Get together to increase awareness in bronchiectasis: a report of the 2nd World Bronchiectasis Conference

Stefano Aliberti1* and James D. Chalmers2

From 2nd World Bronchiectasis Conference
Milan, Italy. 06-08 July 2017

Bronchiectasis is a chronic respiratory disease characterized by a permanent dilation of the bronchi associated with cough, daily sputum production and recurrent episodes of respiratory infections [1]. Bronchiectasis pathophysiology starts with a structural airway damage leading to an impaired mucociliary clearance, subsequent chronic bacterial infection and neutrophilic inflammation, which perpetuates this vicious cycle [1]. For a long period of time, bronchiectasis has been recognized as an isolated radiological finding, while now we understand it as a chronic and debilitating disease which requires specific management to improve patients’ outcomes.

A renewed interest in bronchiectasis has characterized the field of respiratory medicine over the past few years. When bronchiectasis has been the main theme of scientific sessions at national or international meetings, rooms are packed with colleagues sharing their intellectual interest in this disease. The multidisciplinary approach which characterizes the management of bronchiectasis naturally predisposes physicians to share their experiences and work together to define common pathways to optimize patients’ care [1, 2]. This is something that rarely happens in our field and is the reason why we have seen such a substantial increase in the awareness of bronchiectasis which has, in turn, led to a better knowledge on different aspects of this disease.

Bronchiectasis is no more a neglected disease, nor even less a rare one. Recent data suggest a prevalence up to 566/100,000 subjects in the general population with an increase of approximately 40% in the past decade [3]. Bronchiectasis significantly impacts healthcare systems worldwide with an annual exacerbation frequency up to 3 per patient per year and has a clear attributable mortality [4]. However, real-life data suggest that, to date, quality of bronchiectasis care remains poor [5]. A new and emerging interest has been promoted on this condition as demonstrated by an increase in research activity and the development of international registries over the past few years [6]. New areas of the disease management have been recently considered and different research and clinical gaps have been identified [7].

This special issue of Multidisciplinary Respiratory Medicine is following up the largest meeting ever held exclusively focused on bronchiectasis, the 2nd World Bronchiectasis Conference (WBC, Milan, Italy, July 2017; www.world-bronchiectasis-conference.org). This congress, like the previous one held in Hannover, Germany, in 2016 (world-bronchiectasis-conference.com), represents the clear evidence of the emergence of bronchiectasis as a key clinical entity. Meetings like the WBCs offer the possibility for different healthcare professionals working on bronchiectasis to get together from different continents and actively discuss fundamental aspects of the disease management. These meetings also offer unique opportunities to strengthen collaborations with patients and regulatory agencies, and for young investigators to publish their work with an exceptional international visibility [8]. Important steps towards a better understanding of bronchiectasis have been taken during both WBCs. In Hannover, an international group of investigators developed a consensus definition of exacerbation for clinical trials involving patients with bronchiectasis [9]. In the same line, a discussion started at the 2nd WBC in Milan between bronchiectasis patients and healthcare professionals on the risk of cross-infection which ended up with the first position...
statement from the European Bronchiectasis Network, EMBARC/ELF patient advisory group and European Reference Network Bronchiectasis Network on this topic [10].

Bronchiectasis is a heterogeneous respiratory disorder and represents the final pathway of several infectious, genetic, immunologic or allergic diseases [11, 12]. An accurate and prompt identification of the underlying cause is a key recommendation of several international guidelines, including rare diseases such as cystic fibrosis (CF) and primary ciliary dyskinesia (PCD) [13, 14]. The diagnosis of PCD and CF has relevant clinical, socioeconomic and psychological implications, which affect patients’ life, including the possibility to have a specific and multidisciplinary team approach in referral centres, as well as a genetic and fertility counselling and special legal aspects in some countries. Both PCD and CF have been hot topics discussed during the 2nd WBC and are arguments of three different papers published in the present issue [15–17]. The papers by Gramegna and Contarini review current literature on PCD and CF and offer interesting insights on why, when and how to investigate both conditions in bronchiectasis, while Robinson and co-workers summarize structural findings of PCD and highlight the radiological differences between PCD and other causes of bronchiectasis [15–17]. An entire session of the 2nd WBC has been dedicated to non-tuberculous mycobacteria (NTM) as one of the most difficult-to-treat pathogens in bronchiectasis [18]. Several open questions exist on NTM from the microbiological diagnosis to the identification of NTM lung disease in the context of a chronic respiratory disease such as bronchiectasis [19]. Real life data coming from international registries are valuable to understand current practices and design translational research [6]. This is the case of the Italian Registry of Pulmonary NTM (IRENE; www.registroirene.it) whose protocol has been reported in the present issue of the journal [20].

New diagnostic techniques have been presented and discussed at the conference such as shotgun metagenomic sequencing which provides an alternative approach that allows the microbial composition of clinical samples to be described in detail, including the prevalence of resistance genes and virulence traits. Rogers and co-workers described a novel, cost-effective strategy for screening patient cohorts for changes in resistance gene prevalence and identified population-level changes in the relative abundance of specific macrolide resistance genes [21]. This is just one of the examples of how new technologies are advancing our understanding of bronchiectasis and how future multi-omics approaches performed across large multicentre studies might help us in interpreting the heterogeneity of this disease [22]. The 2nd WBC has been one of the most productive meeting exclusively focused on bronchiectasis and helped to pave the way towards an increase in bronchiectasis knowledge which will bring us to Washington DC, USA, where the 3rd WBC will be held next July.

Funding

Publication costs of this article will be funded by Novamedia and Italian Respiratory Society (IRS).

About this supplement

This article has been published as part of Multidisciplinary Respiratory Medicine Volume 13 Supplement 1, 2018: Bronchiectasis: still an orphan disease?. The full contents of the supplement are available online at https://mrmjournal.biomedcentral.com/articles/supplements/volume-13-supplement-1.

Authors’ contributions

Both authors read and approved the final manuscript.

Ethics approval and consent to participate

Not applicable.

Consent for publication

Not applicable.

Competing interests

Speakers’ Advisory boards on bronchiectasis in the past two years for Bayer; Insmed; Chiesi, Menarini, Mundipharma, Glaxosmithkline, Astrazeneca, Grifols, Guidotti, Malesci, Zambon, Actavis, Novartis, Bi, Raptor, Horizon Pharma. In addition. SA and CJD are Guest Editor of the Supplement on Bronchiectasis, whereas SA is Associate editor of Multidisciplinary Respiratory Medicine.

Publisher’s Note

Springer Nature remains neutral with regard to jurisdictional claims in published maps and institutional affiliations.

Author details

‘Department of Pathophysiology and Transplantation, University of Milan, Internal Medicine Department, Respiratory Unit and Cystic Fibrosis Adult Center, Fondazione IRCCS Ca’ Granda Ospedale Maggiore Policlinico, Via Francesco Sforza 35, 20122 Milan, Italy. ‘Tayside Respiratory Research Group, University of Dundee, Dundee DD1 9SY, UK.

Published: 9 August 2018

References

1. Chalmers JD, Aliberti S, Blasi F. Management of bronchiectasis in adults. Eur Respir J. 2015 May;45(S):1446–62.
2. McDonnell MJ, Aliberti S, Gomimine PC, Restrepo MI, Finch S, Pesci A, Dupont LJ, Fardon TC, Wilson R, Loebinger MR, Skrbic D, Obradovic D, De Soyza A, Ward C, Laffey JG, Rutherford RM, Chalmers JD. Comorbidities and the risk of mortality in patients with bronchiectasis: an international multicentre cohort study. Lancet Respir Med. 2016;4(12):969–79.
3. Quint JK, Millett ER, Joshi M, Navaratnam V, Thomas SL, Hurst JR, Smeeth L, Brown JS. Changes in the incidence, prevalence and mortality of bronchiectasis in the UK from 2004 to 2013: a population-based cohort study. Eur Respir J. 2016 Jan;47(1):186–93.
4. Loebinger MR, Wells AU, Hansell DM, et al. Mortality in bronchiectasis: a long-term study assessing the factors influencing survival. Eur Respir J. 2009;34:843–9.
5. Aliberti S, Hill AT, Mantero M, Battaglia S, Centannini S, Lo Cicero S, Lacedonia D, Saetta M, Chalmers JD, Blasi F, SIP Bronchiectasis Audit Working Group. Quality standards for the management of bronchiectasis in Italy: a national audit. Eur Respir J. 2016 Jul;48(1):244–8.
6. Chalmers JD, Aliberti S, Polverino E, Vendrell M, Crichton M, Loebinger M, Dimaou K, Clifton I, van der Eerden M, Rohde G, Murris-Espin M, Masefield S, Gerada E, Shteinberg M, Ringshausen F, Haworth C, Boersma W, Rademaker J, Hill AT, Aksamit T, O’Donnell A, Morgan L, Milenkovic B,
Tramma L, Neves J, Menendez R, Paggiaro P, Botnaru V, Skrgat S, Wilson R, Goeminne P, De Soyza A, Welte T, Torres A, Elborn JS, Blasi F. The EMBARC European bronchiectasis registry: protocol for an international observational study. ERJ Open Res. 2016;2(1).

7. Aliberti S, Masefield S, Polverino E, De Soyza A, Loebinger MR, Menendez R, Ringshausen FC, Vendrell M, Powell P, Chalmers JD, EMBARC Study Group. Research priorities in bronchiectasis: a consensus statement from the EMBARC clinical research collaboration. Eur Respir J 2016;48(3):632–47.

8. Chalmers JD, Timothy A, Polverino E, Almagro M, Ruddy T, Powell P, Boyd J. Patient participation in ERS guidelines and research projects: the EMBARC experience. Breathe (Sheff). 2017;13(3):194–207.

9. Hill AT, Haworth CS, Aliberti S, Barker A, Blasi F, Boersma W, Chalmers JD, De Soyza A, Dimakou K, Elborn JS, Feldman C, Flume P, Goeminne PC, Loebinger MR, Menendez R, Morgan L, Murris M, Polverino E, Quitter A, Ringshausen FC, Tino G, Torres A, Vendrell M, Welte T, Wilson R, Wong C, O’Donnell A, Aksamit T, EMBARC/BRD definitions working group. Pulmonary exacerbation in adults with bronchiectasis: a consensus definition for clinical research. Eur Respir J. 2017;49(6).

10. Chalmers JD, Ringshausen FC, Harris B, Elborn JS, Posthumus A, Haworth CS, Pilkington N, Polverino E, Ruddy T, Aliberti S, Goeminne PC, Winstanley C, De Soyza A. Cross-infection risk in patients with bronchiectasis: a position statement from the European Bronchiectasis Network (EMBARC), EMBARC/ELF patient advisory group and European Reference Network (ERN-Lung) Bronchiectasis Network. Eur Respir J. 2018;51(1).

11. Aliberti S, Lonni S, Dore S, McDonnell MJ, Goeminne PC, Dimakou K, Fardon TC, Rutherford R, Pesci A, Restrepo MI, Sotgiu G, Chalmers JD. Clinical phenotypes in adult patients with bronchiectasis. Eur Respir J. 2016;47(4):1113–22.

12. Lonni S, Chalmers JD, Goeminne PC, McDonnell MJ, Dimakou K, De Soyza A, Polverino E, Van de Kerkhove C, Rutherford R, Davison J, Rosales E, Pesci A, Restrepo MI, Torres A, Aliberti S. Etiology of non-cystic fibrosis bronchiectasis in adults and its correlation to disease severity. Ann Am Thorac Soc. 2015;12(2):1764–70.

13. Araújo D, Shteinberg M, Aliberti S, Goeminne PC, Hill AT, Fardon T, Obradovic D, Dimakou K, Polverino E, De Soyza A, McDonnell MJ, Chalmers JD. Standardised classification of the aetiology of bronchiectasis using an objective algorithm. Eur Respir J. 2017;50(6).

14. Polverino E, Goeminne PC, McDonnell MJ, Aliberti S, Marshall SE, Loebinger MR, Murris M, Cantón R, Torres A, Dimakou K, De Soyza A, Hill AT, Haworth CS, Vendrell M, Ringshausen FC, Subotic D, Wilson R, Vilaró J, Stallberg B, Welte T, Rohde G, Blasi F, Elborn S, Almagro M, Timothy A, Ruddy T, Tonia T, Rigau D, Chalmers JD. European Respiratory Society guidelines for the management of adult bronchiectasis. Eur Respir J. 2017;50(3).

15. Robinson P, Morgan L. Bronchiectasis in PCD looks different to CF on CT scan. Multidiscip Respir Med. 2018;13(Suppl 1). https://doi.org/10.1186/s40248-018-0139-2.

16. Contarini M, Shoemark A, Rademacher J, Finch S, Gramegna A, Gaffuri M, et al. Why, when and how to investigate primary ciliary dyskinesia in adult patients with bronchiectasis. Multidiscip Respir Med. 2018;13(Suppl 1). https://doi.org/10.1186/s40248-018-0143-6.

17. Gramegna A, Aliberti S, Seia M, Pocar O, Bianchi V, Castellani C, et al. When and how ruling out cystic fibrosis in adult patients with bronchiectasis. Multidiscip Respir Med. 2018;13(Suppl 1). https://doi.org/10.1186/s40248-018-0142-7.

18. Faverio P, Stainer A, Bonatti G, Zucchetti SC, Simonetta E, Lapadula G, Marruchella A, Gori A, Blasi F, Codecasa L, Pesci A, Chalmers JD, Loebinger MR, Aliberti S. Characterizing non-tuberculous mycobacteria infection in bronchiectasis. Int J Mol Sci. 2016;17(1).

19. Bonatti G, Pesci A, Marruchella A, Lapadula G, Gori A, Aliberti S. Nontuberculous mycobacteria in noncystic fibrosis bronchiectasis. Biomed Res Int. 2015;2015:197950.

20. Aliberti S, Ruffo Codecasa L, Gori A, Sotgiu G, Spotti M, Di Biagio A, et al. The Italian registry of pulmonary non-tuberculous mycobacteria - IRENE: the study protocol. Multidiscip Respir Med. 2018;13(Suppl 1). https://doi.org/10.1186/s40248-018-0141-8.

21. Taylor SL, Leong LE, Mobegi F, Choo JM, Burr LD, Wesselingh S, et al. Understanding the impact of antibiotic therapies on the respiratory tract resistome: a novel pooled-template metagenomic sequencing strategy. Multidiscip Respir Med. 2018 (Suppl 2:xx):1-30. In press.

22. Chalmers JD, Chotirmall SH. Bronchiectasis: new therapies and new perspectives. Lancet respir med. 2018; 10.1016/S2213-2600(18)30053-5.