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Title: Bronchoscopic findings in children with primary ciliary dyskinesia: Most but not all patients have bacterial bronchitis

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Body: 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 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.