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

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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 evolution 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 repeating course and a second scan was not considered necessary (2nd group). Severity of involvement was assessed with Bhalla score and presence of bronchectasis, 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 bronchectasis: 10 and 27; p=0.9 in the 1st and 2nd group, respectively). In 1st group 10 children had bronchectasis 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:isd: 2.7±0.2 and 2.9±0.4, respectively, p=0.60). There was no difference in bronchoscopic or bronchovascular lavage findings.

Conclusion: Radiological findings do not suffice to predict the clinical course of protracted bacterial bronchitis. Although the literature suggests that radiological findings are reversible, this is not the rule. Apart from the intensity of treatment, there are probably other - yet unidentified- factors that determine the final outcome of the disease.

P3335 Bronchoscopic findings in children with primary ciliary dyskinesia: Most but not all have bronchial malacia
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Chronic bacterial infections of the lower respiratory tract (LRTI) are a relevant problem in patients with primary ciliary dyskinesia (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 bronchovascular 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). Sinus inversion 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 (>10^5 CFU/ml) have been confirmed. Haemophilus influenzae (n=10) and Strep. toccoccus 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. Further, these findings correlated with a worse lung function before bronchoscopy (FEV1 <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
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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 "bassy","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/transverse 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±0.12 and 0.98±0.12, p<0.001, for patients and controls, respectively, at the thoracic inlet and in 1.1±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 extrathoracic tracheomalacia as it is not influenced by the effects of general anaesthris.

P3337 Bronchoscopic findings and interventions in patients with long-term tracheostomy
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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 interve- nions (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 transtracheally, 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 74 (83%) surveillance FBs were performed. The mean frequency of FBs was 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
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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 chemilumines- cence 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, 4.3-43.2) measured nNO with a hand-held device.

Results: Median (range) nNO values were 12.5 (6-2) 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.

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<th>Cut-off points (ppb)</th>
<th>Sensitivity (%)</th>
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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
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The effect of air pollution can play a role in the development and/or the exacerbations of chronic lower airway diseases (CLADs). Our aim is to investigate the macrophages in the airways of children affected by CLADs. We aimed to quantify and analyze the immune-inflammatory activity of AMs and to determine different biomarkers of inflammation. We also aimed to quantify the content of airway macrophages and to correlate them with the air pollution.

We enrolling 50 children under 12 years of age with a diagnosis of asthma or chronic obstructive pulmonary disease (COPD) and 50 healthy children aged 12-17 years old. The children were divided into two groups: the control group and the group with CLADs. The children were divided into two age groups: 0-9 years and 10-17 years old. We collected blood samples and analyzed the content of ABP in AMs, sampled by bronchoalveolar lavage, was determined 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 statistically significant (0.671 m2 vs 0.352 m2, p<0.05). In conclusion the median area of AM in CMS is correlated to air pollution exposure and children affected by CLADs, but it seems not to correlate to underlying disease, sex and age.

P3340 Foreign body aspiration in Tunisian children: Clinical, radiological and endoscopic features and outcome
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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 (2002 to 2011) we performed 101 flexible bronchoscopies for suspicion of foreign body aspiration. A FB inflammation 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 FB was not 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 poly is seen in 28 children. The most common FB seen was seeds and nuts, followed by carrots in 25 cases and almonds in 5. Six children inhaled a scarf 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 poly was 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.
P3344 Pulmonary alveolar proteinosis due to a novel mutation in CSF2RA
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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 qualitative and quantitative alteration in alveolar macrophages. We present the case of a 3.5-year-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 bronchiolar/alveolar 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.

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 adrenaline. 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 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 tracheal biopsy was performed, and a pleomorphic 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 I was made.

Conclusion: Neurofibroma in NF1 may occur in the laryngeal area, presenting early in infancy mimicking common croup.

Type IV laryngotracheoesophageal cleft: A case of success
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Introduction: Laryngotracheoesophageal clefths (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 preliminary intubation, the baby developed severe acute respiratory distress. Diagnose of a type IV LTEC, extended to carina, was made by emergent rigid laryngoscopy 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 examination showed a partial LTEC dehiscence. Successful transcutaneous 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 clefs. This was an atypical presentation without any severe respiratory distress 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
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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 worldwide. The estimated annual incidence is 0.30 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 it 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 PAP. CT showed diffuse ground-glass opacity. Child tolerated the procedure well. She was kept in the ventilator for one day and exubrated. 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 asymptomatic for 2 years now.

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).
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
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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 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 biopsy is an effective and safe procedure to obtain lung tissue for histologic diagnosis in a variety of conditions such as tumors, infections or pulmonary infiltrates and that can be considered as first-line diagnostic procedure in pediatric patients.

References: