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Impact of underlying cause of bronchiectasis on clinical outcome: A comparative study on CF and Non-CF bronchiectasis in Egyptian children

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ABSTRACT

Background: Bronchiectasis represents an important cause of chronic lung disease in children in developing countries and continues to be one of the leading causes of morbidity and mortality with worsening quality of life in these children.
Aim: To compare the clinical course and outcome in Cystic Fibrosis (CF) and non CF bronchiectasis in children.
Patients and method: This cross sectional observational study included 50 children with bronchiectasis; all were followed up at pulmonology Unit, Cairo University Children Hospital, Egypt. Diagnosis of bronchiectasis was confirmed by high-resolution computed tomography (HRCT). Forced expiratory volume in the first second (FEV_1) was recorded for all patients, severity classification of the FEV_1 was according to the interpretive strategy set out by the ATS/ERS task force. All enrolled cases were followed up for 2 years to record clinical outcome (hospitalization, exacerbation, and mortality).
Results: There was a significant association between CF bronchiectasis and very severe FEV_1 impairment (p value 0.002). A significant proportion of children had hospitalization with severe exacerbation in 72%, and 16% of children died during the period of follow up. There was a strong relationship between the duration of bronchiectasis and hospitalization with severe exacerbation (*p value 0.01*). Also, there was a strong positive correlation between the severity of impairment of FEV1 and both hospitalization with severe exacerbation and mortality (*p value < 0.001*).
Conclusion: Although, the majority of bronchiectasis cases in this study were due to non-CF bronchiectasis causes. CF bronchiectasis was associated with significant very severe impairment of FEV1, also with the greatest proportion of deaths compared to non-CF bronchiectasis.

Introduction

Bronchiectasis, a disease characterized by irreversible abnormal dilatation and anatomical distortion of the bronchial tree, likely represent a common end stage of a number of nonspecific and antecedent events [1] Bronchiectasis is a significant problem in developing countries where overcrowding and poor hygiene practices, poverty, malnutrition, lack of healthcare facilities and poor adherence to vaccination programs still common.[2] While the prevalence of bronchiectasis is probably low and decreasing in developed countries, it is still an important cause of chronic lung disease in developing countries, however adequate statistical data is not available because in many cases the etiological diagnosis is not established.[3]

Mortality rates have been reported at 2–5% over 2 years, 10–30% at 5 years.[4,5] Patients may experience 0–12 exacerbations per year, with the average in most series being between one and three exacerbation per year.[6–8] Severe exacerbations have a greater impact in terms of healthcare costs and prognosis as they are associated with more significant lung function decline and increased mortality.[5]

Bronchiectasis is a morphological term used to describe abnormal irreversibly dilated and often thick walled bronchi, the airways become colonized with bacteria, which promotes recruitment of neutrophils and other inflammatory cells leading to damage the airway, airway remodeling and failure of mucociliary clearance and a vicious cycle.[9–11]

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Cystic fibrosis (CF) is the most common cause of clinically significant bronchiectasis. Repeated and persistent infections in CF result in chronic airway inflammation and destruction of lung architecture. Other conditions associated with bronchiectasis includes ciliary dyskinesia, immunodeficiency syndromes and infection especially pertussis, measles and tuberculosis. Bronchiectasis can be also due to congenital disorder or associated with other disorders such as Right middle lobe syndrome and Yellow nail syndrome.[12]

High-resolution computed tomography (HRCT) scanning is now well established as the primary imaging tool for bronchiectasis investigation, it had made the diagnosis of bronchiectasis considerably easier and there has been a renewed awareness of this disease, which remains a significant cause of respiratory morbidity. CT is now being increasingly incorporated into investigative algorithms to assess bronchiectasis treatment effects.[13]

Pulmonary function tests (PFTs) of bronchiectasis patients are usually abnormal, impaired pulmonary function is of prognostic importance in bronchiectasis, the degree of impairment depends not only on the nature and extent of the morphologic abnormalities of bronchiectasis, but also on the presence or absence of associated chronic bronchitis, emphysema, and so on. Persistent inflammation plays an important role in the deterioration of lung function in bronchiectasis.[14]

The present study was performed to evaluate bronchiectasis in a group of Egyptian children through comparing the clinical course of bronchiectasis in CF and non CF bronchiectasis, and studying its impact on the outcome.

Materials and methods

Study design and subjects

This cross sectional observational study was conducted during the period from 1 September 2014 to 1 March 2017, it included 50 patients with bronchiectasis; all were followed up at CF, and Pulmonology clinics, Cairo University Children Hospitals, tertiary hospitals that receive patients from all over Egypt. These patients were diagnosed based on the results of HRCT of the chest, and a clinical history consistent with bronchiectasis.[15] Eligible patients were consecutively recruited over a period of six months and each child was followed up regularly every two months for 2 years starting from the date of first patient enrollment, the end point of this study was either completing the duration of 2 years follow up or patient death.

Ethical considerations

This study was approved by the Scientific Ethics Committee of the Department of Pediatrics, Faculty of Medicine of Cairo University. The study design complies with the Revised Helsinki Declaration of Bioethics. All participant's caregiver were informed about the study and their consent was obtained.

Inclusion criteria were all patients proved to have bronchiectasis by HRCT. The study enrolls patients above 5 years (For better co-operation during performing spirometry; including forced expiratory maneuvers to assess FEV1), both males and females with no gender differentiation. Exclusion criteria were age below 5 years, progressive neuromuscular disease (For being unable to perform spirometry), and history of lobectomy.

Methods

Data was collected by full history taking and thorough analysis of medical records of the studied population. All selected patients were also subjected to clinical examination and selected laboratory testing. The following basic data was recorded for each patient: age, sex, consanguinity, history of bronchiectasis (onset, course, duration, and underlying cause of bronchiectasis). The underlying cause of bronchiectasis was determined according to the recommendation of the British Thoracic Society (BTS) guidelines.[16]

Spirometry was done and FEV1 was recorded for each patient. Severity classification of the FEV_1 according to the interpretive strategy set out by the ATS/ERS task force (mild abnormalities being >70%, moderate 60% to 69%, moderately severe 50% to 59%, severe 35% to 49%, and very severe <35% of predicted values).[17]

Follow up for 2 years to record clinical outcome (hospitalization, exacerbation, and mortality). Acute exacerbation of bronchiectasis was defined as either a change in one or more of the common symptoms of bronchiectasis including sputum volume or purulence, dyspnea, cough, and fatigue or the onset of new symptoms including fever, pleurisy or hemoptysis.[18]

Bacteriological status of the patients was assessed on spontaneous early-morning sputum samples. Chronic colonization was considered by isolation of potentially pathogenic bacteria in sputum culture on at least two occasions in a period of 3 months.

Statistical analysis

Data was statistically described in terms of mean ± standard deviation (±SD), or frequencies (number of cases) and percentages when appropriate. Comparison of numerical variables between the study groups was done using Student *t* test for independent samples. For comparing categorical data, Chi-square (χ^2) test was performed. Exact test was used instead when the expected frequency is <5. *P* values <0.05 was considered statistically significant. All statistical calculations were done using computer program SPSS (Statistical Package for the Social Science; SPSS Inc., Chicago, IL, USA) release 15 for Microsoft Windows (2006).

Results

Among 73 children who were assessed for eligibility, 50 children were eligible to be included in this study; however 23 were excluded for being ineligible. The underlying cause of bronchiectasis was CF in 16/50 children (32%) followed by interstitial lung diseases (ILDs) in 8/50 (16%), then immunodeficiency and idiopathic causes 6/50 (12%) for each, details of underlying causes were given in Table 1.

In the current study, a total of 48/50 eligible patients were capable of performing spirometry, but unfortunately there were two missed patients among the non CF bronchiectasis group who couldn't perform the test.

On comparing clinical, functional, and radiological outcome in CF and non-CF bronchiectasis in this study we noticed that the radiological extent of bronchiectasis was more frequently obvious among patients with non-CF bronchiectasis (*p* value 0.01). Although, very severe impairment of FEV1 was only observed among CF bronchiectasis group, there was a significant FEV_1 impairment among non-CF bronchiectasis group (*p* value 0.002). Results of PFTs showing much more airflow obstruction pattern of lung involvement among the non- CF group (*P* value 0.034). Chronic colonization with *Pseudomonas aeruginosa* was significantly recorded in 13 patients (81.3%) of the CF group (*p* value < 0.001), as were shown in Table 2.

A significant proportion of children enrolled in this study had history of hospitalization with severe exacerbation (72%); the mean number of hospitalization for severe exacerbation/year was 0.9 ± 1.1 in CF bronchiectasis group and 1.5 ± 1.0 in non-CF group. Mortality was the end point of study in eight (16%) children, who

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