



Title	The clinical profile and respiratory ciliary assessment in Kartageners syndrome
Author(s)	Tsang, KWT; Ip, MSM; Lam, WK; Ho, CS; Tam, CY; Wong, MB
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GASTRO-OESOPHAGEAL REFLUX IN BRONCHICTASIS.

K.W.T. Tsang, W. Hu, W.K. Lam, M. Ip, S.K. Lam. *University Dept of Medicine, University of Hong Kong, Hong Kong.*

Bronchiectasis is a chronic inflammatory disease of the tracheobronchial tree and affected patients suffer from recurrent sputum production and exacerbations. Micro-aspiration of gastric acid into the respiratory tract, a result of gastro-oesophageal reflux (GOR), plays a significant role in the pathogenesis of some cases of asthma, another chronic inflammatory disease of the tracheobronchial tree. There is anecdotal experience that treatment of GOR is beneficial to both of these conditions. We have therefore performed this preliminary study to evaluate gastro-oesophageal reflux in 8 patients (5M; mean age \pm SE 51.1 \pm 7.2; mean FEV₁/FVC=1 \pm 0.1/1.8 \pm 0.31) with stable idiopathic bronchiectasis but no upper gastrointestinal symptoms. A two channel Altimony esophageal pH catheter was passed into the esophagus nasally and kept *in situ* for 24h. The upper channel, situated at the upper esophagus, recorded a mean (\pm SE) of 22.9 \pm 10.3, 1.1 \pm 0.5, 31 \pm 14.5min, 2.2 \pm 1.1%, and 9.4 \pm 4.2 as the no. of reflux episodes (RE), no. of reflux $>$ 5min, time pH $<$ 4, fraction time pH $<$ 4, and DeeMeester Score respectively. For the lower esophageal channel, these were 60.8 \pm 21.3, 3.8 \pm 1.8, 98.1 \pm 43.8, 6.8 \pm 3.0, and 24.8 \pm 11.1 respectively. The results from this study suggest that GOR is relatively common in idiopathic bronchiectasis and might have a role in the pathogenesis of chronic tracheobronchial inflammation in bronchiectasis. Controlled studies should be performed to evaluate this further.

THE CLINICAL PROFILE AND RESPIRATORY CILIARY ASSESSMENT IN KARTAGENER'S SYNDROME.

KWT Tsang, M Ip, WK Lam, CS Ho, *CY Tam, +MB Wong. *University Depts of Medicine and +Pathology, University of Hong Kong and *Tuen Mun Hospital, Hong Kong.*

Respiratory epithelium contains cilia on its surface and synchronized ciliary beating is responsible for establishing mucociliary clearance which keeps the lower respiratory tract sterile. Bronchiectasis, defined as pathological dilatation of the tracheobronchial tree, is a disabling disease and affected patients may have abnormal ciliary structure and function. Kartagener's syndrome, originally described in infertile men who were severely affected, is characterized by the presence of situs inversus, sinusitis and bronchiectasis. The clinical features, ciliary function and ultrastructure of this very rare disease in Chinese have never been studied in a systematic manner. To our knowledge, only 7 cases (4M) of Kartagener's syndrome have ever been diagnosed in Hong Kong. Mean age of symptom onset and at assessment was 15.9 and 34.9 yrs respectively and these patients expectorated a mean of 26.6ml sputum/day and suffered from a mean of 2.9 exacerbations/yr. Lung function assessment showed a mean FEV₁/FVC of 95.6/95.6 (% predicted) with little reversibility. HRCT revealed bronchiectatic involvement of the lower lobes in 7/7 and middle lobe/lingular in 4/7 of cases. Assessment of respiratory cilia, obtained by brushing the inferior turbinate under direct vision and then re-suspending the brushing in medium 199, revealed that most of the mucosa was unciliated and the mean ciliary beat frequency was 9.1 Hz (range 0-14, median 12; normal range 12-18Hz). Only 1/7 patient (F,13yr) had immotile/dyskinetic whilst 6/7 patients had normal ciliary movement. Transmission electron microscopy using conventional methods to assess cross sections of the ciliary shafts showed that the outer dynein arms were completely absent in 3/5 patients assessed. Only two cases (1M) were married and both had normal fertility. The results of this study suggest that patients with Kartagener's syndrome may have normal ciliary ultrastructure although the absence of dynein arms might carry a prognostic value as these patients appear to have more active disease. Our experience of Kartagener's syndrome in Chinese patients should help other clinicians on the investigation and management of these patients.