468. Rare lung diseases in children

P4568
Pulmonary function and quality of life in children and adolescents adenovirus bronchiolitis obliterans
Stefhanie Nayar, Ana María Escobar, Monica Saavedra, Alberto Vidal.
Broncopulmonar, Hospital Roberto del Río, Region Metropolitana, Santiago, Chile

Introduction: There are few studies assessing pulmonary function and quality of life of PIBO patients.
Objective: Assess the pulmonary function and the quality of life of patients with PIBO and the correlation between both variables.
Methods: 14 Children with PIBO in follow up at the pediatric pulmonology, were included in this study. Study period: April 2009 - April 2010. Pulmonary function was assessed with spirometry, flow/volume curve and intrathoracic gas volume. A survey of self-administered Quality of Life was applied at the visit to investigate their global, physical and psychosocial quality of life. The Pearson linear correlation between quality of life and the pulmonary function test parameters was assessed, for statistical analysis $p<0.05$ was considered significant.
Results: The mean age of our patients was 12.4. The functional alterations were characteristics of an obstructive respiratory disorder in 85.7% and only 14.3% showed normal pulmonary functionof the patients. The quality of life parameters were in average 58.6% of overall quality of life, 60.9% of physical quality of life and 57.9% of psychosocial quality of life. We found a positive correlation between global quality of life and VEF1, FVC and FEF25-75% (correlation index 0.54, 0.53 and 0.53 respectively) as well between physical quality of life and VEF1, FVC, FEV1/FVC and FEF25-75% (correlation index 0.86, 0.81, 0.70 and 0.74 respectively).
Conclusions: Most patients with PIBO showed pulmonary function impairment characterized by an obstructive respiratory pattern. Better quality of life correlates with better values in spirometric parameters.

P4569
Obliterative bronchitis verified by bronchoscopic visualisation
Petr Pohunek[1], Karolina Vávrová[1], Tamara Svobodová[1], Magdalena Havlišová[1], Ludmila Hornofová[2]. [1]*Division of Paediatric Pulmonology, Paediatric Department, University Hospital Motol, Praha, Czech Republic;* [2]*Institute of Pathology and Molecular Medicine, University Hospital Motol, Praha, Czech Republic*

Background: Chronic bronchial obstruction in children is mostly associated with bronchial asthma. In those with irreversible obstruction the most documented reason is a post-infectious obliterative bronchiolitis.
Case history: We report a 13 years old patient with Tetralogy of Fallot corrected at the age of 3 years. She suffered from respiratory infections since early childhood, in addition four pneumonias at the age of 10, 11 and 12 years. Since age of 10 on anti-asthma medication for frequent wheezing. She was referred to us for poor response to anti-asthma therapy.
Summary of findings: No dyspnoea, bilateral variable quiet wheezing. FVC 53%,FEV_1 34%,MEF 25/75 12%. TLC normal, severe hyperinflation. No immune deffect, no atopy. GOR excluded. On HRCT diffuse bilateral non-homogeneous air-trapping. On bronchoscopy normal anatomy, non-inflammatory mucosa. Bronchial biopsy with no inflammation, no remodelling.
Using 2.9 mm flexible bronchoscope and aiming for the areas with prominent air-trapping, we found at the level of 7th to 9th bronchial branching circular narrow stenoses and obliterative lesions completely closing the lumen. Biopsy revealed normal bronchial epithelium, mild thickening of the basement membrane and mild hypertrophy of smooth muscle.
Conclusion: We confirmed existence of obliterative lesions causing irreversible obstruction in the bronchi of about 2 to 3 mm in diameter. This fits into the diagnosis of obliterative bronchitis, different from obliterative bronchiolitis. This should be suspected in children with irreversible bronchial obstruction and appropriate investigations initiated. Confirmation of this diagnosis can prevent patients from unnecessary burden of anti-asthma therapy.
Supported by MZ0FNM2005.

P4570
Flexible bronchoscope in pediatrics: Other uses
Pilar Caro-Aguilera[1], Estela Perez-Ruiz[1], Javier Perez-Frias[1], Lorena Moreno-Requena[1], Javier Rivas[2], Dolores Argos[3]. [1]*Paediatrics, HRU Carlos Haya, Malaga, Spain;* [2]*Neonatology, HRU Carlos Haya, Malaga, Spain;* [3]*Paediatrics Surgery, HRU Carlos Haya, Malaga, Spain*

Introduction: Nutrition supplied through enteral route is of greater benefit to preterm newborn than that supplied through parenteral administration. The insertion of a nasogastric tube (NGT) can occasionally be problematic. Several methods and maneuvers have been described for difficult insertion, but all of them refered to anaesthetized or unconscious patient. We present the case of a NGT insertion assisted by a flexible bronchoscope in a preterm infant.
Case report: A seven-days old female infant, born at 29 weeks gestation and 979 gr of weigh, was hospitalized in the neonatal intensive care unit (NICU) as she suffered an hyaline membrane disease. She needed mechanical ventilation and enteral nutrition through a NGT. It was not possible to introduce the NGT with the standard procedure. After repeated failures of NGT placement and a mayor complication –pneumothorax-, the neonatologist asked the paediatric pulmonology unit for help as the gastroenterology unit could not help them because their smallest gastroscope was 4.9mm diameter. We used the 2.8mm diameter bronchoscope to go through the esophagus to the stomach. Then we passed a guide through the working channel. After that we removed the bronchoscope maintaining the guide in the stomach. Finally the guide was used to introduce the NGT. We checked the NGT placement with a X-Ray and it was in the correct position.
Conclusion: NGT is commonly used in NICU. Accurate tube placement is important to ensure a safe and effective enteral feeding. Literature describes misplacement of NGT including its location in the respiratory tract and pleural space, upper oesophagus, or past the pylorus. The introduction aided with an endoscopy visualization can avoid these complications.

P4571
Propanolol treatment in pulmonary capillary hemangiomatosis
Francesco Paolo Rossi[1], Paola Francalanci[2], May Chebl El Hachem[3], Teresa Salerno[1], Maria Giovanna Paglietti[1], Laura Menchini[4], Renato Cutrera[1]. [1]*Department of Pediatrics, Respiratory Unit, Bambino Gesù Children Hospital, Rome, Italy;* [2]*Department of Pathology, Bambino Gesù Children Hospital, Rome, Italy;* [3]*Department of Pediatrics, Dermatology Unit, Bambino Gesù Children Hospital, Rome, Italy;* [4]*Department of Radiology, Bambino Gesù Children Hospital, Rome, Italy*

Pulmonary Capillary Haemangiomatosis (PCH) is a rare disease with broad symptoms, from dyspnoea to pulmonary hypertension. Oral steroids are considered the first line treatment for severe haemangioma. Other therapeutic options are less used for side effects and toxicity. Propanolol has been discovered by chance in 2 children who showed rapid regression of skin haemangiomas when treated for cardiopulmonary conditions.
We describe a case of a 8 year old boy with persistent respiratory failure, in whom open lung biopsy demonstrated diffuse proliferation of capillary vessels consistent with PCH. L. began propanolol, with increasing dose up to 2 mg/kg/day. Respiratory symptoms and general conditions immediately improved without side effects and steroid treatment discontinuation was achieved uneventfully. Propanolol therapy was effective in resolving respiratory symptoms and avoiding new lesions growth without any observed side effect. In our patient we achieved lesions stability, excellent general and respiratory conditions, satisfactory growth; the future most likely therapeutic approach may be surgical to completely remove the lesion. A control MRI, performed after 6 months, showed that no new lesions appeared after the treatment, due to the anti-neoangiogenic effect of propranolol, which avoids new vessels growth.
The patient is still on propranolol and in regular follow up. The lesions are stable, his general and respiratory conditions excellent, growth is satisfactory; the future most likely therapeutic approach may be surgically to completely remove the lesion.
The excellent results that we reached with lack of adverse effects confirm propanolol a promising therapy for difficult haemangiomas, including PCH.

P4572
Study of auditory function in children with chronic lung diseases
Hanan Mohamed Ibrahim[1], Terez Boshra Kamel[1], Nithreen M.S. Abdel-Salam[2], Salah R. Abu-Ata[1]. [1]*Pediatrics Department, Ain Shams University, Cairo, Egypt;* [2]*Audiology Unit, Otolaryngology Department, Ain Shams University, Cairo, Egypt*

Objective: Chronic hypoxia has an evident effect on cochlear function and hearing sensitivity. Otoacoustic emissions' testing is efficient in detecting subtle cochlear dysfunction. This cross sectional study was designed to assess the cochlear function in children with chronic lung diseases who were exposed to prolonged hypoxia and prolonged use of ototoxic drugs (as aminoglycosides) using basic audiological evaluation and transient evoked otoacoustic emissions testing.
Methods: The study was carried out on 30 Egyptian children with chronic lung disease recruited from the Pediatric Chest Clinic, Children's hospital, Ain Shams University. Twenty normal children were included as control.
Results: Six patients (20%) showed abnormal otoacoustic emissions. A significant

effect of hypoxia on otoacoustic emissions findings was found ($p<0.05$). However, there was no significant effect of inhaled aminoglycosides on auditory functions whether pure tone audiometry, speech audiometry and transient evoked otoacoustic emissions testing.
Conclusions: Children with chronic lung diseases are liable to cochlear dysfunction due to prolonged hypoxia. Inhaled aminoglycosides in chronic lung diseases is relatively safe on auditory functions.

P4573
Health-related quality of life in non-cystic fibrosis bronchiectasis children
Ameer Hamzah[1], Yasemin Gokdemir[2], Cagatay Cimsit[3], Refika Ersu[2], Fazilet Karakoc[2], Bulent Karadag[2]. *[1]Department of Paediatrics, Marmara University Faculty of Medicine, Istanbul, Turkey; [2]Department of Paediatric Pulmonolgy, Marmara University Faculty of Medicine, Istanbul, Turkey; [3]Department of Radiology, Marmara University Faculty of Medicine, Istanbul, Turkey*

Aim: Bronchiectasis is still an important problem in developing countries. The aim of this study was to evaluate the health-related quality of life and associated factors in children with non-cystic fibrosis bronchiectasis.
Method: Children with non–cystic fibrosis bronchiectasis followed at Marmara University Paediatric Pulmonology Clinic were included to the study. Age of symptom onset, age of diagnosis, frequency of presenting symptoms, physical examination findings, pulmonary function tests, and affected lobes in high resolution computed tomography (HRCT) were evaluated; in addition, HRCT was scored according to modified Bhalla score system to obtain a bronchiectasis distribution severity score. St George's Respiratory Questionnaire (SGRQ) and the Medical Outcomes Study Short Form-36 were completed by patients and parents.
Results: The median age of the patients (48.9% male) was 12.1 years (25%-75% 10.2-14.1years). The median age at diagnosis was 7 years (25%-75% 5-10years) and follow up period was 4 years (25%-75% 2-5 years). The patients' HRCT score did not correlate with the SGRQ score and FEV1 ($p>0.05$). However, a significant correlation was found between the SGRQ scores and FEV1 ($p<0.05$).No correlation was found between SF-36 and FEV1 ($p>0.05$).
Conclusion: Patients with worse pulmonary functions have lower quality of life scores. Early diagnosis and treatment may increase the quality of life and survival of patients with bronchiectasis, which has irreversible and progressive complications if untreated.

P4574
Pediatric tracheostomy: Indications and complications rate. An experience from a semi-intensive pediatric respiratory unit
Alessandra Schiavino, Francesco Paolo Rossi, Maria Giovanna Paglietti, Maria Beatrice Chiarini Testa, Teresa Salerno, Renato Cutrera. *Department of Pediatrics, Respiratory Unit, Bambino Gesù Children Research Hospital, Rome, Italy*

Tracheostomy in children can be used to provide respiratory support as long-term ventilation/pulmonary toilet, or to bypass proximal airway obstruction. Tracheostomy is burdened by early and late complications which could make difficult hospital and home management.
We performed a single centre Survey from September 2008 to September 2010, identifying early and late complications with the aim to assess the most frequent problems related to tracheostomy management in children.
We administered to the parents a questionnaire in which the different topics were analyzed (indications to tracheostomy, operational tecnique, occurrence of intraoperative, early postoperative (within 24 - 48 hours) and late (after 48 hours) complications and the home care management).
In the 2 years period we evaluated 66 patients (M/F 36/30, mean age 7 years). Indications to tracheostomy were prolonged endotracheal intubation followed by tracheal malacia or stenosis, chronic respiratory failure, central apnoeas and vocal cord paralysis. Patients were affected by genetic or metabolic disorders, cerebral palsy, neuromuscular diseases, congenital heart diseases, tracheal diseases and tumors of the nervous system.
In 65% of cases no complications occurred, while in 5% early complications and in 35% late complications occurred. Only 4 patients died during the Survey period, but not due to tracheostomy complication.
Tracheostomy allows to manage different severe disease in children. The indications for its use have changed over the past decades. Late-onset complications are the most frequent and their knowledge allows to manage the trachestomized patient from hospital staff and home caregivers.

P4575
Comparison of conventional chest physiotherapy and high-frequency chest wall oscillation in primary ciliary dyskinesia
Yasemin Gokdemir, Refika Ersu, Bulent Karadag, Fazilet Karakoc. *Paediatric Pulmonology, Marmara University, Istanbul, Turkey*

Background: Early diagnosis and enhancement of mucociliary clearance by chest physiotherapy is important to prevent the development of bronchiectasis in primary ciliary dyskinesia (PSD). Percussion and postural drainage are the most commonly used methods. Vest® is a new chest physiotheraphy method which creates oscillation on the chest wall and move the secretions from small airways to the large airways. There is no study evaluating the efficacy of Vest® in PSD. The aim of this study was to evaluate the efficacy and safety of Vest® in PSD patients.
Material-methods: Both conventional chest physiotheraphy and Vest® were applied randomly to 7 PSD patients older than 6 years of age. Oxygen saturation was monitored and pulmonary function tests (PFT) were performed on the first and 5th day of both therapies. Patients were questioned about the efficiency and comfort level of the both methods.
Results: Forty-two percent of patients were male. Mean age was 9.9±3.4 years and the patients were followed up for 7.2±2.3 years. Dextrocardia was present in all of the patients and situs inversus totalis was observed in 71%. There were no desaturation during sessions. Although day 5 PFT were better in both groups, this did not reach statistical significance. There was no difference between the two groups in terms of PFT. Both physiotherapy methods were found effective and comfortable and there was no significant difference between the two groups.
Conclusion: Vest® was comfortable and tolerated well by the patients. There was no difference in PFT's between the conventional chest physiotheraphy and Vest® group. We presented the preliminary results of ongoing the study.

P4576
Impact of the date of diagnosis on the clinical course of children with primary ciliary dyskinesia
Folke Brinkmann, Nicolaus Schwerk, Theresa Emmrich, Gesine Hansen. *University Children's Hospital, Department of Pulmonology, Allergology and Neonatology, Hanover Medical School, Hanover, Germany*

Objective: Up to now, little is known about the impact of the date of diagnosis on the clinical course of children with primary ciliary dyskinesia (pcd).
Methods: Over a ten year period all paediatric patients with confirmed pcd in a tertiary centre who could be followed up for more than 12 months were retrospectively included in the study.
Results: Out of 19 patients 63% had a situs inversus, the median age a diagnosis was 95 months. There was a significant correlation between the date of diagnosis and development of bronchiectasis (p=0,04), but no effect on lung function. 12 months after diagnosis 75% of patients received professional physiotherapy, 88% were on continuous antibiotic therapy, 50% on inhaled β2 agonists and 13% on inhaled corticosteroids.
After starting the treatment, all patients improved clinically, the number of pulmonary complications decreased, especially pneumonias (p=0,03) and bronchitis (p=0,01). Chronic wet cough and rhinitis often persisted and lung function measurements did not improve permanently.
Conclusions: Late diagnosis of pcd carries an increased risk for developing bronchiectasis. Pulmonary exacerbations decrease with appropriate treatment whereas there is no significant effect on lung function parameters.

P4578
The follow-up of children with tracheobronchial foreign body aspiration
Sinan Tas[1], Zeynep Seda Uyan[2], Nihal Uyar[1], Yonca Anik[3], Tuba Liman[4], Metin Aydogan[5], Salih Topcu[4], Emin Sami Arisoy[6]. *[1]Pediatrics, Kocaeli University Hospital, Kocaeli, Turkey; [2]Pediatric Pulmonology, Kocaeli University Hospital, Kocaeli, Turkey; [3]Radiology, Kocaeli University Hospital, Kocaeli, Turkey; [4]Thorax Surgery, Kocaeli University Hospital, Kocaeli, Turkey; [5]Pediatric Allergy, Kocaeli University Hospital, Kocaeli, Turkey; [6]Pediatric Infectiosus Diseases, Kocaeli University Hospital, Kocaeli, Turkey*

Background: Foreign body aspiration (FBA) is an important cause of morbidity and mortality in children
Objectives: Retrospective analysis of cases who arrived Kocaeli Universtiy Hospital with suspected FBA and underwent bronchoscopy
Methods: Forty children who underwent bronchoscopy for FB removal from February 2005 to January 2010 at Kocaeli Universtiy Hospital were reviewed retrospectively.We recorded the patients' presenting symptoms,physical and radiological findings,the time elapsed from aspiration to presentation,type and location of FB,acute and long-term complications,follow up period and hospitalisation period
Results: There were 26 male patients.The mean age was 59,8±34,2 months (range 29-195 months).FBs were removed within 1-184 days following the aspiration (median: 2 days).During presentation;38 cases had clinical symptoms and wheezing was the most common one.Thirty-three FBs were organic and hazelnuts were the most common. Twenty-three (57.5%) cases had radiological findings during presentation and 10 (25%) had acute complications (pneumonia).Patients were hospitalized for 1-17 days (median 2days) and followed up for 34,5±31,3 months. Seventeen patients had persistent clinical symptoms.There were chronic radiological findings in 5 patients and fibrotic sequelaes in 11 patients.Although statistically not significant; chronic radiological findings were more common in patients in whom bronchoscopy was performed lately
Conclusions: Follow up period and methods of chidren with tracheobrachial FBA are controversial. Additionally, there are not enough studies about its long term complications.Some patients cannot be followed up for a sufficient period and this can lead to the development of chronic changes.

P4579

Evaluation of respiratory symptoms in children with post-infectious bronchiolitis obliterans

Neslihan Baranoglu Him[1], Ihsan Turan[1], Yasemin Gokdemir[2], Refika Ersu[2], Bulent Karadag[2], Fazilet Karakoc[2]. *[1]Paediatrics, Marmara University, Istanbul, Turkey; [2]Paediatric Pulmonology, Marmara University, Istanbul, Turkey*

Background: Bronchiolitis obliterans (BO) is an uncommon and severe form of chronic obstructive lung disease in children that results from an insult to the lower respiratory tract. The aim of this study was to evaluate the respiratory symptoms of BO patients and determine related factors.

Methods: Retrospective evaluation of BO patients who were followed-up from pediatric pulmonology department.

Results: Total of 22 patients (63% male) were included to the study. Mean age was 9.1±3.8 years. During first lower respiratory tract infection, mechanical ventilation and 0_2 was required in 32% and 9% of patients, respectively. Eighty percent of patients were treated with inhaled or oral steroids in the early period. Respiratory symptoms were recurrent in 35% and persistent in 65% of patients after the first lung infection. The age of 11 (50%) patients were above six currently and performed spirometry; mean FEV_1 was 51.-00±16.5% predicted. Right upper lobectomy was performed in one patient. Four (18%) patients were using 0_2 and one patient was using non-invasive ventilation (NIV) at the last visit. The rate of intubation during the first lung infection was 80% in the patients who required respiratory support in the follow-up period. There was a significant relationship with intubation during the first lower respiratory infection and requirement of 0_2 ve NIV (p=0.001) in the follow-up period.

Conclusion: BO is a disease of childhood with high morbidity and mortality. The severity of the respiratory symptoms of patients during follow-up is associated with the requirement of mechanical ventilation during the first lower respiratory infection.