372. Paediatric bronchology

P3334

HRCT features do not predict the clinical course in children with protracted bacterial bronchitis

Konstantinos Douros[1], Efthimia Alexopoulou[2], Aggeliki Nicopoulou[1], Michael Anthracopoulos[3], Vasilios Grammeniatis[1], Marios Papadopoulos[1], Konstantinos Priftis[1]. *[1]3rd Department of Paediatrics, Attikon Hospital, Athens, Greece; [2]Second Department of Radiology, Medical School, University of Athens, Attikon Hospital, Athens, Greece; [3]Respiratory Unit, Department of Pediatrics, University Hospital, Rio - Patras, Greece*

Background/Aim: Chronic wet cough suggests endobronchial infection. We

aimed to investigate the relation between the initial radiological findings and clinical course as well as the evolvement of radiological findings in patients whose clinical findings persisted despite treatment.
Methods: We retrospectively reviewed 90 patients aged 0.6 to 16.4 years, with chronic endobronchial infection. In 25 (27.8%) patients follow up HRCT scan was performed (1st group), 6-38 months apart (median 13 mo) based on clinical grounds, mainly on the duration of symptoms despite long courses of antibiotic treatment and physiotherapy; the remaining 65 (72.2%) had a remitting course and a second scan was not considered necessary (2nd group). Severity of involvement was assessed with Bhalla score and presence of bronchiectasis, *per se*.
Results: Radiological findings on the first HRCT did not differ between the 2 groups (Bhalla scores: 2.7±0.3 and 2.6±0.3, p=0.80; presence of bronchiectasis: 10 and 27, p=0.9 in the 1st and 2nd group, respectively). In 1st group 10 children had bronchiectasis in the 1st HRCT scan compared with 14 in the second scan (p=0.10). Bhalla scores did not differ between the 1st and the 2nd CT-scan (mean values±sd: 2.7±0.2 and 2.9±0.4, respectively, p=0.60). There was no difference in bronchoscopic/bronchoalveolar lavage findings.
Conclusion: Radiological findings do not suffice to predict the clinical course of protracted bacterial bronchitis. Although the literature suggests that radiological findings may be reversible, this is not the rule. Apart from the intensity of treatment, there are probably other - as yet unidentified- factors that determine the final outcome of the disease.

P3335
Bronchoscopic findings in children with primary ciliary dyskinesia: Most but not all patients have bacterial bronchitis
Helen Burmester, Folke Brinkmann, Nicolaus Schwerk, Gesine Hansen.
Pediataric Pneumology, Medical University of Hannover, Germany

Chronic bacterial infections of the lower respiratory tract (LRTI) are a relevant problem in patients with primary ciliary dyscinesia (PCD) and therefore a continuous antibiotic treatment regime is often used. However, it remains unclear if all of these children indeed have relevant chronic LRTI and thus profit from antibiotics. Data on bronchoalveolar lavage fluid (BALF) with cytological and microbiological analysis in PCD patients are still lacking.
In a retrospective study BALF findings of 19 children with PCD were analyzed and the clinical course of patients treated with antibiotics has been investigated.
Median age of the patients at the time of bronchoscopy was 10 year (range 4-17 years). Situs inversus was seen in 10 patients. The main symptoms leading to bronchoscopy were chronic cough, recurrent bronchitis or pneumonia. Lung function was performed in 17 patients before bronchoscopy. FEV1 ranged between 62-114% (median 80%). In the BALF of 12 patients significant bacterial counts ($>10^4$ CFU/ml) have been confirmed. *Haemophilus influenzae* (n =10) and *Streptococcus pneumoniae* (n= 3) were the most frequent isolated species. In all of the cases where a BALF-cytology was performed a granulocytic inflammation was detected. Furthermore, these findings correlated with a worse lung function before bronchoscopy (FEV-1 <80%) and a significant improvement of lung function under antibiotic therapy.
Bronchoscopy is a safe diagnostic method in children with PCD to distinguish between patients with LRTI from these without infections. This has an extensive clinical relevance because we could show that patients with a proven infection profit from an antibiotic therapy.

P3336
Computed tomography and flexible bronchoscopy techniques for assessment of tracheomalacia in children
Gerasimos Kremmydas[1], Vasilios Grammeniatis[2], Konstantinos Douros[2], Michaeil Anthracopoulos[3], Marios Papadopoulos[2], Argyro Mazioti[2], Konstantinos Priftis[2], Efthimia Alexopoulou[1]. *[1]Second Department of Radiology, Medical School, University of Athens, General University Hospital 'Attikon', Athens, Greece; [2]3rd Department of Paediatrics, Attikon Hospital, Athens, Greece; [3]Respiratory Unit, Department of Pediatrics, University Hospital, Rio - Patras, Greece*

Background/Aims: Tracheomalacia is not an unusual diagnosis in pediatric respiratory clinics. We tried to find the most suitable approach to the evaluation and diagnosis of "brassy","*barking*"cough.
Methods: We performed a dynamic helical CT scan (HCT) in 15 patients (aged 5 to 14 years) suspected of tracheomalacia on the grounds of clinical symptoms (barking cough ± recurrent chest infections). All patients underwent flexible bronchoscopy (FB) under deep sedation and spontaneously breathing. Four children who suffered from various parenchymal lung diseases but no "*barking*" cough were used as controls. The ratio of anteroposterior/tranverse diameter was measured in the thoracic inlet and the carina level, in full inspiration and end expiration.
Results: Flexible bronchoscopy confirmed the existence of tracheomalacia in all patients; in 5 out of 15, lesions were located in the upper part of the trachea as shown by FB. HCTs showed localized narrowing of the *intrathoracic trachea in 7 patients*. The measured ratios were lower in the patient group as follows: at the carina level in inspiration 0.81±0.08 and 0.97±0.08, p=0.001, and at expiration 0.66±0.13 and 0.89±0.12, p<0.001, for patients and controls, respectively; at the thoracic inlet in inspiration 0.96±0.13 and 1.12±0.11, p=0.023, and at expiration 0.77±0.25 1.02±0.10, p=0.033, for patients and controls, respectively.
Conclusions: FB is valuable in the assessment of patients with extrathoracic tracheomalacia since HCTs cannot be recommended as a safe approach (radiation of the thyroid). However, CT scans provide a more accurate estimation of endothoracic tracheomalacia as it is not influenced by the effects of general anaesthesia.

P3337
Bronchoscopic findings and interventions in patients with long-term tracheostomy
Andreas Pfleger, Ernst Eber. *Department of Paediatrics and Adolescence Medicine, Medical University of Graz, Austria*

Aims: To describe airway abnormalities identified by flexible bronchoscopy (FB) in patients with long-term tracheostomy (LTT) and interventions as a consequence of FB findings.
Methods: Records of patients with LTT followed from Jan 08 to Dec 11 were reviewed. FBs were performed as routine surveillance FBs, additional scheduled FBs, or because of disease or tracheostomy related complications. Resulting interventions (ventilator -, cannula -, or medication changes, and surgical interventions) and extra caregiver trainings were recorded.
Results: In 52 patients (20 f, 32 m) 210 FBs were performed. 30 patients had LTT for long-term ventilation, 22 as a bypass for upper airway obstruction. Median age was 4.5 yrs (0.1-32.7). In 97 instances FBs were performed transnasally, in 93 via the cannula, and in 20 via both routes. In 13 instances (6%) complications led to FBs; in 23 (11%) additional scheduled FBs, and in 174 (83%) surveillance FBs were performed. The mean frequency of FBs was 1.1/patient and year (0.25-2.7). The most common findings were airway malacia in 38%, clinically relevant granulation tissue in the suprastomal region in 8%, at the end of the cannula in 7%, and in other regions in 13%. Cannula changes were performed in 21%, ventilator changes in 4%, and surgical interventions in 3%. 12% of the caregivers received extra training on correct suction techniques.
Conclusions: In this series of patients with LTT we found a high incidence of airway abnormalities. As FB findings resulted in interventions in a quarter of our patients we recommend that FB should be performed at least once a year. Patients with significant airway pathology, however, may benefit from more frequent endoscopic evaluations.

P3338
Nasal nitric oxide measurement using continuous aspiration by hand-held device discriminates patients with primary ciliary dyskinesia from healthy subjects
Silvia Montella[1], Kjell Alving[2], Sara De Stefano[1], Laida Lisa Di Micco[1], Angela Di Giorgio[1], Francesca Santamaria[1]. *[1]Department of Paediatrics, Federico II University, Naples, Italy; [2]Department of Women's and Children's Health, Uppsala University, Uppsala, Sweden*

Background: Low nasal nitric oxide (nNO) has been reported in subjects with primary ciliary dyskinesia (PCD). Thus, nNO measurement has been proposed as a diagnostic tool to screen for PCD. The best validated method for nNO assessment is aspiration at a constant flow rate from one naris (Am J Respir Crit Care Med 2005;171:912-30). Traditionally, nNO is obtained using stationary chemiluminescence analysers. Hand-held electrochemical devices have been used in PCD for nNO analysis using the nasal exhalation method (silent and humming exhalation). No study compared nNO measured using continuous nasal aspiration in PCD and healthy subjects by a hand-held device.
Aim: To find out whether nNO measured by a hand-held analyser using the continuous aspiration method discriminates PCD from healthy subjects.
Methods: Twenty-three PCD patients (median age, 15.8 yrs; range, 4.6-32.8) and 23 healthy controls (age, 15.7 yrs; range, 4.3-32.1) measured nNO with a hand-held electrochemical device (NIOX MINO®, Aerocrine AB) during oral breathing through a mouthpiece.
Results: Median (range) nNO values were 12 (5-62) and 506 (215-777) ppb in PCD and controls, respectively ($p<0.001$). Sensitivity and specificity at different cut-off points for nNO are reported in the Table.

Cut-off points (ppb)	Sensitivity (%)	Specificity (%)
36	78	100
44	83	100
51	87	100
53	91	100
58	96	100
138	100	100

Conclusion: Measurement of nNO by the hand-held device using the continuous aspiration method has an excellent sensitivity and specificity in distinguishing PCD from healthy subjects. Its wider use might result in an increased number of detected individuals suspected to have PCD.

P3339

Carbon in the airway macrophages of children affected by chronic lower airway disease

Deborah Snijders[1], Laura Bottecchia[1], Veronica Storer[1], Erika Bazzan[2], Elena Mutti[2], Serena Calgaro[1], Angelo Barbato[1]. *[1]Department of Pediatrics, University of Padova, Italy; [2]Department of Cardiac, Thoracic and Vascular Sciences, University of Padova, Italy*

The effect of air pollution can play a role in the development and/or the exacerbations of chronic lower airway diseases (CLADs) in children.
The aim of this study is to investigate the association between the major component of inhalable particulate matter (PM10), black carbon, contained in lower airway macrophages (AM) of children with CLADs and variables that may affect individual exposure.
We studied 24 children undergoing FOB for CLADs. For each child the area of the black material (BM) in AMs, sampled by bronchoalveolar lavage, was determined by analysis of digitized light microscopic images of random AMs; median carbon content of AMs was obtained calculating the area (μm^2) of BM.
Children had a median age of 6 years (range 2,9-16,6); they were affected by recurrent lower airways infections in 12 cases (PCD in 2), by asthma in 7, by chronic cough in 3 and by laryngospasm in 2 cases. There was a correlation between the median area of BM and air PM10 levels during the week before FOB ($p<0,001$). No significant correlation was found between FEV1 measured in 14 children. No significant difference in black area size in AMs was detected comparing children based on sex, underlying disease or household. Preschool-children had a larger median black area in AMs than school-children, even if the difference was not significant ($0,671 \mu m^2$ vs $0,352 \mu m^2$, $p=0,35$).
In conclusion the median area of BM in AMs is correlated to air pollution exposure in children affected by CLADs, but it seems not to correlate to underlying diseases, age, sex and FEV1. A larger study group is necessary to better define the correlation of PM10 and CLADS in children.

P3340

Foreign body aspiration in Tunisian children: Clinical, radiological and endoscopic features and outcome

Anissa Berraies, Jamel Ammar, Hanadi Abid, Essaied Wafa, Adel Marghli, Taher Mestiri, Agnès Hamzaoui. *Pavillon B, Abderrahmen Mami Hospital, Ariana, Tunisia Pavillon B, Abderrahmen Mami Hospital, Ariana, Tunisia Pavillon B, Abderrahmen Mami Hospital, Ariana, Tunisia Pavillon B, Abderrahmen Mami Hospital, Ariana, Tunisia Thoracic Surgery, Abderrahmen Mami Hospital, Ariana, Tunisia Anesthesia, Abderrahmen Mami Hospital, Ariana, Tunisia Pavillon B, Abderrahmen Mami Hospital, Ariana, Tunisia*

Foreign body aspiration is a frequent accident among children under 3 years. Delay in diagnosis may lead to serious complications. The aim of this study is to describe the experience of a pediatric respiratory department in managing foreign body aspiration and to identify predictive factors of complications.
In 10 years (from 2002 to 2011) we performed 101 flexible bronchoscopies for suspicion of foreign body aspiration. A FB inhalation was confirmed in 75 children with a mean age of 32 months and a sex ratio of 1.8. The FB was visualized in 64 cases and in three cases the the FB was'nt seen in first-line rigid bronchoscopy. Twenty three percent of children consult after 1 month. The common clinical manifestations of FB aspiration were persistent cough (66%) and history of choking (81%). The FB is in the right bronchial tree in 49.33% of cases and an inflammatory polyp was seen in 28 children. The most common FB was seeds and nuts: sunflower seeds in 12 cases and almonds in 5. Six children inhaled a scarf pin all of them in the last two years because of the change of clothing habits in our society. Fourteen Children developed bronchiectasis and 6 of them required resection. The organic nature of the FB is associated with this complication ($p=0.01$). In the group of children who developed complications, the time to diagnosis was longer 78 vs 43 days (NS). Inflammatory polyps were not predictors of complications.
Diagnosis of FB aspiration in children is difficult and misdiagnosis as respiratory infections may delay treatment and cause morbidity. Flexible bronchoscopy is a safe procedure and should be performed in suspicious cases.

P3341

Removal of foreign bodies by rigid bronchoscopy technique in children – The end of an era?

Dirk Schramm, Julia Delißen, Antje Schuster. *Department of Paediatric Cardiology and Pneumology, University Hospital Düsseldorf, Düsseldorf, Germany*

BACKGROUND/HYPOTHESIS: Recommendations regarding management of foreign body (FB) aspiration in childhood have not substantially been modified since 1897 when Gustav Kilian first described the technique how to extract FBs by rigid bronchoscopy. Many paediatric pulmonology centers have adapted a two-step-procedure: First, to explore the airways by flexible bronchoscopy in case of suspicion of a FB aspiration, and then, if confirmed, to switch to rigid bronchoscopy for FB removal. In recent years, instruments have been developed especially for FB removal by flexible bronchoscopy. We hypothesize that it is no longer necessary to routinely pursue the two-step procedure.
Case: We present the case of a 2-year-old boy who aspirated a peanut. In order to exemplarily share our experiences regarding the modern technique, we display serial photographs from the video taken during the procedure of FB removal by specialized instruments for flexible bronchoscopy. In our center, this flexible technique has developed into the usual method of choice; only in rare cases it is necessary to switch to a rigid bronchoscopy procedure.
Discussion: There are only few reports on the novel flexible method for FB removal in children. A safe one-step procedure with flexible bronchoscopy offering both a diagnostic and a therapeutic potential appears to be an advantageous method for the management of our young patients. We suggest to compile and analyze paediatric pulmonologists' experiences with the new technique, in order to finally formulate recommendations for the most advisable first-line procedure of bronchial FB removal in children.

P3342

A new mutation of surfactant protein C gene causing severe respiratory insufficiency and pulmonary fibrosis

Francesca Petreschi[1], Teresa Salerno[1], Donatella Peca[2], Maria Beatrice Chiarini Testa[1], Alessandra Schiavino[1], Maria Giovanna Paglietti[1], Olivier Danhaive[3], Renato Cutrera[1]. *[1]Department of Paediatrics, Bambino Gesù Children's Hospital, IRCCS, Rome, Italy; [2]Research Laboratory, Bambino Gesù Children's Hospital, IRCCS, Rome, Italy; [3]Department of Neonatal Medicine and Surgery, Bambino Gesù Children's Hospital, IRCCS, Rome, Italy*

The pulmonary surfactant is an unique phospholipid and protein complex that is synthesized, packaged and secreted by alveolar type II cells. The phospholipid compontents constitute approximately 90% weight of pulmonary surfactant, while the remaining 10% is constituted by protein components, including surfactant-associated proteins SP-A, B, C and D. Genetic mutations in surfactant production and function are associated with different clinical phenotypes and can cause significant primary lung disease in full-term infants, older children and adults.
Mutations in the surfactant protein C gene have been recently associated with the development of diffuse lung disease, particularly sporadic and familial interstitial lung disease.
SC is a one year old girl, who presented acute respiratory insufficiency at 6 months of age during an acute bronchiolitis, requiring intubation and mechanical ventilation. CT scan showed bilateral ground glass appearance and siderocytes were found in BAL fluid. She was referred 6 months later in poor general conditions, failure to thrive and oxygen dependent. Cystic fibrosis was excluded by negative sweat tests and CFTR mutations study. CT scan confirmed diffuse interstitial lung disease and "honeycombing" appearance. Pulmonary biopsy showed an interstitial lung disease with lamellar bodies anomalies typical of surfactant proteins deficiency.
The genetic mutations analysis showed a new mutation in the exon 3 of the gene encoding SP-C.Patients with severe respiratory failure and/or interstitial lung disease should be always investigated for surfactant protein deficiency, particularly if other more common diseases are excluded.

P3343

Identification of KCNRG, a bronchial autoantigen, in a children with IPEX syndrome

Véronique Houdouin[1], Didier Chevenne[2], Cecile Raverdy[3], Souhir Zaoui[2], Bénedicte Neven[4], Nadia Tubiana[3], Mohammad Alimohammadi[5], Jean Claude Carel[3]. *[1]Department of Pediatric Pulmonology, Hôpital Robert Debrè, Paris, France; [2]Laboratory of Biochemistry and Hormonology, Hôpital Robert Debrè, Paris, France; [3]Department of Pediatric Endocrinology, Hôpital Robert Debrè, Paris, France; [4]Department of Pediatric Immunology and Rheumatology, Hôpital Necker-Enfants Malades, Paris, France; [5]Department of Medical Sciences, University Hospital, Uppsala, Sweden*

IPEX syndrome is a rare disorder of immune regulation caused by mutations in the FOXP3 gene, which is required for the suppressive function of naturally arising CD4 + CD25 + regulatory T cells. It is associated with the presence of common autoantibodies associated with autoimmune disorders.
We report an infant, who manifested at first week after birth a type 1 diabetes mellitus and eczema. IPEX syndrome was confirmed by proved V408M mutation in the FOXP3 gene. At one month old he developped wheezing, cough, respiratory distress without infection. Despite treatment with salbutamol and budesonide nebulized, he was still symptomatic. High-resolution tomography scan showed hyperinflation, trapping, and ground-glass opacities. Analysis of bronchoalveolar lavage showed 1 350 000 cells/ml, with 65% alveolar macrophages without infection. Autoantibodies to KCNRG were present in serum at one month and still persist at nine months. Treatment with mycophenolate mofetil was started because of combination of severe diarrhea, persistent respiratory symptoms with an optimal nebulized corticotherapy. One month after the begining of immunosuppresive therapy the corticotherapy was reduced.
KCNRG, is a potassium channel regulating protein expressed in bronchial epithelial cells. The presence of this antibody was first decribed in the polyendocrine syndrome type 1. The presence of this antibody in an other autoimmune syndrome confirms the fact that KCNRG is a major bronchial autoantigen.The recognition of pulmonary autoimmunity, and its distinction from asthma is important because the autoimmune bronchiolitis in this case respond well to immunosuppression.

P3344

Pulmonary alveolar proteinosis due to a novel mutation in CSF2RA

C. Happle, M. Wetzke, A.M. Dittrich, G. Hansen, N. Schwerk. *Department of Paediatric Pneumology, Allergology and Neonatology, Hannover Medical School, Hannover, Germany*

Pulmonary alveolar proteinosis (PAP) is a rare disease characterized by pulmonary accumulation of surfactant protein. Congenital forms can result from mutations in granulocyte macrophage-colony stimulating factor (GM-CSF) receptor genes, leading to a terminal differentiation block of alveolar macrophages.
We present the case of a 3yr-old girl born to consanguineous parents presenting with progressive dyspnoea, cough and failure to thrive. Her arterial oxygen saturation was 80% while breathing ambient air and dropped to 50% during agitation. Chest radiographs showed bilateral opacities, and high-resolution computed tomography (CT) revealed interlobular densification with typical "crazy paving" pattern. Due to a milky, opaque appearance of bronchioalveolar lavage fluid (BALF) and a strongly PAS-positive staining in histology, the diagnosis of PAP was suspected. After whole lung lavage (WLL), significant clinical improvement occurred. Oxygen saturation increased to >90% and follow-up chest radiographs showed partial clearance.
Currently, the patient is undergoing WLL every 4-6 weeks. After 10 months of treatment, she has gained 9 kg of weight, visits kindergarten, and has a good quality of life.
The patient's serum- and BALF- GM-CSF concentrations were significantly elevated. Functional analyses of neutrophils and monocytes showed significantly reduced GM-CSF responsiveness. Sequencing revealed a novel mutation in exon seven of the GM-CSF receptor alpha chain gene (CSF2RA). Regarding long-term perspectives, hematopoietic stem cell transplantation (HSCT) has to be considered. However, only one case HSCT in paediatric PAP has been described, with fatal outcome due to a transplantation-associated infection.

P3345

An unusual development in a girl with recurrent croup – Case report

Diana Reppucci, Juerg Hammer, Daniel Trachsel. *Pulmonology and Intensive Care, University Children's Hospital, Basel, Switzerland*

Background: Recurrent croup is common in childhood. Rare congenital and acquired pathologies may mimic viral croup.
Case: A girl with previously suspected laryngomalacia was admitted with a first episode of croup at 8 months of age, only partially responding to inhaled adrenalin. Laryngoscopically, the dorsal tracheal mucosa bulged into the subglottic area causing a 50% narrowing of the airway, and a CT showed a soft tissue mass between trachea and esophagus that was not suggestive of a hemangioma. In view of the inconspicuous appearance, the rapid recovery from the croup, the young age, and the location it was decided not to biopsy the lesion. After an uneventful observational period with decreasing symptoms the girl presented again at 4 years of age with a typical OSAS. With respect to the personal history, a bronchoscopy was made that confirmed significant adenotonsillar hypertrophy, but also revealed marked growth of the subglottic mass. A transtracheal biopsy was performed, and a plexiform neurofibroma was found. Due to the infiltrative growth of the neurofibroma, extensive surgery including partial tracheal resection became necessary. Eventually, the diagnosis of neurofibromatosis (NF) type 1 was made.
Conclusion: Neurofibroma in NF1 may occur in the laryngeal area, presenting early in infancy mimicking common croup.

P3346

Type IV laryngotracheoesophageal cleft: A case of success

Manuel Ferreira-Magalhães[1], Inês Azevedo[1], Susana Pissarra[2], Jorge Spratley[3], Jorge Correia-Pinto[4], Venceslau Hespanhol[5], Jose Matute[6]. *[1]Paediatric Department, Centro Hospitalar S. João, Porto, Portugal; [2]Neonatology Departement, Centro Hospitalar S. João, Porto, Portugal; [3]Otorhinolaryngology Department, Centro Hospitalar S. João, Porto, Portugal; [4]Paediatric Surgery Department, Centro Hospitalar S. João, Porto, Portugal; [5]Pulmonology Department, Centro Hospitalar S. João, Porto, Portugal; [6]Paediatric Surgery Department, Hospital Virgen del Rocío, Sevilla, Spain*

Introduction: Laryngotracheoesophageal clefts (LTEC) are extremely rare congenital anomalies. Mortality and morbidity is high due to difficulty in assure newborn stability, surgical approach and associated comorbidities.
Clinical case: A 12 days male newborn, without relevant prenatal history, was admitted in NICU with feeding problems and suspected tracheoesophageal fistula. Pulmonary parenchyma was preserved. After initiation of ventilatory support he developed severe acute respiratory distress. Diagnose of a type IV LTEC, extended to carina, was made by emergent rigid laryngotracheoscopy with immediate selective intubation of right bronchus. Multidisciplinary surgical correction was performed 48 hours after, with international expertise collaboration. Tracheoesophageal separation was made by median sternotomy, requiring cardiopulmonary bypass. Few days after surgery the newborn underwent gastrostomy, fundoplication and tracheostomy because of severe tracheomalacia causing difficulties in ensure noninvasive ventilation. At 3 months a small bleeding granuloma near carina was coagulated with YAG laser. Consecutive endoscopic evaluations showed a partial LTEC dehiscence. Successful transcervical surgical correction and definitive tracheostomy decannulation was made at 10 months. By 20 months he had a normal growth and development, with little hoarseness, sporadic cough and almost full oral feeding.
Conclusion: Type IV LTEC is the rarest and severest form of laryngeal clefts. This was an atypical presentation without any severe respiratory distress in the first hours of life and any prenatal or pulmonary findings. These aspects and the prompt expertise intervention assured an excellent outcome without major comorbidities.

P3347

Whole lung lavage with PAP in a 12 year old female using ET tube under GA

Mohankumar Thekkinkattil[1], Menon A.R. Narendra[2]. *[1]Pulmonology, Sri Ramakrishna Hospital, Coimbatore, Tamilnadu, India; [2]Cardiac Anaesthesia, Sri Ramakrishna Hospital, Coimbatore, India*

Alveolar proteinosis is a rare disease in children as the rare cause of severe respiratory failure. In 2002 report at least 410 cases were reported world-wide. The estimated annual incidence is 0.36 and the prevalence 3.76 cases/million population. In neonates the mortality is 100% Lung transplantation improves survival. In adults, PAP is mainly seen in males at the age of 40-55. In female children PAP is rare. PAP is characterized by intra-alveolar accumulation of proteinaceous material. It is not associated with inflammation and lung architecture is preserved. Case study: A 12 year girl was admitted with breathlessness, cough, with two weeks duration. She was referred from a private hospital in Salem with X-ray and CT features of? Alveolar Proteinosis. O/E patient restless,Sao2 83%, tachypneic and tachycardia present. CT scan repeated showed features of alveolar proteinosis. After stabilizing her with o2, bronchodilators and antibiotics she was taken up for whole lung lavage on the right side. Since the double lumen catheter of 26 size could not permit insertion of bronchoscope, through 6.5 ET tube under general anaesthesia Right lung was lavaged with about 1700 ml of normal saline. The child tolerated the procedure well. She was kept in the ventilator for one day and extubated. The saturation went up to 90%. The left lung was washed after one week. The saturation and clinical signs and symptoms improved. She is again admitted on 22/12/2011 for the next lavage session.
The treatment for alveolar proteinosis is whole lung lavage in adults. We report the case in a young girl (rarely reported)treated with whole lung lavage using ET tube (rarely tried).

P3348

Endoscopic laser-assisted management of pediatric airway lesions: A single institution experience

Juan L. Antón-Pacheco, Carmen Luna, Gloria García-Hernández, Francisca Gómez-Acebo, Miguel A. Villafruela. *Pediatric Airway Unit & Division of Pediatric Surgery, Hospital Universitario 12 de Octubre, Madrid, Spain*

Background: Laser endoscopic surgery is increasingly being used in the treatment of pediatric airway disorders.
Aims: We evaluated the outcomes of laser therapy in a selected group of patients with congenital and acquired airway lesions.
Methods: The medical charts of patients who were treated for airway disorders with diode laser between January 2008 and December 2011 at a tertiary care children's hospital were reviewed. All the procedures were performed endoscopically using rigid or flexible instruments. Data included relevant history and physical examination, diagnostic work-up, number of laser procedures, complications, and outcomes.
Results: 37 patients who had laser surgery were identified,13 girls and 24 boys with a mean age of 20 months (range, 3 days-12 years). 47 laser surgeries were performed, representing 1.2 procedures per patient. Indications for laser therapy were: Laryngomalacia 11 cases (29%), granulation tissue 8 (21%), vocal cord paralysis 7 (19%), laryngeal stenosis 5 (13%), vascular-lymphatic malformation 3 (8%), subglottic cyst 1 (2.7%), congenital tracheal stenosis 1, and papillomatosis 1 case. In 7 patients (19%), other concomitant endoscopic procedures were performed. Two complications were identified: Postlaser bleeding and aspiration in one patient, and subcutaneous emphysema and pneumothorax in another patient. In 89% of patients, partial or complete clinical improvement occurred after laser therapy.
Conclusions: Endoscopic laser surgery is a safe and effective technique when treating airway disorders in children. It is a good alternative to open surgery in selected patients either used alone or in combination with other endoscopic procedures.

P3349

Endobronchial blockers: A tool in the interventional flexible bronchoscopy in children

Borja Osona, Jose Antonio Peña, Jose Antonio Gil, Joan Figuerola. *Pediatrics, Son Espases Univertsitary Hospital, Palma Mallorca, Balearic Island, Spain*

Introduction: Techniques for selective lung ventilation usually involves the use of large devices or tools not designed for the airway. Placement in children is difficult and a variety of complications have been reported.
Objectives: To evaluate the efficacy and safety in selective ventilation and selective lobar blockade of a recently developed tool, the Arndt endobronchial blocker.
Methods: A prospective study was performed analyzing all children who underwent a bronchial blockade between 2008 and 2011. We use the 5 Fr Arndt blocker

and a pediatric fiberoptic bronchoscope (2.8mm). The following variables were recorded: age, indication, location, duration and number of insertion attempts, other techniques performed, complications, and effectiveness assessed by the surgeon or pediatric intensivist.
Results: Blocker placement was successful in 17 of 18 patients. Median (range) age was 37 (14-76m) months, 9 cases < 2 years old. The main indications were thoracic surgery, pulmonary bleeding and persistent bronchopleural fistula. Number of attempts needed was 1 to 4 (median 1). Average time for positioning the blocker was 7.5 minutes. In 3 procedures, we injected sealant (Tissucol) through the 2 Fr central lumen of the blocker. The most frequent complication encountered was dislodgement toward main bronchi or trachea in 5 cases, related to turning the patient into a lateral position. They were managed deflating the cuff and placing the blocker again. Average satisfaction after the procedure (assessed from 1 to 5) was 4.7.
Conclusion: Arndt endobronchial blockers are useful. Our initial experience has shown that placement with a fiberoptic bronchoscope is easy, acceptably rapid and effective.

P3350
The use of transbronchial biopsies in pediatric lung diseases in a tertiary care hospital in Bogota, Colombia
Patricia Panqueva, Fernando Polo, Jorge Carrillo, Alvaro Morales. *Pediatrics, Hospital Universitario San Ignacio, Bogota, Colombia Pathology, Hospital Universitario San José, Bogota, Colombia Radiology, Hospital Universitario San José, Bogotá, Colombia Epidemiology, Hospital Universitario San Ignacio, Bogota, Colombia*

A total of 12 Transbronchial Biopsies (TBB) were performed in pediatric patients during the last year at the Hospital Universitario San Ignacio, using an adult small bronchoscope (5.3 mm outer diameter Olympus).
Results: In 9 patients adequate lung tissue for histological diagnosis was obtained. In two patients with HIV infection, respiratory symptoms and chest x-ray abnormalities, pathogens were identified and successfully treated: positive ZN (13 y.o. female) and P. Jiroveci (12 y.o. female). Several patients presented with dyspnea and mosaic patterns in the chest x-ray: in three cases the information was useful, and all of them improved after treatment. 12 y.o.male in whom lung metastasis were found (successful chemotherapy initiated), 14 y.o. male in whom capillaritis was found (improvement after treatment). In an 8 y.o. male insufficient sample was reported. In patients with malignancies, useful information was provided: in patients with lymphomas and lung infiltrates CMV was identified and treated (10 y.o. male) and P. Jiroveci was found in two patients with leukemia. The only complications were a small pneumothorax that did not require intervention and hypotension in a septic HIV patient that was sent to Intensive Care for management.
Conclusion: The Transbronchial biopsie is an effective and safe procedure to obtain lung tissue for histologic diagnosis in a variety of conditions such a tumors, infections or pulmonary infiltrates and that can be considered as first –line diagnostic procedure in pediatric patients.
References:
[1] Visner GA, Faro A, Zander DS. Role of transbronchial biopsies in pediatric lung disease.Chest 2004; 126:273-280.