after Nissen fundoplication with endoscopical negative BE we did not use any additional procedures, because they did not have any visible signs of the BE.
Conclusions
In patients who had endoscopical positive BE we achieve successful results due to electro and crio coagulations in all cases.
Esophageal replacement indicated in patients who had severe esophageal stricture and unsuccessful Nissen fundoplication.
The patients with endoscopical negative BE has been observed after antireflux surgery, because they did not any visible signs of the BE.

A022 – Routine use of ph study in young children to evaluate gastro-esophageal reflux following esophageal atresia and tracheo-esophageal fistula repair.

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Purpose
The incidence of GER following EA-TEF repair is high. Early GER diagnosis and treatment is essential to prevent its complications. The usefulness of routine pH study in young EA-TEF patients was evaluated.
Methods
Retrospective review of children with EA-TEF who underwent a pH study at CHU Ste-Justine from 2004 to 2008. Demographics, atresia type, associated anomalies, pH study, symptoms and complications were collected. Descriptive analysis of the association between GER and those factors was done.
Results
Twenty-one EA-TEF patients underwent a pH study. Median gestational age and birth weight were 38 weeks (33-40) and 2530g (1577-3720). Fourteen patients (67%) had associated anomalies. The first pH study was performed at a median of 9 months (5-24), with 12 (57%) positive results. A higher rate of reflux was found in babies born under 2000g (75% vs 53%). Gestational age, associated anomalies and Waterston/Spitz status did not impact GER incidence. All patients with GER were symptomatics. All patients (N=10;48%) with a history of vomiting/regurgitations had GER on pH study. From sixteen patients with pulmonary symptoms (76%), only 9 (56%) had GER on pH study. Complications included anastomotic leak (N=7;33%), fistula recurrence (N=1;5%), pneumonia (N=5;24%) and stenosis (N=10;48%). The incidence of positive pH study in children with pneumonia and stenosis was 40% and 60% respectively. One patient (5%) required fundoplication.
Conclusions
Young EA-TEF children have a 57% incidence of GER on pH study and are symptomatic. GER was higher in children born under 2000g and in those with stenosis, gastro-intestinal or respiratory symptoms. The pH study guided therapeutic decisions to change or discontinue reflux therapy and should be keep in the routine follow-up of children with EA-TEF before the year of age. However, despite empiric therapy, complications linked to GER developped. Usefulness of pH study while on therapy at a younger age to assess treatment efficacy could be evaluated.

A023 – A comparison of short and long-term feeding outcomes in infants with esophageal atresia.

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Introduction
Esophageal Atresia (EA) is a common congenital defect affecting between 3,000 and 4,500 live births. It prevents infants from early oral feeding exposure which may play a role in long-term feeding outcomes.
Aims
We evaluated early and long-term feeding outcomes of infants born with esophageal atresia.
Methods
Records of infants born between Jan 2004-Jan 2010 with esophageal atresia at a single tertiary care medical center (Children's Hospital of Wisconsin, Milwaukee, WI, USA) were reviewed. Demographic, surgical, and nutrition outcomes were abstracted. The percentage of oral feedings at discharge and 1 year follow-up were compared between two groups, based on the type of surgical repair.
Results
16 infants (7 females, mean weight of 2.17 kg, mean gestational age of 35.7 weeks) with esophageal atresia were identified. 10 of 16 patients had an accompanying tracheo-esophageal fistula. 1 patient died, 1 was lost to follow-up, and 3 did not meet criteria for 1 year follow-up. 10 patients had esophago-esophagostomy (EEA) and 4 had esophago-gastrostomy anastamosis (EGA). Infants with EEA were older (mean gestational age of 36.3 vs 34.8 weeks), larger (mean weight of 2.3 vs 2.1 kg), had an earlier surgical repair (median age at repair of 2 vs 63 days), and earlier initiation of oral feedings (mean days to 1st oral feed post repair of 17 vs 34 days) compared to infants with EGA. A greater percentage of infants with EEA achieved full oral feeds at discharge (60% vs 25%) and at 1yr follow-up (88% vs 67%) compared to infants with EGA.
Conclusions
Type of surgical repair plays a role in short and long-term feeding outcomes in infants with esophageal atresia.

A024 – Dumping syndrome after esophageal atresia repair without antireflux surgery.

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Surgical treatment of gastroesophageal reflux (GER) is the main cause of dumping syndrome in children. Other causes include congenital microgastria, partial or total gastrectomy, accidental intraduodenal or jejunal administration of bolus feeding, inadequate meals or generalized autonomic dysfunction. In children with esophageal atresia, all reported cases of dumping syndrome have so far been related to the surgical treatment of associated GER.
We report 3 children with esophageal atresia (EA) with a distal tracheoesophageal fistula (type A) (n=3), and associated congenital malformations (VACTERL syndrome) (n=2). They underwent primary esophageal anastomosis during the neonatal period simultaneously with colostomy for the 2 patients with VACTERL syndrome. Two patients presented with postoperative GER treated medically; one patient required repeated endoscopic bougienage because of recurrent anastomotic esophageal stenosis. No patient had GER surgery or microgastria. The 3 patients were all diagnosed with dumping syndrome at a median age of 4 months (range: 3-6 months), revealed through various symptoms: feeding difficulties, n=1; malnutrition, n=3; diarrhea, n=1; postprandial seizures, n=1; postprandial discomfort, n=1. Dumping syndrome was confirmed by a glucose load test and the treatment consisted in dietary measures (uncooked starch) and nocturnal enteral nutrition. Weight gain and growth were normal on follow-up for 2 patients (treatment could be stopped respectively at 12 and 17 months of age), whereas one is still treated at 5 months of age.
Dumping syndrome is responsible for non-specific symptoms that are frequent in patients with esophageal atresia and may be attributed to several different causes, such as anastomotic stenosis, gastroesophageal reflux, esophageal dysmotility, and/or oral abnormal behavior.
Conclusion
Our data show that dumping syndrome: 1) can occur after primary anastomosis of esophageal atresia without GER surgery; 2) should be considered in every child treated surgically for esophageal atresia presenting with digestive symptoms, malaise, failure to thrive or refusal to eat.

A025 – Is inlet patch more frequent in patients with esophageal atresia ?

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Objectives
An inlet patch (IP) is defined as heterotopic gastric mucosa located in the proximal esophagus. IP pathogenesis could be congenital and/or acquired. Prevalence of IP is unknown in childhood even if case reports suggest that it could be more frequent in patients with esophageal atresia (EA). The aim of this retrospective multicenter study was to evaluate the clinical characteristics and evolution of IP in children.
Methods
All cases of IP recorded in 7 tertiary pediatric gastrointestinal centers. Information about demographics, clinical symptoms, endoscopic characteristics, histology, treatment and evolution were collected. Results: Fifteen children were included (8 boys, 7 girls). Median age at diagnosis was 9.5 years (range, 3.3–15 years). Six children presented congenital abnormalities (5 with EA = 33% of the patients) and 9 had gastroesophageal reflux. Only 1 child was asymptomatic. Digestive symptoms (dysphagia, food impaction) were noted in 14 patients and respiratory or ear, nose, and throat symptoms in 6. All 5 children with EA complained of digestive symptoms and 4 of them presented cough or globus sensation. Proton pump inhibitor treatment was initiated in 14 children for a mean duration of 4.7 months (range: 1–12 months). Evolution was good in 8 patients. Clinical symptoms disappeared in 2 patients with EA, improved in 1 and evolution of symptoms could not be determined in the 2 remainers.
Conclusions
IP is a rare, although probably underdiagnosed, pathology in children undergoing endoscopy but seems to be more frequent in patient with EA. IP could be responsible of digestive and respiratory symptoms in these patients, provided that esophageal stenosis and peptic esophagitis have been ruled out. Whether the association of IP and EA is fortuitous or not remains to be determined and long-term longitudinal studies are required to assess the outcome.

A026 – Medium-term efficacy and safety of mitomycin-c for recurrent esophageal stricture in children.

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(Hôpital Jeanne de Flandre, Lille, France)

Mitomycin-C is an antiproliferative agent that has been used successfully as a local treatment in ophthalmologic procedures, for laryngeal and tracheal stenosis, and more recently to prevent recurrence of esophageal stricture in children. The aim of this study was to assess the medium-term efficacy and safety of local application of mitomycin-C to refractory esophageal stricture in children.
We performed a cross-sectional study of children (n=6; median age, 7 years; range: 5.0–9.5 years) who received at least one local application of mitomycin-C for refractory esophageal stricture (caustic stenosis, n=3; esophageal atresia, n=3) and were followed up for at least 2 years in our center. Clinical evaluation, radiological control and esophagoscopy with multilevel esophageal biopsies at the site of stenosis were performed.
The mean follow-up period after mitomycin-C application was 4.3 years (range: 3.3–4.7 years). The frequency of dilation decreased from 0.32 per child per year before mitomycin-C application to 0.04 per child per year after the procedure ($p<0.05$). Four patients required a second application of mitomycin-C (15 days, 2 months, 3 months, and 13 months after the first application, respectively) and one required a third application (12 months after the first application). Digestive symptoms improved in all patients. None of the children presented with esophageal stenosis according to radiology and endoscopy at follow-up. Biopsies revealed the presence of Barrett's esophagus at the site of the stenosis in two children but neither intestinal metaplasia nor dysplasia was detected.
Our study shows that mitomycin-C has beneficial effects that persist for several years after its application to children presenting with recurrent esophageal stenosis and that it is not associated with complications in the medium term. However, the risk–benefit ratio of this treatment should be weighed by considering possible long-term complications, especially neoplasia. These patients require a long-term follow-up.

A027 – Esophageal peristaltic response to meal and acid reflux in operated esophageal atretic children.

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Long standing ambulatory esophageal pH-manometry (LSEpH-M) allows the recording of contractions appearing during different periods such as meals (M) and pH falls below 4 (GER). Frequency of contractions and their complete transmission (23% of total recording) are normally increased during M (30%) due to primary peristalsis and in reaction to GER (28%) due to primary and secondary peristalsis. In patients operated of esophageal atresia (EA), the percentage of completely propagated contractions (CPC) is significantly lowered.
Aim
Compare esophageal motility patterns during M and GER stimulations after repair of EA.
Methods
18 patients (mean age 5 4/12 y, SD: 4y) operated upon for an EA of the Ladd- Gross type C without further surgical complication, were investigated by LSEpH-M during a systematic survey. ESpH-M was performed with a μ Digitrapper 4MB. Controlled by fluoroscopy, the tip of the antimony pH-metry probe was in the standard position whereas that of the solid state pressure sensors of the manometry probe was controlled by fluoroscopy and endoscopy: the proximal transducer (P4) above while the two others (P5 - P6) below the level of the anastomotic scar. The percentage of CPC from P4 to P5 and P6 during the total duration of the recording (T), M and GER are compared (2-test).
Results
In EA children, the percentage of CPC is significantly increased during M (8,4%) compared to T (6.5%) $p< 0,05$, but not during GER (4,7%).
Conclusion
GER initiates less often CPC than swallow in EA patients suggesting a perturbation of the esophageal sensitivity as well as of its motility.

A028 – Are multichannel intraluminal impedance measurements useful in esophageal atresia.

M. MORRIS, A. ASPIROT, M. POMERLEAU, G. LAPIERRE, C. FAURE

Objective
Patients with esophageal atresia (EA) frequently have esophageal dysmotility and gastroesophageal reflux which is often hard to characterize as these patients are often asymptomatic. Combined multichannel intraluminal impedance (MII) and pH measurement is a novel method detecting intraesophageal bolus movement. Our aim was to determine if MII is able to effectively detect the retrograde bolus movement (i.e. reflux) in patients with EA.
Methods
We retrospectively reviewed MII-pH tracings and medical records of 20 patients at our institution. Patients were categorized into 3 groups: patients with repaired EA (6), patients with documented gastroesophageal reflux disease (GERD) without EA (7) and patients with normal studies and without EA (7). Sensitivity and diagnostic accuracy of MII-pH monitoring and symptom association were measured in the three groups. The baseline amplitudes of all 6 channels of the catheter were compared between the three groups at rest and in the recumbent position. In the subgroup of patients with EA, analysis of the impedance measurements in the 6 individual channels was performed, when the patient was upright.
Results
Symptoms of reflux captured by MII-pH measurements were higher in patients with GERD (50%) vs. pts with repaired EA (10%) which was statistically significant ($p<0.001$). The baseline amplitude of the esophageal at rest in the recumbent position was significantly lower in patients with repaired EA when compared with patients with GERD and the control group ($p<0.001$).
Conclusion
The low baseline amplitude observed in esophageal atresia appears to impair the capacity of MII-pH to capture the changes associated with reflux in esophageal atresia patients. The subtle changes in the esophagus in EA patients not acquired by MII-pH is postulated to be secondary to the poor esophageal function and/or stasis of liquid observed in these patients as suggested by the significant difference in impedance values in the upper esophagus vs. the lower esophagus in these patients .

A029 – HIgh resolution esophageal manometry in patients with operated esophageal atresia: a preliminary experience.

PARIS, L. GIGUÈRE, C. FAURE
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Objective
High resolution esophageal manometry (HREM) has revolutionized the evaluation of esophageal motility. We hypothesized that HREM will permit a better understanding of the esophageal motility in patients with esophageal atresia (EA). We assessed the esophageal body peristalsis, the upper (UES) and the lower (LES) esophageal sphincters.
Methods
HREM Manoscan 360 (36 channels with 12 pressure sensing points/channel) was performed as an outpatient procedure in patients with operated EA. Patients were asked to perform at least 10 swallows. Tracings were reviewed and analyzed by a paediatric gastroenterologist and a paediatric surgeon using the software provided by the company. Esophageal body peristalsis, sphincter basal pressure and relaxation were calculated. Charts were reviewed for past medical /surgical histories and symptoms to compare the results with the clinical history.
Results
Eight patients (7 boys;1 girl) with a median age of 8 years (range: 2.3-13.5yo) had a HREM. Seven patients had a type C EA (with tracheoesophageal fistula) and 1 had type A EA (without fistula). Compliance was good in all but one patient refused to take more than 4 swallows. Aperistalsis was found is 6 patients with only 2 complaining of dysphagia. Only 1 patient had antegrade contractions of low amplitude (42 ± 12 mmHg) in the distal esophagus and this patient reported intermittent dysphagia. All patients except 1 had normal UES. The patient with abnormal UES presented with cough associated with liquids and an HREM showing intermittent incomplete relaxation. Five patients had a hypotensive LES (range: 0.7-6.2 mmHg) with complete relaxation. Two patients had a normotensive LES with one of them having intermittent abnormal relaxation. The last patient had a hypertensive LES (66.9mmHg) that did not relax. This patient had a Toupet fundoplication. He was asymptomatic at the time of the study.
Conclusion
HREM permits a better evaluation of the esophageal motility in patients with operated EA. Esophageal motility seems to be severely affected by EA even though most patients had few symptoms. The UES does not seem to be affected and the LES was almost uniformly low. More patients will be needed to draw stronger conclusions.

A030 – Conservative treatment of anastomotic stricture in patients with esophageal atresia: a 10 years italian experience.

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Background
Esophageal stricture is a common early sequel following repair of esophageal atresia (EA). Need for dilations of the strictured anastomosis has been reported in up to 80% of the patients particularly for long gap atresia (LGA).
Aim
To demonstrate the efficacy of conservative treatment (endoscopic dilations and OPBG dynamic stent placement) in pediatric patients (pts) with esophageal atresia and anastomotic stenosis.
Patients
We reported data from a 10 year follow up of 173 (93M) pts with repaired EA. 127 (73,5%) had a non long gap (NLG) EA and 46 (26.5%) a LGA (LG: distance between the two pouches >3 cm). 108 pts developed stricture of the esophageal anastomosis (NLG:LGA=76:32). At 1 month from surgery or if severe dysphagia appeared, pts underwent to upper gastrointestinal endoscopy under general anaesthesia and, if esophageal lumen was < 5 mm, dilations using Savary-Gilliard were performed. OPBG stent≥was placed after at least 4 dilations. The custom silicone stent OPBG is built on a nasogastric tube and its diameter ranged from 5 to 12 mm; the two pointed ends and the central «filling», are designed to allow the passage of food bolus between them and the esophageal wall with dynamic effect. Difference between NLG and LGA groups in terms of number of dilations (<4 ; 4) was evaluated according to Fisher's exact test. p value < 0.01 was considered significant.
Results
Total dilations were 364: NLG pts 208 dilations (mean 1,67± 2,37 SD), LGA pts 156 dilations (mean 4,46 ± 3,33 SD); p value is statistically significant (p<0.0011). Mean age at first dilation after surgery was in NLG population 2.53 months and in LGA population 2.39 months. We observed no complications due to endoscopic procedure. Nine pts (NLG:LGA=1:8) needed OPBG stent placement: in 7 pts effective. In 2 pts with LGA, multiple OPBG stenting failed and they underwent surgery for stricture resection and T-T anastomotic re-do.
Conclusions
Endoscopic dilations with Savary and use of OPBG stent represent a safe and effective therapeutic approach in patients with esophageal stenosis. Surgery must be considered if conservative treatment fails.

A031 – Treatment of refractory esophageal strictures using removal endoluminal stenting.

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Aim
Esophageal stenting is used in adults with stenosis resistant to repeated dilations, but is rarely reported in children. The aim of this study was to report the French pediatric experience.
Methods
A questionnaire was sent to the members of the French Speaking Group for Pediatric Hepatology, Gastroenterology and Nutrition. Five centres reported esophageal stenting in 9 children.
Results
Children, aged of 36 months (16 months – 12.5 years), had caustic (n=7), peptic (n=1) or post-anastomotic (n=1) stenosis which were refractory to 2 to 16 dilations. A total of 20 procedures (1 – 4 stents / child) were performed: 7 non-expanding silicon stents were placed for a mean duration of 34 days (range: 21 – 109 d), 5 self-expanding metallic stents were placed for a mean duration of 63 days (range: 13 – 93 d), 8 self-expanding silicon stents were place for a mean duration of 27 days (range: 7 – 42 d). The correct position was difficult to obtain for 5/15 silicon stents (2 non-expanding and 3 self-expanding). Complications were encountered in 5/7 procedures with non-expanding stent: secondary migration (n=1), secondary stenosis (n=1), ulcerative esophagitis (n=2), difficulty of removal (n=1). Complications were present in all procedures using self-expanding stent: migration (n=1), difficulty of removal (n=2; one stent could not be removed, requiring esophagoplasty), and secondary stenosis (n=5) using the 5 metallic stents; migrations (n=5; with tracheal compression in one case), difficulty of removal (n=2), and dysphagia (n=1) using the 8 silicon stents. Esophageal stenosis recurred in all cases of non expanding stents, and in 11/13 cases of self-expanding stents.
Conclusion
Esophageal stenting for refractory stenosis is associated with numerous complications, limiting its use in pediatric patients.

A032 – Clinical characteristics and management of congenital esophageal stenosis alone or associated with esophageal atresia.

F. COUTTENIER, F. GOTTRAND, G. PODEVIN, A. BONNARD, A. SCHNEIDER, N. KHEN-DUNLOP, F. AUBER, A. MAUREL, T. GELAS, M. DASSONVILLE, C. BORDERON, A. DABADIE, D. WEIL, C. PIOLAT, A. BRETON, A. LEKE, A. MORALI, F. BASTIANI, T. LAMIREAU, L. MICHAUD

(Hôpital Jeanne de Flandre, Lille, France)

Congenital esophageal stenosis (CES) is a rare clinical condition in childhood and is frequently associated esophageal atresia (EA). The purpose of this study was to present the French experience of CES in terms of diagnosis, management and outcome.

Methods
Medical records of all the patients with CES from the French network on congenital and malformative esophageal diseases were reviewed retrospectively with regard to diagnostic method, therapy and outcome.

Results
During the last 18 years, 61 patients (30 boys) were found to have CES. CES was associated with esophageal atresia (EA) in 29 patients. The average age at diagnosis was 24 months (from 1 day to 14 years). Patients with associated EA were younger at the time of diagnosis than patients with isolated CES (7 vs. 126 months, $p<0.05$). 24 of 61 CES had no clinical symptom: 6 were found fortuitously, 18 (of 29 with associated EA) were diagnosed at the time of surgical repair of EA or during post operative systematic esophageal Xray opacification. For the 37 remainders initial symptoms were dysphagia (54%), vomiting (43%), and food impaction (54%). 15/37 patients presented with respiratory distress and/or dyspnea. 14/37 presented with impaired growth at diagnosis. Diagnosis of CES was confirmed with barium esophagram (56/61) and/or esophageal endoscopy (50/61) in all patients. Of the 61 patients, 16 had tracheobronchial remnants (TBR), 40 had a fibromuscular stenosis (FMS) and 5 had a membrane stenosis (MS). None had multiple stenosis; MS was never found in patients with EA (0/29, $p<0.05$). 34 patients were treated by dilatation only (13/34 were asymptomatic a follow-up), 15 patients were treated by dilatation first but required surgery (4/15 remained asymptomatic at follow up), whereas 9 patients had immediate surgical intervention (4/9 were asymptomatic at follow up). Dilations were complicated by esophageal perforation in 2 patients. At follow-up, dysphagia was present in 28% of patients (not different in EA group compared to isolated CS; 10/29 vs. 7/32, $p=0.27$). All patients with TBR underwent operative repair (resection and anastomosis).

Conclusion
Dilation may be effective for treating patients with FMS and MS while surgical repair is the preferred treatment for TBR. CS associated with EA can be missed at the time of initial esophageal surgical repair.

10:00-10:30 Pneumony, ENT and Fistula posters

A033 – «Dying spells» in association with oesophageal atresia.

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Introduction
Tracheomalacia may exist in association with oesophageal atresia (OA), and may cause significant respiratory distress in early infancy. It can cause serious apnoeic episodes and 'dying spells'. We report our experience with the condition and a review of the literature.
Methods
Retrospective case note review of all cases of OA treated at our centre from October 2003, studying in-particular those who had severe tracheomalacia.
Results
Three out of 38 infants with OA had significant tracheomalacia. Two of the three infants had anastomotic stricture that required dilatation. All presented early in infancy with blue episodes. They had recurrent episodes of apnoea and cyanosis. One infant remained on tube feeds for 5 months because of recurrent episodes of desaturations. All three had serious apnoeas with even 'dying spells', and required cardiopulmonary resuscitation. Bronchoscopy (MLB) demonstrated severe tracheomalacia, and all had aortopexy. All symptoms disappeared following aortopexy. Recent literature suggests that aortopexy is the main stay of treatment for tracheomalacia, curing near fatal episodes. It surprisingly, shows that «dying spells» occur in 22% to 93% of cases before an aortopexy was performed.
Conclusion
Three infants with severe tracheomalacia had dying spells prior to surgery thar required cardiopulmonary resuscitation. We recommend that any infant with persistent symptoms of apnoea and cyanosis after excluding post anastomotic stricture and treatment of gastro-oesophageal reflux should have the diagnosis confirmed early by MLB.Early aortopexy, probably, within a cardiothoracic unit, should be performed for all severe cases. A single «dying spell» should prompt urgent admission and

A034 – Long term evaluation of respiratoy status after oesophageal atresia.

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Initial prognosis of oesophageal atresia is nowadays excellent, due to neonatal and surgical support progress. This study aimed to evaluate long-term respiratory status of these children.

Method
Thirty eight children (23 boys) aged 8.2 y (3.5 to 15), operated between 1990 and 2004 have been evaluated. Neonatal data have been identified in medical records. Respiratory and digestive symptoms have been listed by standardized interview of parents and child. Each child received a clinical examination and a chest x-ray. Thirty-five children performed complete respiratory functional test. Cardiorespiratory response to effort was evaluated in 30 children from a test on ergometric bicycle.
Results
The most common respiratory symptoms were early and repeated respiratory infections (79%) with trend to improvement with age, clinical signs of bronchial hyper-responsiveness (63%), occasional hoarse cough (58%), effort dyspnea (24%). History of gastro-oesophageal reflux, dysphagia and food blocking had high frequency (58 %, 63 % and 55 %) but were still persisting at the time of evaluation in only 16 %, 21 % and 34 %. Eight children received treatment for asthma. The lung parenchyma was normal on chest x-ray in 35 (92%). Eight respiratory functional tests (23 %) were strictly normal. Bronchial obstruction, essentially for distal airways, was found in 17 cases (49 %). Eighty children (51 %) had bronchial hyper-responsiveness. A moderate restrictive syndrome was found in 5 cases (14 %). CO diffusing capacity value was reduced in 7 cases (18 %). All children had a normal cardio-respiratory response to effort but 13 showed a limited ventilatory response (43 %). This respiratory evaluation was helpful to adapt or initiate an antiasthmatic therapy in 14 children and educate parents, children, and house doctors for preventive measures of respiratory disease.
Conclusion
Due to frequent, although moderate, respiratory functional abnormalities and respiratory symptoms, a specialized, early and long-term respiratory follow-up of this population seems desirable. This monitoring will ideally include regular pediatric pneumology consultations, respiratory functional test and exercice-test.

A035 – Respiratory complications in children with oesophageal atresia disease.

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Background
Oesophageal atresia (OA), with or without tracheo-oesophageal fistula is a congenital disease that involves a group of foregut abnormalities, and may be complicated with a number of long-term respiratory complications and potentially fatal conditions.
Aim
The aim of the study is to explore the magnitude of respiratory complications in children with OA disease.
Methods : A case-note review of all cases of oesophageal atresia who were admitted at our centre over the last 6 years; 2003-2009.
Results
They were 44 children, 23 boys and 21 girls. There was no mortality. Surgery was complicated with oesophageal anastomotic leak in 2 children (4.5%), recurrent fistula in one (2%) and oesophageal stricture that required an average of 4 dilatations in 16 (36%). Twenty six children (59%) continued to receive antireflux medications for assumed or proved gastro-oesophageal reflux.
Respiratory complications were found in 29 patients (66%). Twelve children (27%) had tracheomalacia. Three of them had severe apnoeic episodes and "dying spells", and required aortopexy. One child had tracheal stenosis that required tracheoplasty. Eighteen children (41%) had recurrent chest infection that required a number of hospital admissions. Five children had pneumonia, 2 had asthma and 4 had prolonged episodes of cough.
Conclusion
Two thirds of the children with OA disease suffered from respiratory complications, mainly recurrent chest infections and aspiration. Extensive work is needed in order to find out why such complications arise. Which is to blame; the gastro-oesophageal reflux, the antireflux medications, the oesophageal dysmotility, an inherent abnormality of the tracheo-bronchial tree or a combination of these factors? Such significant complications need to be discussed with the parents on antenatal counselling and in an early visit to respiratory medicine outpatients clinic.

A036 – Persistent airflow obstruction and respiratory morbidity after repair of esophageal atresia.

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Background
Respiratory morbidity has been reported in patients with repaired esophageal atresia (EA) and trachea-esophageal fistula (TEF) in the first years of life. Respiratory tract infections (RTI), wheezing, chronic cough, stridor, dyspnea, apnea attacks and gastro-esophageal reflux (GER) are common problems.
Aim
We perform a standardized follow-up program including medical history and infant lung function test in the first year of life after repair of EA-TEF.
Method
Flow-volume loops, maximal expiratory flow at functional residual capacity (VmaxFRC) and lungvolume (FRC) were measured in 37 sedated patients with EA-TEF at 6 and 12 months. At measurement, patients were without symptoms of a RTI. Z-scores were calculated for VmaxFRC (Hoo 2002).
Results
Patients were born after a median of 38 weeks (IQR 36-40 weeks), 12 patients were born prematurely. Patients were mechanically ventilated for a median of 2 days (IQR 1-2.75 days). At 6 and 12 months patients showed evidence of mild airflow obstruction. There was no significant change in time (p=0.8). Median FRC expressed in ml/kg was within the normal range (Hülskamp 2003).

Median/IQR	6 months	12 months
VmaxFRC z-score	-2.3 (-3/-1.6)	-2.2 (-3.2/-1.5)
FRC ml/kg	21.4 (19.4/23.9)	24.6 (20.9/25.9)

15 patients (41%) had been treated with antibiotics for RTI at 6 months and 14 (38%) between 6 and 12 months. Antibiotic prophylaxis was given in 3 and 2 patients at 6 and 12 months, respectively. VmaxFRC z-score and FRC (ml/kg) were similar in patients with or without RTI (p=0.5 and 0.9 respectively). Only 3 patients had pathological GER (shown by pH-metry) at time of assessment.
Conclusion
In patients with repaired EA-TEF airflow obstruction and respiratory symptoms are common during the first year of life. Patients with a history of RTI had similar VmaxFRC as those without RTI. We assume that abnormally low VmaxFRC reflects tracheal malacia rather than peripheral airway obstruction. FRC is within the normal range, indicating normal lung volumes. Close follow-up of EA-TEF patients is needed to treat airway infections adequately and further elucidate the mechanisms of respiratory morbidity.

A037 – Laryngotracheal malformations associated with esophageal atresia.

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(CHRU Lille, Lille, France)

Purpose
Death from airway problems may be preventable by its early recognition, adapted assessment, and aggressive management. Airway problem is one of the major causes of morbidity and late death in patients with EA/TEF. This study aims to evaluate the incidence, the type and the consequences on the management of laryngotracheal malformations associated with oesophageal atresia.
Setting
Pediatric tertiary care center
Methods
Retrospective study including 72 consecutive patients referred for EA management between september 2003 and January 2010. Airway morphology was assessed using flexible and /or rigid laryngotracheal endoscopy.
Results
Population included 27 females and 45 males with a mean gestational age of 37 GA.
The repartition of EA according to Gross classification reported type A in 7 cases, type B in 2 cases, type C in 61 cases, type D in 2 cases.
Laryngotracheal endoscopy revealed significant tracheomalacia in 48 cases with a mean airways collapse around 65% (50 to 95%) and allowed to diagnose types B and D by identification of proximal fistula. Moreover laryngotracheal assessment showed associated upper airways anomalies in 24 patients (33%) including laryngomalacia in 8 cases, laryngeal clefts in 10 cases (grade 0 in 7 cases, grade 1 in 1case and grade 2 in 2 cases), unilateral vocal fold paralysis in 5 cases, and subglottic stenosis in 5 cases (grade 1 in 2 cases and grade 2 in 3 cases).
Postextubation respiratory distress was observed in 11 cases, related with airways obstruction in 9 cases, cardiac failure in 1 case and pneumothorax in 1 case. In the 9 patients with airways obstruction all of them presented tracheomalacia associated with subglottic stenosis in 3 cases and/or laryngeal cleft in 4 cases and unilateral vocal fold paralysis in 1 case.
Follow-up revealed persistent respiratory symptoms in 52 patients including chocking or cyanosis in 8 cases, leading to death in 4 cases. In these 8 patients, severe symptoms were attributed to cardiac cause in 2 cases, gastroesophageal reflux in 2 cases and airways lesions in 4 cases.
Finally, laryngotracheal malformations required a specific surgical management in 9 cases including aortopexy in 2 cases, proximal TEF closure in 2 cases, laryngeal cleft closure in 3 cases and laryngotracheoplasty in 2 cases.
Conclusion
In our experience, laryngotracheal malformations are frequently observed in patients with EA/TF. Some of these malformations represent a significant factor of morbidity and mortality in the management of EA. In this case, the early diagnosis of these lesions should be recommended in order to propose a planned management of the laryngotracheoesophageal tract.

A038 – Bronchoalveolar lavage fluid findings in children with congenital esophageal atresia.

TTD NGUYEN, M. POMERLEAU, C. FAURE, A. ASPIROT, S. LABERGE, J. LAPIERRE
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Introduction
Respiratory complications are common in patients with repaired esophageal atresia. Bronchoalveolar lavage (BAL) findings have not yet been reported in patients with this disorder.
Methods
We reviewed BAL findings obtained from 25 patients aged 0.5-15.7 years with congenital esophageal atresia with or without tracheoesophageal fistula. Elective flexible bronchoscopy was performed under general anesthesia as part of the clinical workup. Seven out of 25 had chronic cough and 14 out of 25 had recurrent respiratory infections. BAL was done with 1 to 3 ml/kg of normal saline instilled and suctioned. The samples were sent for bacterial culture, BAL cytology and determination of lipid-laden macrophages index (LLMI) based on the methods outlined by Colombo and Hallberg.
Results
The mean percentage of neutrophils (32.9±33.1%) was increased compared to published normal values. Patients with recurrent respiratory infections had higher percentage of neutrophils (40.7±32.6%) compared to patients without recurrent respiratory infections (23.1±32.5%) but the association was not statistically significant using the Mann Whitney U-test (p=0.058). The LLMI were variable ranging from 0 to 209. Bacterial cultures were positive in 18 samples: 1 bacterium in 9, 2 bacteria in 7 and 3 bacteria in 2 samples. Varying degrees of tracheomalacia were observed in 80% of patients.
Conclusion
BAL neutrophilia is often found in patients with repaired esophageal atresia with and without recurrent respiratory infections. BAL may add information to the physiopathology of respiratory complications in this population.

A039 – Isolated congenital tracheosophageal fistula.

G. HOSANA, N.-X. BONNE, A. BIECHLIN, P. FAYOUX
(CHRU Lille, Lille, France)

Objectives
Isolated congenital tracheoesophageal fistula ("H-type fistula") is a rare malformation that represents 3 to 6% of the oesophageal atresia / tracheo-oesophageal fistula spectrum. The aim of this study was to evaluate clinical presentation, diagnostic and treatment modalities of isolated congenital tracheoesophageal fistula
Methods
We report a retrospective analysis of 10 patients treated between 1998 and 2009 in a tertiary care centre.
Results
The mean gestational age was 38 GA (36 to 20 GA). The mean age at time of diagnosis was 20 days (1 to 116 days). Symptoms included episodic cyanosis in 10 cases, coughing with feeding in 8 cases, Gastric distension in 4 cases and recurrent pneumonia in 4 cases.
Radiological opacification was performed in 6 cases and confirm an airways opacification in 4 cases but located the fistula in only 2 cases.
Flexible laryngotracheal endoscopy confirmed the diagnosis in all cases. Rigid tracheoscopy under general anaesthesia was performed only preoperatively to conform the location of the fistula.
Associated malformations was present in 6 patients, including VACTER association, Carbimazole embryopathy , deletion of chromosome 2, laryngeal stenosis and laryngeal cleft.
Treatment was based on surgical closure by right cervicotomy. No per or post-operative complication was observed. Extubation was achieved after 3.2 days and oral feeding was reintroduced after 5.4 days. The mean follow-up was 51 months (4 months to 9 years). Follow-up revealed one case of tracheomalacia and two patients required a gastrostomy. Recurrence was observed in one case after 16 postoperative months and required a second surgical procedure with fistula closure associated with periosteum interposition.
Conclusion
The tracheoesophageal congenital isolated fistula is a rare malformation. Recurrent respiratory symptoms should alert to this diagnosis. The simplest and more efficient diagnosis method seems to be flexible laryngotracheal endoscopy. Surgical treatment by cervicotomy must be performed without any delay. A long-term follow-up is necessary because of the recurrence risk.

A040 – Conservative management of an esophageal fistula after recurrent trache-sophageal fistula in an esophageal atresia type iii: patience is needed!

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Background
Treatment of oesophageal atresia (EA) with tracho-esophageal fistula (TEF) is often associated with anastomotic complications. Short- and long-term morbidity is common in these patients and recurrent TEF represents 10% of the complications. Postoperative leaks of the esophagus are a dangerous complication after esophageal surgery. Open surgical approaches have been associated with substantial rates of morbidity, mortality, and repeated recurrences.
Case report
A premature boy weighting 2 kg was operated at birth for a type III EA associated with tracheomalacia, duodenal stenosis due to annular pancreas and right renal agenesis. We performed a successful open repair of the EA with an uneventful postoperative course and good thriving. Three months later, after a considerable Valsalva manoeuvre, the baby acutely presented with a recurrent tracheoesophageal fistula without having had any prodrome. Because of the size of this recurrent fistula, we performed an open repair by thoracotomy and placed a chest tube. Three days postoperatively we observed an important leakage in the pleural space from the oesophagus. Conservative management with parenteral nutrition, antibiotic treatment and thoracic and naso-gastric drainage was started. Complete resolution of the leak was achieved after 36 days of treatment. An upper-GI series confirmed closure of the oesophageal fistula. Per oral nutrition was reintroduced and well tolerated. Three months later an upper-GI series as well as an oesphagoscopy was repeated, showing no anastomotic stenosis.
Conclusion
In case of recurrent TEF and/or oesophageal leakage the choice between surgical, interventional (i.e. fibrin glue) and conservative treatment might be difficult. Anastomotic leakage can resolve with a conservative management, avoiding the potential complications associated with different surgical approaches. However, patience is needed!

A041 – Arterio-esophageal fistula from an aberrant right subclavian artery after esophageal stenting for stenosis after atresia repair.

R. BAIRD, D. LÉVESQUE, V. MORINVILLE, J.-M. LABERGE
(Montreal Children's Hospital, Montreal, Canada)

Background/Purpose
Post-operative management of long-gap esophageal atresia frequently mandates multiple dilations and application of local agents to control recalcitrant anastomotic strictures. Recently, temporary esophageal stenting has been employed to maintain patency. We report a significant complication from stent erosion into an aberrant right subclavian artery.
Case Presentation
A child delivered at 33 weeks, birth weight 1.3 kg, was diagnosed with long-gap atresia and underwent initial gastrostomy tube insertion followed by delayed anastomosis at two months of life. Initial post-operative esophagram revealed no evidence of leak, but a stricture at the site of anastomosis. Over the next year, the child underwent multiple upper endoscopies to manage the anastomotic stricture, including local courses of Triamcinolone and Mitomycin C. At one year of age, an esophageal stent was placed, which was changed or upsized every 1-3 weeks for three months.
The child then developed massive upper GI haemorrhage resulting in cardiorespiratory arrest. CT angiography after stabilization demonstrated extensive inflammatory changes around the stent which was abutting a previously unrecognized aberrant right subclavian artery – coursing behind the esophagus. Rebleeding in the PICU required urgent tamponade via antegrade and retrograde bougienage, and emergent angiography confirmed bleeding from the aberrant vessel (See Figure 1). This was embolized, and the esophageal stent was retrieved via flexible esophagoscopy. While haemostasis was achieved and maintained in the intensive care unit, the child manifested devastating neurologic injuries and expired 3 days after presentation.
Discussion
Aberrancy of the right subclavian artery is the most frequent vascular abnormality of the great vessels (0.5% to 1.0%). Based on our experience with this case, we advocate mandatory CT or MR angiography to rule out vascular abnormality should the utilization of esophageal stenting be considered in the treatment of esophageal strictures.

A042 – Tracheoesophageal fistula care at the alberta children's hospital: past, present and future.

L. FAIRSERVICE, L.A. SCHULTZ, M. BAILEY, C. BJORNSON, I. MITCHELL
(Alberta Children's Hospital, Alberta, Canada)

The Tracheoesophagel Fistula (TEF) Clinic at the Alberta Children's Hospital (ACH) is a unique clinic in Canada. A retrospective data review of 18 children with TEF, surgically repaired at ACH from 1994-1999 revealed numerous inpatient admissions and high healthcare needs that involved multiple programs. Coordination of services was recommended to enhance family centred care.
The first clinic began in the spring of 2005. The clinic operates one half day per month with 4-5 children seen per clinic by a multidisciplinary team. All newly diagnosed infants are seen within 1-2 months and then an average of 6 visits up until 2 years of age. Toddlers and older children are seen yearly. Twenty –two new cases have been seen since 2005. Thirty-six children are followed in the clinic.
Analysis of this population reveals that the majority of these children have oesophageal atresia with a distal TEF (type C). Twenty-three children (64%) were born premature at <37 weeks gestation, 8 (22%) have been diagnosed with VACTERL Syndrome, 3 (0.1%) have other syndromes. There are 5 sets of twins (14%).
TEF is not a simple "fixable" congenital abnormality. Gastroesophageal reflux, oesophageal dysmotility and stricture formation impact feeding and growth and require management of texture progression by the occupational therapist and dietician. Tracheomalacia complicates childhood respiratory illness, necessitating management by Respirology. The general paediatrician provides education, support and a specialized plan of care to the family and community physician. General surgery provides regular consultation to the clinic. The nurse clinician is the key contact for families. Early identification, team collaboration, family education / support and coordination of care are essential for the provision of high quality multidisciplinary care.
Future directions for the clinic include improved integration/involvement during hospitalization; establishing family connections, improved family education, and expansion of the clinic to include older children.

A043 – Recurrence of trachea-esophageal fistula with respiratory and diges tive pattern in a 6-month old boy.

S. BOILEAU, C. THUMERELLE, R. SFEIR, P. FAYOUX, L. MICHAUD, C. SANTOS, A. DESCHILDRE
(CHRU Lille, Lille, France)

We report the case of a 6-month old boy with history of esophageal atresia (EA) (Gross classification type C), who developed a recurrence of trachea-esophageal fistula (TEF) revealed by respiratory and digestive pattern. Recurrence of fistula is a rare event after EA repairs, concerning about 3% of patients. This boy had no choking or cyanosis during feeding but minimal and intermittent symptoms. Abdominal distension and pain were principal features with chronic constipation. Diagnosis of milk cow allergy or Hirschprung disease was suspected when the child was admitted for pneumonia. At admission, X-rays revealed pneumonia of the right upper lobe, and a marked gaseous abdominal distension with interposition of the right colic angle between the liver and thoracic wall without signs suggestive mechanical obstruction of the colon. Classical symptoms of the triad of Helmsworth and Pryles were present with rare coughing during feeding, pneumopathy and abdominal distension. The parents reported also an important hypersialosis for two months which is one frequent symptom in little child with TEF.

Severe abdominal distension has been reported in patients with congenital TEF of any age favoured by positive pressure ventilation during anaesthesia. Tracheal inhalation of oesophageal content may also generate high positive pressure in airways. So airflow through TEF increases gaseous in upper portion of gastrointestinal tract and may generate important bowel distension. Practitioners usually focused on respiratory symptoms, in particular cyanosis or coughing during meals, for investigation and diagnosis of a TEF. But if abdominal meteorism is extremely frequent in young child, recurrent gaseous bowel distension may evoke a TEF when associated with hypersialosis and respiratory infections, in particular if history of EA.

A044 – Use of covered adult biliary and tracheobronchial metallic stents in the management of oesophagobronchial fistulae in 2 infants and 1 toddler.

P. BICHARD, N. THOMASSIN, K. DILWORTH, C. PIOLAT
(CHU Grenoble, Grenoble, France)

Introduction
Covered self-expanding metal stents (CSEMS) have been used to manage esophageal disease in adults (strictures, leaks, fistulae). Such stents are rarely used in children.
Patients and Method
Three children who had neonatal surgery for esophageal atresia (EA) and developed a resistant anastomotic stricture with esophagobronchial fistula (EBF) were treated: 2 girls had type 1 EA (cases 1 and 2), the remaining girl had type 3 EA (case 3). The EBF in case 1 manifested at the age of 3 years following a post-operative sepsis after gastroplasty. In the other 2 children, EBF was diagnosed during an endoscopic dilatation of the anastomotic stricture (at the age of 4 and 8 months respectively). CSEMS were placed and removed under general anaesthesia with radiologic and endoscopic control. Adult biliary and tracheal stents were used.
Results
Ten CSEMS were inserted during the following period. 6 stents in case 1 (from 3.5 to 4.9 years old), 3 in case 2 (from 8 to 19 months) and 1 in case 3 (at 3 months). There was no technical failure related to stent placement or removal. CSEMS were easily removed after a mean of 2,1 months (1-6 months). The success rate of fistula closure with the stent in place was 90%. Case 3 required no further intervention and had no residual fistula or stricture. In the other 2 cases, the EBF and the stricture are well controlled when the stent was in place but recurred rapidly after his removal. After optimisation of pulmonary function, surgical resection of the anastomosis was undertaken in case 2 with successful fistula closure. The remaining child is still at home with tracheal stent in place meanwhile awaiting final surgery. 3 complications occurred (30%): pneumonia, minor haematemesis and transient dysphagia. No mortality was observed.
Conclusion
The use of CSEMS in children is a reliable therapeutic option for the management of EBF and resistant anastomotic stricture complicating EA. Type 1 EA may be associated with a higher rate of recurrence of EBF and stricture after stent removal. Yet CSEMS allow to stabilize the respiratory status in order to program esophageal surgery in better conditions.

13:00-13:30 Outcome posters

A045 – Oesophageal atresia research auxiliary (oara) – an essential contribution to follow-up care.

K. GAWNE, L. OVERDYK
(Victoria, Australie)

Oesophageal Atresia Research Auxiliary (OARA) is the first and only Australian oesophageal atresia support and research-based group. OARA was established in 1980 by a group of parents of children with oesophageal atresia and is based at the Royal Children's Hospital (RCH), Melbourne, with the aim to:
1.Raise funds for continued research into TOF/OA
2.Provide parental support to new families and existing members.

OARA have raised funds for over 30 years. Projects have included

- Adriamycin Treated Fetal Rat Model (5 yr funded project) under the guidance of Prof. Spencer Beasley.
- The set up, management and use of the Nate Myers Database which includes all RCH patients with TOF/OA since 1948.
- Funding of the employment of a RCH dedicated TOF/OA liaison and specialist nurse.
- Involvement in the formation and the ongoing support of the adult clinic at St Vincent's Hospital Melbourne, Australia.
- Respiratory medicine funding for Forced Oscillation Technique (FOT) specifically measuring lung function in young TOF/OA children.
- Funding for 6 month employment for a Research Fellow to utilise the Nate Myer's Database for research.

OARA Parental Support has included

- Publication of a biannual newsletter
- Establishment and ongoing support of a playgroup for new families
- The birthday card initiative.
- The inception of an Australian-based online social network forum for TOF/OA families
- Annual Morning tea
- Visits by committee members to RCH Neonatal intensive care unit, to offer support to new parents.
- Production of a discharge package including pamphlets and information, as well as funding for equipment and/or Pharmaceutical items.

OARA's assistance helped establish the Adult Review Clinic at St Vincent's Hospital, Melbourne.

- Dr Nate Myers' vision for establishment of an adult clinic was to allow for ongoing care of RCH patients into adulthood and help improve long-term quality of life of patients.
- Launched in April 2000, jointly with surgeons, gastroenterologists, patients and OARA committee members.
- Paper published in 2007 as an outcome of the clinic - "Gastroesophageal Reflux and Related Pathology in Adults Who Were Born With Esophageal Atresia: A Long-Term Follow-Up Study"1.

OARA are very proud to have been involved in the establishment of this clinic, which is still ongoing for current and future patients.

1. Taylor, A, Breen, K, Auldist, A, Catto–Smith, A, Clarnette, T, Crameri, J, Taylor, R, Nagarajah, S, Brady, J & Stokes, K 2007. 'Gastroesophageal reflux and related pathology in adults who were born with esophageal atresia: a long-term follow-up study'. Clinical Gastroenterology and Hepatology, Vol. 5, No. 6, pp 702-706.

A046 – The impact of oesophageal atresia disease on neonatal health service.

P. CHARLESWORTH, S. KAMALEDEEN, H. WARD
(The Royal London Hospital, London, UK)

Introduction
Oesophageal atresia (OA) is a common case of admission and referral to neonatal surgery service. Associate conditions are usual and the clinical course varies.
Aim
We aim to review the impact of OA disease on neonatal surgery service.
Methods
A retrospective review of all neonates admitted with the diagnosis of OA to our neonatal unit over the last 6 years. Data are presented as median (range)
Results
There were 47 neonates (27 male). The gestation age was 38 weeks (27-41) and birth weight was 2360 grams (998-4080). Thirteen cases had history of polyhydraminos, and in two cases OA was suspected on antenatal scans. Other anomolies were detected in 5 infants antenatally. One infant was born at our hospital and 46 infants were brought by neonatal transport service. Thirteen of these infants were referred from outside our network. Four infants had long gap atresia, and three have had delayed primary repair. One infant had both proximal and distal fistulae and one had an isolate fistula with no atresia. The remaider all had OA with a distal tracheo-oesophageal fistula. Associated anomolies were found in 61% of infants. Cardiac anomolies, on routine echo were seen in 29%, renal anomlies in 21% and spinal anomolies in 10%.
Eighteen patients required further surgery. One pateint had a recurrent fistula.
Thirteen had surgery for an additional morbidity. One infant had tracheal stenosis that required tracheoplasty. Seven patients had tracheomalcia, three of which required aortopexy. Seventeen patients developed a stricture requiring 4 (1-12) dilatations. Two developed post operative leak which were managed conservatively. Gastro-oesophageal reflux was present in 20 patients. Feeding was established by 12 (6 to 90) days. The length of hospital stay was 18 (8 to 235) days. Twenty-one patients have suffered respiratoy complications. Follow up is 25 (1 to 78) months. There were no deaths.
Conclusion
OA has a significant impact on the neonatal services. A considerable number of infants with OA have associated anomalies, require additional surgical procedures and have long hospital stay.

A047 – Ten years of longitudinal follow-up of esophageal atresia patients: what have we learned so far?

H. IJSSELSTIJN, P. MAZER, MHM VAN DER CAMMEN, CJHM MEEUSSEN, GC MADERN, D. TIBBOEL, SJ GISCHLER
(Sophia Children's Hospital, Rotterdam, The Netherlands)

Background
In 1999, we initiated a prospective longitudinal follow-up study for infants born with anatomical congenital anomalies. Amongst those were children with esophageal atresia (EA). Follow-up encompassed evaluation of medical condition and development of the child and wellbeing of parents and child by a multidisciplinary team. This team includes pediatricians, pediatric surgeons, developmental psychologists, a pediatric physiotherapist, a clinical geneticist, a social worker and ICU nursing staff taking care of a 24-hr telephone helpline.
Aim
To share experiences and emphasize the need for longitudinal multidisciplinary evaluation of EA patients.
Methods
All infants born with EA were included. Evaluation was performed at 6,12, 24 months, and 5 and 8 years.
Results
Since 1999, 122 infants were included, 11 died within the first months of life. Of 111 participants, 9 refused to join the program. Since only 12 children have been studied at 8 years so far, we focussed at evaluation of the first 5 years. Recurrent respiratory tract infections were reported in 74%, 25% of children had abnormally low FEV1. Nissen fundoplication was performed in 35%. Decreased weight and height were observed at all time points. Gross motor function was at risk or impaired in 36% of children and maximal exercise capacity was below the norm. At 24 months psychomotor and mental development were within the normal range, and results were positively related with outcome at 5 years. In 51% of EA patients parents used the telephone helpline outside office hours, mainly for feeding problems and respiratory problems.
Conclusion
With low mortality rates, a high morbidity rate is observed in surviving EA patients Since morbidity includes not only gastrointestinal and respiratory problems we assume that a multidisciplinary approach aimed at early intervention is important. Therefore, our follow-up program was extended until 18 years of age.
Ref: thesis Gischler SJ, Mazer P.

A048 – Impaired motor function and exercise capacity in children born with esophageal atresia; an evaluation at 5 years of age?

CAMMEN-VAN ZIJP, SJ GISCHLER, CJHM MEEUSSEN, D. TIBBOEL, H. IJSSELSTIJN
(Sophia Children's Hospital, Rotterdam, The Netherland

Background
Children with esophageal atresia (EA) often need prolonged hospitalization with (multiple) surgical interventions. Improvement of intensive care treatment has reduced mortality rates, but at the cost of more morbidity. Multidisciplinary follow-up is important to detect developmental and physical problems early.
Aim
To evaluate motor function and exercise capacity in children with EA at 5 years.
Methods
Survivors of EA treated between 1999 and 2003 were eligible (n=43). Exclusion criteria: severe psychomotor delay, neurological impairments, and inability to understand instructions (n=5). Lost to follow-up n=9. Thus, 29 (67.4%) children were included. The Movement Assessment Battery for Children (M-ABC) was used to evaluate motor function, the Bruce treadmill test to evaluate exercise capacity. Data shown are median (range).
Results
Gestational age 38.4 (28.6-42.0) weeks; hospital stay first 6 months 50 (11-168) days; surgical interventions in first 24 months 6 (1-18). Motor function was normal in 19 children (65.5%), borderline in 7 (24.1%) and 3 (10,4%) had a motor problem (distribution differs from the reference population and points at more serious problems in motor performance; Chi square p=0.05). Most problems were encountered in the ball and balance scales. Total percentile score of M-ABC correlated negatively with hospital stay first 6 months and with surgical interventions in first 24 months (Spearman's rho: rs = -0.44 and -0.43 respectively, $p < 0.05$). Twenty-two children performed a reliable exercise test. They performed worse than children from the reference population; SDS endurance time (mean (SD) = -0.8 (1.1); $p < 0.01$).
Conclusion
Children with EA - even without neurological impairment - are at risk for delayed gross motor function and decreased exercise capacity at age 5 years. We recommend a standard evaluation by a pediatric physical therapist at an early stage; so intervention can be started as soon as necessary.

A049 – Persistent impaired physical growth in esophageal atresia patients: a longitudinal cohort study up till 8 years.

SJ GISCHLER, CJHM MEEUSSEN, GC MADERN, D. TIBBOEL H. IJSSELSTIJN
(Sophia Children's Hospital, Rotterdam, The Netherlands)

Background
Esophageal atresia (EA) patients suffer from respiratory morbidity and gastro-esophageal reflux within the first years of life. We previously observed that EA-patients have impaired growth at the age of 5 years (Gischler, 2009)
Aim
To evaluate longitudinally physical growth in EA-patients up till 8 years.
Methods
Height, weight and body mass index (BMI) were determined at 1, 2, 5 (n=29) and 8 years (n=12) of age as part of a standardized follow-up program. Z-scores were calculated with reference data from the Dutch population and compared using a one-sided t-test (Z=0). Growth was considered abnormally low with Z-scores < -1.96.
Results
Gestational age 38 (29-42) weeks; surgical interventions in first 2 years 6 (1-18); major congenital anomalies 1 (1-4); total anomalies 2 (1-9); ventilation 3 (1-44 days). Nissen fundoplication in 9/29 (31%) patients.

Mean (SD) Z-score	height	weight	BMI
1 year (n=28)	-0.84 (1.12)*	-0.90 (0.92)*	-0.36 (1.03)
2 years (n=28)	-0.77 (1.14)*	-0.76 (0.77)*	-0.32 (0.98)
5 years (n=29)	-0.44 (1.13)*	-0.45 (0.87)*	-0.22 (0.86)
8 years (n=12)	-0.65 (1.03)*	-0.74 (1.17)*	-0.51 (1.22)

*: significantly below the norm ($p < 0.05$) Abnormally low height in 5/28 (18%) and 1/29 (3.4%) patients at 2 and 5 years, respectively. Decreased weight in 2 patients (6.9%) only at all time points. Similar growth in EA patients with or without Nissen fundoplication.
Conclusion
Although the proportion of EA patients with abnormal physical growth seems to decline after 2 years, height and weight are significantly below that of the reference population in EA patients up till 8 years. We assume that not only gastrointestinal morbidity but also respiratory problems may contribute. Prolonged multidisciplinary follow-up with early intervention is needed.

A050 – Quality of life and behaviour in children with esophageal atresia.

P. MAZER, A. VROEGINDEWEIJ, H. IJSSELSTIJN, CJHM MEEUSSEN, D. TIBBOEL SJ GISCHLER
(Sophia Children's Hospital, Rotterdam, The Netherlands)

Background
We examined quality of life and behaviour in children with esophageal atresia (EA) at age 5 years.
Methods
Mothers and fathers of 21 five-year-old children with EA completed the Dutch version of the Pediatric Quality of Life Inventory (PedsQl) and the Child Behavior Checklist (CBCL). Dutch norm scores were used.
Results
Twenty-one children, of whom 71,4% were boys, participated in this study. Total number of major and minor anomalies per child ranged from one to three (IQR) with a median of two. Median duration of admission in the first 6 months was 40 days. Children underwent a median of four surgical interventions in the first 24 months with a range from 1 to 8 (IQR).
One mother and 1 father, from a different family, refused to fill in the PedsQl. In three children one or both parents refused to fill in the CBCL. With respect to health related quality of life, children with EA scored significantly lower than norm scores on the subscales social functioning and school functioning ($p<0.05$). On the other subscales: psychosocial health, physical and emotional functioning children with EA did not differ from norm scores. Paired measurements (n=19) showed no significant differences between scores reported by mothers and fathers. All mothers reported behaviour in the normal range. Fathers in 5 children reported behavioural problems: 3 children with internalizing problems within the borderline range and 1 child with externalizing problems within the clinical range. In one child internalizing and externalizing problems within the clinical range occurred.
Conclusion
At 5 years of age, social and school functioning in children with EA is significantly lower than in healthy children. Mothers reported no behavioural problems, whereas fathers reported internalizing and externalizing problems in 23.8% of the children. Internalizing problems occurred slightly more than did externalizing problems.

A051 – Prevalence of gastroesophageal reflux disease (gerd) and associated complications in adults born with esophageal atresia.

S. MAYNARD, M. POMERLEAU, C. FAURE, M. BOUIN
(Hôpital Sainte-Justine, Montréal, Canada)

Background
GERD is one of the most frequent digestive complications in adult born with esophageal atresia. However, only few studies exist on the outcome and long term consequences of GERD in this population.
Aim
Assess the prevalence of GERD and its complications in adults who underwent surgical treatment for EA at birth.
Methods
Patients over 18 years operated for EA at Ste-Justine's hospital (paediatric tertiary center, Québec, Canada) from 1951 to 1991 were contacted by mail. Respondents were invited for a consultation with a specialized physician. The protocol included standardized questionnaires on gastrointestinal symptoms, an oesophagoscopy and a 24-h pH-metry.
Results
We received 56 responses (20%), out of the 283 letters that were sent (381 pts operated less 98 deceased/incorrect addresses). 33 subjects (16 women) were included (23 refused) with a mean age of 27±7 yrs. The prevalence of GERD symptoms was 61% (n=20). Proton pomp inhibitors were used by 10 pts (33%). Upper gastrointestinal endoscopy was performed in 22 patients (67%). No strictures were found, but scar of surgical anastomosis was still recognizable in most cases of primary anastomosis. The other findings were: hiatal hernia in 36% (n=8), oesophageal diverticulum in 11% (n=3), oesophagitis in 27% (n=6), Barrett endoscopically suspected in 23% (n=5). Systematic biopsies confirmed intestinal metaplasia in 4 cases. There was no dysplasia or adenocarcinoma. In the stomach, aberrant pancreas was found in 18% (n=4) and H. Pylori was present in one patient. A 24-h pH-metry was performed in 13 pts. Seven (54%) had a pathological pH measurement (Meester's score $\geq$ 14,72). Of these abnormal pH-Metry, the percentage of recording time with pH <4 was 12,9% (117 mn/24 h) with a range of 3,9%–45,7%. Five of the 7 patients with abnormal pH-metry did not reported symptoms of GERD.
Conclusion
Adults who underwent surgical treatment in childhood for EA have a high prevalence of GERD. This study also shows a high prevalence of complications of GERD like oesophagitis and Barrett oesophagus. These findings suggest that a special monitoring including endoscopic screening may be beneficial in this population.

A052 – Upper gastrointestinal symptoms and health-related quality of life in adults born with esophageal atresia.

S. MAYNARD, M. POMERLEAU, C. FAURE, M. BOUIN
(Hôpital Sainte-Justine, Montréal, Canada)

Background
EA is the most common congenital anomaly of the oesophagus. However, only few studies exist on the outcome and long term consequences of this malformation in adulthood.
Aim
Assess the prevalence of upper GI symptoms in adults who underwent surgical treatment for EA at birth. The secondary objective was to measure the long-term quality of life (QOL) of these adults.
Methods
Patients over 18 yrs old operated for EA at Ste-Justine's hospital (paediatric tertiary center), from 1951 to 1991, were contacted by mail. Respondents were invited for a consultation with a specialized physician. Subjects answered standardized questionnaires of GI symptoms and QOL (SF-12 questionnaire).
Results
We received 56 responses (20%), out of the 283 letters that were sent out (381 patients operated less 98 deceased/incorrect addresses). 33 subjects (16 women) were included (23 refused), with a mean age of 27±7 yrs. Proton pomp inhibitors were used by 10 pts (33%).
The prevalence of symptoms found in our cohort is the following: pyrosis 42% (n=14), regurgitation 48% (n=16), pyrosis or regurgitation 61% (n=20), dysphagia 67% (n=22), food impactions 48% (n=16), odynophagia 21% (n=7), post-prandial fullness 52% (n=17) and post-prandial abdominal distensions 33% (n=11). Adaptative dietary habits were noted among 64% (n=21) of patients who needed to drink fluids to facilitate swallowing (1/2 to all meals), 42% (n=14) ate more slowly than their surroundings and 12% (n=4) avoided certain foods.
The average scores of QOL were compared to those of the general population. With a 95% confidence interval, no difference was found for both the Physical health Composite Score (50,33±2,8 vs. 50,8) and the Mental health Composite Score (52,03±3,33 vs. 52,4).
Conclusion
Adults who underwent surgical treatment in childhood for EA have a high prevalence of GI symptoms, in particular gastroesophageal reflux and dysphagia. However, QOL scores are not different from those of general population. Our study highlights adaptative dietary habits in 2/3 of patients and is in favour of the need to develop specific questionnaires adapted to this population. Complimentary studies are necessary to assess complications of long-term GERD and to evaluate the existence of associated oesophageal motility disorders.

A053 – Quality of life following surgery for esophageal atresia: once upon a time the gap.

L. SPAGNOL, F. MORINI, B. D IACOBELLI, P. ATZORI, A. TRUCCHI, A. BRAGUGLIA, P. BAGOLAN

Aim of the study
Children operated on for long gap esophageal atresia (LEA) are reported to experience a higher prevalence of complications than their "normal" gap counterparts. No data are available on the long term quality of life (QOL) of children operated on for long gap EA with preservation of the native esophagus. Our aim was to compare the QOL of children with long gap EA with that of children with "normal" gap EA, after primary esophago-esophageal anastomosis.
Methods
A questionnaire including a modified version of the Gastrointestinal Quality of Life Index (GIQLI) and the Pediatric Quality of Life Inventory (PedsQL™ 4.0, a standardized instrument to assess health related QOL in pediatric patients with chronic health conditions) was sent to the parents of all patients consecutively treated for EA with or without tracheo-esophageal fistula between 1995 and 2003. Patients were divided based on the presence of a long (>3 cm or 3 vertebral bodies) or "normal" gap esophageal atresia, and differences in QOL between the 2 groups were evaluated by Mann-Whitney test. Results are medians (interquartile ranges).
Results
In the study period, 99 consecutive patients (29 with long gap) were treated for EA. Seven died and 13 were lost to follow-up. The questionnaire was sent to 79 (with 5 to 13 years of follow-up) and returned by 59 families (75% response rate). Both the GIQLI and PedsQL™ 4.0 showed no significant differences in the QOL between the 2 groups (table).
Conclusion
Children operated on for long gap EA with primary esophago-esophageal anastomosis have a QOL similar to their "normal" gap counterparts. Long gap EA should be treated by primary/shortly delayed repair and anastomosis in tertiary-level centres. Esophageal substitution should be reserved for cases with failed previous attempts to preserve the native esophagus.

	"Normal" gap (42 patients)	Long gap (17 patients)	p
GIQLI (120 points)	106.0 (101.0-114.0)	111.0 (91.5-114.0)	0.87
PedsQL™ 4.0 (100 points)	89.0 (81.5-93.6)	86.2 (69.2-93.8)	0.45

A054 – Symptomatic vocal cord paralysis in infants operated on for esophageal atresia and/or tracheo-esophageal fistula.

L. VALFRÈ, F. MORINI, B. D. IACOBELLI, A. CROCOLI, S. BOTTERO, M. TROZZI, A. BRAGUGLIA, A. CONFORTI, P. BAGOLAN.

Aim of the study
Temporary dyspnea and stridor are common after esophageal atresia (EA) repair. Although a number of causes have been implicated in their pathogenesis, the role of vocal cord paralysis (VCP) has been rarely studied. Aim of our study was to describe the prevalence and pathogenesis of symptomatic VCP in a large cohort of patients (pts) operated on for EA and/or tracheo-esophageal fistula (TEF).
Methods
Retrospective review of all consecutive pts treated for EA and/or TEF between August 1995 and June 2009. Symptomatic VCP suspected on the basis of persistent stridor, dyspnea, and/or dysphonia/aphonia, was confirmed by flexible laryngoscopy. Pts with and without VCP were compared for several variables (table) by Mann-Whitney or Fisher's exact test as appropriate. Results are medians (interquartile ranges) or prevalence, $p<0.05$ was considered significant. Results: During the study period 174 consecutive pts with EA and/or TEF were treated; symptomatic VCP was detected in 7 (4%).
Conclusions
VCP is a rare but might be an important cause of respiratory morbidity in patients treated for EA and/or TEF, that should be ruled out in case of persisting symptoms. Surgical factors that may increase the risk of recurrent laryngeal nerve injury, such as cervical esophagostomy, should be limited as much as possible. Further systematic studies are needed to define the prevalence of acquired asymptomatic and congenital VCP in pts with EA and/or TEF.

	VCP +	VCP -	p
GA (wks)	37 (32-38)	38 (36-40)	0.36
BW (kgs)	2.6 (2.5-3.4)	2.5 (2.1-3.0)	0.22
Other anomalies (N)	4	118	0.43
Long gap (N)	5	41	0.02
Esophagostomy (N)	5	7	<0.01
Anast. Leak (N)	3	10	<0.01
Azygos ligation (N)	0	48	0.19
Major cardiac surg (N)	0	4	1.00

A055 – Long term outcome of children with esophageal atresia.

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The aim of the present study was to assess the outcome of patients with esophageal atresia (EA), with a special emphasis on quality of life and the presence of late sequelae.

Methods

One hundred and three patients with EA were treated in our institution during a 10-year period (January 1989-December 1998). Study parameters included patients' demographics, type of EA, associated malformations, gastroesophageal reflux (GER), nutritional status, presence of digestive, respiratory and orthopedic symptoms, pulmonary function tests (PFT) and quality of life (PedsQoL 4.0 questionnaire).

Results

Sixty-three patients (mean age: 13.4 years; range: 2.7) accepted to participate to the study. Ten had died between 1 to 510 days of life. Thirty patients were not included (23 were lost to follow-up, 7 refused to participate). Non participants (30%) presented the same characteristics than the 70 % participating patients. Eighty-two percent of the patients had EA type III; 35 % underwent fundoplication and 45 % presented with anastomotic stenosis. At he last follow-up, 73% had a normal nutritional status whereas 16 % were obese and 11% undernourished). Only 13 % of them were free of any digestive symptoms: 65 % had dysphagia and 35 % had symptoms of GER at last follow-up. Main respiratory symptoms were chronic cough (19 %) and dyspnea on exertion (33 %). Only 38 % of patients had no respiratory symptoms. PFT showed that 50 % of patients had proximal obstruction and/or pulmonary distension whereas 11 % had restriction syndrome. The quality of life score was rather good but lower than controls (80 vs 84, $p<0.05$). QoL scores were lower when the patient had associated congenital heart disease (71 vs 85, $p=0.01$) or respiratory symptoms (75 vs 87, $p=0.04$).

Conclusion

The high frequency of late sequelae in EA justifies regular and multidisciplinary follow-up throughout adulthood.

A056 – Long term follow up for patients with livaditis procedure.

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Introduction

The long gap esophageal atresia (EA) remains a challenging problem. Livaditis (proximal pouch esophagomyotomy) introduced the operation of relaxing circular myotomy as an effective means of bridging such a wide gap.

Methods

We reviewed the data of 4 patients with Livaditis procedure from 1993 till 2000 and their follow up. All patients were still followed by our team.

Results

Three patients had EA with distal fistula and one patient without fistula. Livaditis procedure was done as first procedure in all type III atresia, and at 1 month for the type I. All patients had multiple dilatations for esophageal stenosis and all patients had a severe GER and Nissen procedure was done between 5 mo and 5 yo. At the time of the follow up, all patients presented variable degree of dysphagia. In one case, endoscopy showed a heterotopic gastric mucosa in the upper esophagus and symptoms relieved after proton pump inhibitors treatment. Mean follow up was 13 years (10-17 yo). Body Mass Index of patients were 12.6 at 10 yo, 20 at 13 yo, 35.6 at 14 yo, 25 at 17 yo respectively. All patients are still under respiratory and GER medications.

Conclusions

There are very few long follow up of these patients in literature. Our small series suggest that this lengthening procedure may be a solution for the long gap problem but on the long term persistent gastrointestinal and respiratory suggest a longitudinal follow up with adult medicine passing on.

A057 – An innovation to a multidisciplinary follow up clinic for esophageal atresia patients.

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Objective

In order to improve the long term care of patients with operated esophageal atresia (EA) a multidisciplinary clinic was created. This was done to provide a favourable environment where parents could learn about the disease of their child and where the child can be assessed by multiple specialists on the same visit. Subsequent evaluation of the clinic was performed by the parents.

Methods

An outpatient clinic was established, taking place every two weeks, where the child is assessed by a gastroenterologist, pulmonologist, surgeon, and nurse specialized in the treatment of EA. Following each clinic a multidisciplinary meeting is held where individual cases are discussed and a treatment plan is formulated. A follow up phone call his made to the parents by the nurse to convey the treatment plan. Patients are followed up until the age of 18yrs, when a transition to adult care is done in collaboration with adult gastroenterologists. A survey was mailed to families in order to evaluate their satisfaction with regards to the structure and functioning of the clinic.

Results

The survey was distributed to 60 families, with a 75% response rate. The response was of a overwhelming success with 91% rating their care with the clinic as very good or excellent. 84% felt the environment was excellent, 87% consider the participation of the team was outstanding and 94% thought the attitude of the team was tremendous. Only 67% felt that they had received sufficient information at the clinic and most would have requested more information. The major complaint was suboptimal access to the clinic, parking and administration.

Conclusion

A collaborative approach is an excellent way to follow patients with complex medical conditions. The multidisciplinary clinic runs efficiently and the results of the survey demonstrate a very high level of satisfaction of parents with children with EA. A nutritionist was added to the team after receiving feedback from the parents from the survey, which was widely appreciated. We are currently developing an information brochure for parents to optimize distribution of information.

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