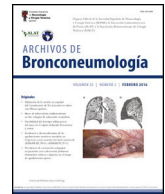




ARCHIVOS DE Bronconeumología

www.archbronconeumol.org



Editorial

Implementation of the Spanish Paediatric Bronchiectasis Registry (Child-BEAR-es Registry)

Childhood bronchiectasis is defined as a clinical syndrome characterized by persistent or recurrent episodes of chronic wet or productive cough and the presence of abnormal bronchial dilatation on chest high-resolution computed tomography (HRCT) with increased broncho-arterial ratio using paediatric data (abnormal when >0.8).¹ Although the adult definition includes irreversible dilatation of the bronchial lumen,² paediatric data have consistently shown that radiographic bronchiectasis, especially when mild and cylindrical, is reversible with early diagnosis and optimal treatment in until 40% of children.^{3,4}

Bronchiectasis is the common denominator of numerous aetiologies, all characterized by failed mucus clearance, chronic bacterial infection, and inflammation. Other than cystic fibrosis, the main aetiologies in children are postinfectious, immunodeficiencies, primary ciliary dyskinesia, aspiration, congenital malformations, and obstructive airway lesions. The prevalence of idiopathic bronchiectasis is still high, between 18 and 55%.^{1,5} While the basics of treatment, improving airway clearance or controlling infections, are the same, irrespective of the underlying cause, there are specific treatments for some aetiologies, and others will have genetic implications for the family.⁶ A precision medicine approach is increasingly possible, and for cases of bronchiectasis without a treatable cause, phenotypes and endotypes may facilitate the recognition of problems and related pathophysiological mechanisms that indicate treatable traits.⁷

Only two decades ago, no patient registries or guidelines on bronchiectasis existed, and this disease's knowledge was limited. The Spanish Society of Pneumology and Thoracic Surgery (SEPAR) promoted the world's first adult national bronchiectasis registry in 2002, which was followed in 2015 by the Computerized Registry of Bronchiectasis (RIBRON), which is collecting cross-sectional and prospective data.^{8,9} Together with the European Multicentre Bronchiectasis Audit and Research Collaboration (EMBARC),¹⁰ these adult registries have made significant contributions to understanding better the natural history of bronchiectasis and various dimensions of the disease, including comorbidities, aetiological, clinical-functional, pathophysiological, diagnostic, microbiological, nutritional, prognostic and therapeutic aspects.⁹

However, insufficient worldwide data exists on paediatric bronchiectasis. Bronchiectasis in children is considered an understudied and underdiagnosed syndrome requiring more attention from the medical community to ensure the correct diagnostic approach and effective treatment.¹¹ Although there seem to be geographical variations, there is hardly any published Spanish

paediatric data regarding its prevalence and incidence in children. Monteagudo et al. described the prevalence and incidence of bronchiectasis in primary care in 2012 in Catalonia. They found a prevalence of 36.2 cases per 10,000 inhabitants and an incidence of 4.81 cases per 10,000 inhabitants, noting that prevalence and incidence rose with age, being lower than 5/10,000 and 1/10,000 in <14 years.¹²

Early childhood factors are mediators of future lung disease, poorer lung function and premature mortality in adults. Proper childhood attention is important for reducing adult lung disease. King et al. reported a high proportion of adult patients with onset of symptoms during infancy, and these patients had more severe disease and worse prognosis than those whose symptoms begin in adult life.¹³ Therefore, the profile of bronchiectasis in children must be studied further to enable its prompt recognition and improved management.¹¹

International guidelines exist for managing bronchiectasis in children, including the European Respiratory Society guidelines.¹⁴ Children's Bronchiectasis Education, Advocacy and Research Network (Child-BEAR-Net) is a European Respiratory Society (ERS) Clinical Research Collaboration whose overall aim is to improve the knowledge about paediatric bronchiectasis and the quality of life of children and their families. They have elaborated quality standards of care, clinical and research priorities, the definition of bronchiectasis exacerbations for clinical practice and for clinical trials, and a bronchiectasis action management plan, all of them through an international point of view.¹⁵

Currently, there are no Spanish guidelines for paediatric bronchiectasis in our country, mainly due to the lack of a proper characterization of our population. It is essential to describe our population and understand its specificities while also considering the functioning of our national healthcare system and its referral process. Describing our population may be the first step towards creating these resources at a national level.

Child-BEAR-Net has promoted the creation of an International Paediatric Bronchiectasis Registry (Child-BEAR-Net Registry). Before this, the Australasian Bronchiectasis Registry was the only bronchiectasis registry that included children. The registry aims to evaluate the underlying aetiology, treatments, and outcomes, assess long-term outcomes that are important to patients, providers, and other decision-makers, and facilitate patient recruitment for clinical studies. It will also be very important to link children's data with adult-based registries to understand the importance of continuity of care throughout life and the impact of early interventions in adult life.

<https://doi.org/10.1016/j.arbres.2024.05.003>

0300-2896/© 2024 SEPAR. Published by Elsevier España, S.L.U. All rights reserved.

Every country that participates in the Child-BEAR-Net Registry can create its national registry using a standard structure. Information collected can be merged, allowing the study of clinical and medical practices among countries. It is important that every country has its own registry, as there may be differences among countries in forms of presentations, etiological spectrum, and management.⁸

We have created the Spanish Paediatric Bronchiectasis Registry (Child-BEAR-es Registry) to provide a tool for generating this needed information. The Child-BEAR-es Registry is part of the Child-BEAR-Net Registry,¹⁵ sharing the same structure and agreeing to data transfer.

Establishing this national registry will help gather data on the epidemiology, aetiology, morbidities, symptoms, diagnosis, and management of paediatric bronchiectasis in Spain. It will enable us to determine the prevalence and incidence of this syndrome and enhance the knowledge of its particularities within our country. In addition, analysing the annual follow-up can help us identify clinical predictors and monitor improvements and deficiencies in the standard of care; furthermore, it will allow us to identify the needs of our paediatric bronchiectasis patients. The registry also establishes a framework for future research projects at a national level and collaborations at an international level.

This project was promoted jointly by SEPAR and the Spanish Society of Paediatric Pulmonology (SENP) and has been created on SEPAR's REDCap platform. The inclusion criteria for this registry are patients aged between 0 and 17 years and diagnosed with bronchiectasis and whose informed consent has been obtained. Patients with a history of lung or heart transplant or those diagnosed with cystic fibrosis are excluded. The registry will include cross-sectional and prospective data; all Spanish centres are invited to collaborate. Our goal is to involve all centres, regardless of their complexity level, to obtain a characterization of the population that is as similar as possible to the reality in Spain. The initial acceptance has been exciting, with positive feedback. Today, 40 Spanish centres are willing to participate in the registry.

We hope that the Spanish Paediatric Bronchiectasis Registry will be as successful as the previous Spanish bronchiectasis registries in providing evidence that improves the care of our children and adolescents.

Funding

There are no funders for this article.

Conflict of Interests

AM-G has received fees for lectures from Abbvie, Sanofi-Pasteur, Janssen and Astra-Zeneca and fees for participation in Advisory Boards sponsored by AbbVie, Sanofi-Pasteur and Astra-Zeneca. VS-S has received fees for lectures from GlaxoSmithKline and Vertex and fees for participation in Advisory Boards sponsored by Novartis. LG-G states that she has no conflict of interests.

References

1. Chang AB, Bush A, Grimwood K. Bronchiectasis in children: diagnosis and treatment. *Lancet*. 2018;392:866–79, [http://dx.doi.org/10.1016/S0140-6736\(18\)31554-X](http://dx.doi.org/10.1016/S0140-6736(18)31554-X).

2. Martínez-García MA, Máiz L, Olveira C, Girón RM, de la Rosa D, Blanco M, et al. Spanish guidelines on the evaluation and diagnosis of bronchiectasis in adults. *Arch Bronconeumol*. 2018;54:79–87, <http://dx.doi.org/10.1016/j.arbres.2017.07.015>.
3. Chang AB, Kantar A, Redding GJ, Hill AT. Is bronchiectasis a reversible disorder? *Arch Bronconeumol*. 2023;59:199–200, <http://dx.doi.org/10.1016/j.arbres.2022.11.007>.
4. Mills DR, Masters IB, Yerkovich ST, McEniery J, Kapur N, Chang AB, et al. Radiographic outcomes in paediatric bronchiectasis and factors associated with reversibility. *Am J Respir Crit Care Med*. 2024, <http://dx.doi.org/10.1164/rccm.202402-04110C>.
5. Brower KS, Del Vecchio MT, Aronoff SC. The etiologies of non-CF bronchiectasis in childhood: a systematic review of 989 subjects. *BMC Pediatr*. 2014;14:4, <http://dx.doi.org/10.1186/s12887-014-0299-y>. PMID: 25492164; PMCID: PMC4275950.
6. Bush A, Floto RA. Pathophysiology, causes and genetics of paediatric and adult bronchiectasis. *Respirology*. 2019;24:1053–62, <http://dx.doi.org/10.1111/resp.13509>.
7. Choi H, Chalmers JD. Bronchiectasis: from orphan disease to precision medicine. In: Wagner TOF, Humbert M, Wijsenbeek M, et al., eds. *Rare diseases of the respiratory system (ERS monograph)*. Sheffield, European Respiratory Society, 2023; pp. 150–164. doi:10.1183/2312508X.10018122.
8. Martínez-García MÁ, Oscullo G, Gómez-Olivas JD, Olveira C, Girón R, García-Clemente M, et al. Bronchiectasis: changes in the characterization of patients during 20 years of follow-up. Data from the Spanish bronchiectasis registries. *Arch Bronconeumol*. 2023;59:688–90, <http://dx.doi.org/10.1016/j.arbres.2023.07.023>. English, Spanish.
9. de la Rosa-Carrillo D, Máiz-Carro L, Martínez-García MA. What have we learned about bronchiectasis from RIBRON (Spanish bronchiectasis registry)? *Arch Bronconeumol*. 2023;59:625–6, <http://dx.doi.org/10.1016/j.arbres.2023.07.010>.
10. Chalmers JD, Polverino E, Crichton ML, Ringshausen FC, De Soyza A, Vendrell M, et al. Bronchiectasis in Europe: data on disease characteristics from the European Bronchiectasis registry (EMBARC). *Lancet Respir Med*. 2023;11:637–49, [http://dx.doi.org/10.1016/S2213-2600\(23\)00093-0](http://dx.doi.org/10.1016/S2213-2600(23)00093-0).
11. Chang AB, Dharmage SC, Marchant JM, McCallum GB, Morris PS, Schultz A, et al. Improving the diagnosis and treatment of paediatric bronchiectasis through research and translation. *Arch Bronconeumol*. 2024, <http://dx.doi.org/10.1016/j.arbres.2024.03.003>. S0300-2896(24)00062-0.
12. Monteagudo M, Rodríguez-Blanco T, Barrecheuren M, Simonet P, Miravittles M. Prevalence and incidence of bronchiectasis in Catalonia, Spain: a population-based study. *Respir Med*. 2016;121:26–31, <http://dx.doi.org/10.1016/j.rmed.2016.10.014>.
13. King PT, Holdsworth SR, Farmer M, Freezer N, Villanueva E, Holmes PW. Phenotypes of adult bronchiectasis: onset of productive cough in childhood and adulthood. *COPD*. 2009;6:130–6, <http://dx.doi.org/10.1080/15412550902766934>.
14. Chang AB, Fortescue R, Grimwood K, Alexopoulou E, Bell L, Boyd J, et al. European Respiratory Society guidelines for the management of children and adolescents with bronchiectasis. *Eur Respir J*. 2021;58:2002990, <http://dx.doi.org/10.1183/13993003.02990-2020>. PMID: 33542057.
15. Chang AB, Boyd J, Bush A, Grimwood K, Hill AT, Powell Z, et al. Children's Bronchiectasis Education Advocacy and Research Network (Child-BEAR-Net): an ERS Clinical Research Collaboration on improving outcomes of children and adolescents with bronchiectasis. *Eur Respir J*. 2021;58:2101657, <http://dx.doi.org/10.1183/13993003.01657-2021>. PMID: 34675034.

Laura Garriga-Grimau^a, Verónica Sanz-Santiago^b,
Antonio Moreno-Galdó^{a,c,*}

^a Paediatric Pulmonology Section, Department of Paediatrics,
Hospital Universitari Vall d'Hebron, Vall d'Hebron Barcelona Hospital
Campus, Universitat Autònoma de Barcelona, Spain

^b Paediatric Pulmonology Section, Hospital Universitario Infantil Niño
Jesús, Madrid, Spain

^c Centro de Investigación Biomédica en Red (CIBER) de Enfermedades
Raras (CIBERER), Instituto de Salud Carlos III, Madrid, Spain

* Corresponding author.

E-mail address: antonio.moreno@vallhebron.cat
(A. Moreno-Galdó).