

## CILIARY ABNORMALITIES IN IDIOPATHIC BRONCHIECTASIS

KWT Tsang, J Sun, R Leung, JCM Ho, B Lam, TF Cheung, M Ip, WK Lam. *University Department of Medicine, The University of Hong Kong, Hong Kong SAR, China.*

Bronchiectasis is defined as pathological dilatation of the bronchial tree and affected patients suffer from regular sputum production and recurrent exacerbations. We have performed this cross sectional clinico-pathological study on 100 consecutive Chinese patients with bronchiectasis. Respiratory mucosa was obtained from the inferior turbinate of 85 Chinese bronchiectatic (mean age $\pm$ SD, 53.8 $\pm$ 18.78 years) and 52 normal subjects for transmission electron microscopy assessment. Transmission electron microscopy revealed that bronchiectatic and control subjects (n=85, 52) displayed very frequent ultrastructural abnormalities (91.8, 55.8%;  $p<0.001$ ) including presence of compound cilia (68.2, 34.6%;  $p<0.001$ ), single (23.5, 17.3%;  $p=0.37$ ) or extra-microtubule (34.1, 23.1%;  $p=0.17$ ), extra central pair (7.1, 1.9%;  $p=0.19$ ), matrix abnormalities (10.6, 19.2%;  $p=0.16$ ), dynein arm deficiency (5.9, 0%;  $p=0.08$ ), absence of one or both (8.3%, 0%;  $p=0.034$ ) of central pair, and abnormal ciliary membrane (2.4, 9.6%;  $p=0.27$ ). 56.6 and 28.8% of the patients and control subjects had more than one defects ( $p=0.001$ ). The results of our study show that ciliary defects appear to have important aetiological relevance in bronchiectasis. Further clinico-pathological correlations should be evaluated to delineate the clinico-pathological value of ciliary ultrastructural assessment in bronchiectasis.

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## CILIARY CENTRAL MICROTUBULAR ORIENTATION IN STABLE BRONCHIECTASIS

KWT Tsang, J Sun, \*KN Chan, R Leung, TF Cheung, JCM Ho, B Lam, WK Lam. *University Department of Medicine and \*Paediatrics, The University of Hong Kong, Hong Kong, China.*

Cilia are minute hair-like structure which exist on the surface of respiratory mucosa and beat continuously to maintain the sterility of the lower respiratory tract. Ciliary beating and ultrastructural abnormalities are well described in bronchiectasis in whom chronic infection and inflammation occur in a pathologically dilated tracheobronchial tree. The central microtubules of cilia have received much attention recently as some patients with bronchiectasis have normal ciliary ultrastructure and beating but abnormal orientation of the central microtubules. This central microtubular orientation can only be studied with transmission electron microscopy and has never been correlated with clinical parameters. We have therefore assessed this in 100 consecutive Chinese patients with steady state bronchiectasis. Respiratory mucosa was obtained from the inferior turbinate of 100 Chinese bronchiectatic (mean age $\pm$ SD, 55 $\pm$ 17 years; 24h sputum 24.7 $\pm$ 26.1ml; FEV<sub>1</sub>% 66.6 $\pm$ 29.4%; FVC 74.7 $\pm$ 25.9%; number of bronchiectatic lobes 2.9 $\pm$ 1.5; ciliary beat frequency 11.1 $\pm$ 3.0 Hz; and exacerbation frequency 3.0 $\pm$ 3.5/annum) for transmission electron microscopy assessment. The acute angle formed between a line joining the pair of central microtubules and the horizontal axis was determined for each cilium in each case. The mean $\pm$ SD of orientation angle were 20.6 $\pm$ 15.0). Patients with *Pseudomonas aeruginosa* in their sputum displayed a significantly higher angle than those without ( $p=0.01$ ). There was, however, no significant association between the orientation angle and age, 24h sputum volume, FEV<sub>1</sub>%, FVC%, number of bronchiectatic segments, ciliary beat frequency, or exacerbation frequency ( $p>0.05$ ). Our study is the first to identify the correlation between *Pseudomonas aeruginosa* infection and ciliary disorientation *in vivo* and the lack of correlation between ciliary orientation and disease parameters. Our results are of major clinical and scientific significance.

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