'World Bronchiectasis Day': The Indian perspective

Global lung health organisations from the USA, Europe, Australia and Asia joined together to declare 1 July 2022 as the first 'World Bronchiectasis Day'to raise awareness about bronchiectasis. The motive of the day is to emphasise on the increased prevalence of bronchiectasis globally and to increase awareness and promote effective diagnosis and management of this rather neglected disease. Proposed to be celebrated each year, the day is intended to improve the lives of patients and their families worldwide.^[1]

Bronchiectasis is a chronic respiratory disease characterised by a clinical syndrome of cough, sputum production and bronchial infection, and radiologically by abnormal and permanent dilatation of the bronchi.^[2] Bronchiectasis imposes a significant burden on patients with respect to symptoms, lung function, exacerbations and quality of life. Although there is currently no cure, detecting and treating bronchiectasis early can improve the quality of life and may impact the longevity of those affected.^[2] The disease is broadly categorised as cystic fibrosis (CF) bronchiectasis and non-cystic fibrosis bronchiectasis (NCFB). CF is inherited in an autosomal recessive manner. It is caused by the presence of mutations in both copies of the gene for the CF transmembrane conductance regulator (CFTR) protein. This disease affects mostly the lungs, but also the pancreas, liver, kidneys and intestine. The emphasis has always been on the management of CF, even though the morbidity with NCFB is no less profound. 'World Bronchiectasis Day' is, however, more about NCFB, which is widely recognised both in the developed and the developing world.

In the last decade, awareness of bronchiectasis in adults and children has grown due to an increase in clinical research in the field of bronchiectasis. The research was initially mainly confined to the developed countries, but has embraced the developing world including India as well. An important milestone in the field was the foundation of the European Multicentre Bronchiectasis Audit and Research Collaboration (EMBARC), which promotes multidisciplinary collaborative research in NCFB with substantial patient and physician involvement.^[3] The EMBARC family now embraces virtually all of Europe, USA, parts of Latin America, India, Korea and so on. The Indian registry was designed to utilise identical definitions and data fields as the European registry, which in turn was designed to mirror data from the US registry. This is true of all data sets collected throughout the world.^[4]

Comparison of data showed that Indian patients with bronchiectasis were found to differ from those in the developed world (West Europe and USA) in many ways. Patients in India were younger (by about 10 years; mean age 56 years) and more likely to be men (56.9%). Previous tuberculosis (35.5%) was the most frequent underlying cause of bronchiectasis. While Pseudomonas aeruginosa was the most common organism (similar to global data) in sputum culture (13.7%), culture positivity was far lower in the Indian registry. The Indian patients were also different in growing Enterobacteriaceae (esp. Klebsiella) in 10% of cases. Organisms commonly reported in Western Europe and US cohorts, such as Haemophilus influenzae (11 [0.5%] of 2195 patients), Moraxella catarrhalis (22 [1.0%]), Streptococcus pneumoniae (18 [0.8%]), Staphylococcus aureus (50 [2.3%]) and non-tuberculous mycobacteria (8 [0.4%]), were uncommon in the Indian cohort.^[5] The follow-up results showed that on multivariate analysis, factors like Enterobacteriaceae infection, previous exacerbations, cardiovascular disease, diabetes, MRC dyspnoea score and current smoking were all predictors of mortality.

In light of its prevalence globally, bronchiectasis should no longer be considered an 'orphan lung disease'. For instance, the prevalence of bronchiectasis has been found to range from 67 to 566.1 per 100,000 inhabitants in Europe and North America^[6-9] and has reached 1200 per 100,000 inhabitants among those aged 40 years or older in China.^[10] In fact, bronchiectasis is one of the three most common chronic airway inflammatory diseases (along with chronic obstructive pulmonary disease [COPD] and asthma) that are gaining global attention. Understanding the disease burden is imperative for the improvement of the global management of bronchiectasis.

Bronchiectasis imposes a substantial economic burden, which although well recognised in the western populations, is relatively unaddressed in developing countries like India. The healthcare expenditure of these patients is higher than that of matched controls, the cost of hospitalisation is 56% higher and the cost of mucoactive drugs, inhalation and home ventilation devices and antibiotics is three to five times higher.^[11] These patients also have a higher length of hospital stay and 5.8% higher annualised mortality compared to matched controls, putting a substantial economic burden on the patients and the healthcare system. Against the backdrop of sparse data, it is important to recognise that the true economic burden of bronchiectasis is likely to be underestimated because most studies have been retrospective, have used ICD-9-CM coding to identify patients and have often ignored outpatient burden and cost.

The aetiology of bronchiectasis in developing countries is mostly idiopathic, which requires appropriate attention as it might reflect a rather inadequate inquiry into the aetiologies. The EMBARC India registry demonstrated that 50% patients evaluated for ABPA had the disease (indicating likely high prevalence) and a very small minority had their immunoglobulin levels assessed, facts indicative of less-than-adequate assessment. Such data call for an emphasis on adequate workup of the patients. Although advances in the management of bronchiectasis are positive developments, there remain persistent ethnic, socioeconomic and geographical disparities in the diagnosis and management of bronchiectasis. Genomics, metabolomics and microbiomics can be used to predict the risk of disease and treatment response, and advances will no doubt continue to improve their accuracy, with the ultimate goal of the achievement of personalised approaches based on an individual's biology and not their ethnicity. The EMBARC India registry also showed that the management of the disease was suboptimal with more than 60% patients being treated in line with the management of OAD with an ICS or an ICS + LABA. This was despite the fact that only 35% of patients showed an obstructive pattern on spirometry. More evidence-based treatment, like low-dose macrolides, inhaled antibiotics like tobramycin and colistin, was used in less than 10% of cases. No airway clearance technique was prescribed in around 60% of patients. Hence, the management protocols for the disease also need to be far more standardised in our country.

The emerging data about the increasing prevalence of the disease call for significantly higher awareness and investments in research in the field, so that we understand the disease better, as it encounters us in the setting of developing countries, and inform our policies accordingly. With a large deficit in our understanding of the epidemiology, risk factors, causative factors, diagnosis and treatment of this rather uncommon disease, the coming together of multiple lung health organisations needs to percolate done to the physician bodies in developing countries too for a proper and deserved emphasis to the disease. Comprehensive guidelines need to be developed and modified based on the requirements of the local geographical area and updated regularly. Public health authorities could consider the distribution of HRCT scanners and microbiology labs in accordance with the prevalence and geographical distribution of bronchiectasis in various regions of the country. Educational programmes are needed to raise the awareness of the physicians and the public alike regarding bronchiectasis and the importance of continuous monitoring and maintenance therapy for patients with moderate or severe disease. 'World Bronchiectasis Day' represents a major step in our endeavours towards understanding of the disease and may be just the reminder that the time is now just opportune for major steps forward in the field, so that the 'orphan' disease does not stay 'orphan' anymore.

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