

SUNDAY, OCTOBER 5TH 2008

---

## 41. The role of flexible bronchoscopy and bronchoalveolar lavage in lower airway disease

---

178

**A new ABCA3 gene mutation in a now 2-yr-old boy**

Ernst Eber<sup>1</sup>, Gabriela Thalhammer<sup>1</sup>, Gerfried Zobel<sup>1</sup>, Matthias Griese<sup>2</sup>, Charalampos Aslanidis<sup>3</sup>, Gerd Schmitz<sup>3</sup>, Helmut Popper<sup>4</sup>, Maximilian Zach<sup>1</sup>.  
<sup>1</sup>Paediatrics and Adolescence Medicine, Medical University of Graz, Graz, Austria; <sup>2</sup>Pediatrics, Dr. von Hauner Kinderspital, Medical University of Muenchen, Muenchen, Germany; <sup>3</sup>Institut fuer Klinische Chemie und Laboratoriumsmedizin, University of Regensburg, Regensburg, Germany; <sup>4</sup>Institute of Pathology, Medical University of Graz, Graz, Austria

Adenosine triphosphate (ATP)-binding cassette transporter A3 (ABCA3) plays an important role in surfactant homeostasis. A number of mutations in the ABCA3 gene have been identified, primarily presenting as acute respiratory failure in newborns. We report on a now 2-yr-old boy born as the third child of healthy unrelated parents. On day 2, mild respiratory distress developed; the clinical condition improved with O<sub>2</sub> and antibiotics, and the boy was discharged at day 11. He was readmitted at 3 mo with chronic cough and failure to thrive, and then was O<sub>2</sub> dependent. At 5 mo he was referred to our department for further evaluation. HRCT scan showed disseminated ground-glass opacities and irregular thickening of interlobular septa in both lungs. Lung biopsy revealed interstitial lung disease (ILD) with infiltrates of lymphocytes and plasma cells, proliferation of myofibroblasts, and a proteinaceous exudate in the alveoli, compatible with some type of surfactant deficiency. With clinical deterioration, treatment trials with steroids, hydroxychloroquine, therapeutic BAL, surfactant and GM-CSF were performed; at 8 mo the boy had to be intubated and ventilated with 0.5 FiO<sub>2</sub>. At this time, surfactant protein B and C deficiencies were ruled out, but analysis of the ABCA3 gene revealed compound heterozygosity for the already known nucleotide insertion c.4311-4312InsG and the so far unknown amino acid change c.2905C>T (Pro969Ser). The boy is still ventilator-dependent via tracheostomy with FiO<sub>2</sub> between 0.4 and 0.5, with normal psychomotor development. This case confirms that, while a majority of the currently known mutations of ABCA3 are fatal within 3 months of life, some mutations result in a paediatric ILD compatible with prolonged survival.

179

**Safety of double endoscopy in children with gastroesophageal reflux and difficult-to-treat respiratory symptoms**

Nicola Ullmann<sup>1</sup>, Michela Silvestri<sup>1</sup>, Oliviero Sacco<sup>1</sup>, Angela Pistorio<sup>2</sup>, Serena Panigada<sup>1</sup>, Arrigo Barabino<sup>3</sup>, Giovanni A. Rossi<sup>1</sup>.  
<sup>1</sup>Pulmonary Disease Unit, G. Gaslini Institute, Genoa, Italy; <sup>2</sup>Clinical Epidemiology and Biostatistic Unit, G. Gaslini Institute, Genoa, Italy; <sup>3</sup>Paediatric Gastroenterology Unit, G. Gaslini Institute, Genoa, Italy

Double flexible endoscopy (laryngo-tracheal-bronchoscopy and oesophago-gastroscopy, DE) may be useful in management of children with gastroesophageal reflux (GER) and difficult-to-treat respiratory symptoms (DTTRS) refractory to medical treatment. Case notes of children with DTTRS and typical GER symptoms, who underwent DE under weak general anaesthesia, were reviewed to evaluate the safety of this procedure. During the procedure and in the following 72hrs, complications (i.e. wheezing, cough, fever) were recorded in 60 patients (pts) [38m, 22f; mean age: 5.4 yrs (8.5 mo-17.5 yrs)]. During DE, mucosal biopsy (MB) and bronchoalveolar lavage (BAL) were performed respectively in 80% and 86% of the pts. The prevalence of complications was 10.0% during the procedure, 15.0% in the following 24hr, and respectively 8.3% and 1.7% at 48hr and

SUNDAY, OCTOBER 5TH 2008

72hr. The most common complications were fever (24hr: 6.7%, 48hr: 5.0%, 72hr: 1.7%) and/or cough (at endoscopy time: 5.0%, 24hr: 1.7%), never related to signs of bronchopulmonary infection. Interestingly, no 'new onset' complication was observed after 48hr. Onset of DTTRS early in life ( $\leq 2$  yrs of age) was significantly associated with the presence of any DE complication at any observation time ( $p=0.046$ ). Endoscopic, MB and BAL findings, showing congenital malformations or acquired disorders of the gastroesophageal or of the respiratory tract not leading to GER-induced DTTRS, were detected in 11 pts. DE results supporting an association between GER and DTTRS were found in 49 pts. Thus, in children with DTTRS and GER, DE procedure with MB and BAL may be helpful in the diagnosis and is associated with a low prevalence of complications.

180

#### Inflammatory mediators in the BAL of children in relation to asthma and atopy

Deborah Snijders<sup>1</sup>, Paola Dalla Via<sup>1</sup>, Eva Michelin<sup>1</sup>, Thea Tagliaferro<sup>1</sup>, Simonetta Baraldo<sup>2</sup>, Graziella Turato<sup>2</sup>, Kim Lokar-Oliani<sup>2</sup>, Marina Saetta<sup>2</sup>, Angelo Barbato<sup>1</sup>. <sup>1</sup>Department of Pediatrics, University of Padua, Padova, Italy; <sup>2</sup>Department of Cardiology, Thoracic and Vascular Sciences, University of Padua, Padova, Italy

**Introduction:** Inflammatory cells and mediators have been described in bronchoalveolar lavage (BAL) of asthmatic children.

The aim of this study is to compare the BAL cell morphology and cell mediators (ECP, IL-8) in atopic and non-atopic children with asthma and in atopic children without asthma to evaluate the possible differences between the different phenotypes.

**Methods:** We recruited 81 children (aged 1-15.5 years) undergoing fiberoptic bronchoscopy (FOB) for appropriate clinical indications: 19 with asthma and atopy (AAS), 20 children with asthma without atopy (NAAS), 23 atopic children without asthma (ANAS) and 19 non-asthmatic non-atopic control children (C).

In BAL samples, total and differential cell counts and inflammatory mediators; including eosinophil cationic protein (ECP) and interleukin 8 (IL-8) were analyzed.

**Results:** ECP- BAL levels were increased in both AAS and NAAS children ( $p=0.037$  and  $p=0.053$ ), as compared to controls. Moreover, ECP levels were also increased in ANAS as compared to controls ( $p=0.009$ ). Conversely, the percentage of eosinophils was increased only in NAAS children as compared to controls ( $p=0.025$ ), while AAS children and ANAS groups did not show any difference. No correlation was found between serum eosinophils and ECP- BAL levels in the different groups. IL-8 and neutrophil numbers did not show any significant difference among the four groups.

**Conclusion:** These results suggest that BAL cytokines are more useful markers than BAL differential cell counts in demonstrating the recruitment and activation of eosinophils, which occur in asthmatic children, irrespective of the presence of atopy and, unexpectedly, even in atopic children without asthma.

181

#### Bronchological examinations in the neonatal intensive care unit

Lajos Kovacs. *1st Department of Pediatrics, Semmelweis University, Budapest, Hungary*

Indications of bronchoscopies in NICU were localized hyperinflation (5 cases), difficulty in extubation (14), stridor (38), persistent atelectasis (10), suspicion of tracheoesophageal fistula (9) and presumed iatrogenic tracheal injury (2). Bronchial stenosis at the corresponding localization was found in four cases of localized hyperinflation. One of them was a severe right main stem bronchus stenosis which was successfully treated with stent implantation. In patients with extubation failure 3 patients with glottic-, 3 with subglottic-, and 2 with tracheal stenoses were treated conservatively and eventually successfully extubated, while three neonates with glottic stenosis needed tracheostomy tube placement and one newborn with subglottic stenosis was cured with cricoid split surgery. In the stridor group 12 patients with soft larynx, 10 with glottic stenosis, 4 with subglottic stenosis, 3 with vocal cord paralysis and one with big tongue (Beckwith-Wiedemann syndrome) did not require intervention. Two neonates with stridor had mucous pellicle in the glottis, which were bronchoscopically sucked. In six cases with stridor surgery was not avoidable including 3 tracheostomy tube placements, 2 cricoid split operations and one tracheal resection due to tracheal web. In the group of persistent atelectasis mucous plug was discovered and bronchoscopically removed in 5 cases, severe bronchial stenosis was confirmed in 4 children and no alteration was found in one baby. We diagnosed two congenital tracheoesophageal fistulas and detected one tracheal rupture in a child with suspected tracheal injury. In our presentation we will show the most interesting video records of the past 5 years.

182

#### Multiple bronchoalveolar lavage as an important tool for treating lipid pneumonia associated with ascariasis

Selma Sias<sup>1</sup>, Pedro Dalro<sup>2</sup>, Edson Marchiori<sup>1</sup>, Regina Caetano<sup>1</sup>, Thereza Quirico-Santos<sup>1</sup>. <sup>1</sup>Materno Infantil-Faculdade de Medicina, Universidade Federal Fluminense, Niterói-Rio de Janeiro, Brazil; <sup>2</sup>Instituto Fernandes Figueira, FUICRUZ, Rio de Janeiro, Brazil

This work describes the development of lung inflammation and lipid pneumonia

(LP) in 4 children with mean age of 25 months, undergoing treatment with mineral oil for partial bowel obstruction due to A. Lumbricoides. The mean of oil ingestion duration was 3 days. All children had received antibiotics. The symptoms were respiratory distress, cough, fever, failure to thrive, hypoxemia ( $n=3$ ) besides anemia, marked neutrophilia and increased erythrocyte sedimentation rate. Major radiologic chest findings were consolidation in both lower lobes especially in the right lung and in the upper lobe ( $n=2$ ). Chest high-resolution computer tomography showed bilateral and multifocal consolidation involving 3 to 5 lobes and mild ground-glass opacities near areas of consolidation. Bronchoalveolar lavage (BAL) microbiologic analysis for current infectious agents (bacteria, fungi, M.tuberculosis) were negative, but marked pleocytosis ( $1320$  to  $3200$  cells/ $mm^3$ ) and showed alveolitis with predominance of lymphocytes, eosinophils and macrophages Sudam positive laden with oil particles. All patients were treated with multiple bronchoalveolar lavage (mean 6 procedures), though one patient received up to 8 lavages. Such procedure restored clinical, radiologic and BAL normal parameters without any interurrences. The present data indicate that multiple BAL procedure is an efficient tool for removing the alveolar oil and reducing the risks of sequels in the long run.

183

#### The role of bronchoscopy in children with atelectasis

Sofija Galbova, Biljana Dimceva, Blagica Manceva, Lidija Petrushevska, Katerina Obocki. *Bronchology Unit, Institute for Respiratory Diseases in Children-Kozle, Skopje, Fyrom (Macedonia)*

Atelectasis is not uncommon in infants and children because of the small tracheo-bronchial tree and it is one of the many indications for bronchoscopic examination.

**Aim:** The aim of the study was to evaluate the bronchoscopic findings in children with atelectasis.

**Material and method:** During a period of 5 years, in our Bronchology Unit, 975 children were underwent to bronchoscopy (702 flexible and 273 rigid). From them, 53(5.4%) had massive, long-term or recurrent atelectasis on the chest X-ray. The bronchological findings we classified in 4 groups.

**Results:** **group-1**-the most common reason for atelectasis were mucous and pus plugs usually processed by reactive mucosa and located in the middle right bronch -21(39.6%); **group- 2**-foreign bodies 11(20.75%), **group -3**-normal findings 9(16.98%), in **group -4** were the rare conditions: plastic endobronchitis 5(9.4%), granulomatous tissue 3(5.7%), tumors 2(3.8%), extramural compression 2(3.8%). The most common atelectasis localization were in the middle right bronchus (23 or 43.4%), lingula left (12 or 22, 6%), right upper bronchus (7 or 13.2%), left basal bronchus 4 (7.55%), right main bronch 3 (5.7%), bilateral 3 (5.7%), left main bronchus -1 child. All children from the 1<sup>st</sup> and the 3<sup>rd</sup> group and the most of those in the 2<sup>nd</sup> group improved their clinical and X-ray finding after the first bronchoscopy. Same of the children in the 4<sup>th</sup> group needed repeated bronchoscopic interventions (plastic endobronchitis), or surgery.

**Conclusion:** As the intraluminal obstructions are the most common reasons for the atelectasis, bronchoscopic procedure should be performed for diagnostic and therapeutic purpose in every child with massive, long-term or recurrent atelectasis.

184

#### Role of temporary bronchial endoscopic occlusion (TBEO) in surgical pulmonology

Paata Gvetadze<sup>1</sup>, Nino Adamia<sup>2</sup>. <sup>1</sup>Department of Pediatrics, M. Iashvili Pediatric Clinic, Tbilisi, Ghana; <sup>2</sup>Department of General Pediatrics, Tbilisi State Medical University, Tbilisi, Ghana

The work describes our experience of application of TBEO to 188 children of age under 15 with various lung pathologies.

**The method:** at a time of bronchoscopy, under general anesthesia, damaged bronchial lobe is occluded with special foam rubber ball with the diameter 2-3 times greater than the one of the bronchus, to ensure tight adhesion and prevent its migration. As a result of occlusion of the bronchial opening the damaged lobe is not supplied with air and air is not discharged into the cave of pleura. Healthy lung lobes can expand - pneumatothorax is closed. Through repeated bronchoscopy foam rubber ball is removed.

Major group consisted of 127 children with pyopneumothorax; their treatment with puncture and drainage method did not yield any results. In 110 cases stable expansion of the lung occurred after the initial occlusion. 17 children required secondary occlusion by the reason of pneumothorax recurrence after removal of the bronchial blocker. In 4 cases the necessity of surgical treatment emerged. 2 cases were lethal, by the reason of progressing of the pyoseptal process.

TBEO was applied as a supplementary method in case of surgical treatment of pneumonia complicated by an abscess in 40 patients. Unlike the standard methods, after opening and sanitation of the lung, temporary occlusion performed immediately after completion of operation ensures impermeability.

TBO was also successfully applied as pre-surgical preparation method for various purposes.

Experience showed that TBEO is characterized with wide range of opportunities not only in treatment of pleuro-pulmonary suppurative diseases it is highly effective for elimination of number of urgent situations for preparation before surgery.

SUNDAY, OCTOBER 5TH 2008

185

**Primary bronchomalacia successfully treated with bronchopexy**

Srdjan Banac<sup>1</sup>, Vladimir Ahel<sup>1</sup>, Vojko Rozmanic<sup>2</sup>, Koraljka Manestar<sup>2</sup>, Aldo Ivancic<sup>3</sup>, Vladimir Curuvija<sup>3</sup>. <sup>1</sup>Department of Paediatrics, Clinical Hospital Centre Rijeka, Rijeka, Croatia; <sup>2</sup>Department of Paediatrics, University of Rijeka, School of Medicine, Rijeka, Croatia; <sup>3</sup>Department of Surgery, Clinical Hospital Centre Rijeka, Rijeka, Croatia

A two-year old, otherwise healthy girl having normal growth and development, was brought to a paediatrician office due to a six-day history of high fever and cough. Besides the signs of an acute infection of upper airways, auscultation revealed diminished breath sounds over her left lung. Acute phase reactants showed to be quite high (CRP 163.5 mg/L). Surprisingly, chest radiography, performed for the first time in her life, did not confirm an expected presence of leftsided pneumonia. Instead, a significant hyperinflation of the left lung was noticed. In spite to the successful treatment of respiratory infection for more than a week, the hyperinflation persisted. Bronchoscopy was performed and severe bronchomalacia (BM) of the left main bronchus followed with consecutive chronic inflammatory changes of respiratory mucosa were visualised. Since CT examination showed that it was going for the primary BM, and had confirmed chronic changes of the left lower airways, the girl underwent bronchopexy (BPX). BPX entails surgical suspension of the affected left main bronchus superiorly to the ligamentum arteriosum. The intervention provided a very good suspending effect which was confirmed by an almost normal visual finding on control bronchoscopy. Six months later chest radiography was normal, with no signs of lung hyperinflation. In this case BPX showed to be the treatment of choice for isolated severe primary BM. We believe that suspending effect of BPX will last long enough to allow the affected bronchus to grow and reach a size at which BM will no longer produce problems.