Phenotyping Adults with Non-Cystic Fibrosis Bronchiectasis: A 10-Year Cohort Study in a French Regional University Hospital Center

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Key Words
Bronchiectasis · Epidemiology · Phenotypes

Abstract
Background: Data concerning phenotypes in bronchiectasis are scarce. Objective: The aim of this study was to describe the clinical, functional and microbiological phenotypes of patients with bronchiectasis. Methods: A monocentric retrospective study in a university hospital in France was conducted over 10 years (2002–2012). Non-cystic fibrosis patients with tomographic confirmation of bronchiectasis were included. The clinical, functional and microbiological data of patients were analyzed relying on the underlying etiology. Results: Of the 311 included patients, an etiology was found for 245 of them. At the time of diagnosis, the median age was 61 years and the mean FEV₁ was 63% of predicted. The main causes of bronchiectasis were post-infectious (50%, mostly related to tuberculosis), chronic obstructive pulmonary disease (COPD; 13%) and idiopathic (11%). Other causes were immune deficiency (6%), asthma (4%), autoimmunity (3%), tumor (2%) and other causes (4%). The comparison of phenotypic traits shows significant differences between COPD, congenital and idiopathic groups in term of sex (p = 0.0175), tobacco status (p < 0.0001), FEV₁ (p = 0.0412) and age at diagnosis (p < 0.001), Pseudomonas aeruginosa (PA) colonization (p = 0.0276) and lobectomy (0.0093). Functional follow-up was available in 30% of patients with a median duration of 2.7 years. Presence of PA was associated with a lower median FEV₁ at diagnosis (43% p < 0.003) but not with a faster rate of decline in FEV₁. Conclusion: Distinctive clinical, functional and microbiological features were found for idiopathic, congenital and COPD-related bronchiectasis. A prospective follow-up of these subgroups is necessary to validate their relevance in the management of bacterial colonization and specific complications of these bronchiectases.

Introduction
Bronchiectasis is considered to be the most common outcome of airway injury leading to irreversible dilated bronchi with poor mucus clearance [1]. This results in a vicious circle of bacterial colonization, inflammation and persistent lung tissue destruction. The population of patients with bronchiectasis is extremely heterogeneous, with a wide range of etiologies with clinical, microbiological, functional and radiological features.
Scientific societies have very recently issued or updated statements or guidelines to help clinicians manage patients with bronchiectasis [2]. Moreover, various large-scale registries and consortiums have been created to describe more accurately the patients’ phenotypic and genetic features [3, 4].

Yet, the extreme heterogeneity of this condition remains a challenge for the clinicians. In the era of personalized medicine, it is of great interest to define homogeneous phenotypes of patients sharing similar physiological mechanisms, phenotypes and prognosis. This approach could help define more appropriate therapeutic targets for each group of patients and better understand the physiopathology of bronchiectasis.

Different methods have been proposed to identify discrete groups of bronchiectasis patients, such as the one based on the severity index [4] or the underlying etiology. Indeed, some bronchiectasis factors are thought to specifically trigger the vicious circle of bronchiectasis. It thus seems legitimate to postulate that a homogenous group of patients could share the same etiology and physiopathology. As a matter of fact, some specific phenotypes such as chronic obstructive pulmonary disease (COPD)-associated bronchiectasis have already been described in the literature [3].

In this context, we tried to determine whether specific clinical, functional and bacteriological features could be identified in each bronchiectasis etiological group in our cohort of patients.

Methods

A retrospective monocentric study was conducted in the Pulmonology Department of the University Hospital of Nice, France. The computerized database of the hospital was queried for the diagnosis of bronchiectasis from January 2002 until December 2012 (ICD-10: J47, Q33.4).

Inclusion criteria were: (1) presence of bronchiectasis on a chest computed tomography (CT), and (2) exclusion of any cystic fibrosis transmembrane conductance regulator (CFTR)-related disease. Each selected hospital chart was then carefully reviewed. Age at diagnosis, gender, smoking status, bronchiectasis etiology, bronchial bacteriological status and pulmonary function tests (PFT) were recorded.

The different etiologies of bronchiectasis were classified into ten nosological entities according to their pathophysiological mechanisms (table 1). Bronchiectasis was considered idiopathic when no etiology could be found, provided the following etiologies were ruled out: immune deficiency, congenital disorder (including cystic fibrosis [CF]), and medical history of respiratory infection (measles, tuberculosis, pertussis). When no appropriate diagnostic work-up had been carried out, the etiology was considered as undetermined. Since the aim of this work was to analyze the phenotypes of patients according to their etiologies, we decided to exclude patients with bronchiectasis of unknown origin from the phenotype analyses.

The microbiological status was determined based on bacteriological analysis (sputum or bronchoalveolar lavage). When no laboratory report was available, the information was sought in the medical report.

Respiratory function values at rest were extracted from the medical files. In order to assess the functional decline rate, values over the longest gap of time were considered for each patient. This period stared with the oldest PFT available and lasted until the most recent one was recorded, consistent with the end of the study or the patient being lost to follow-up. For each patient, the availability and the duration of the functional follow-up were recorded.

Respiratory complications [hemoptysis, bronchial artery embolization, lobectomy, pneumothorax, pulmonary hypertension (PH)] and comorbidities (gastroesophageal reflux disease, hiatus hernia, chronic sinusitis) were recorded. Finally, we assessed whether particular features were more likely to be associated with an etiology.

Normality of variables was assessed with the Shapiro-Wilk test. Variables with normal distribution were expressed as mean ± standard deviation. Variables with non-normal distribution were expressed as median with interquartile range (IQR). Nominal values were expressed as a ratio or percentage. The χ² test was used to assess for the difference between frequencies in unpaired groups. The Fisher test was used when samples where smaller than 5. The Student’s t test was used to test for the difference of continuous variables between unpaired samples greater than 30

| Table 1. Classification of non-CF bronchiectasis into nine nosological entities |
|------------------|-------------------------------|
| Idiopathic       | exclusion of all the other etiologies |
| Post-infectious  | tuberculosis, whooping cough, measles, non-tuberculosis mycobacterial infection, Swyer-James syndrome, other unspecified pathogen |
| COPD             | GOLD 1, 2, 3 or 4 unknown |
| Asthma           | asthma, allergic bronchopulmonary aspergillosis |
| Congenital       | primary ciliary dyskinesia, young syndrome, fragile-X syndrome, α1 antitrypsin deficiency |
| Immune deficiency| primitive or acquired |
| Autoimmune       | rheumatoid arthritis, hemorrhagic rectocolitis, Sjögren’s syndrome, granulomatosis with polyangiitis |
| Tumoral          | pulmonary cancer, adenopathy, tracheal papillomatosis, cystic adenomatoid malformation, foreign body |
| Other            | Gastroesophageal reflux disease, radiotherapy, bronchial stenosis, sarcoidosis, tracheomalacia |
| Undetermined     | no etiological exploration, or lacking information |
or following a normal distribution. Otherwise, a non-parametric Mann-Whitney test was used to assess for differences between two groups. The Kruskal-Wallis test was used if more than 2 groups were compared. A p value <0.05 was considered as significant. All statistical analyses were performed using StatView 5.0 software.

Results

Characteristics of the Population

Three hundred and eleven patients were included in the analysis (fig. 1; table 1). Women represented two thirds of the population. Half of the diagnoses were made in the second half of life, with a median age of 61 years (IQR: 41; 73) (table 2). Sixty-six patients were considered as having 'undetermined etiology' as no complete etiological work-up was available in their medical files. Figure 2 shows the distribution of the main etiological groups. For readability, etiologies were grouped as follows: post-infectious, non-post-infectious and idiopathic (see online suppl. table S1, www.karger.com/doi/10.1159/000446923 for details). The largest group consisted of patients with post-infectious bronchiectasis (50%), mostly related to tuberculosis (more than half of the cases). Within the non-post-infectious group, COPD was the most frequent etiology encountered (about one third of the patients). Idiopathic bronchiectasis was found in 9% of the cohort.

Characterization of Phenotypes in Each Etiological Subgroup

A respiratory function test at diagnosis was available in 57% of the patients. Functional follow-up was available for 30% of the patients with bronchiectasis of known etiology for a median period of 2.6 years. The median rate of decline of forced expiratory volume in 1 s (FEV₁) was –30 ml/year (IQR: –95; 10) in the cohort. There was a trend towards a slower decline in patients with idiopathic and immunodeficiency-related bronchiectasis, although no statistical significance could be found for this variable in these populations.

The clinical, functional and microbiological characteristics of the patients are shown in table 2. COPD-related bronchiectasis predominated in men (M/F ratio = 2.22) and was significantly associated with tobacco use (94% were former or active smokers). In this group, the patients were the oldest and had the lowest FEV₁ at diagnosis and Pseudomonas aeruginosa (PA) was the most frequently found microorganism in their sputum (65%).

Patients with congenital bronchiectasis were the youngest at diagnosis [median age: 20 years (9; 55)] and had the lowest rate of smoking (13%). Almost all of them (93%) had potential pathogen microorganisms in their sputum, and PA was found in 57% of the cases.

Distinctive features were found as well in patients with idiopathic bronchiectasis. Women were overwhelmingly represented in this group (M/F ratio = 0.23) and they were rarely smokers (15%). They had the best FEV₁ values
### Table 2. Characteristics of non-CF bronchiectasis patients with known etiologies

<table>
<thead>
<tr>
<th></th>
<th>Total (n = 245)</th>
<th>Post-infectious (n = 123)</th>
<th>Non-post-infectious (n = 95)</th>
<th>COPD (n = 32)</th>
<th>congenital (n = 15)</th>
<th>immune deficiency (n = 14)</th>
<th>asthma (n = 10)</th>
<th>autoimmune (n = 7)</th>
<th>tumor (n = 6)</th>
<th>other (n = 11)</th>
<th>Idiopathic (n = 27)</th>
<th>p</th>
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<tr>
<td>Female</td>
<td>145 (59)</td>
<td></td>
<td>10 (31)</td>
<td>8 (53)</td>
<td>7 (50)</td>
<td>7 (70)</td>
<td>5 (71)</td>
<td>3 (50)</td>
<td>5 (45)</td>
<td></td>
<td>22 (81)</td>
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<td>M/F ratio</td>
<td>0.69</td>
<td>2.22</td>
<td>0.88</td>
<td>1.00</td>
<td>0.43</td>
<td>0.4</td>
<td>1.00</td>
<td>1.00</td>
<td>1.2</td>
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<td><strong>Smoking status</strong></td>
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<td>Former or active</td>
<td>88 (36)</td>
<td>30 (94)</td>
<td>2 (13)</td>
<td>4 (28)</td>
<td>3 (30)</td>
<td>1 (18)</td>
<td>3 (50)</td>
<td>3 (27)</td>
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<td>4 (15)</td>
<td>&lt;0.0001</td>
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<tr>
<td>Never</td>
<td>70 (29)</td>
<td>2 (6)</td>
<td>11 (74)</td>
<td>5 (36)</td>
<td>2 (20)</td>
<td>1 (18)</td>
<td>1 (20)</td>
<td>1 (10)</td>
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<td>16 (59)</td>
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<tr>
<td>Unknown</td>
<td>87 (35)</td>
<td>0 (0)</td>
<td>2 (13)</td>
<td>5 (36)</td>
<td>5 (30)</td>
<td>5 (63)</td>
<td>2 (30)</td>
<td>7 (63)</td>
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<td>7 (26)</td>
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<tr>
<td><strong>Median age at diagnosis (IQR), years</strong></td>
<td>60 (4; 72)</td>
<td>73 (64; 78)</td>
<td>20 (9; 55)</td>
<td>29 (17; 56)</td>
<td>58 (54; 62)</td>
<td>68 (46; 74)</td>
<td>62 (55; 75)</td>
<td>69 (54; 76)</td>
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<td>42 (22; 65)</td>
<td>&lt;0.0001</td>
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<tr>
<td>PA</td>
<td>83 (42)</td>
<td>41 (40)</td>
<td>15 (65)</td>
<td>4 (31)</td>
<td>2 (50)</td>
<td>3 (50)</td>
<td>3 (100)</td>
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<td>S. aureus</td>
<td>43 (22)</td>
<td>16 (36)</td>
<td>7 (30)</td>
<td>2 (15)</td>
<td>– (0)</td>
<td>3 (50)</td>
<td>– (0)</td>
<td>2 (25)</td>
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<td></td>
<td>8 (35)</td>
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<tr>
<td>MACs</td>
<td>7 (4)</td>
<td>5 (5)</td>
<td>– (0)</td>
<td>– (0)</td>
<td>– (0)</td>
<td>– (0)</td>
<td>– (0)</td>
<td>– (0)</td>
<td></td>
<td></td>
<td>2 (9)</td>
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<tr>
<td>H. influenza</td>
<td>62 (32)</td>
<td>30 (29)</td>
<td>8 (35)</td>
<td>6 (46)</td>
<td>1 (25)</td>
<td>2 (33)</td>
<td>1 (33)</td>
<td>1 (13)</td>
<td></td>
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<td>6 (26)</td>
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<tr>
<td>M. catarrhalis</td>
<td>10 (51)</td>
<td>2 (2)</td>
<td>3 (13)</td>
<td>2 (15)</td>
<td>– (0)</td>
<td>– (0)</td>
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<td>– (0)</td>
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<td></td>
<td>1 (4)</td>
<td></td>
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<tr>
<td>No pathogen</td>
<td>54 (27)</td>
<td>30 (29)</td>
<td>4 (17)</td>
<td>3 (23)</td>
<td>1 (25)</td>
<td>1 (16)</td>
<td>– (0)</td>
<td>6 (63)</td>
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<td>7 (30)</td>
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<td>Pulmonary function values</td>
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<tr>
<td>Mean FEV1 at diagnosis ± SD, % pred</td>
<td>62±43</td>
<td>55±46</td>
<td>42±18</td>
<td>45±33</td>
<td>72±53</td>
<td>84±19</td>
<td>52±12</td>
<td>68±40</td>
<td></td>
<td></td>
<td>74±57</td>
<td>0.0412</td>
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<td>Median follow-up duration (IQR), years</td>
<td>2.1 (1.7; 4.8)</td>
<td>3.25 (1.6; 4.9)</td>
<td>4.5 (2.2; 8.9)</td>
<td>2.6 (2.2; 4.2)</td>
<td>1.5 (1.4; 1.6)</td>
<td>2.7 (2.2; 3.9)</td>
<td>3.5 (2.1; 5.5)</td>
<td>2.8 (1.2; 2.7)</td>
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<tr>
<td>Median ΔFEV1/Δt (IQR), ml/year</td>
<td>-30 (–95; 10)</td>
<td>-22 (–58; 9)</td>
<td>-58 (–11; –20)</td>
<td>-107 (–120; –2)</td>
<td>-8 (–26; 94)</td>
<td>-50</td>
<td>-37 (–215; 27)</td>
<td>-99 (–107; –87)</td>
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<td>Complications</td>
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<tr>
<td>GERD/HH</td>
<td>25 (10)</td>
<td>12 (10)</td>
<td>1 (3)</td>
<td>– (0)</td>
<td>1 (7)</td>
<td>2 (20)</td>
<td>1 (14)</td>
<td>1 (17)</td>
<td>1 (14)</td>
<td></td>
<td>2 (7)</td>
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<tr>
<td>Sinusitis</td>
<td>18 (7)</td>
<td>8 (7)</td>
<td>– (0)</td>
<td>1 (7)</td>
<td>2 (14)</td>
<td>1 (10)</td>
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<td>– (0)</td>
<td></td>
<td></td>
<td>6 (22)</td>
<td></td>
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<tr>
<td>Hemoptysis</td>
<td>55 (22)</td>
<td>37 (30)</td>
<td>5 (16)</td>
<td>3 (20)</td>
<td>1 (7)</td>
<td>2 (20)</td>
<td>1 (17)</td>
<td>3 (27)</td>
<td></td>
<td></td>
<td>3 (11)</td>
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<tr>
<td>Embolization</td>
<td>10 (4)</td>
<td>7 (19)</td>
<td>– (0)</td>
<td>– (0)</td>
<td>– (0)</td>
<td>– (0)</td>
<td>1 (17)</td>
<td>1 (33)</td>
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<td>1 (33)</td>
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<tr>
<td>Lobectomy</td>
<td>27 (11)</td>
<td>10 (8)</td>
<td>1 (3)</td>
<td>6 (40)</td>
<td>2 (14)</td>
<td>1 (10)</td>
<td>1 (9)</td>
<td>5 (19)</td>
<td></td>
<td></td>
<td>0.0093</td>
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<tr>
<td>PH</td>
<td>13 (5)</td>
<td>4 (3)</td>
<td>5 (16)</td>
<td>– (0)</td>
<td>1 (7)</td>
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<td>– (0)</td>
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<td></td>
<td>2 (7)</td>
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</table>

Values are shown as n (%), unless otherwise indicated. In each group, the number of patient included in the analysis is indicated in parentheses in italics; when no precision is given, the full effective of was used. Underlined figures indicate extreme values in the row when the variable shows a statistically different repartition between the compared groups. COPD = Chronic obstructive pulmonary disease; FEV1 = forced expiratory volume in 1 s; GERD = gastroesophageal reflux disease; HH = hiatus hernia; PH = pulmonary hypertension; SD = standard deviation; IQR = interquartile range.
at diagnosis (median FEV$_1$ = 74% of predicted, standard deviation = 57) and were less likely to carry PA (30%).

No distinctive phenotypic trait could be observed in the group of patients with bronchiectasis of undetermined etiology (data not shown).

**The Impact of PA**

Microbiological documentation could be found in 57% of patients. To address the impact of PA we compared patients with PA to those with normal flora and those with other pathogens than PA (table 3). PA was significantly associated with a lower FEV$_1$ at diagnosis (p = 0.003). PA was not significantly associated with the annual decline rate of FEV$_1$, though a trend towards a more rapid degradation could be observed compared to patients with normal flora and no potential pathogen microorganisms.

Complications Associated with Bronchiectasis

Hemoptysis was encountered in about 1 out of 5 patients. Twenty-four percent of them required arterial embolization. The hemoptysis rate tended to be lower (10%) in autoimmune, immune deficiency and idiopathic bronchiectasis.

Lobectomy for bronchiectasis was performed in 10% of patients, with a significant variation between groups (p = 0.0093). Surgical treatment was more prevalent in idiopathic and congenital bronchiectasis.

PH was found in 5% of patients without significant variation between the etiological subgroups. PH was found in 16% of COPD-associated bronchiectasis cases.

**Discussion**

This series reports 311 consecutive patients with non-CF bronchiectasis included over a 10-year period (2002–2012). Distinctive phenotypes of patients with bronchiectasis could be identified according to their etiology. As a matter of fact, specific clinical, functional and microbiological features could be identified in idiopathic, congenital and COPD-associated bronchiectasis. Interestingly, post-infectious bronchiectasis showed no distinctive features compared to the other non-post-infectious subgroups.
Despite the growing number of reported cohorts of non-CF bronchiectasis, information regarding phenotypes remains scarce or contradictory. Thirty [5–34] of the thirty-six reported series [4–39] are smaller than ours. Half of them were retrospective. Five characterized the etiology of bronchiectasis. Notably, only one [31] was aimed at phenotyping a prospective cohort of 189 patients.

Six cohorts larger than ours have been described [4, 35–39]. They were mainly prospective and were designed for epidemiological purposes or for the construction and validation of a severity score and not with the intent to phenotype. The rate of etiology repartition was comparable to that previously described. The reported proportion of idiopathic bronchiectasis is highly variable, ranging from 6 to 77%. We found a 11% rate of idiopathic bronchiectasis, which is low but consistent with other cohorts using the same stringent definition criteria as we used. In other cohorts, the rate of idiopathic forms may be falsely increased by the confusion between idiopathic and unknown etiology.

We report a 50% rate of infectious bronchiectasis, which is more than what is usually described (20–30%). The high proportion of post-tuberculosis bronchiectasis in our cohort can explain this discrepancy. Indeed, for historical and demographic reasons, our center serves a population with a high incidence of tuberculosis [40].

COPD-associated bronchiectasis is our third etiologic group (13%), a proportion that is consistent with other reported rates (10–20%) [4, 32]. This specific phenotype has been previously identified and has been shown to be associated with a higher morbidity and mortality rate than COPD alone [30, 41]. As expected, these patients featured the poorest prognostic factors: the most severe obstruction, the most frequent presence of PA in the sputum and the highest proportion of PH (16%).

Recently, Aliberti et al. [42] developed a non-etiologic cluster-based approach to identify new phenotypes in bronchiectasis. Variables, such as sputum production and chronic infection, were implicated in the definition of new homogenous entities of bronchiectasis, independently of the underlying etiology. However, the analysis detected significant associations between some clusters and etiologies, especially in idiopathic, post-infectious and COPD-related bronchiectasis. These data confirm the relevance of our results, particularly concerning the identification of the COPD and the post-infectious phenotype.

One limitation of our work is its retrospective nature. Missing data limits the significance of some results, especially concerning the functional follow-up of patients. Thus, we show a dramatically low rate of sinusitis. This could nevertheless be explained by the fact that patients with ear, nose and throat (ENT) disorders are managed in an independent hospital with no shared medical file and database. Some relevant phenotypic information was also not retrospectively available in the medical files, such as time since first diagnosis. Moreover, our cohort partially consists of in-patients referred to a tertiary referral university hospital, which implies an inclusion bias. Our patients were more likely to be referred for a severe disease or be in exacerbation. However, despite all these limitations, the structure of our cohort remains comparable to what has previously been reported.

The high rate of unknown etiology (21%) is a debatable point. This rate corresponds to patients for whom no extensive diagnostic work-up could be found in the medical file. We chose not to include for phenotype analysis patients for whom we could not assess with certainty the etiology, since they represented a highly heterogeneous population with no clinical relevance. This methodological option was not chosen in some previous studies, being potentially responsible for an overestimation of the idiopathic cause.

This latter point highlights the fact that no proper recommendations were available before the publication of the first British Thoracic Society guidelines for bronchiectasis in 2010 [43]. This publication marked a turning point in medical practice of patients with bronchiectasis. As a matter of fact, a systematic etiological work-up allowed a decrease in the rate of unknown etiologies in our center (data not shown). These recommendations also led to dramatic changes in infectious management. Since 2010, thanks to a more appropriate use of antibiotics, we observed that lobectomy in congenital bronchiectasis had virtually disappeared.

Another limitation was the absence of discrimination between chronic colonization and primary infection with PA. However, if we consider that the presence of a mucoid stain indicated chronic colonization, we can assume that at least 19% of patients had chronic colonization by PA, which is consistent with the Belgian cohort by Geminne et al. (13%) [36].

PA colonization was not associated with a decline in FEV₁ in this retrospective cohort. However, the low rate of patients with available PFT makes the assessment of the relationship between PA and the decline of FEV₁ not reliable. Yet, this result is consistent with a previous study by Davies et al. [20]. Conflicting results have been reported in other cohorts that showed a faster decline in FEV₁.
in PA-colonized patients [7, 22]. However, the causal link between the rate of decline in FEV\textsubscript{1} and PA remains controversial. Only a prospective study can validate such a link. However, PA must be actively sought, as it was identified as a risk factor for pulmonary exacerbation in non-CF bronchiectasis [25].

**Conclusion**

This study provides an overview of patients treated for bronchiectasis in a tertiary center. Specific phenotypes were identified in idiopathic, congenital and COPD-related bronchiectasis. The presence of PA in sputum was not associated with an accelerated decline in function. This work highlights the need for a standardized etiological diagnostic approach for bronchiectasis. Recommendations regarding eradication and background treatment against PA are needed, given the high prevalence and the morbidity involving this pathogen, especially in patients with COPD. This study underlines the need for setting up longitudinal cohorts including well-phenotyped patients. The European EMBARC (European Multicentre Bronchiectasis Audit and Research Collaboration) network is currently being developed to achieve this goal [44].

**References**