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LETTER TO THE EDITOR

Should there be a tailored guided management plan for children with post-infectious bronchiolitis obliterans and bronchiectasis?

Dear editor,

Current guidelines for chronic lung diseases such as cystic fibrosis (CF), Primary Ciliary Dyskinesia (PCD) and Bronchiectasis in children recommend regular and multidisciplinary monitoring to delay long—term pulmonary complications.[1-3]

Post-infectious Bronchiolitis Obliterans (PIBO) with or without Bronchiectasis is heterogeneous with diverse clinical expression and severity,[4-6] which contributes to the lack of specific guidelines for monitorization of disease progression and precludes the design of management plans after diagnosis is established.

We aimed to describe the management plans of children with PIBO and/or Bronchiectasis in a tertiary care hospital and analyse whether the variability of care depends on the severity of the obstructive ventilatory defect as determined by FEV_1.

A retrospective chart review of children with PIBO and/or bronchiectasis followed in a tertiary care paediatric hospital was undertaken. Diagnosis was based on clinical history and thoracic computed tomography (CT) findings. All patients followed for two consecutive years (2018-19) were included. Children with otherwise etiological diagnosis like CF or PCD were excluded. The frequency (average per year) of respiratory physician visits, physiotherapy prescription (including education in the management of airway clearance), number of spirometric evaluations, anthropometric assessments and number of microbiological samples taken for bacterial culture were registered.

At our centre, spirometry is performed in cooperative patients older than 4 years of age in accordance with ERS/ATS guidelines. Analysis of height, weight, and BMI z—scores (standard scores) taken from the spirometry records and standardized using WHO reference values was completed. The best FEV_1 and FEV_1/FVC results for the calendar year were collected for all patients who performed good quality tests and averaged for analysis.

Respiratory microbiology surveillance (frequency of testing) was determined from medical record as was referral rates to physiotherapy services. The number of samples (sputum or cough swabs) analysed in a calendar year was counted and averaged.

A descriptive analysis for continuous variables was done and described as median (min and max). Linear regression analysis between average number of physician visits per year and FEV_1 z-score was performed.

During the study period, 28 children were observed (Table 1). Only four had respiratory samples collected (all with bronchiectasis). During this period, three non-residents in Portugal were observed, seven patients initiated follow-up, four were transferred and three lost to follow-up.

Fourteen (50%) had been prescribed mucus clearance devices or educated on respiratory clearance manoeuvres. Three patients with PIBO (plus bronchiectasis in two) are currently on long term oxygen (FEV_1 z-score ranged from -5.0 and -5.7).

No association between clinic visits, clearance methods or other follow-up measures described and FEV_1 z-score was found.

This study shows that patients with PIBO and/or bronchiectasis had an average of 2.7 physician appointments and performed 2/1.5 spirometries per year. Respiratory microbiology surveillance was low and only half had some record for respiratory rehabilitation or use of mucus clearance devices.

In this sample, lung function alone did not affect follow-up. However, the lack of association may be due to small sample size, presence of outliers and shared follow-up with local hospitals.

Despite the limitations and biases of our retrospective analysis, we can assume that the management plan of these children is heterogeneous, with regular physician appointments and spirometry, but low respiratory microbiology surveillance and no standard physiotherapy or nutritional consultations.

In our setting, patients with PCD, PIBO and/or bronchiectasis are managed by respiratory physicians in a general respiratory clinic without a formal multidisciplinary team. Allied health services are available on request and require a separate appointment. There are no internal or national guidelines for the management of these conditions concerning frequency of visits, lung function testing, collection of respiratory samples for bacterial culture, nutritional assessment, timing of imaging by CT or referral to respiratory physiotherapists. By comparison, at the same centre, patients with CF are seen at least every three months by a multidisciplinary team according to international recommendations.

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Table 1 Description of Patients' characteristics.

	Post-Infectious Bronchiolitis Obliterans n=16 (PIBO plus bronchiectasis 6)	Bronchiectasis n=12
Median age at last appointment (in years)	12.5 (7.1, 18.2)	14.1 (8.2, 17.9)
Male gender (n)	12	8
Median duration of follow-up (in years)	10.0 (2.5, 15.4)	8.1 (2.5, 14.4)
Average number of physician visits per year (range)	2.66 (1, 6)	2.67 (1, 6)
Average number of spirometries performed per year	2.00 (1, 4)	1.46 (1, 4)
Average BMI z-score	−0.69	0.36
Average FEV_1 z-score	−3.24	−0.54
Average FEV_1/FVC z-score	−2.57	−0.11

Legend: results are shown as median (min. and max.) or average (range).
PIBO: Post-Infectious Bronchiolitis Obliterans; BMI: Body Mass Index; FEV_1: Forced Expiratory Volume in 1 second; FVC: Forced Vital Capacity; z-score: standard score.

The clinical course of children with PIBO and/or bronchiectasis can be less predictable and differs from CF and PCD. While some children have serious structural and functional lung disease, others experience more subtle effects.[5-7] These heterogeneous characteristics are due to aetiology and age at diagnosis/referral to tertiary centres. Furthermore, some of these children tend to stabilize or even improve their overall status, including lung function results, over time.[5,6]

For adult patients with Chronic Obstructive Pulmonary Disease, routine follow-up is essential. It focuses on symptoms, exacerbations, objective measures of airflow limitations and identifying complications and/or comorbidities.[8] However, for children it can be on reversing the disease when possible and halting its progress.[3] Furthermore, integrated care needs to be individualized to the developmental stage of the child and the family's health literacy.

Extrapolating from standards of care for other complex chronic lung diseases could preserve lung function, reduce exacerbations, improve quality of life, prevent nutritional decline, and enhance survival for patients with PIBO and/or bronchiectasis. Since these are rare entities, there is little evidence to advise on their management. Future directions should dictate more precise standards of care tailored by severity, rate of exacerbations, decline of lung function and minimal follow-up requirements.

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