Short Report

A comparison of spirometric variables in cystic fibrosis and bronchiectasis over 4 years

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Introduction

Bronchiectasis, which is a sequel to childhood infection, is characterized by lung function decline akin to that in the general population (1,2). Bronchiectasis in cystic fibrosis is a consequence of altered mucus rheology, which predisposes the airway to mucoid impaction and recurrent lower respiratory infection. The pathology in cystic fibrosis is widespread and constantly active compared to bronchiectasis. This can cause a faster decline of lung function in cystic fibrosis. The goal of this report was to compare lung function decline in cystic fibrosis and bronchiectasis.

Materials and Methods

Twenty-eight cases of cystic fibrosis (Cystics), documented by raised sweat chloride and sodium, were studied. Age- and sex-matched patients with bilateral bronchiectasis and comparable lung functions (FEV₁ and FVC post-bronchodilator ± 50 ml that of Cystics) were Controls. Spirometry was performed 6 h after bronchodilator medication, and approximately 2 h after eating and chest physiotherapy, using the modified Steadwell's water-sealed spirometer. Lung functions were repeated annually during stable state for 4 consecutive years.

Subjects were treated conventionally (3), and admitted for any increase in sputum over stable state, fever, leucocytosis or radiological evidence of deterioration, which could not be managed on outpatient basis. Sputum was cultured for organisms and antibiotic sensitivity during each admission, and also twice every year. Subjects with proven fat malabsorption were treated with pancreatic supplement and vitamins.

Lung function parameters used for analysis were FVC and FEV₁, expressed in absolute values (1). Regression coefficients for serial FVC and FEV₁ were derived for each subject. Comparison of initial and final lung function within groups, as well as comparison of function and regression coefficients between groups, was done using the paired t-test (2).

Results

Each group comprised 20 male and eight female subjects. The mean age was 13 ± 4.2 years (range 7–21 years).

Mean baseline FVC for Controls and Cystics were 1680 ± 268.9 ml and 1677.9 ± 271.0 ml (P = 0.52). It was respectively 1622.7 ± 271.5 ml and 1622.4 ± 242.8 ml (P = 0.98) in the first year, 1637.9 ± 258.7 ml and 1609.9 ± 271.3 ml (P = 0.041) in the second, 1606 ± 261.2 ml and 1565.1 ± 242.2 ml (P = 0.005) in the third and 1585.9 ± 257.7 ml and 1551.3 ± 261.2 ml (P = 0.005) in the last year studied (Fig. 1). The mean annual decline in FVC was 23.9 ml and 31.6 ml, respectively.

Mean baseline FEV₁ for Controls and Cystics were 1428.6 ± 283.2 ml and 1456.8 ± 286.9 ml (P = 0.2) respectively. The corresponding consecutive annual means were 1411.6 ± 258.9 ml and 1412.1 ± 259.0 ml (P = 0.98) in the first year, 1419.2 ± 254.5 ml and
1376.8 ± 266.3 ml (P = 0.052) in the second, 1407.9 ± 254.8 ml and 1333.9 ± 261.9 ml (P = 0.0009) in the third, and 1389.3 ± 262.2 ml and 1323.9 ± 253.3 ml (P = 0.001) in the final year (Fig. 1). The mean annual decline in FEV₁ was 13.1 ml and 44.3 ml, respectively.

FVC in both groups was significantly lower than baseline from the first year (mean fall 57.9 ml and 55.4 ml, respectively, for the first year). FEV₁ was more stable in Controls. It dipped significantly below baseline only by the fourth year (P = 0.013). In Cystics FEV₁ declined significantly within the first year.

Regression coefficients for FVC and FEV₁ were lower in Cystics. One control subject had a positive coefficient for FVC compared to none from Cystics. Ten Controls as compared to one cystic had a positive coefficient for FEV₁. Of the 28 controlled pairs, Cystics had a lower coefficients in 22 and 24 instances respectively for FVC and FEV₁.

Discussion

In bronchiectasis as in chronic bronchitis the bulk of sputum is produced in the large airway, as only about 10% of mucus glands are in the small airway. Since airway obstruction begins in the small airway, the pathology of bronchiectasis would contribute little to obstruction (4). This could explain why lung function is stable in Controls.

A subgroup of bronchitis has been described with rapid decline in lung function. This is due either to secretion, mucus gland hypertrophy or smooth muscle hyperplasia in the small airway (4). The altered mucus in Cystics impacts the small airway easily. This can cause lung function declines in Cystics just like in the susceptible bronchitic population. In Controls the pathology is localized to the abnormal segment which takes little part in ventilation. The remaining lung is relatively normal. Hence lung function declines slowly as reported before (1). The universal abnormality of the exocrine gland in Cystics, compared to the patchy pathology in Controls, could add to the greater decline in cystic fibrosis.

Since lung function declines despite optimal management, the stress should be on prevention of bronchiectasis among Cystics detected when the lungs are still normal. Lung transplantation seems to be the only long-term solution for cystic fibrosis with established bronchiectasis.

Acknowledgement

We thank the Dean, K.E.M. Hospital, Bombay for permitting this study.

References