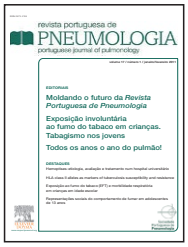
Rev Port Pneumol. 2011;**17(1):**32-40

revista portuguesa de
PNEUMOLOGIA
portuguese journal of pulmonology
www.revportpneumol.org



SPECIAL ARTICLE

# Bronchiectasis: do we need aetiological investigation?

**A. Amorim[a,]* and J. Gracia Róldan[b]**

[a]*Pneumology Department, Hospital São João-EPE, Faculdade de Medicina, Universidade do Porto, Porto, Portugal*
[b]*Pneumology Department, Hospital Universitari Vall d'Hebron, Universidad Autónoma de Barcelona, Barcelona, Spain*

Received August 26, 2010; accepted September 28, 2010

**KEYWORDS**
Bronchiectasis;
Aetiology;
Investigation

**Abstract**
Bronchiectasis (BE) is characterized by irreversible dilation and damage to the bronchial walls. It is a morphological expression of a large variety of pathologies.

The true prevalence of BE is unknown. It is known, however, that in Third World countries it is common due to lung infections. Its current prevalence in developed countries is rather significant due to a greater capacity for diagnosis, its association with highly prevalent diseases and to a greater chronicity of some of the underlying pathologies.

Over the last few decades there has been little interest in the investigation of BE, unless it is associated with cystic fibrosis. One of the reasons is the presupposition that treatment is the same for all patients diagnosed with BE, regardless of the underlying aetiology. Several works carried out over the last decade show that a diagnosis based on aetiology changes both the approach and the treatment of BE within a relevant percentage of patients, with a consequent change in the prognosis.

Currently, systematic investigation into the aetiology of BE is recommended, particularly in those disorders that respond to specific treatment.


**PALAVRAS-CHAVE**
Bronquiectasias;
Etiologia;
Investigação

**Bronquiectasias: será necessária a investigação etiológica?**

**Resumo**
As bronquiectasias caracterizam-se por uma dilatação e destruição irreversível das paredes brônquicas. São a expressão morfológica de uma grande variedade de patologias.

A verdadeira prevalência das BE é desconhecida, contudo sabe-se que são frequentes nos países de Terceiro Mundo devido a infecções pulmonares. A sua prevalência nos países desenvolvidos é também significativa em consequência da maior capacidade de diagnóstico, da sua associação com doenças de prevalência elevada e da maior cronicidade de algumas das patologias subjacentes.

*Corresponding author.
*E-mail:* adelinamorim@gmail.com (A. Amorim).

Durante as últimas décadas tem havido pouco interesse na investigação das BE, exceptuando as consequentes à fibrose quística. Uma das razões relaciona-se com o pressuposto de que o tratamento é comum a todos os doentes com BE, independentemente da etiologia subjacente. Vários trabalhos nesta última década demonstraram que a obtenção de um diagnóstico etiológico modifica a abordagem e o tratamento numa percentagem relevante de doentes e consequentemente o seu prognóstico.

Recomenda-se actualmente a investigação sistemática da etiologia, principalmente das doenças que poderão responder a um tratamento específico.


## Introduction

Bronchiectasis (BE) is characterized by irreversible dilation and damage to the airways, associated with a vicious cycle of inflammation, recurrent infection and bronchial impairment.

BE is frequently described as a lung disease but it is more appropriate to say that it is the pathological expression of a large variety of disorders. However, its aetiology is still unknown in many cases.

Practically two centuries have gone by since René Laënnec described a patient with BE[1] for the first time, more than 80 years since Jean Athanase Sicard introduced the bronchography as a diagnostic tool,[2] more than 50 years since Reid described and classified BE based on histologic and bronchographic findings[3] and approximately 20 years since high resolution computed tomography (HRCT) has progressively replaced bronchography as a diagnostic tool.

Over the last century several syndromes/pathologies associated with BE have been described: the Mounier-Kuhn syndrome in 1932,[4] cystic fibrosis (CF) in 1944,[5] the Williams-Campbell syndrome in 1960,[6] the yellow nail syndrome in 1964,[7] diffuse panbronchiolitis in 1969,[8] the Young's syndrome in 1970,[9] the Lady-Windermere syndrome in 1992.[10]

During the last two decades of the 20th century an apparent correlation was noted between BE and several systemic disorders, such as rheumatoid arthritis, inflammatory bowel disease and AIDS.[11] Just recently an association between the autosomal dominant polycystic kidney disease and BE[12] has also been described.

Towards the end of the 80's, Barker defined BE as "an orphan disease",[13] referring to the definition used to describe diseases that had been forgotten by the scientific community and were of no interest for therapeutic research due to the supposedly low prevalence.[14]

## Epidemiology

There are very few studies on the true prevalence of BE. In the past, tuberculosis, whooping cough and measles were seen as the significant causes of BE. Better social conditions, the development of broad spectrum antibiotics, the appropriate treatment of pulmonary tuberculosis and vaccination during childhood have most certainly contributed toward a decrease in postinfectious BE in developed countries. On the other hand, various factors have contributed to an increased prevalence of BE in these same developed countries: greater diagnostic capability through the use of HRCT, insight into new causes of BE that are highly prevalent, such as AIDS, recognising that BE can be a possible complication of heart, lung and bone marrow transplants and the higher survival rate of patients with CF and immunodeficiency disorders.

In the 40's and 50's in the United Kingdom, there was described a prevalence of 77-130/100,000 inhabitants.[15,16] A recent study, carried out in the USA, calculated a prevalence of 52/100,000 adults. Prevalence was higher in women of all ages, a fact which is in accordance with many other studies.[17]

There was considerably higher prevalence noted in some communities: 16/1,000 in children from a region in Alaska, 15/1,000 in aboriginal children from Australia and 1/6,000 in children from New Zealand.[18-20] These differences are essentially due to social and economic conditions, although other possible underlying genetic factors are also being considered.

There are only 2 studies that estimated incidence. In Finland there was an annual incidence of 0.5/100,000 for children under 15 years of age and 3.9/100,000 for the total population and in New Zealand it was estimated an incidence of 3.7/100,000 for children under 15.[21,22]

A study carried out in the USA revealed that treatment costs per patient were higher than those spent on heart disorders and COPD.[17] Data provided by the United Kingdom indicate that 78% of patients afflicted by BE that use the emergency services are hospitalized and one third have at least one annual exacerbation that requires hospitalization, lasting for an average of 10.5 days, higher than that estimated for other pathologies such as asthma and COPD.[23] To conclude, in another study it was noted that 25% of the patients died within the 9 years following the first hospitalization during which BE was detected.[24]

## The importance of diagnosis and of aetiological investigation

Despite the increasing interest in this pathology, BE continues to be underdiagnosed and given little importance. Many cases are misdiagnosed as asthma or COPD.[25]

Most cases are diagnosed when the illness is already in an advanced stage even though symptoms have been present