The prognosis of PAH has altered since the advent of the prostanoids. These drugs induced a rapid improvement in hemodynamic parameters and exercise capacity, and increased survival. However, their short half-life and mode of administration (epoprostenol, intravenous; iloprost, inhaled; treprostinil, subcutaneous) significantly limit their use. Shortly afterwards, beraprost, an orally active prostanoid, proved beneficial, but its effects were not maintained over time.

Studies with sildenafil provided similar results to those of prostacyclin derivatives. Recent publications report improved functional class, response to effort, and a tendency toward decreasing systolic pulmonary pressure. These findings are associated with those reported for endothelin-1 receptor antagonists.

Platelet-derived growth factor acts as a potent mitogen and chemoattractant in vitro, and it intervenes in the distal extension of the pulmonary vascular smooth muscle cells into small nonmuscular arterioles. Hence current research into imatinib, a platelet-derived growth factor antagonist, for use in this condition. García et al. present 4 cases of patients with severe PAH (functional class IV) whose treatment with prostanoids, sildenafil, or bosentan was combined with imatinib. All the patients in this small series died after starting the drug, although their disease was advanced and their prognosis was very poor.

Based on results published to date on treatment of PAH, bosentan continues to be a valid option. Therefore, we feel that it is important to provide information on cases of patients from other population groups who have been treated with this drug.

References


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### A United Airway: Bronchiectasis Is Also Associated With Chronic Rhinosinusitis and Nasal Polyps

**Una vía respiratoria unificada: las bronquiectasias también se asocian a rinosinusitis crónica y pólipos nasales**

**To the Editor:**

In November 2008, *Archivos de Bronconeumología* published the guidelines of the Spanish Society of Pulmonology and Thoracic Surgery (SEPAR) on the diagnosis and treatment of bronchiectasis. Although the document is extremely important for the diagnosis and treatment of bronchiectasis, it makes no mention whatsoever of the impact of this disease on the upper airway in the form of chronic rhinosinusitis with or without nasal polyps, a fact which was pointed out in a review article published in this journal in 2006.

The concept of a united airway has been demonstrated through epidemiologic, pathophysiologic, diagnostic, and therapeutic associations between bronchopulmonary and sinonasal disease. Reported associations include asthma with allergic rhinitis, asthma with nasal polyposis, and nonallergic asthma and chronic obstructive pulmonary disease (COPD) with chronic rhinosinusitis. Several years ago, prompted by our clinical experience, the otorhinolaryngology and pulmonology departments at Hospital Clínic de Barcelona, Spain started to investigate the association between sinonasal disease and bronchiectasis.

Our research has resulted in the publication of 2 articles with a third in press on the association between bronchiectasis and chronic rhinosinusitis with or without nasal polyposis. Interestingly, 1 of the studies found that 3 out of every 4 patients with bronchiectasis (77%) met clinical and radiologic criteria for chronic rhinosinusitis while 1 out of 4 (25%) had visible nasal polyps on nasal endoscopy. These results support the concept of a united airway and indicate that there could be a yet unknown process in postinfective bronchiectasis that may affect the entire airway. Furthermore, patients with bronchiectasis and chronic rhinosinusitis have a poorer quality of life, measured by both generic and specific questionnaires, than those with bronchiectasis but without chronic sinonasal disease.

From these results we can conclude that all patients with bronchiectasis should be clinically assessed by nasal endoscopy and/or sinus computed tomography for the purpose of confirming a possible diagnosis of chronic rhinosinusitis with or without nasal polyps. Consequently, all patients with bronchiectasis, or indeed with asthma and COPD, should undergo an ear, nose, and throat examination.

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**Table**

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<th>Change in Functional Parameters Between Starting Treatment With Bosentan and Follow-up</th>
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<td>Mean</td>
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<td>6-MWT</td>
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<td>NYHA</td>
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<td>SPP measured by echocardiography</td>
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Abbreviations: NYHA, New York Heart Association; SPP, systolic pulmonary pressure; 6-MWT, 6-minute walk test.
examination, ideally by a multidisciplinary team, with a view to improving the diagnosis, treatment, and follow-up of both bronchopulmonary and sinonasal disease.

References


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Authors’ Reply to “A United Airway: Bronchiectasis Is Also Associated With Chronic Rhinosinusitis and Nasal Polyps”

Respuesta de los autores a la carta: Una vía respiratoria unificada: las bronquiectasias también se asocian a rinorriosis crónica y pólipos nasales

To the Editor:

We would like to thank the authors for their comments and the contributing data from their article,1 which had not been published at the time the guidelines for the diagnosis and treatment of bronchiectasis were drawn up.2 We would like to point out that the guidelines have not been published as a supplement. Moreover, these guidelines are the first to be presented in Spain and indeed internationally. They were designed to improve, facilitate, and unify the approach to managing patients with bronchiectasis, whatever the etiology, by prioritizing interest in managing lung involvement, which is what determines prognosis. Unfortunately, the space available did not allow us to focus on issues, as we would have liked, other than the respiratory monitoring of these patients. Neither is there any mention of the impact that bronchiectasis might have on other organs, on other sites, or on the underlying disease that causes it.

Bronchiectasis is not a disease in itself like asthma or chronic obstructive pulmonary disease. Rather it is a lesion of the bronchial structure that is the end result of varying causes or diseases that may or may not produce alterations in other areas of the airway or in other organs. The guidelines mention that “(s)inusitis may be present, especially if there is cystic fibrosis, primary ciliary dyskinesia, primary immune deficiency, young syndrome, yellow nails syndrome, or diffuse panbronchiolitis” and Figure 1 also mentions ear infection.3 We believe that it would be more appropriate to talk about causes of bronchiectasis that can affect the upper airway rather than bronchiectasis in general.

The only original article on the subject published before the guidelines by the authors of the letter compares patients with bronchiectasis and nasal symptoms with the general population.4 They show that patients with bronchiectasis have a poorer quality of life but do not clarify how bronchiectasis and nasal involvement affect it. Their study only includes nasal endoscopy but not a computed tomography scan of paranasal sinuses and concludes that the presence of nasal polyps has no additional impact on quality of life.5 Therefore, with reference to this study, it cannot be said that patients with bronchiectasis and chronic rhinosinusitis have a poorer quality of life than those with bronchiectasis in the absence of chronic sinonasal disease.

It is always desirable to rely on various specialists interested in cooperating in the management of patients with bronchiectasis. We look forward to reading the authors’ forthcoming publication (still in press at the time of writing this letter), and taking it into consideration if we have the opportunity to participate in future guideline updates.

References


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